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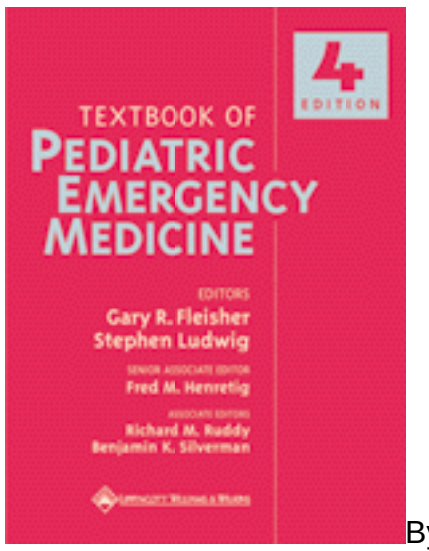

By OkDoKeY

Textbook of Pediatric Emergency Medicine

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Preface

With age comes stability. Relatively profound structural changes occurred nationally during the years of development of the three prior editions of the *Textbook of Pediatric Emergency Medicine*, including burgeoning numbers of practitioners, the establishment of a specialty board, and most important, recognition and broadening of the mission for pediatric emergency medicine. This fourth edition arrives on the scene less than 20 years after the first, which essentially coincided with the birth and initial recognition of the subspecialty. Although growth and refinement of the art proceed at a gratifying pace, this edition appears at a time of consolidation of the gains of the prior two decades--from infancy to maturity.

Throughout the text, we have updated extensively but innovated selectively. We have continued to recruit authors who are fellowship-trained, board-certified and board-qualified pediatric emergency physicians. Fortunately, this task grows easier each time around. We believe you will agree that the chapters written by these new contributors reflect the skills that they acquired in the trenches, and emphasize the perspective of pediatric emergency medicine.

We have made some important additions. You will find six new chapters. The first, in the section on “Life-Threatening Emergencies,” is “Myocardial Infarction.” We have asked Baruch Krauss, MD, board-certified in family practice as well as pediatric emergency medicine, to discuss the immediate treatment of ischemic cardiac catastrophes, both to prepare us for the rare occurrence of this event during childhood and for the occasional parents and grandparents who show up in pediatric emergency departments, clutching their chests in pain. Then, under the section of “Signs and Symptoms” Anne Stack, MD and Mark Roback, MD provide an initial approach to two new problems, “Cyanosis” and “Oral Lesions,” the first quite serious, and the second less ominous but common, and at times confusing. Following, in the section on “Medical Emergencies,” you will find a chapter on “Neonatal Problems.” Our experience over the last five years, with the entrenchment of early discharge for newborns, is that parents are bringing their infants to the emergency department in many cases where they suddenly notice a color change, a rash, cranial swelling, jitteriness, or some other finding that represents an issue previously handled prior to discharge from the nursery on day 3 or day 4 of life. To address this topic, Benjamin K. Silverman, MD, shares his clinical acumen from 3 decades of pediatric practice and 15 years in pediatric emergency medicine. Also, we have identified an emerging area where parents call upon the pediatric emergency physician for unique expertise to manage the “Approach to the Care of the Technology-Dependent Child.” To address the diverse issues--many related to implanted hardware--that occur in these patients, Joel A Fein, MD, Kate Cronan, MD, and Jill Posner, MD, have written a new chapter for the section on “Surgical Emergencies.” Lastly, events around the world in the last five years have forced us to confront the medical consequences for children of chemical and biological terrorism. To address this, Chapter 132 reviews emergency department preparedness for such terrorism with an emphasis on pediatric issues.

Although the majority of the chapters are carried over from the third edition, you will find that they contain updated information and, not infrequently, address new diseases and treatments. For example, the chapter on “Infectious Diseases” discusses babesiosis and ehrlichiosis, two tick-borne infections that only recently have been well-described in children. In “Hematologic Emergencies,” a new option, using an anti-D antibody preparation instead of prednisone or intravenous gamma-globulin, is offered to simplify the treatment of the majority of cases of idiopathic thrombocytopenia, and 4-methylpyrazole is introduced in “Toxicologic Emergencies” for the management of poisoning with ethylene glycol. Thus, we believe you will encounter the newest and very latest information with each turn of a page in this fourth edition.

As a consequence of the aforementioned essential transition to authors trained in pediatric emergency medicine, some close friends and talented colleagues who served well as valued collaborators previously have not joined us in the current edition. We wish to thank each of these individuals for their immense contributions to the success and maturation of the prior editions. In many of the chapters that have changed hands, the new authors have followed our suggestions to retain those portions of the original framework, tables, and illustrations that have worked so well in the past and remain relevant. As editors, we wish to acknowledge our gratitude and debt to the former authors of these revised chapters.

Despite our protestations of constancy, you will encounter immediately one major change on the cover and title page. Jack Templeton, MD, who has labored with some of us since residency, and with the entire group throughout three editions, has relinquished his role as an editor. Several years ago Jack made the decision to retire from pediatric surgery at the peak of his illustrious career so that he could devote his full energy to the philanthropic efforts of the Templeton Foundation. Having chosen to work toward improving the health and well-being of humankind through a venue other than medicine, Jack asked us to reassign his responsibilities. Steve, Fred, Rich, Ben, and I cannot thank Jack enough for his tremendous assistance in sustaining the textbook throughout all these years.

Finally, we want to pay special recognition to two authors from the third edition, John Duckett, MD, and Edward B. Charney, MD, both now deceased. These two colleagues helped us to launch the textbook, and they would once again be among our contributors, were it not for their untimely deaths.

As editors, we rely on support from many sources to help us put forth a vital new edition of the textbook each time around. At home, our families tolerate our long absences during which we do our writing and fill us with the joy that motivates us in all our endeavors. At work, our colleagues, as well as our residents and fellows, allow us to spend some time in the “back office” and also offer up an endless flow of great ideas. Finally, at national meetings and through their correspondence, our readers have inspired us, by sharing their boundless enthusiasm for the textbook and their creative suggestions for future topics and alternative approaches. We have benefitted greatly from this feedback and hope that each of you will think about areas for expansion and/or revision whenever you consult the *Textbook of Pediatric Emergency Medicine*. You can rest assured that we have begun to plan the fifth edition and will look forward, once again, to receiving your input.

Acknowledgments

As we complete the final pages of the fourth edition of *Textbook of Pediatric Emergency Medicine*, it is more than appropriate to acknowledge and sincerely thank those around us who have made it all possible.

The writing and editing of this edition, like that of its predecessors, is a process that does not take place in the workplace. Most of the work was done during early morning hours, nights, and weekends. When deadlines drew close, it was done on vacations and on holidays. The precious commodity of time has been the very time that we have taken from our families. Their commitment to us and to our objectives and aspirations has been strong. Their donation of their own time has been magnanimous. Without their love and support our work would not have meaning. It is the treasure of our own families that drives us to help other children and families. To Jan and Zella, our love and gratitude. To Daniel, Carl, and Madeline Fleisher, and to Susannah, Elisa, and Aubrey Ludwig, our thanks for your sharing and understanding. Your only payback can come through the health and well-being of strangers of your generation and from those to come.

To our coeditors—our closest colleagues—we extend our greatest respect and thanks. You are the unsung heroes of this work. Fred Henretig's contributions have been so great that we have elevated his title to Senior Associate Editor. To Associate Editors Rich Ruddy and Ben Silverman, there are no appropriate accolades. We hope that your level of personal satisfaction fills the void of our inability to say enough about your skills and dedication to the project. May our careers continue to grow side by side.

During the years of the four editions there have been so many colleagues and coworkers, that it is impossible to name them all without forgetting some and thereby inadvertently offending them. They have worked by our side in the emergency department. They have taught with us in lectures, conferences, and workshops. They have served on committees, task forces, and boards. They have written chapters and rewritten them based on our whims and notions. They have covered some of our nights and weekends. They have taught us, questioned us, stimulated us, and always supported us. Many remain at our sides at Children's Hospital in Boston and the Children's Hospital of Philadelphia. But equally valuable are those who have moved to other centers of pediatric emergency care around the United States and around the world. Those who have moved beyond the "nests" are not forgotten. We continue to appreciate and acknowledge all.

We offer a special note of acknowledgment to our trainees. We appreciate all those we have encountered as medical students, residents, fellows, and continuing medical education students. We thank you all. It is you who have asked the questions. It is you who have longed for the information. You have held out the expectation that we provide the answers in an accurate and available form. We have also learned from our many thousands of our patients and their parents. They too have kept the bar of expectation high, forcing us to try to meet those expectations.

In each of our offices there has been a coworker of special patience and extraordinary skills. For this edition, Cindy Chow and Carolyn Trojan have gone above and beyond the call. We have not forgotten those who were there for other editions: Rose Beato, Pat Parkinson, and Carmen Christmas. Work without these devoted women would be unimaginable.

To our coworkers at Lippincott Williams & Wilkins, our thanks also. For this edition we appreciate your consistency and dedication to the project. In particular, we thank Tanya Lazar, Elizabeth Greenspan, Sonya Seigafuse, Lisa Franko, and Jeff Gruenglas.

A final note goes to our teachers, chairmen, and mentors. There are and have been many, and we thank you all. As Jean Cortner, MD, is about to retire, a special note of thanks goes out to him. He was there when it all began. He bet on two young faculty members. I hope he feels that his bet has had adequate pay out. And for each of us, there has been a special David in our careers. To the late David Cornfeld and the very active David Nathan, we often think about how you would have done it. It is the true sign of a mentor.

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Gary R. Fleisher, MD

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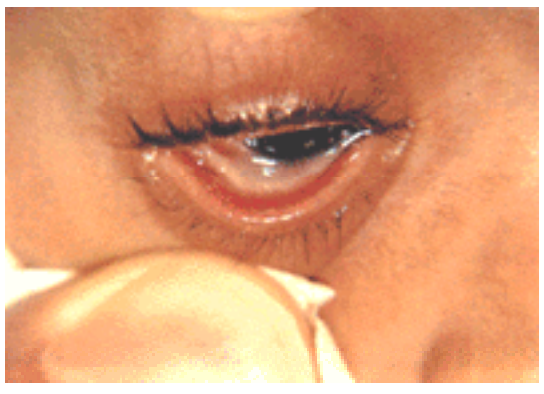
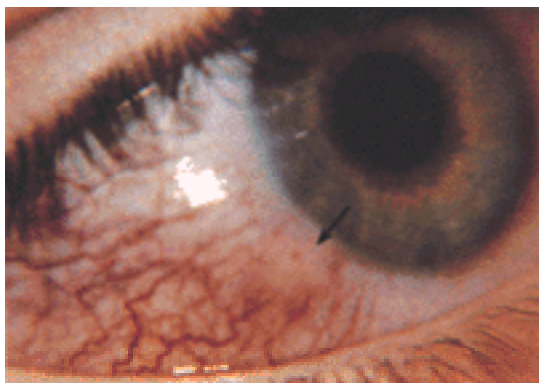
For their love and support, we dedicate this book to
our wives,
our children,
and our parents.

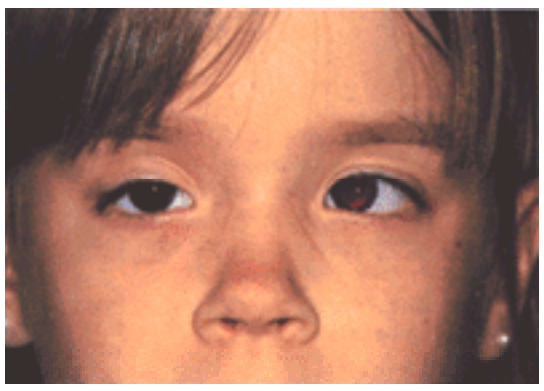
Color Plate



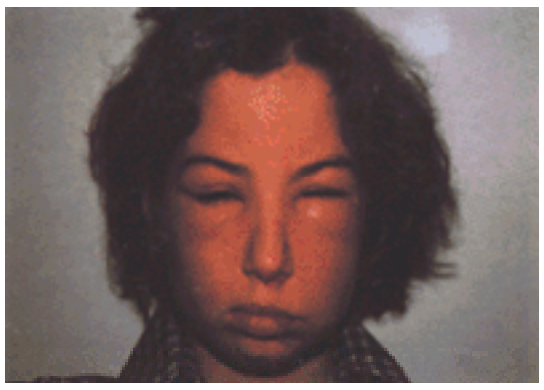
COLOR PLATE 24.1. Pseudomembrane on lower lid palpebral conjunctiva and extending into inferior fornix in patient with epidemic keratoconjunctivitis. (This figure is printed in black and white as FIGURE 24.1.)



COLOR PLATE 24.4. Red eye caused by chickenpox (varicella) involvement of conjunctiva. Note sectorial injection of conjunctiva. White area *(arrow)* at junction of conjunctiva and cornea is the pox lesion. (This figure is printed in black and white as FIGURE 24.4.)

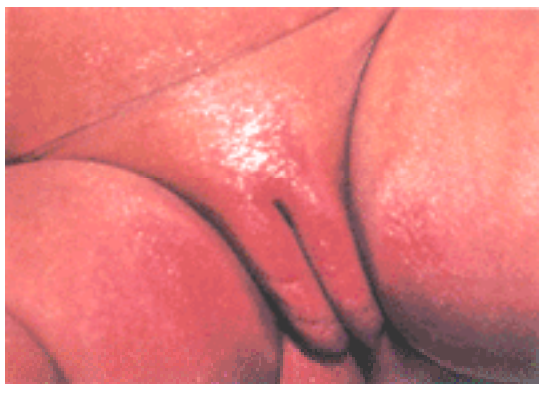


COLOR PLATE 25.6. Left esotropia. Note lateral displacement of Hirschberg light reflex in the left eye. Photograph demonstrates right ptosis and orange-red reflex in the left eye with black reflex in the right eye. Pupils are pharmacologically dilated. Asymmetry of reflex is caused by misalignment of the eyes. (This figure is printed in black and white as FIGURE 25.6.)

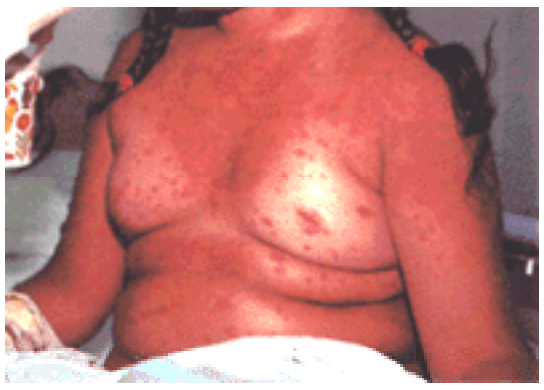


COLOR PLATE 99.11. Facial edema and inflammation in response to exposure to airborne contact allergen (e.g.,

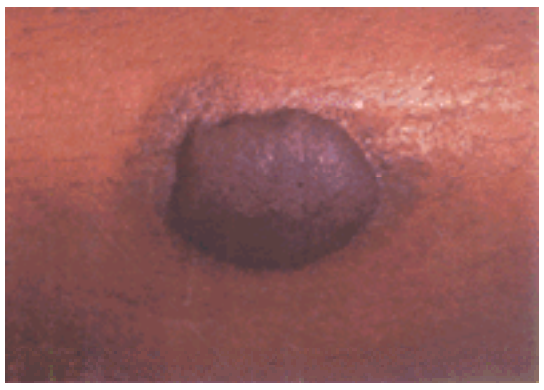
vaporized oil in smoke of burned poison ivy plants). (This figure is printed in black and white as FIGURE 99.11.)



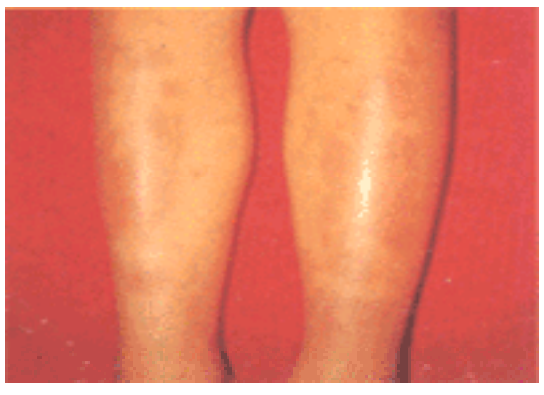
COLOR PLATE 99.14. Infant with occlusion diaper dermatitis. (This figure is printed in black and white as FIGURE 99.14.)



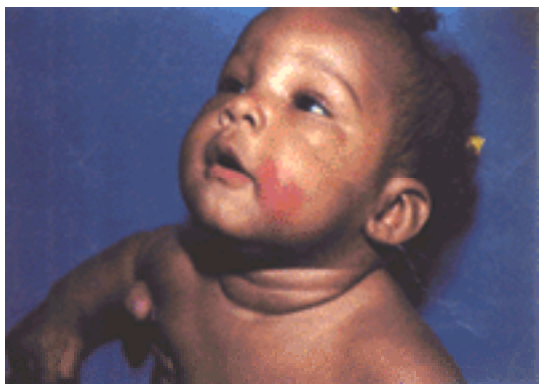
COLOR PLATE 99.19. Adolescent with Stevens-Johnson syndrome secondary to sulfonamides. Note involvement of mucous membranes of the mouth. (This figure is printed in black and white as FIGURE 99.19.)



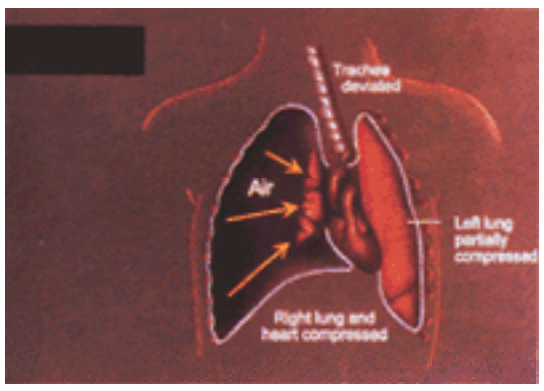
COLOR PLATE 99.20. Hemorrhagic bulla in patient with vasculitis. (This figure is printed in black and white as FIGURE 99.20.)



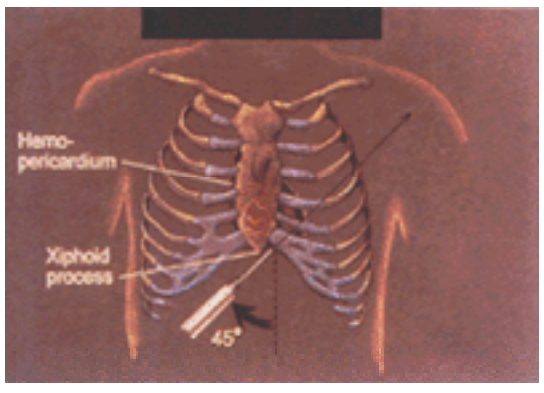
COLOR PLATE 99.21. Extensor surface involved with lesions of erythema nodosum. (This figure is printed in black and white as FIGURE 99.21.)



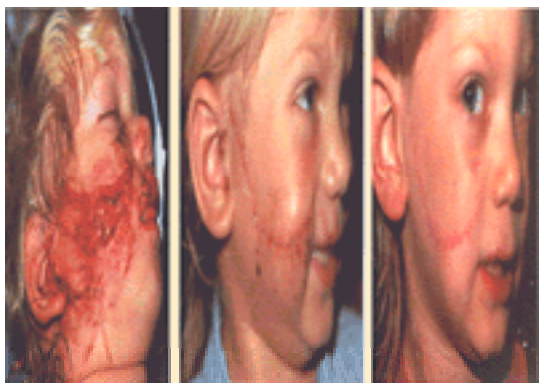
COLOR PLATE 99.41. Infant with popsicle panniculitis of the cheek. (This figure is printed in black and white as FIGURE 99.41.)



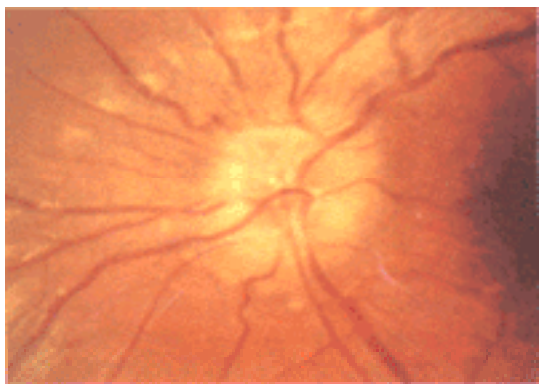
COLOR PLATE 107.1. Tension pneumothorax with a mediastinal shift. (This figure is printed in black and white as FIGURE 107.1.)



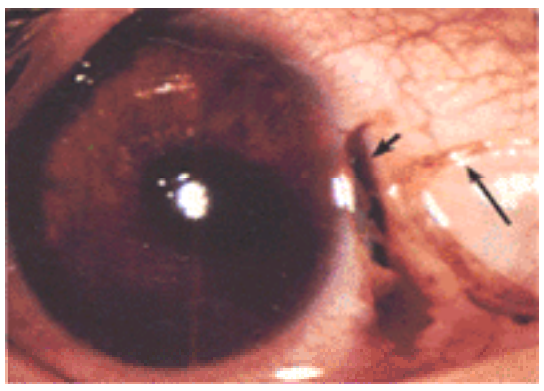
COLOR PLATE 107.20. Pericardiocentesis is performed by inserting a 20-gauge spinal needle below the xiphoid process at a 45-degree angle toward the left shoulder. (This figure is printed in black and white as FIGURE 107.20.)



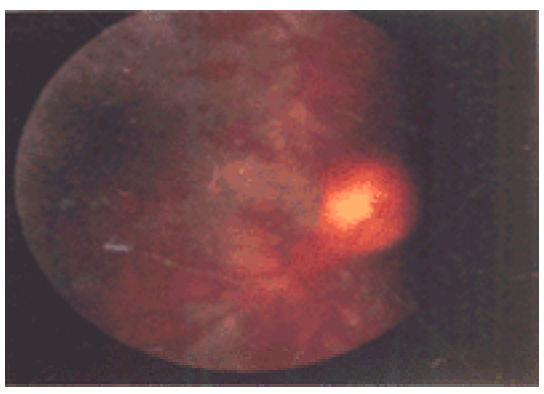
COLOR PLATE 110.13. Photographs of a 3-year-old boy after an attack by a dog. Child was evaluated in the pediatric emergency room, intravenous antibiotics were given, his facial wounds were irrigated, and a plastic surgery consultation was made. The **left** photograph shows the child in the operating room before sharp debridement, facial nerve exploration, and an exacting layered closure of his complex wound. The **middle** panel pictures the child 1 week after his repair and demonstrates the precise reapproximation of the facial soft tissues. The **right** photograph was taken 8 months after the attack and demonstrates a nicely healing facial scar that will continue to fade and soften. (Courtesy of David W. Low, MD.) (This figure is printed in black and white as FIGURE 110.13.)



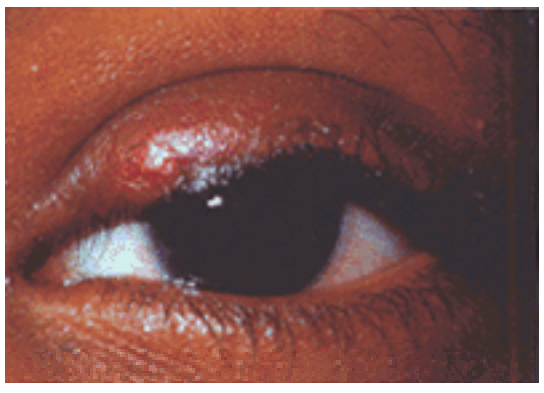
COLOR PLATE 111.3. Papilledema. Note blurred disc margins and loss of view of blood vessels on disc. (This figure is printed in black and white as FIGURE 111.3.)



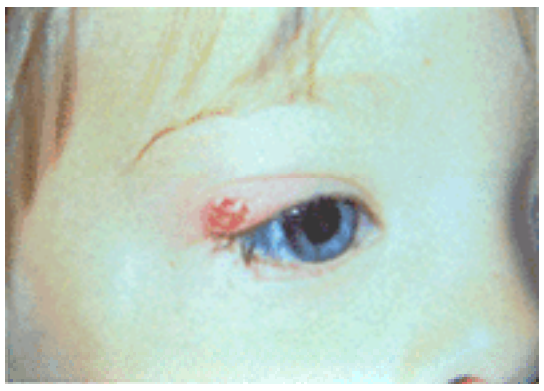
COLOR PLATE 111.7. Ruptured globe. The scleral laceration *(short arrow)* appears as a linear brown line on the white of the eye. The pupil has a teardrop shape, the apex of which points in the direction of the rupture. The *long arrow* points to the upper border of a large conjunctival laceration. Note that the underlying sclera is intact. There is a diffuse hyphema in the anterior chamber, which partially obscures the pupil. (This figure is printed in black and white as FIGURE 111.7.)



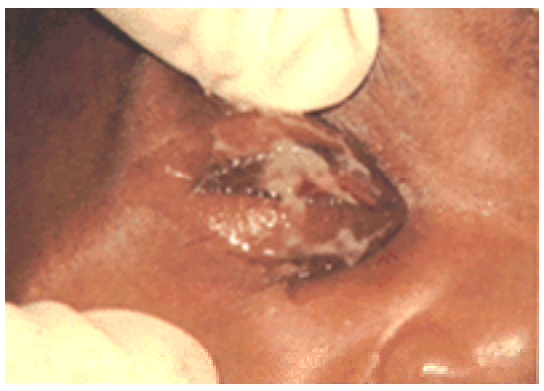
COLOR PLATE 111.14. Retinal hemorrhages in shaken baby syndrome. (This figure is printed in black and white as FIGURE 111.14.)



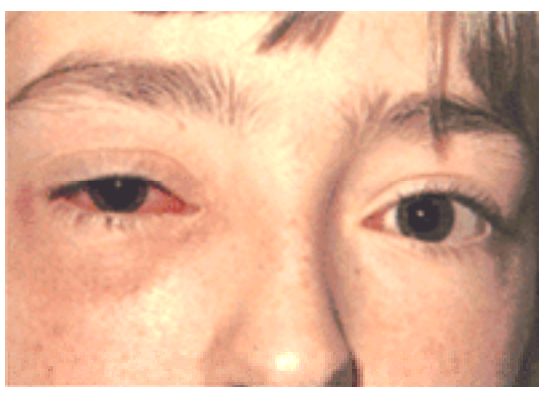
COLOR PLATE 120.4. Acute sty (hordeolum). (This figure is printed in black and white as FIGURE 120.4.)



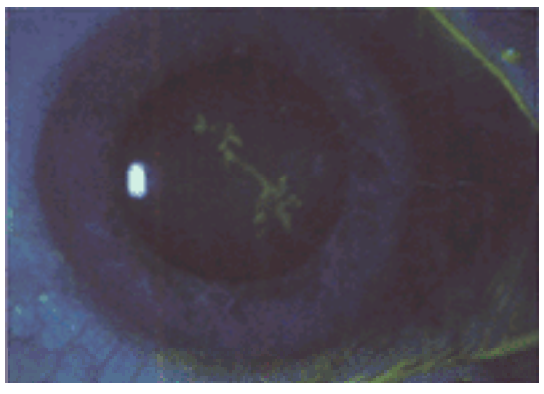
COLOR PLATE 120.5. Chalazion draining spontaneously via skin. (This figure is printed in black and white as FIGURE 120.5.)



COLOR PLATE 120.7. Neonatal gonorrheal conjunctivitis. Note the dramatic lid swelling and severe purulent discharge. (This figure is printed in black and white as FIGURE 120.7.)



COLOR PLATE 120.8. Patient with right epidemic keratoconjunctivitis infection. Note the lid swelling, red eye, and absence of purulent discharge. Patient also has right preauricular adenopathy (not visible). Note the early injection of left eye, representing sequential involvement. (This figure is printed in black and white as FIGURE 120.8.)



COLOR PLATE 120.10. Fluorescein staining pattern of herpes simplex virus corneal infection. Eye is illuminated with blue light to demonstrate yellow/green branching staining pattern of herpetic dendrite. (This figure is printed in black and white as FIGURE 120.10.)

CHAPTER 1

Resuscitation—Pediatric Basic and Advanced Life Support

STEPHEN LUDWIG, MD

Departments of Pediatrics and Emergency Medicine, The University of Pennsylvania School of Medicine, and The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania

Cardiopulmonary resuscitation (CPR) is a series of interventions aimed at restoring and supporting vital function after apparent death. The urgent and immediate goal of resuscitation is to reestablish substrate delivery to meet the metabolic needs of the myocardium, brain, and other vital organs. The overall goal is to return the child to society without morbidity related either to the underlying disease process or to the resuscitation process.

Instruction in CPR techniques has come from two widely disseminated national courses: Pediatric Advanced Life Support (PALS) and Advanced Pediatric Life Support (APLS). Both of these courses have been successful in training health care providers in the appropriate resuscitative techniques. Both courses stress the early recognition of the child who is in need of resuscitative efforts. The outcome of resuscitation in situations in which there has been prolonged asystolic arrest is poor. As in many conditions we manage in pediatric patients, primary prevention, or at least early recognition, is the most successful strategy.

Pediatric CPR has been based largely on the adult model of resuscitation. There is an orderly progression through the assessment and management of the ABCs—airway, breathing, and circulation. Like its adult counterpart, pediatric CPR is best performed by a well-coordinated team of physicians, nurses, respiratory therapists, and other support personnel.

BACKGROUND

Incidence

There are no incidence data for pediatric resuscitations performed annually in the United States. We can estimate the incidence of potential resuscitations by examining the data on infant and childhood mortality. Table 1.1 shows the childhood mortality rates and the leading causes of death for children. Table 1.2 shows the leading causes of death in the United States in different age groups in 1996 and the relative rates per population. Note that there is a relatively higher mortality rate for young children. Note also that, of the causes of death listed in Table 1.1, most are potentially reversible. Trauma is the leading cause of death in childhood. Special techniques of trauma management are presented in Section IV. We believe that the techniques of basic and advanced life support, when readily available and skillfully applied, contribute to the significant reduction in childhood mortality.

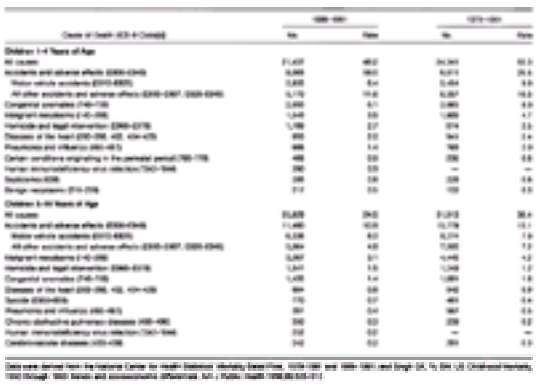


Table 1.1. U.S. Deaths and Death Rates per 100,000 Population for Children 1 to 4 and 5 to 14 Years of Age, for

the 10 Leading Causes of Death, 1979 through 1991

Birth–1 (70)*	1–4 (3)	5–9 (1.5)	10–14 (2.2)	15–19 (7)
Congenital anomalies	Injury	Injury	Injury	Injury
Prematurity	Congenital anomalies	Neoplasm	Neoplasm	Homicide
SIDS	Malignant neoplasm	Congenital anomalies	Homicide	Suicide
Respiratory distress syndrome	Homicide	Homicide	Suicide	Neoplasm
Newborn affected by maternal complications	Heart disease	Heart disease	Heart disease	Heart disease

*Rate per 100,000 population in specified group.

Table 1.2. Causes of Death: Five Leading Causes in United States, 1996

Patient Characteristics

Age

In many series of CPR cases, most children were at the younger end of the pediatric age range. In a series published from The Children's Hospital of Philadelphia, the mean age was 1.98 years and the median was 5 months. The age range was between 2 weeks and 16 years (Fig. 1.1). Although pediatric CPR education generally should be tailored to the anatomic and physiologic characteristics of the young child, emergency department staff must be prepared to cope with the full spectrum of age and size.

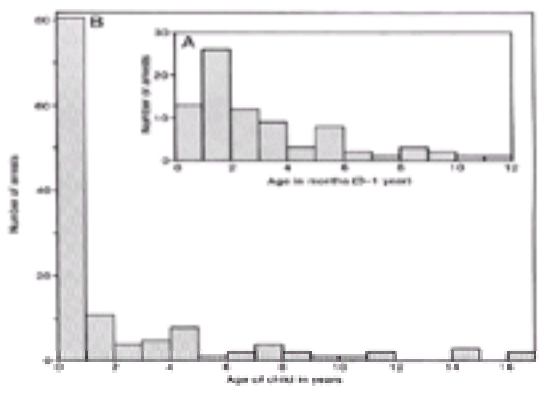


FIGURE 1.1. Histogram showing frequency of cardiac arrest as related to age in months **(A)** and years **(B)** from The Children's Hospital of Philadelphia survey.

Etiology

The most common primary diagnoses of hospitalized pediatric patients requiring resuscitation involve the respiratory system (Table 1.3). Conditions such as pneumonia, bronchiolitis, asthma, aspiration, and respiratory distress syndrome account for the largest group of diagnoses. Cardiac diagnoses and central nervous system (CNS) disorders occur in roughly equal frequency, but half as often as respiratory diagnoses. Common cardiovascular diagnoses include congenital heart disease, septic shock, and severe dehydration. CNS diagnoses include hydrocephalus (ventricular shunt failure), meningitis, seizure, and tumor.

Respiratory	*Central Nervous System*
Pneumonia	Acute hydrocephalus
Aspiration	Head trauma
Asthma	Seizure
Epiglottitis	Tumor
Laryngotracheobronchitis	Meningitis
Respiratory failure/chronic lung disease	Hemorrhage
Bronchiolitis	*Gastrointestinal*
Botulism	Trauma
Primary apnea	Enterocolitis
Bronchopulmonary dysplasia	Bowel perforation
Cardiovascular	Bowel obstruction
Congenital heart disease	Tracheoesophageal fistula
Septic shock	*Miscellaneous/Multisystem*
Dehydration	Sudden infant death syndrome
Pericarditis	Drug ingestion
Congestive heart failure	Tumors (non-CNS)
Myocarditis	Multiple trauma

From The Children's Hospital of Philadelphia 1979–1980.

Table 1.3. Diagnoses of Children Requiring Life Support by Body System

In the emergency department, the physician is more likely to encounter children whose cardiac arrest results from trauma, sudden infant death syndrome (SIDS), or unknown causes. Children with congenital anomalies, chronic sequelae of prematurity, and birth trauma, and those with chronic relapsing disease are also seen in the emergency department, as increasing numbers of children have survived the neonatal period, transplantation, complex surgery, and cancer therapy and have been discharged from the hospital. The broad range of diagnoses encountered in our review of resuscitation is noted in Table 1.3. This clearly differs from the adult circumstance of resuscitation, in which case most arrests are related to myocardial infarction secondary to coronary artery disease.

The PALS course teaches that the many etiologies of arrest follow one of two pathways: respiratory distress to respiratory failure to arrest or circulatory compromise to circulatory failure to arrest. In our experience, 80% of children who have arrested have followed the first pathway (Fig. 1.2). Twenty percent of patients follow the circulatory failure pathway to arrest. It is difficult in some cases to determine which mechanism was primary.

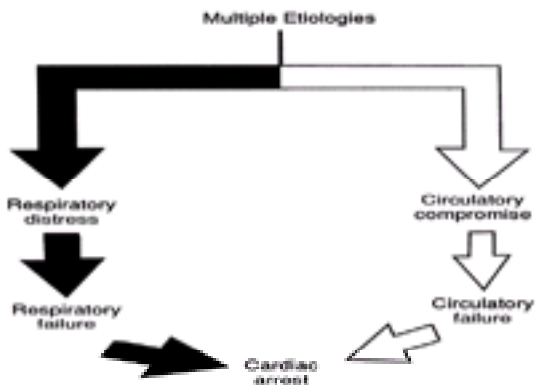


FIGURE 1.2. Pathophysiologic pathways from etiologies to cardiac arrest. (Adapted from P.A.L.S., American Heart Association.)

Demographics

There are no national demographic studies to identify socioeconomic, racial, familial, or community characteristics of the pediatric patient who requires life support intervention. Such studies would be important for developing profiles of the high-risk patient population for subsequent development of surveillance or prevention programs. It is most important to study such factors on a local level, where local solutions may be implemented. Some states have begun child death review efforts.

Treatment

Pediatric CPR presents the emergency physician with several complexities and frustrations. The first difficulty often encountered is that the patient has received only minimal prehospital care. Although this is changing with enhanced development of emergency medical services for children (EMS-C), pediatric patients are likely to be brought to the emergency department without the same field treatment that adult patients receive routinely. In many locales, paramedics are trained and equipped primarily to provide adult life support; however, they may be forced to initiate pediatric life support at the most basic level. In other areas, paramedics are limited by laws, regulations, or negative attitudes. Absent or inadequate prehospital care leads to longer periods of hypoxia and hypoperfusion, which directly affect prognosis and CNS morbidity (see Chapter 6). Seidel has documented the difference in survival rates between traumatized adults and children who require prehospital care as well as the lack of adequate pediatric equipment in prehospital care systems. Other investigators have documented the deficiencies in emergency care provided in free-standing emergency care centers and in primary care providers' offices.

The wide spectrum of age and diagnoses adds additional complexity. The resuscitation team must provide an array of technical skills, drugs, and equipment. Without delay, the team must have the flexibility to adjust to the correct sizes and drug dosages for children.

Our experience shows that careful management of airway and breathing is extremely important. Because the cause of the arrest is often related to respiratory failure and because the child's myocardium is relatively resilient to hypoxemia, the rapid correction of hypoxemia may be all that is necessary to effect resuscitation. In one-third of the resuscitations we reviewed, resuscitation was effected with only oxygen and positive-pressure ventilation.

For those patients who do not respond to airway and breathing management alone, life support will be significantly more difficult. In the emergency department, the lack of an immediate patient response usually predicts a need for multiple drug interventions. Our patients received a mean number of 4.5 drugs per patient.

One of the common frustrations when administering drugs is the establishment of an intravenous line. This technical skill continues to be the most common obstacle toward achieving successful CPR. However, the use of intraosseous technique and central line placement have been great advances in the solution of the access problem.

Arrhythmia management is generally not a problem in pediatric life support. The absence of atherosclerotic vascular disease makes the child's myocardium less susceptible to arrhythmia. As a result, antiarrhythmic medications and defibrillation are used infrequently. The most common cardiac rhythms to be recognized and managed are sinus bradycardia and asystole. The expectations to this are those children with congenital heart disease (preoperative and

postoperative) and those who have sustained direct myocardial trauma (see Chapter 82). These children may have unusual and difficult arrhythmias that require esoteric management to achieve a successful outcome.

Perhaps the greatest difficulty comes not with specific knowledge or technical skill, but with attitude. Many emergency departments are unaccustomed to resuscitating children and will become immobilized when faced with the task. There is a fear that the child is somehow more fragile. In other circumstances, there is overcompensation to the point that the resuscitation of a child is prolonged beyond an optimal point for either the child or the family.

On the other hand, many pediatric emergency physicians feel uncomfortable with the adult patient who may be a visitor at the pediatric hospital yet is brought to the emergency department with acute chest pain and possible myocardial infarction. Chapter 8 serves as a brief review of this important topic.

The emergency department team should review the effectiveness of each individual resuscitation effort as well as the emergency department's collective effort. This audit may be accomplished using one or more of the following approaches: 1) postresuscitation conference; 2) review of the videotape recording of resuscitation; 3) monthly morbidity mortality conferences; 4) chart audit; 5) review of resuscitation database (e.g., cross-referencing morbidity and mortality with various shifts, personnel teams, and prehospital treatment); and 6) performance on practice codes. When the team recognizes that its efficiency and effectiveness with children are less than ideal or less than they are with adults, specific remedial education should be undertaken.

Prognosis

The outlook for survival after CPR is very good for pediatric patients. In our experience, if the arrest is recognized rapidly and managed skillfully, immediate survival may be as high as 90% and the survival to discharge rate 60%. These figures are based on a hospitalized population of children who require resuscitation. For patients in the emergency department, the outcome is not as good. Children who arrive in the emergency department in asystolic arrest have a poor prognosis. In our small prospective series, patients who had no cardiovascular response to two standard doses of epinephrine were unable to be resuscitated. In the emergency department, the immediate survival may be as high as 60%, whereas the survival to discharge rate is 40%. The poorer prognosis for patients in the emergency department may be attributed to delayed recognition of the arrest and to limited prehospital care. Other series have documented an even more grim prognosis as shown in Table 1.4.

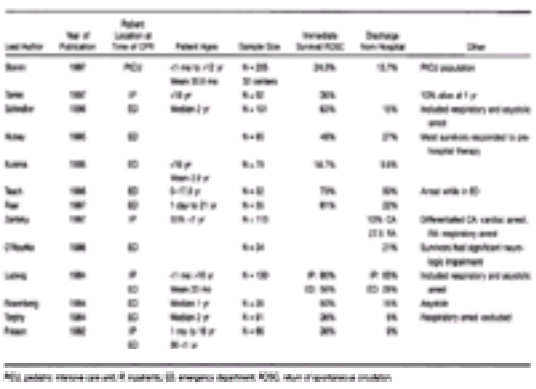


Table 1.4. Case Series and Outcomes of Pediatric Cardiopulmonary Resuscitation

Research

CPR research is extremely difficult to perform. Most of our information is based on retrospective studies like those reported in Table 1.4. Patient populations, characteristics, terminology, and methodology vary among studies, making it difficult to compare one investigator's work with another's. Performing a prospective study is challenged by legal and ethical considerations in enlisting patients at a time when “informed consent” is impossible. A recent conference was held and a Special Report issued to bring uniformity to the terminology of CPR research. This important report may bring more clarity to the CPR research of the future.

CLINICAL MANIFESTATIONS

Infants and children who have experienced disruption of oxygen or glucose delivery to the brain may benefit from the various elements of basic or advanced cardiac life support. The clinical manifestations of those requiring immediate life support are most often related to failure of oxygen delivery to the skin, brain, kidneys, and cardiovascular system. Cutaneous manifestations of oxygen deprivation include circumoral pallor, grayish hue, cyanosis, diaphoresis, mottling, and poor capillary refill. Manifestations of CNS hypoxia include irritability, confusion, delirium, seizures, and unresponsiveness. Cardiovascular manifestations include tachycardia, diaphoresis, bradycardia, and hypotension. Figure 1.3 shows the sequential development of signs and symptoms when there is failure of substrate delivery to different oxygen systems.

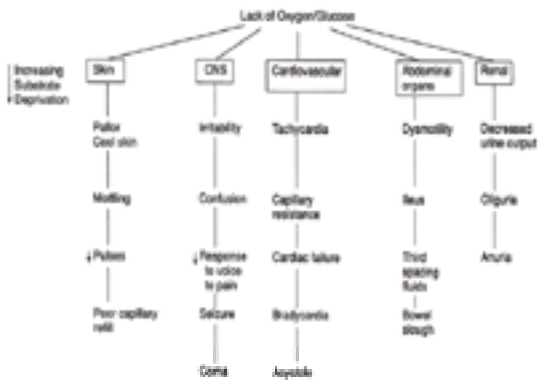


FIGURE 1.3. Signs and symptoms of lack of substrate delivery to vital organ systems.

Glucose is the second essential substrate necessary for maintenance of CNS integrity. Severe hypoglycemia may be just as devastating as severe hypoxemia. Clinical manifestations are often similar to hypoxemia because the primary effect on the CNS is coma. In addition, the effect of hypoglycemia on the cardiovascular system may lead to a secondary failure of oxygen delivery because of hypotension and related hypoperfusion.

A patient who has experienced a failure of substrate delivery to the central circulation must be resuscitated or supported until more specific diagnosis and management can be determined. It is also essential to identify patients who are *at risk* for failure of substrate delivery. This can be accomplished by a physical examination with emphasis on evaluation for airway patency, gas exchange, and cardiovascular integrity. In detecting those at risk, pulse oximetry, if available, may be useful in identifying mild degrees of hemoglobin desaturation. In addition, the laboratory may be helpful because patients with a low partial pressure of arterial oxygen (Pa_{O_2}), pH, glucose, hemoglobin, hemoglobin saturation, or high Pa_{O_2} are at risk. Also, recognition of certain disease entities allows early intervention, careful monitoring, and prevention of cardiovascular collapse. Examples include croup, airway foreign body, meningitis, and increased intracranial pressure (ICP).

MANAGEMENT

Management Sequence

Once a child has been identified as requiring life support, a sequence of evaluations and interventions should be accomplished (Fig. 1.4). Initially, CNS integrity must be evaluated: Is the patient alert? Does he or she respond to a shout or painful stimulus? If there is no response, the physician assumes that the brain is no longer receiving an adequate amount of oxygen, and the three basic sequences of evaluation and management are initiated.

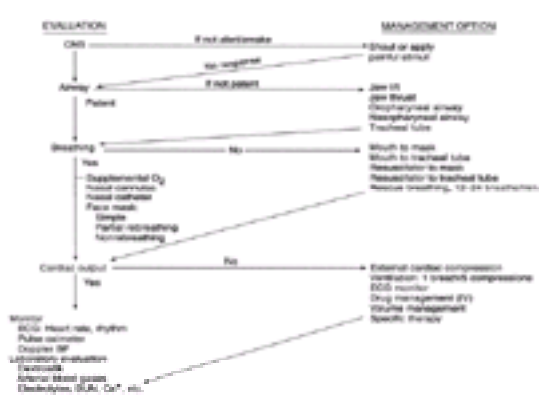


FIGURE 1.4. Management sequence for pediatric life support.

First, the airway is maneuvered to move the mandibular block of tissue up and off the posterior pharyngeal wall. The physician places his or her cheek next to the mouth and nose while listening and feeling for movement of air. At the same time, the physician is watching the chest for any evidence of chest wall movement. If the patient is moving air independently, the physician simply continues to support the airway and looks to provide a mechanism for delivering supplemental oxygen. If the patient is not breathing spontaneously, the physician must breathe for the victim, using an expired air technique when a manual resuscitator is not available. As soon as advanced life support breathing technology is available, it should be used. With the recognition that the airway is open and ventilation is occurring, the third phase of oxygen delivery is evaluated by feeling for arterial pulsations. The physician should palpate the brachial, carotid, or femoral arteries. If palpable pulses are not present after a 15-second evaluation, external cardiac compression (ECC) is initiated to provide a circulation. The adequacy of ECC is initially determined by feeling for pulses. In determining whether the oxygen delivery system has been reestablished, the physician should look for improvement in the level of consciousness or a return to spontaneous breathing or an inherent cardiac rhythm.

More specific management sequences are offered at the end of this chapter.

Airway

Evaluation

The first priority in the sequential evaluation and management paradigm of basic and advanced life support is evaluation and treatment of the airway. The physician should look, listen, and feel for evidence of gas exchange. The physician should *look* at the chest to see whether there is chest wall or abdominal movement suggestive of breathing effort. The physician should *listen* over the mouth and nose for the sound of air movement. With a stethoscope, the physician should listen over the trachea and the axilla for air entry. The physician should *feel* with his or her cheek for evidence of air movement. If there is evidence of spontaneous breathing and no evidence of gas movement through the central airway, the presumptive diagnosis is that of airway obstruction.

Management

If trauma is suspected, the head and cervical spine must be stabilized during evaluation and management of the airway. Someone must be assigned to hold the head in the midline position while applying gentle cephalad traction. The most effective noninvasive maneuver for clearing an obstructed airway involves tilting the head back slightly and lifting the chin forward by pulling or pushing the mandibular block of tissue forward (Fig. 1.5). The traditional mechanism of gentle flexion of the cervical spine on the thoracic spine may open the airway, but it provides less efficient ventilation and is hazardous if cervical spine trauma has occurred.

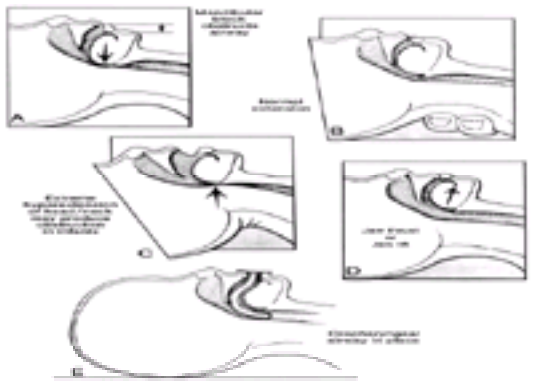


FIGURE 1.5. A. Upper airway obstruction related to hypotonia. **B.** Partial relief of airway obstruction by means of head extension (danger of cervical spine injury in cases of trauma). **C.** Extreme hyperextension causing upper airway obstruction. **D.** Fully open airway through use of jaw thrust or jaw lift. **E.** Oropharyngeal airway stenting mandibular block off of posterior pharyngeal wall.

Most airway obstruction is related to the mandibular block of tissue falling posteriorly and lying against the posterior wall of the hypopharynx. This can be relieved by physically grasping the mandibular block and pulling it forward so that the lower anterior central incisors are anterior to the maxillary central incisors. The same result can be obtained by pushing the mandibular block of tissue forward. The fingers should be placed behind the angle of the jaw and the jaw pushed forward so that the lower central incisors are in a plane anterior to the upper central incisors (Fig 1.5). These noninvasive maneuvers should be attempted before any of the more invasive airway adjuncts are tried. Table 1.5 lists airway equipment and respiratory monitoring equipment that should be available for pediatric life support. Table 1.6 lists rates of respiration to be achieved by providing CPR for pediatric life support.

1. Pocket mask
2. Laryngoscope handle with knurled finish
3. Laryngoscope blades:
   - Miller 0, 1, 2, 3
   - Macintosh 2, 3, 4
   - Wis-Hipple 1.5
4. Oropharyngeal airways: Guedel or Berman type, all sizes
5. Nasopharyngeal airways: French sizes 12, 16, 20, 24, 28
6. Endotracheal tubes: I.D. sizes
   - Uncuffed—2.5 to 7.5 mm in 0.5-mm increments
   - Cuffed—5.0 to 10 mm in 0.5-mm increments
7. Stylet: infant, adult
8. Magill forceps: child, adult
9. Extra batteries and laryngoscope lamps
10. Suction catheters: French sizes 6, 8, 10, 12, 14
11. Yankauer suction tip
12. End-tidal CO_2 monitor
13. Pulse oximeter

Table 1.5. Airway Equipment Kit for Pediatric Resuscitation

Infant:	20-24 breaths/min
Child:	16-20 breaths/min
Adolescent:	12-16 breaths/min

Table 1.6. Rate of Respiration

Artificial Airways

Oropharyngeal Airways

Oropharyngeal airways are used when manual manipulation of the airway cannot maintain airway patency. The purpose of the oropharyngeal airway is to stent or support the mandibular block of tissue off of the posterior pharyngeal wall. There are three basic parts to this airway (Fig. 1.6). The flange is used to prevent the airway from falling back into the mouth. It also serves as a point of fixation for adhesive tape. The bite block portion is designed to prevent approximation of the central incisors. A forceful bite may produce obstruction of an oral tracheal tube. The stent of the oropharyngeal airway is designed specifically to hold the tongue away from the posterior pharyngeal wall. Secondarily, the stent may provide an air channel or suction conduit through the mouth. The proper size oropharyngeal airway can be estimated by placing the airway alongside the face so that the bite block portion is parallel to the palate. The tip of the airway should just approximate the angle of the mandible.

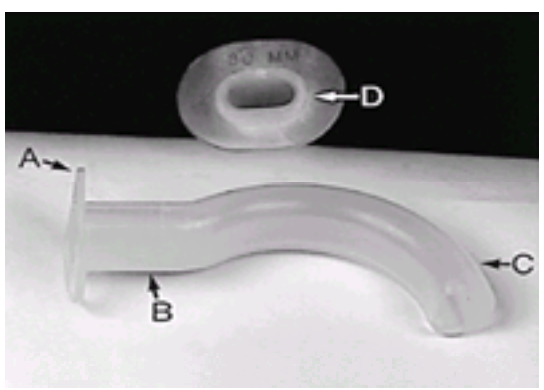


FIGURE 1.6. Oropharyngeal airway: flange *(A),* bite block *(B),* stent *(C),* and gas exchange or suction conduit *(D).*

The primary use of the airway is in the unconscious patient. The airway should be placed by using a wooden spatula or tongue depressor to press the tongue into the floor of the mouth. The airway is then passed so that the stent conforms to the contour of the tongue. If the oropharyngeal airway is not inserted properly, it may push the tongue backward into the posterior pharynx, aggravating or creating upper airway obstruction. If the airway is too long, it may touch the larynx and stimulate vomiting or laryngospasm.

Nasopharyngeal Airways

The purpose of nasopharyngeal airways is to stent the tongue from the posterior pharyngeal wall (Fig. 1.7). It may also be used to facilitate nasotracheal suctioning. The length of the nasopharyngeal airway is estimated by measuring the distance from the nares to the tragus of the ear. The outside diameter of the airway should not be so large that it produces sustained blanching of the skin of the ala nasae. The nasopharyngeal airway is inserted through the nares and passed along the floor of the nostril into the nasopharynx and oropharynx so that it rests between the tongue and the posterior pharyngeal wall. Nasopharyngeal airways may lacerate the vascular adenoidal tissue found in the nasopharynx of children. Therefore, adenoidal hypertrophy and bleeding diatheses are relative contraindications to the use of these airways.

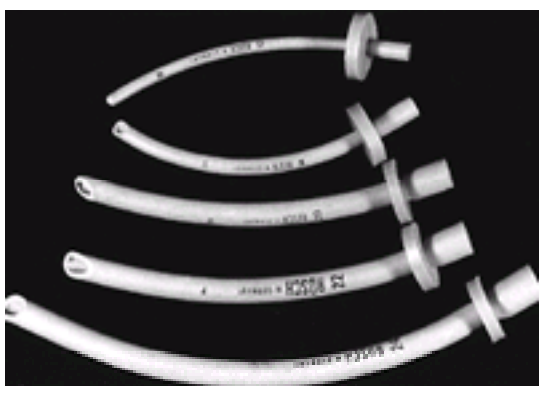


FIGURE 1.7. Nasopharyngeal airways in a variety of sizes.

Esophageal Obturator Airways

Esophageal obturator airways have not been designed or tested for use in children. Their use should be avoided.

Endotracheal Tubes

The purpose of the endotracheal (ET) tube (Fig. 1.8) is to supply a stable alternate airway. ET tubes are used to 1) overcome upper airway obstruction, 2) isolate the larynx from the pharynx, 3) allow mechanical aspiration of secretions from the tracheal bronchial tree, and 4) facilitate mechanical ventilation or end-expiratory pressure. The correct tube size can be approximated by using a simple formula based on the patient's age:

$$\text{Inside diameter (ID) in mm} = \frac{16 + \text{Age in years}}{4}$$

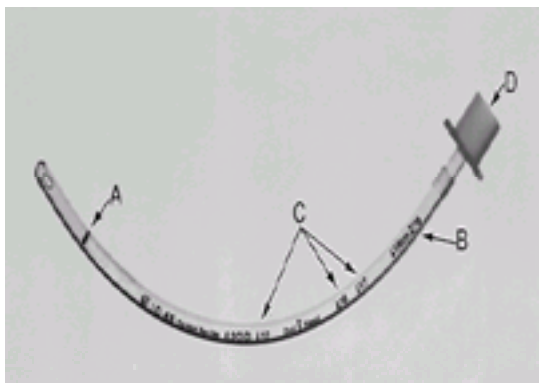


FIGURE 1.8. Oral tracheal tube: vocal card marker *(A),* manufacturer's indication that tube meets ANSI-Z79 subcommittee standards *(B),* distance in centimeters from tip of tube *(C),* and standard 15-mm connector *(D).*

Because this is an estimate, it is prudent to have the next smaller and larger size ET tube available. Estimation of tube size based on the size of the patient's fifth finger is not accurate. Tube size may also need to be modified based on the cause of the arrest (e.g., croup). In the pediatric patient, uncuffed tubes are used and are compatible with positive-pressure ventilation. This is because in children, there is a normal narrowing of the trachea at the level of the cricoid ring (Fig. 1.9). With proper tube selection, this narrowing serves as a functional seal. With patients 10 years of age and older, cuffed ET tubes should be used. By using a cuffed tube, one essentially adds 0.5 mm to the tube size.

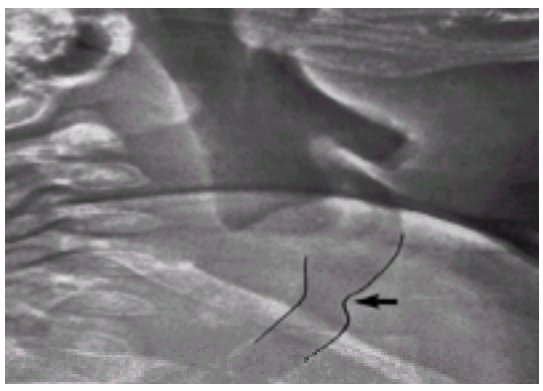


FIGURE 1.9. Lateral neck xeroradiograph showing narrowing at level of cricoid ring.

A variety of ET tubes are available. Tracheal tubes (Fig. 1.8) should be translucent to facilitate inspection of internal debris or occlusion, have a radiopaque tip marker, have the internal diameter noted proximally so that it is visible after intubation, have a distal vocal cord marker so that when the marker is placed at the level of the vocal cords the tip of the tube is in a midtracheal position, have centimeter markings along the course of the tube to be used as reference points for detecting tube movement, and meet the American National Standard Institute Z-79 guidelines for tracheal tubes and cuffs.

Other Techniques

Alternative airway management systems, including esophageal/tracheal tubes, laryngeal mask airways, and transtracheal ventilation systems, have all been used with adult patients with varying degrees of success. All of the methods have been approved by governmental agencies and professional societies, but their use in children has not been well tested or

researched. Thus, they remain as techniques worth consideration in a setting where standard methods have failed.

Laryngoscopy and Intubation (See Procedure in Section VII)

The purpose of laryngoscopy is to create a spatial plane through the mouth to the larynx through which an ET tube can be passed into the trachea. The laryngoscope consists of a blade and a handle. It is used to identify the glottis and to compress the intervening soft-tissue structures into the floor of the mouth. The three components of the laryngoscope blade are the spatula, the tip, and the flange (Fig. 1.10). The spatula may be curved or straight and is used to compress tissue. The tip of the blade is used for positioning the spatula so that an optimal compression of the mandibular block or soft tissue can be achieved. The purpose of the flange is to keep the tongue out of the way of the intubating channel. The laryngoscope is introduced into the mouth so that the tip of the blade slides down the right side of the tongue. As the tip of the blade follows the tongue posteriorly, it bumps into the anterior pillars of the tonsils. The tip is moved around the pillars of the tonsils until it bumps into the epiglottis. When using a curved spatula, the tip is placed in the vallecula, the space between the tongue and epiglottis. When using a straight spatula, the tip is placed under the epiglottis with the leading edge resting on the aryepiglottic folds. Once the tip is properly placed, the spatula is shifted from the right side of the mouth to the middle of the mouth. This left lateral movement of the spatula allows the flange to push the tongue ahead of it so that the tongue eventually occupies the middle third of the mouth. The right one-third of the mouth is then available as a channel through which the tracheal tube can pass. Once the tip of the blade is properly positioned and the flange has moved the tongue into the left corner of the mouth, the full surface of the spatula is used to compress the tongue into the floor of the mouth. With compression of the soft tissue of the mouth, the glottis should be exposed and the tracheal tube can be passed. The tracheal tube should be fitted with a stylet. The purpose of the stylet is to provide some degree of curvature to the tube for those circumstances where a totally straight channel cannot be achieved. The tracheal tube is passed through the glottis so that the ring marker near the tip of the tube is aligned with the vocal cords. If the tube selected is the proper size and the ring marker is placed directly at the vocal cords, the tip of the tube should be at a midtracheal position.

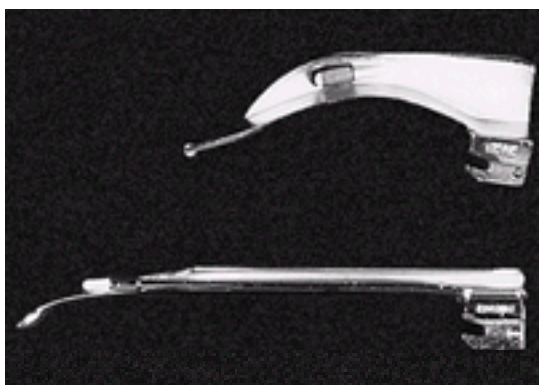


FIGURE 1.10. Laryngoscope blades—straight blade (Miller) and curved (MacIntosh).

Proper positioning of the tube is confirmed most accurately by end-tidal CO_2 monitoring (Fig. 1.11) and by auscultating for breath sounds and observing for symmetric chest movement. The child's small chest wall may transmit sounds widely and thus mislead the physician into thinking that positioning is correct. The physician should listen carefully. He or she should listen over the stomach and both axillas, and look for improved color of the patient. If breath sounds are not equal or end-tidal CO_2 monitoring is not available, the tube should be withdrawn slightly and the breath sounds and chest movement reevaluated. When circumstances allow, tube position should be confirmed with an anteroposterior (AP) chest roentgenogram. On the AP film, the tip of the tracheal tube should be at a T2 to T3 vertebral level or directly between the lower edges of the medial aspect of the clavicles (Fig. 1.12).

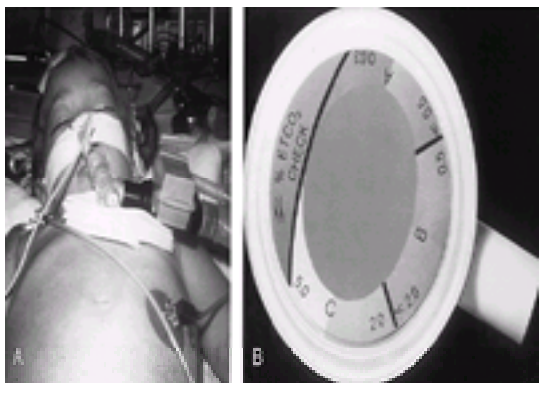


FIGURE 1.11. End-tidal CO_2 monitor. **A.** In-line on patient's endotracheal tube. **B.** Disposable-type monitor.

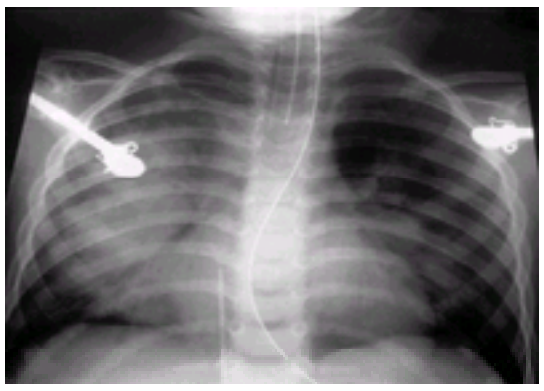


FIGURE 1.12. Chest radiograph showing proper endotracheal tube placement at T2 to T3 vertebral level.

Loss of an established airway is an unnecessary complication. The tracheal tube should be thoroughly secured with adhesive tape. The skin to which the adhesive tape is affixed should be cleansed, dried, and painted with tincture of benzoin (Fig. 1.11)

The management of airway obstruction is detailed in a separate section at the end of this chapter, and Chapter 5 covers other advanced aspects of airway management.

Breathing

Evaluation

When a clear and stable airway has been established, the patient should be reassessed. The physician should look, listen, and feel for evidence of gas exchange. In infants, adequacy of ventilation is assessed by observing free uniform expansion of the lower chest and upper abdomen. This is in contrast to older children and adolescents in whom one looks for uniform upper chest expansion as a sign of adequate ventilation. Gas exchanges should be confirmed by auscultation and by electronic monitoring of end-tidal CO_2 and pulse oximetry. First, the physician should listen over the trachea to establish quickly that gas exchange is occurring through the central airway. Then, he or she should listen to breath sounds bilaterally to assess for peripheral aeration and symmetric lung expansion.

Management

Spontaneous Ventilation

If the airway has been established and the patient is breathing spontaneously, supplemental oxygen should be administered. Although elimination of carbon dioxide is important, it is not nearly as important as delivery of oxygen. Children are quite resistant to the effects of severe hypercarbia and respiratory acidosis. They do not tolerate even short periods of oxygen deprivation.

Oxygen Delivery Devices

A variety of oxygen delivery devices are available for use in patients who have stable airways without ET tubes.

Nasal Cannulas. Nasal cannulas have two hollow plastic prongs that arise from a flexible hollow face piece. Humidified oxygen delivered through the hollow tubing is directed to the nostrils. One hundred percent oxygen is run through a bubbler into the cannula system at a flow of 4 to 6 L/minute. Because of oropharyngeal and nasopharyngeal entrainment of air, the final oxygen delivery is usually 30 to 40%. The advantages of cannulas are that they are easy to apply, lightweight, economical, and disposable. Inefficiency of the bubbler humidifier is compensated for by the fact that the normal humidification and warming systems of the upper airway are not bypassed. The use of this device presumes that the patient's oxygen needs can be met with substantially less than 100% oxygen. This method of oxygen delivery is best tolerated by the older child.

Oxygen Hoods. Oxygen hoods are clear plastic cylinders with removable lids (Fig. 1.13) or clear, soft, plastic tents just large enough to accommodate the infant's head. They are used for delivery of oxygen to infants and come in a variety of sizes. They usually have a gas inlet system for wide-bore tubing and a port for positioning the cylinder across the neck. Their purpose is to maintain a controlled environment for oxygen, humidity, and temperature. This can be done without producing a tight seal at the neck. Hoods are best used for newborns and infants. One can, without difficulty, deliver oxygen concentrations in the 80 to 90% range simply by increasing the oxygen flow to flood the canister. Another advantage is that the oxygen may be well humidified. Because of their potential for delivering concentrations of oxygen that may be toxic to the eyes or lungs of the infant, it is imperative to monitor both the fraction of inspired oxygen (FiO_2) and the PaO_2.

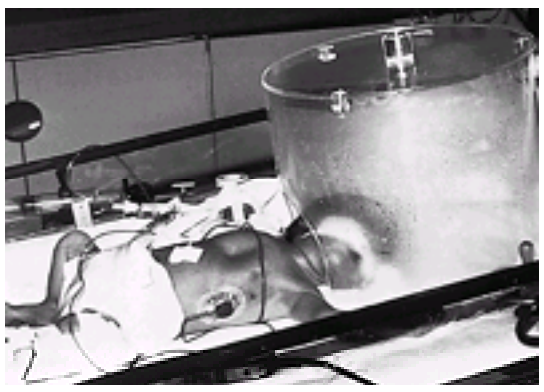


FIGURE 1.13. Infant oxygen hood. Oxygen hoods are clear plastic cylinders with removable lids or clear, soft, plastic tents just large enough to accommodate the infant's head.

Oxygen Tents. The basic purpose of the oxygen tent is to provide a controlled and stable environment for humidity, temperature, and oxygen. Tents are useful for delivery of oxygen between 21 and 50%. Oxygen concentration may be variable because of a poor seal and frequent entry. Therefore, a tight fit and only necessary entry should be allowed. Tents potentially impede access to the patient, and if mist is used, the patient may be hidden in a cloud which makes skin color difficult to evaluate.

Oxygen Masks. The most often used equipment for the spontaneously breathing patient is the oxygen mask. Several types of oxygen masks can be used to offer the patient a wide range of inspired oxygen concentrations. Masks seem to be better tolerated than nasal cannula by the young child, particularly when the mask is held by a calm parent or by emergency department personnel. There are several mask types from which to select. As with all equipment, even masks have associated hazards. In patients prone to vomit, the mask can block the flow of vomitus and increase the risk of aspiration. The obtunded patient wearing a mask must always be observed.

Simple Masks. The purpose of the simple face mask is to deliver a moderate concentration of oxygen. These masks are lightweight and inexpensive. They should be clear to allow observation of the child's color. They can be used in a loose-fitting fashion and are relatively comfortable. If the flow of oxygen is inadvertently disrupted, the child can breathe through side ports. A minimal flow of oxygen is necessary to flush potential dead space. This type of oxygen delivery device does not bypass the upper airway mechanisms for warming and humidification of inspired gas. The disadvantages of the simple mask lie in the fact that it is difficult to provide a known and stable Fi O_2. The FiO_2 will vary with the inspiratory flow rate of the patient and with the oxygen flow into the system. The actual pharyngeal Fi O_2 may be difficult to predict or measure.

Partial Rebreathing Masks. Partial rebreathing masks allow delivery of a higher oxygen concentration than simple masks do. They are also helpful in conserving oxygen. This system is a combined face mask and reservoir bag. When the flow rate into the bag is greater than the patient's minute ventilation and when the oxygen is adjusted so that the bag does not collapse during inhalation, there is negligible CO_2 rebreathing. Partial rebreathing masks are usually used for midrange oxygen delivery. We use one when we are trying to maintain an Fi O_2 between 35 and 60%.

Nonrebreathing Masks. Nonrebreathing masks are combined face mask and reservoir bag devices that have nonrebreathing valves incorporated into the face mask. They are useful for giving oxygen concentrations up to 100%.

Assisted Ventilation

If the airway has been established and the child is not breathing spontaneously or gas exchange is not adequate, artificial ventilation should be started. The recommended rates for rescue breathing in infants and children are shown in Table 1.5. Rates may need modification based on the etiology of the child's arrest (e.g., treatment of ICP may require a faster rate)

If adjuncts for mechanical ventilatory support are not available, an expired air technique may be used. Patient size, type of available airway, and trial will determine which type should be used. Because of risk of human immunodeficiency virus (HIV) transmission, mouth-to-mouth resuscitation is no longer recommended. Instead, rescue breathing should be done with a pocket mask that contains an appropriate millipore filter (Fig. 1.14). Placement of the mask over the mouth alone, over the mouth and nose, or over a tracheostomy site depends on the patient and the equipment available (Fig. 1.14). See Chapter 127 for care of the patient with tracheostomy.

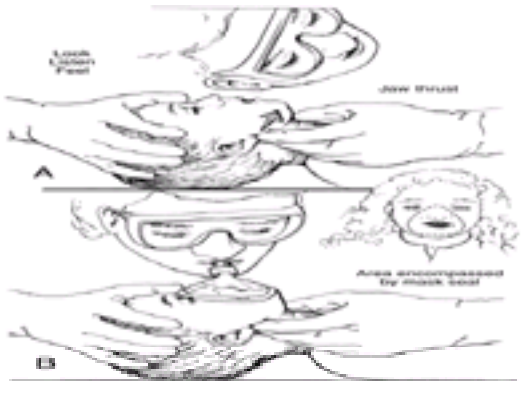


FIGURE 1.14. Basic life support—airway and breathing. **A.** Positioning of head to open airway and evaluation for spontaneous ventilation. **B.** Expired air (mouth to mask) ventilation.

Expired Air Techniques

Hand-Squeezed, Self-Inflating Resuscitators. Hand-squeezed, self-inflating resuscitators are the most commonly used resuscitators for infants and children. The elasticity of a self-inflating bag allows the bag to refill independently of gas flow. This feature makes the self-inflating bag easy to use for the inexperienced operator. Many of the self-inflating bags are equipped with a pressure-limiting pop-off valve that is usually preset at 30 to 35 cm H_2O to prevent delivery of high pressures. Self-inflating bags that are not pressure-limited should have a manometer in line. For gas to flow, the bag must be squeezed. Thus, for the patient who is breathing spontaneously, the operator must time the bag compressions to the patient's efforts. These resuscitators should be adapted to deliver high concentrations of oxygen. In most cases, this involves using an oxygen reservoir adaptation with the unit (Fig. 1.15). Recent research has shown that even with an attached reservoir, only oxygen concentrations of 60 to 90% were obtainable. Units without oxygen reservoir adaptations often deliver low concentrations of supplemental oxygen and therefore should be avoided.

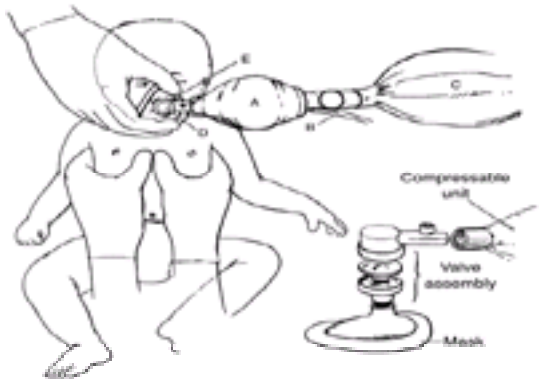


FIGURE 1.15. Self-inflating hand-powered resuscitator: compressible unit *(A),* oxygen source *(B),* oxygen reservoir *(C),* one-way valve assembly *(D),* and mask with transparent body *(E).*

The resuscitator may be used with a mask. When selecting a family of mask sizes, select a mask type that seals a variety of facial contours. Also, the body of the mask should be sufficiently transparent so that vomitus can be recognized easily through the mask. Masks with a pneumatic cuff design allow for easiest and most efficient fit to avoid air leaks around the mask. Resuscitators, masks, and ET tubes should be standardized so that any resuscitator can connect with any mask or ET tube.

Anesthesia Bags. Anesthesia bags depend on an adequate gas flow to maintain a compressible unit that propels gas toward the patient (Fig. 1.16). An exit port must also be present so that the bag does not become a carbon dioxide reservoir. When used with an oxygen blender, any desired concentration of oxygen may be provided for the patient because this system delivers directly the gas flowing into it. When used correctly, this device allows 100% oxygen to be delivered as well as maintaining end-expiratory pressure. However, the major disadvantage of this type of bag is that considerable experience is needed to use it effectively, which has prompted some to recommend the use of the self-inflating bag as the primary mode of ventilation. One must be able to judge accurately the rate of gas flow into the bag and the rate of gas escape from the exit port so that underfilling or overfilling does not occur. If the bag is removed from a leak-tight patient application, it promptly deflates and one must wait for the reservoir to refill. Overfilling the bag is dangerous because high pressures can be transmitted to the lung and stomach (Fig. 1.17).

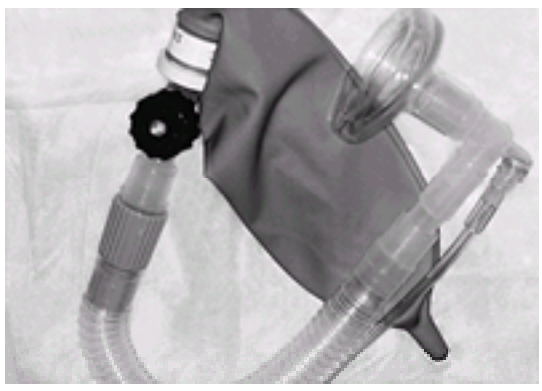


FIGURE 1.16. Family of clear plastic, air-filled collar facial masks.

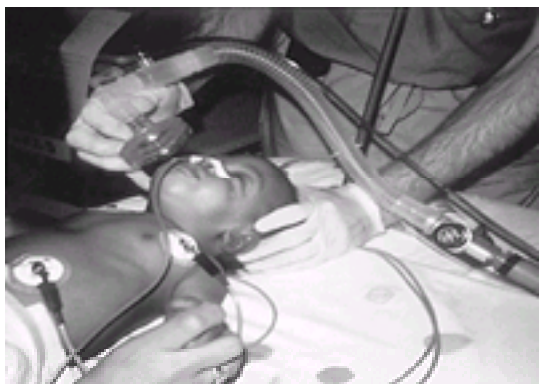


FIGURE 1.17. Anesthesia bag in use.

Mechanical Ventilators. The emergency department should be equipped with a mechanical ventilator as well. This is important in the event that the resuscitation is successful for maintenance of ventilation while the patient awaits transfer or transport to a critical care unit. A mechanical ventilator is also crucial in the event of multiple arrest victims in order to free personnel for other vital tasks.

Circulation

As with the other components of CPR, the circulation must be first assessed and then managed.

Evaluation

Once the airway has been opened and gas exchange ensured, the physician must evaluate the effectiveness of circulation by 1) observing skin and mucous membrane color and 2) palpating a peripheral pulse and checking capillary refill. If the patient's color is ashen or cyanotic, the circulation will need to be treated.

The palpation of a peripheral pulse and assessment of capillary refill is mandatory. Often, ineffective cardiac activity can be palpated over the child's thin chest wall. Thus, the presence of an apical pulse may not be meaningful. The palpation of a strong femoral or brachial pulse (Fig. 1.18) indicates presumptively that the cardiac output is adequate. Capillary refill should be assessed repeatedly (Fig. 1.19).

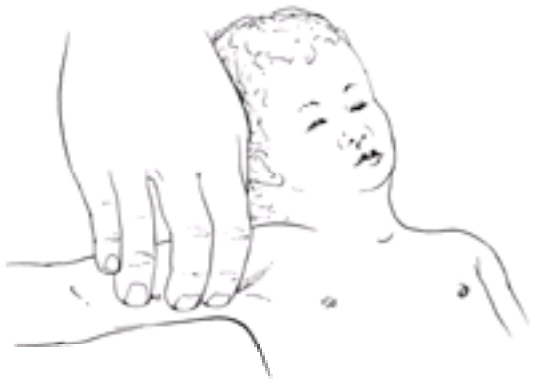


FIGURE 1.18. Palpation of brachial pulse on medial aspect of the upper arm in the subbicep groove.

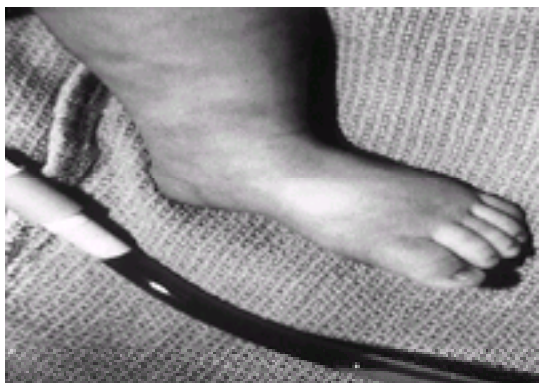


FIGURE 1.19. Delayed capillary refill.

Most modern defibrillators have a “quick-look” paddle configuration that allows a rapid evaluation of cardiac rhythm to be made by placing the defibrillation paddles on the chest and using them as monitoring electrodes.

The resuscitation team will also find it helpful to have continuous blood pressure monitoring. Blood pressure measurements will help quantify the effectiveness of cardiac function. An ultrasound or portable Doppler device may be

necessary to detect systolic pressure at low levels in small infants (Fig. 1.20).

FIGURE 1.20. Portable Doppler device for determining blood pressure during resuscitation.

As soon as possible, the team will also require continuous electrocardiogram (ECG) monitoring to assess the development of arrhythmia as the resuscitation proceeds.

Management

Management may be divided into five phases: 1) cardiac compression, 2) establishment of an intravascular route, 3) use of primary drugs, 4) use of secondary drugs, and 5) defibrillation.

Cardiac Compression

Absence of a peripheral pulse requires immediate institution of cardiac compression (CC) to establish at least a minimum circulation to the brain. ECC has the advantage of immediate applicability. The technique of ECC is also widely known and can be applied in almost any setting. However, several recent reports have pointed to the potential advantages of open cardiac compression (OCC). This technique is obviously more difficult to apply. OCC may produce better coronary and cerebral perfusion pressures. Survival rates may also be higher in studies comparing OCC with ECC. The role of OCC may become more important as future studies evolve. For now, OCC is indicated primarily in the selected traumatized patient (see Chapter 104 and Chapter 107).

The mechanism by which blood moves during ECC has been the subject of ongoing investigation. The traditional view assumes that chest compressions move blood by direct CC. Therefore, it was believed to be important to compress directly over the ventricles that were believed to be located under the middle one-third of a child's sternum. However, another model suggests that the movement of blood is caused by an increased intrathoracic pressure and expulsion of blood from the lungs through the left heart, and simultaneous openings of both the mitral and aortic valves. This proposed thoracic pump mechanism quiets the controversy concerning the relative position of the child's heart within the thorax and its relation to the sternum. The intrathoracic pressure pump model for the production of blood flow has sparked the investigation of several alternative methods for achieving flow. Probably both models of ECC-produced blood flow play a part in the actual mechanism in children.

For now, we continue to recommend that compression be applied evenly over the midsternum of the child. This standard technique has proved effective. Compression over the lower one-third of the sternum should be avoided because of a potential risk of liver trauma.

The midsternal location for ECC can be found by spanning the sternum between the thumb and fifth finger and then measuring or judging the midpoint. Another method used in locating the midsternum is to find the point where the transnipple line intersects the sternum. Finger or hand position should be just caudal (toward the feet) to the transnipple line.

The depth and rate of compression are based on the child's age. Suggested rates and depths are shown in Table 1.7. Compression should be smooth, continuous, and uninterrupted. The compression phase should consume 60% of the time for the compression–release cycle. Jabby, jerky, brief compressions may produce pressure pulses of adequate amplitude on a monitor, but blood flow will be inadequate for cerebral perfusion.

	Infant	Child	Adult[a]
Compression (rate/min)	100	80	60
Depth of compression (inches)	0.5–1	1–1.5	1.5–2
Ventilation (rate/min)	20	16	12

[a]Two-person rescue.

Table 1.7. Ventilation/Compression Schedule for Pediatric Resuscitation

Because the child has a relatively large occiput, neck extension may elevate the shoulders and upper thorax off of the firm resuscitation surface. The resultant wedge-shaped dead space beneath the upper thorax may absorb the force of compression (Fig. 1.21A). The dead space must be filled with a firm substance so that the work of compression will not be lost. A firm towel or the rescuer's hand should be placed beneath the upper thoracic spine (Fig. 1.21B). Compressions may then be applied with one or two fingers in the infant or with one hand in the older child. When using the technique developed by Thaler, the rescuer links his or her fingers beneath the thoracic spine and compresses with the thumbs (Fig. 1.22). This method is comfortable when using external cardiac compression on a newborn patient. With older children, an effort should be made not to allow the encircling hands to limit the respiratory movements of the thorax.

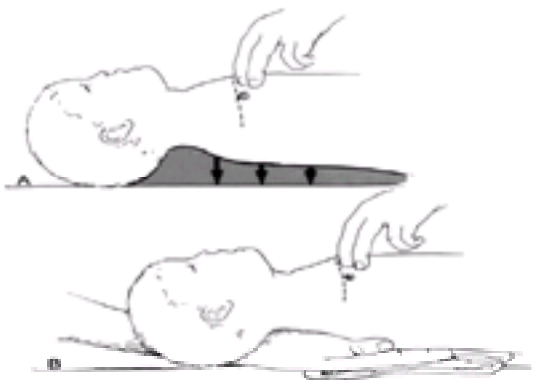


FIGURE 1.21. Thoracic cage support for external cardiac compression. **A.** Dead space created by prominent occiput. **B.** Hand providing thoracic support.

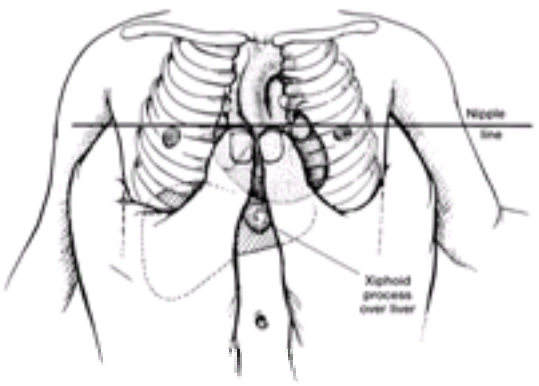


FIGURE 1.22. External cardiac compression—Thaler technique for infants.

Mechanical chest compressors should not be used for children. There is insufficient information about their effectiveness or safety for this patient population.

Intravenous Lines

The placement of an intravenous line is often the most difficult and time-consuming aspect of pediatric life support. The resuscitation team should have a set protocol for the attainment of intravenous access (an example is shown in Table 1.8). This protocol should outline which personnel should attempt which form of access for a finite period as well as when to move on to more advanced forms of access. When possible, a short, large-gauge intravenous line should be obtained. Peripheral sites are an acceptable choice and may be readily available. Central lines are useful for getting drugs and a large volume of fluids into the central circulation. Intraosseous (into the bone) infusion is an old technique that has been revived and widely promulgated (Fig. 1.23). The technique is explained in Appendix D, Section VII. Spivey et al. have shown it to be an excellent route for fluid and drug administration. While waiting for an intravenous line to be secured, some drugs may be given through the ET tube as they are absorbed at an alveolar level and circulated rapidly, the drugs should be administered through a small-gauge catheter which is inserted through the ET tube into the distal trachea or one of the mainstream bronchi and followed by several positive pressure ventilations. Table 1.9 lists the drugs that may be given through this route.

1. First 1.5 min Peripheral IV catheter, two sites 2. 1.5-5 min a. If intubated: Give drugs via endotracheal tube (including epinephrine/atropine/lidocaine) b. If not intubated: Intracsseous—one site Continued peripheral IV—one site 3. Longer than 5 min a. Femoral vein percutaneous b. External/internal jugular percutaneous c. Subclavian vein percutaneous d. Saphenous vein cutdown

Adapted from Kanter RK, Zimmerman JJ, Strauss RH, et al. Am J Dis Child 1986; 140:144.

Table 1.8. Sample Protocol for Intravenous Access

Epinephrine
Atropine
Lidocaine

Table 1.9. Resuscitation Drugs That May Be Given Intratracheally

FIGURE 1.23. A and B. Intraosseous needle being placed in proximal tibia.

When using IV ET route, the physician should dilute the medications with normal saline to a volume of 3 to 5 mL lest they remain on the tube surface and fail to reach the lung.

Intravenous cutdown over the saphenous vein is another acceptable method but seems to be time-consuming in even experienced hands. Intracardiac administration of drugs is dangerous and should be avoided. In the circumstance of CPR, the cannulation of the subclavian vessel may also be difficult and associated with pneumothorax and hemothorax.

Three vessels that are easily cannulated and give access to the central circulation are the femoral, internal jugular, and external jugular veins. Our vein of first choice is the femoral because its cannulation does not interfere with ongoing CPR. The femoral vein is located by palpating the femoral artery pulsation and moving just medial to it. If good external cardiac compression is being accomplished, the pulsation should be present. When the pulse is not palpable, the physician should find the midpoint between the symphysis pubis and the anterior superior iliac spine on the plane of the inguinal ligament. The vein should be approached at a 45-degree angle to the skin 2 to 3 cm below the inguinal ligament at the defined midpoint. The needle must not pass cephalad to the inguinal ligament because it may enter the peritoneal cavity and pierce the bowel (see Appendix D, Section VII)

The internal jugular is best approached by finding the triangle created by the medial one-third of the clavicle and the two heads of the sternocleidomastoid muscle—the sternum and clavicular. The internal jugular vein courses within the triangle. The physician should puncture the skin at the apex of the triangle and direct the needle toward the ipsilateral nipple at a 45-degree angle to the frontal plane. The external jugular vein is located by placing the child in a 20-degree head-down position. With the patient at this angle, the vein will fill and be seen as it courses over the midportion of the sternocleidomastoid muscle (see Appendix D, Section VII)

In cannulating either the femoral or internal jugular vein, the Seldinger technique is recommended (see Appendix D, Section VII). This method uses a 20- to 22-gauge, short-beveled, thin-walled needle for the small child. Larger needles should be used for older children. The needle is attached to a syringe, and the vein is punctured and aspirated so that free blood flow into the syringe is seen. The syringe is then removed, and blood is sent for appropriate laboratory studies.

A braided wire is introduced through the needle and positioned in the lumen of the vein. The needle is withdrawn over the free end of the wire. Next, a flexible Teflon catheter is passed over the wire and into the vein. Finally, the wire is removed from the lumen of the catheter, and the catheter is secured with suture. Although this technique requires several steps, it represents a safe and effective way to secure an intravenous line. It allows a small initial puncture to result in the placement of a larger-bore catheter. It is particularly useful for the femoral and internal jugular veins.

If the Seldinger technique is unsuccessful, an acceptable alternative is to cut down over the lesser saphenous vein just above the anterior and the medial malleolus (see Appendix D, Section VII). In extreme circumstances, cut down over the femoral vein may be attempted. This technique is more difficult and fraught with more complications. Remember also that in newborns during the first 2 weeks, it may be easy to cannulate the umbilical vein (see Appendix D, Section VII).

Primary Drugs

The primary drugs for advanced life support are oxygen, epinephrine, sodium bicarbonate, atropine, and glucose (Table 1.10). Each agent has specific actions, indications, dosages, and untoward effects. The primary drugs are those that are used first in the resuscitation. Our recommended drug dosages are based on kilograms of body weight. The resuscitation team must be able to determine the weight of the child accurately. If estimates prove too difficult, a standardized growth curve should be posted in the resuscitation area so that the 50th percentile weight for the child's age may be used. Another method for establishing the correct dosage schedule has been proposed by Broselow in the invention of the Broselow Tape. This method bases dosage on the patient's length, which is determined by placing the tape along side the child in the supine position (Fig. 1.24A). An appropriate drug dosage schedule has been printed on the tape (Fig. 1.24B) at intervals. Using this method eliminates the estimation of patient weights and the need to cross-reference to a drug dosage chart or to memorize the dosage schedule. If the pediatric drug dosage schedule is not used often, it is advisable to prominently display the schedule on the emergency department wall.

Epinephrine	10–200 μg/kg IV or IT
Sodium bicarbonate	1 mEq/kg IV
Atropine	0.02 mg/kg IV or IT (min 0.2 mg)
Glucose	0.5–1.0 g/kg IV

Table 1.10. Primary Life Support Drugs and Dosages

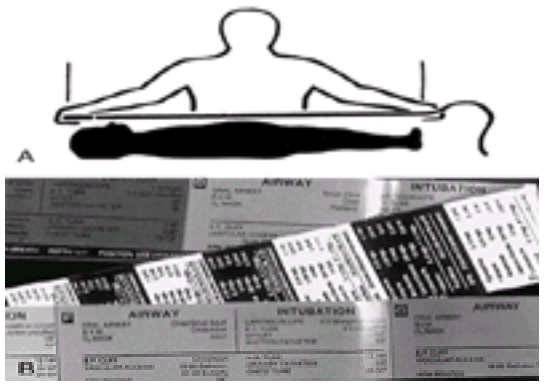


FIGURE 1.24. Broselow Tape for determining drug dosage schedule based on patient length. **A.** Placement of tape. **B.** Equipment size and drug dosage schedule printed on tape.

Oxygen. In the patient in cardiac arrest, there are many factors that contribute to the pathophysiologic disruption of oxygen delivery (i.e., hypoxemia). A fundamental goal of basic and advanced life support is to correct cerebral and myocardial hypoxia before irreversible injury occurs.

Oxygen is indicated for any patient who is having decreased oxygen delivery to the tissues, whether this is due to 1) pulmonary abnormality such as decreased air exchange, intrapulmonary shunting, or ventilation-perfusion abnormality; 2) cardiac dysfunction such as decreased stroke volume or arrhythmia; or 3) oxygen-carrying abnormalities such as low hemoglobin states, methemoglobinemia, or carbon monoxide poisoning. Any patient who is even suspected to be hypoxemic should be given oxygen.

During resuscitation, the initial dosage of oxygen for the patient needing oxygen is 100%. The potential negative effects of high oxygen concentration are not a consideration in the life-or-death setting of CPR.

The physician should be familiar with the different oxygen delivery systems mentioned in the sections on airway and breathing. Oxygen concentrations delivered by mouth-to-mask breathing vary from 17 to 21%. Bag-valve-mask devices deliver 21%. When an oxygen source is attached, bag-valve-mask devices may deliver 30 to 60%, and when a reservoir

is added, 70 to 90% may be achieved. An anesthesia bag may deliver 100%. Oxygen should be ordered like other drugs. The order should clearly specify the dose, mechanism, and duration of treatment.

Epinephrine. Despite the development of many new exogenous catecholamines, epinephrine remains the essential cardiac stimulant. Epinephrine actions include a- and b-adrenergic stimulation. The primary a effect is vasoconstriction and a resultant increase in systolic and diastolic blood pressure. The b-adrenergic action of the drug is also beneficial in producing an increased inotropic (contractile force) and chronotropic (cardiac rate) effect. In addition, the b effect produces vasodilation of the coronary and cerebral vasculature. All of these actions are beneficial to the resuscitative effort.

Indications for the use of epinephrine include asystole, symptomatic bradycardia, and hypotension not related to volume depletion. Epinephrine is also used to try to change a fine fibrillation pattern to a coarse one before a defibrillation attempt. There is belief that a coarse fibrillation pattern is more easily converted.

The recommended initial dose of epinephrine for asystole or electromechanical dissociation is 10 µg/kg intravenously (IV). Recently, several investigators have looked at the scientific basis for current epinephrine dose recommendations and have used doses 10 to 20 times greater (i.e., 0.1 to 0.2 mg/kg). Thus far, higher-dose therapy with epinephrine (HDE) has not been proved to be of greater efficacy. In the experimental situations in which it has been used, it has resulted in no adverse effects. We continue to support a dose-response method of epinephrine administration. We initiate therapy with the currently recommended dose (0.01 mg/kg). Subsequent doses are given by a dose-response method of titration. With this method, the second dose is increased tenfold to 0.1 mg/kg, and the physician observes the patient for the desired response. If an inadequate response occurs, the dose should be doubled to 0.2 mg/kg. In the absence of underlying cardiovascular disease, this is a safe and effective approach. The dose of epinephrine for hypotension is 1 µg/kg IV. An epinephrine infusion may be prepared by adding 6 mg of epinephrine to 100 mL of 5% dextrose in water (D5W). This creates a solution that has 60 µg/mL. When this solution is infused at 1 mL/kg per hour, the patient receives a dose of 1 µg/kg per minute. If an intravenous or intraosseous line has not been established, epinephrine can be given endotracheally. The dose and concentration of the drug (1:10,000) should be the same. The drug is instilled past the tracheal tube via a smaller-gauge catheter and is followed by several positive-pressure ventilations.

Epinephrine is a relatively safe drug, and few untoward effects are seen in pediatric patients. Ischemia of the myocardium rarely occurs and has not been reported even with high-dose epinephrine. There is the hazard of producing supraventricular or ventricular tachycardia, premature ventricular contractions, or ventricular fibrillation. When possible, the physician should avoid intracardiac injection of epinephrine because this route is associated with greater risk and complications. As previously noted, mixing bicarbonate with epinephrine inactivates the epinephrine.

Sodium Bicarbonate. With the onset of respiratory failure, the patient develops respiratory acidosis. Rising levels of carbon dioxide in the blood produce a fall in pH. The immediate treatment for this type of acidosis is adequate ventilation. As the patient's circulation begins to fail, there is production of lactic acid and a metabolic acidosis. Sodium bicarbonate corrects the metabolic acidosis by combining with hydrogen to form carbon dioxide and water. This additional production of carbon dioxide must also be eliminated through ventilation.

Metabolic acidosis is a harmful byproduct of the arrest because acidosis further impairs cardiac and circulatory function. Acidosis depresses spontaneous cardiac activity, decreases the contractile force of the ventricle, and predisposes the patient to ventricular fibrillation. In addition, catecholamines such as epinephrine will be less effective in a patient whose metabolic acidosis is uncorrected.

It is not necessary to prove a diagnosis of metabolic acidosis before beginning the first steps in treatment. Sodium bicarbonate is also indicated for the correction of suspected metabolic acidosis. This includes any patient who has arrested for more than a few minutes.

For the patient whose arrest was not witnessed, the initial dose of sodium bicarbonate is 1 mEq/kg IV. This should be given as full-strength bicarbonate (1.0 mEq/mL) for children more than 6 months of age. For infants less than 6 months of age, we recommend that the same dose be given, but in the form of half-strength bicarbonate (0.5 mEq/mL) to lessen the osmotic load of the drug.

After the initial dose of bicarbonate, subsequent doses are best determined by measuring arterial pH and calculating the dosage using the base deficit.

The factor of 0.4 represents the bicarbonate distribution space, which is 40% of the total body weight. The formula divides the dose by 2 to prevent overcorrection with bicarbonate and produce alkalemia. If blood gas analysis is impossible, doses of 0.5 mEq/kg may be administered every 10 to 15 minutes.

Untoward effects of bicarbonate include alkalosis, hypernatremia, and hyperosmolar states. Each of these effects are significant and can in themselves lessen survival. In the context of inadequate ventilation, bicarbonate administration may result in rapid increase in P_{CO_2} levels. Because carbon dioxide is readily permeable across cell membranes and sodium bicarbonate is not, rapid elevation in carbon dioxide can exacerbate intracellular acidosis. This intracellular acidosis depresses myocardial function in animal studies. Hypernatremia and hyperosmolar states are most easily produced in the young infant. Alkalosis is tolerated poorly by the body. Thus, it is important to determine the exact need for bicarbonate as quickly as possible. Bicarbonate administration must always be secondary to the establishment of an airway and ventilations that are important for correcting the acidosis of respiratory origin.

There are other potential untoward effects of bicarbonate: the precipitation of bicarbonate and calcium in the intravenous line and the inactivation of catecholamines. To avoid these two problems, we discourage the addition of bicarbonate to intravenous fluid reservoirs during the resuscitation. Bicarbonate should be given by direct intravenous administration

and followed with a saline flush solution before giving subsequent medication, such as calcium or epinephrine. ET administration of bicarbonate can be hazardous. Bicarbonate is irritating to the airways, destroys lung surfactant, and can produce massive atelectasis. Also, the large volume of bicarbonate usually required will virtually drown the patient.

Atropine. The actions of atropine are parasympatholytic. Atropine has both peripheral and central effects. The peripheral effect is vagolytic and thus increases heart rate by increasing the rate of discharge from the sinoatrial node, while increasing conduction through the atrioventricular node. The central effect of atropine is to stimulate the medullary vagal nucleus and is produced with low dosage of the drug. The actions are opposite to those desired for resuscitation therapy.

The indication for atropine is bradycardia associated with hypotension, premature ventricular ectopic beats, or symptoms of poor CNS or myocardial perfusion. Atropine may be used for second- or third-degree heart block, although its actions may be only temporary for these arrhythmias.

The dose of atropine is 0.02 mg/kg IV. There is a minimum dose of 0.10 mg repeated every 5 to 10 minutes to a maximum total dose of 1.0 mg in a child and in an adolescent. Atropine may be given intratracheally if an intravenous route is not available.

The untoward reactions associated with atropine include paradoxical bradycardia, atrial and ventricular tachyarrhythmias, and myocardial ischemia. Paradoxical bradycardia is caused by the central action of atropine. This side effect can be avoided by using at least 0.2 mg for any patient being treated with atropine. Tachyarrhythmias occur but are not usually hemodynamically significant in the pediatric patient. Myocardial ischemia is rare in the absence of existing cardiac disease.

Glucose. The action of glucose is to correct hypoglycemia. Glucose should be considered a primary drug. Infants have minimal glycogen stores for rapid conversion to glucose. Moreover, many infants may have had decreased caloric intake and excessive losses (diarrhea and vomiting) in the days before the arrest.

The dose of glucose is 0.5 to 1.0 g/kg IV. Either a 10% or a 25% solution may be used except in the neonatal period when 10% glucose is indicated. Doses of glucose should be based on a rapid serum glucose determination.

Untoward effects include hyperglycemia and hyperosmolality, but these should not occur if the initial dose of glucose is based on a documented need. Some studies have found a correlation between mortality and high serum glucose levels in pediatric head trauma patients.

Secondary Drugs

A number of useful second line drugs should be available in the emergency department for resuscitations. These secondary drugs are listed in Table 1.11.

Drug	Initial	Subsequent
Lidocaine	1 mg/kg IV or IT	10-20 µg/kg/min IV
Bretylium	5 mg/kg IV	10 mg/kg IV
Adenosine	0.1 mg/kg IV bolus	0.2 mg/kg IV
Dopamine	10 µg/kg/min IV	
Isoproterenol	0.1 µg/kg/min IV	
Calcium chloride	10-20 mg/kg/min IV	
Calcium gluconate	30 mg/kg IV	
Furosemide	1 mg/kg IV	2 mg/kg IV
Naloxone	1 mg child/2 mg adolescent	Repeat
Methylprednisolone	30 mg/kg IV	
Dexamethasone	1 mg/kg IV	
Difibrillation current	2 watt-sec/kg	4 watt-sec/kg

Table 1.11. Secondary Useful Life Support Drugs

Lidocaine. Lidocaine works by reducing the automaticity of ventricular pacemakers. Thus, it increases the fibrillation threshold. Ventricular fibrillation is a relatively uncommon event in pediatric resuscitations. However, when it occurs, lidocaine is the drug of choice. The initial dose is 1 mg/kg IV. This dose may be repeated three times at 5-minute intervals. Once the initial bolus has been given, an infusion of lidocaine 20 to 50 µg/kg per minute should be initiated (Table 1.12). If an intravenous line cannot be established, the initial dose of lidocaine may be administered by the tracheal route.

Drug	Add to D5W to Make 100 mL	Infuse	Dosage Delivers
Lidocaine	120 mg (3 mL)	1 mL/kg/hr	20 µg/kg/min
Dopamine	60 mg (1.5 mL)	1 mL/kg/hr	10 µg/kg/min
Epinephrine	6 mg (6 mL)	1 mL/kg/hr	1 µg/kg/min
Isoproterenol	0.6 mg (3 mL)	1 mL/kg/hr	0.1 µg/kg/min

Table 1.12. Drugs for Intravenous Infusion

The adverse reactions of lidocaine include nausea, vomiting, lethargy, paresthesias, tinnitus, disorientation, and seizures. CNS symptoms may be the first to appear. Later, symptoms of cardiac toxicity may appear, including depression of myocardial contractility and ventricular irritability. Heart block and eventual drug-induced asystole may occur. The metabolism of lidocaine depends on normal liver function. Thus, the dose must be modified for children with chronic congestive heart failure, hepatitis, or cirrhosis.

Bretylium. A second line drug for ventricular tachycardia or ventricular fibrillation is bretylium. The mechanism of action of bretylium has not been elucidated fully. It has been noted to have a positive inotropic effect in addition to its antiarrhythmic action.

The initial dose of bretylium is 5 mg/kg IV given rapidly. Then defibrillation is attempted. If defibrillation is not successful, repeat bretylium at 10 mg/kg IV and repeat the defibrillation attempt.

The most common untoward effect of bretylium is hypotension. The patient should be placed in a Trendelenburg position, and the physician should be prepared to support the blood pressure with the administration of fluids. Bretylium is contraindicated in patients with a digitalis-induced arrhythmia. The resuscitation team should be prepared to deal with the side effect of drug-induced vomiting.

Adenosine. Adenosine is an endogenous purine nucleoside. It was first noted to have an effect on cardiac condition in the early part of the 20th century. In the mid-1950s, it was first successfully used for treatment of supraventricular tachycardia (SVT). Adenosine seems to exert a strong but brief depressant effect on the sinus and atrioventricular (AV) nodes, resulting in slowed conduction and interruption of the reentry pathway. The indication for the use of adenosine is SVT with hemodynamic compromise. Other treatments for stable SVT are outlined in Chapter 82

The dose of adenosine is 0.1 to 0.2 mg/kg (maximum dose is 12 mg). The half-life of the drug is estimated to be less than 10 seconds. If an initial lower dose is chosen and is unsuccessful, the dose may be doubled and repeated. The agent must be given in an intravenous bolus.

The untoward effects of adenosine are flushing and dyspnea, but these are short lived. In patients with “sick sinus syndrome,” long sinus pauses may occur. Adenosine has been used successfully in infants and children of all ages.

Dopamine. Dopamine is a precursor of epinephrine. It acts on both a- and b-adrenergic receptors. Dopamine has a unique “dopaminergic” effect that increases blood flow to renal and mesenteric blood vessels. The dopaminergic effect occurs over the low-dose range, 2 to 10 µ/kg per minute. The cardiac actions of dopamine are similar to those of epinephrine and include a positive inotropic and chronotropic effect. There is also an increase in peripheral vascular resistance, which causes an increase in blood pressure at moderate dosages, 5 to 20 µ/kg per minute (Table 1.12). At high dosages, greater than 20 µ/kg per minute, there is a marked increase in peripheral vascular resistance and a decrease in the renal and mesenteric blood flow.

Dopamine is indicated for the patient with hypotension and inadequate renal perfusion. The dosage of dopamine is 10 µg/kg per minute, which is within the range for desired cardiac action. The standard infusion may be made by mixing 60 mg of dopamine in 100 mL of D5W. This solution is infused at 1 µ, 1/kg per hour. Thus, the rate in milliliters per hour is equal to the patient's body weight (e.g., in a 20-kg patient, the solution is infused at a rate of 20 mL/hour) (Table 1.12). This simple method is appropriate for emergency department use because minimal calculation is required. When the patient reaches an inpatient critical care unit, an infusion better suited for precision drug titration can be prepared. Dopamine may produce tachyarrhythmias as an untoward reaction. Ectopic cardiac beats, nausea, and vomiting may occur. Myocardial ischemia is a rare event in children. The drug must be used with careful monitoring. Rapid increases or decreases in the dosage must be avoided. At low dosages, the dopaminergic effect may result in hypotension that must be supported with intravascular volume expansion. Extravasation of the drug into subcutaneous tissue may cause tissue necrosis. Thus, it should be given through a central line when possible. As with other catecholamines, dopamine should not be mixed with bicarbonate because this will inactivate the drug.

Isoproterenol. Isoproterenol is a synthetic catecholamine. The action of this drug is almost entirely through b-adrenergic receptors. The effects on the cardiovascular system are due to an increase in heart rate, an increased contractile force, and an increase in venous return to the heart. Unlike epinephrine, isoproterenol produces peripheral arterial dilation. Despite the decrease in vascular resistance, the drug usually produces an increase in blood pressure due to increased cardiac output.

Isoproterenol can be used for bradyarrhythmias that are not responsive to atropine. Heart block, sinus bradycardia, or nodal bradycardia may be treated while the more definitive therapy of electrical pacing is arranged.

The dose of isoproterenol is variable. A simple infusion can be prepared by adding 0.6 mg of isoproterenol to 100 mL of D5W. Infusing the solution at 1 µg/kg per hour will deliver 0.1 µg/kg per minute (Table 1.12)

The adverse reactions of isoproterenol include tachyarrhythmias and myocardial ischemia. It should be used with extreme caution in children receiving digitalis. In dehydrated or hypovolemic patients, isoproterenol may produce or aggravate existing hypotension. Thus, the physician must be prepared to support a further drop in blood pressure with

intravascular volume repletion.

Calcium. The actions of calcium are to increase myocardial contractibility, increase ventricular excitability, and increase conduction velocity through the ventricular muscle. There is currently a trend against the use of calcium in CPR because of possible cytotoxic effects, but for pediatric patients, the positive cardiac stimulant effects require that it remain on the list of secondary drugs.

The indications for calcium include asystole and electromechanical dissociation, but there is no good scientific support for this. Further indications include documented hypocalcemia, hyperkalemia, hypermagnesemia, and calcium channel blocker overdose.

The dose of calcium depends on the form of calcium used. Calcium chloride should be given as 10 to 20 mg/kg IV (to deliver 3 to 5 mg/kg of elemental calcium). In the chloride form, calcium should be given only through a central venous line. Calcium gluconate has properties that make the calcium ion less available. Thus, the dose is 30 mg/kg IV. Calcium gluconate may be given through a peripheral vein. When giving either form of calcium, the physician should infuse it slowly while watching the cardiac monitor for the appearance of bradycardia. The initial dose of calcium may be repeated once. However, subsequent doses should be guided by a serum ionized calcium level.

The untoward effects of calcium are significant. The patient who is made hypercalcemic may experience an arrest in systole. This is an untreatable situation in which even ECC will not be beneficial. Calcium must always be given slowly, particularly to the patient receiving digitalis who is more prone to develop arrhythmias.

Dembo and others have questioned the use of calcium in the asystolic adult patient. The measured serum calcium levels 5 minutes after infusion of a standard adult dose ranged between 12.9 and 18.2 mg/dL. Other studies show that calcium antagonist drugs appear to be protective against ventricular fibrillation in the ischemic myocardium. However, in our experience, fibrillation is not a common arrhythmia.

Furosemide. Furosemide is fast-acting and thus is the agent of choice for treating acute pulmonary edema and congestive heart failure. Furosemide works by inhibiting reabsorption of sodium in the proximal and distal tubules and in the loop of Henle. In addition to its action as a diuretic, furosemide also acts to increase blood flow to renal vasculature.

The initial dose of furosemide is 1 mg/kg IV. If there is not adequate urine output within 20 to 30 minutes, a repeat dose may be given or the dose may be doubled. If the patient is hypovolemic, the administration of furosemide may result in worsening of hypotension. Other adverse reactions include hypokalemia and hyperosmolality. Hypokalemia in the patient receiving digitalis therapy may result in the development of life-threatening arrhythmias.

Naloxone. Naloxone is a narcotic antagonist. It works to block the action of both synthetic and natural narcotics. Naloxone reverses the actions of codeine, morphine, heroin, hydromorphone, and methadone. Children who have overdosed on these drugs may have signs and symptoms of respiratory or cardiorespiratory arrest. The details of management of these poisonings are discussed in Chapter 88.

If narcotic overdose is suspected as the cause of the arrest, naloxone should be administered at 2 mg in a child older than 5 years of age or an adolescent, or 0.1 mg/kg of body weight in a child less than 5 years of age or an infant. This drug has a rapid onset of action and a short half-life. If a positive reaction to the agent is noted, a repeat dose of 0.1 mg/kg should be given as often as every 3 to 5 minutes up to a total of 10 to 20 mg. After three to five doses, a sustained effect should be apparent. No significant adverse reactions are noted with naloxone.

Corticosteroids. There continues to be great controversy about the specific indications for the use of corticosteroids. There is a general belief that steroids exert a positive effect on the shock state by stabilizing lysosomal membranes and preventing the release of histamine interleukin, tumor necrosis factor, and bradykinin. The possible clinical indications for the use of steroids include adrenal insufficiency either primary or due to adrenal suppression secondary to prolonged steroid use, anaphylaxis, and asthma.

The dose of corticosteroids is also controversial. However, current recommendations include methylprednisolone 30 mg/kg IV, or dexamethasone 1 mg/kg IV. The adverse reactions with short-term administration of corticosteroids are minimal. There may be worsening of hyperglycemia and retention of sodium and water. The worsening of a bacterial infection is a theoretical risk that should not inhibit the short-term use of steroids in a life-threatening situation.

Defibrillation

Defibrillation is a relatively uncommon intervention in pediatric resuscitation. It is unusual for a child's heart to fibrillate, and thus there should be careful confirmation of the rhythm before attempted defibrillation. Unmonitored defibrillation of a child is discouraged. However, if the onset of fibrillation was monitored and the defibrillator is at the bedside, direct defibrillation should be attempted. Precordial thump is not recommended for use in children.

Defibrillation works by producing a mass polarization of myocardial cells with the intent that a spontaneous sinus rhythm returns.

Once the diagnosis of ventricular fibrillation has been made, the patient should be prepared for defibrillation. Acidosis and hypoxemia should be corrected. If the patient was unobserved or if a long interval of poor perfusion has elapsed, 100% oxygen and sodium bicarbonate (1 mEq/kg) should be administered. Coarse (high-amplitude) fibrillation is treated more easily than fine (low-amplitude) fibrillation. Fibrillation can be coarsened with the administration of epinephrine. Defibrillation doses are given two at a time. If the first dose is ineffective, a second dose at the same energy level is

given immediately. The first dose will lessen resistance; thus, the second may be more effective.

Standard adult paddles are 8 cm in diameter. Pediatric paddles that are 4.5 cm in diameter are also available for most defibrillators. The correct size paddle is that which makes complete uniform contact with the chest wall. If the large size paddle fits entirely on the chest wall, it is preferred because the larger the paddle the lower the intrathoracic impedance and the more effective the defibrillation current. The electrodes should be prepared with electrode paste or saline-soaked pads. Placement of the paste or pads must be meticulous. The small size of the child's chest wall predisposes to bridging of the electric current. Electrical bridging will result in ineffective defibrillation and possible burning of the skin surface.

Once the correct paddles are selected and the electrode skin interface carefully prepared, the electrodes are ready to be placed on the chest wall. Both electrodes may be placed on the anterior chest wall, one at the right of the sternum below the clavicle and the other at the level of the xiphoid along the left midclavicular line. An AP placement of the electrodes is also acceptable; however, this is cumbersome in the usual resuscitation situation. Using either method of electrode placement, the physician should apply firm pressure to the paddles to hold them in contact with the skin. Personnel should be cleared from contact with the patient and the bed.

A dose of current for the initial two shocks is 2 J/kg. If the first defibrillation couplet is unsuccessful, CPR is continued for 3 to 5 minutes and then the dose of current is doubled to 4 J/kg and repeated twice if needed. If a third defibrillation round is needed, the dose is again doubled to 8 J/kg. Defibrillation should not be confused with synchronous cardioversion, which is a treatment for ventricular tachycardia or resistant supraventricular tachycardia. With cardioversion, the synchronous switch of the defibrillator must be activated and a low dose of current used (see Chapter 82 for more details)

Adverse reactions from defibrillation include myocardial injury from excessive current or from multiple discharges delivered in rapid succession (less than 3-minute intervals). Another adverse reaction is the damage to skin and subcutaneous tissue that occurs when the electrode skin interfaces are inadequate. Alcohol pads should never be used as an electrode interface because they are flammable.

Specific Resuscitation Situations

Asystole

For the treatment of asystole, we suggest the algorithm presented in Figure 1.25. No algorithm can cover every clinical situation, thus those presented in this chapter represent guidelines for treatment and not absolute treatment regimens. The physician must keep in mind the possible etiology of the arrest in order to modify treatment in some cases. It should be noted that asystole can be diagnosed clinically or by monitor. The monitor tracing may be confused with ventricular fibrillation. If there is a doubt that it is asystole, the resuscitation leader may want to follow the protocol for ventricular fibrillation.

FIGURE 1.25. Management approach to asystole.

Bradycardia

The algorithm for bradycardia management is shown in Figure 1.26. As with other algorithms, this represents only a treatment guideline and not an absolute treatment. Remember that bradycardia may result from hypoxemia and that the patient should first have assessment of the airway and breathing. Other causes of bradycardia include intrinsic node disease, increased intracardiac pressure, hypoglycemia, hypercalcemia, drug effect (e.g., digitalis, propranolol), increased parasympathetic tone (e.g., distended abdomen), or hypothermia. An athletic adolescent may have a resting heart rate of 48 to 60 beats/minute. Treatment must be clinically indicated by signs of cardiovascular failure. There are many forms of bradycardia, including sinus bradycardia, the most common form, and bradycardia from second- and third-degree heart blocks. The differentiation of these forms of heart block is covered in Chapter 79.

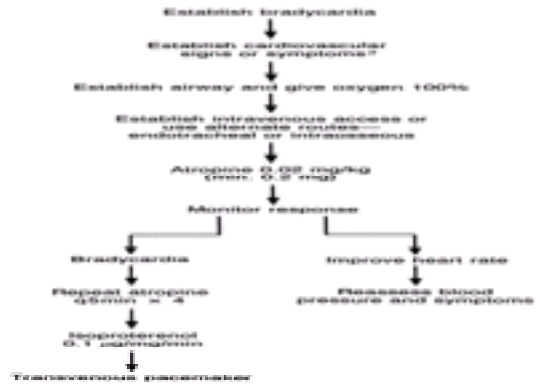


FIGURE 1.26. Management approach for bradycardia. [a]These include hypotension, altered mental status, poor peripheral perfusion, premature ventricular contractions, or chest pain.

Ventricular Fibrillation

Ventricular fibrillation initially may be mistaken for asystole. This depends on the sensitivity setting of the cardiac monitor and the coarseness (amplitudes) of the fibrillation. The treatment protocol for ventricular fibrillation is shown in Figure 1.27. It is essential that fibrillation be diagnosed correctly. Loose electrodes can readily simulate fibrillation; thus it is important to check the leads and correlate the monitor with the clinical appearance of the patient. It may be difficult to differentiate ventricular fibrillation from ventricular tachycardia with hypotension. This differentiation is moot because the same treatment protocol should be followed in either case.

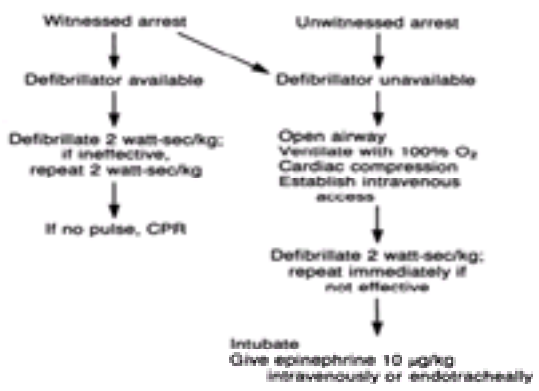


FIGURE 1.27. Management approach for ventricular fibrillation or ventricular tachycardia without pulse.

If the onset of fibrillation is witnessed, the immediate response is to attempt defibrillation. If the arrest was not witnessed, basic CPR must be initiated and the patient prepared before defibrillation is attempted. We do not recommend precordial thump for the pediatric patient.

Rarely is ventricular fibrillation an agonal rhythm in the pediatric patient. Its presence usually indicates some underlying cardiac pathology such as myocarditis, cardiomyopathy, prolonged Q-T syndrome, cardiac trauma, electrical shock, or intoxication with digitalis.

Electromechanical Dissociation

The term *electromechanical dissociation* (EMD), or pulseless electrical activity, refers to a state in which there is an observable cardiac rhythm on a monitor, yet no apparent effective circulation. A number of clinical conditions result in this form of arrest. Figure 1.28 shows the management scheme for EMD. The physician must keep in mind the potential causes of EMD and must assess the patient for each of these and treat specific causes when indicated.

FIGURE 1.28. Management approach to tachycardia with poor perfusion (for electromechanical dissociation.)

Tachycardia with Poor Perfusion

Tachycardia with poor perfusion is a condition that requires resuscitative effort. The underlying rhythm is usually SVT caused by aberrant reentry pathways due to defects in the normal cardiac conduction system. Most often, SVT is reasonably well tolerated by the infant or young child whose only manifestation may be increased restlessness, pallor, sweating, or poor feeding (see Chapter 82). Sometimes, however, SVT can result in cardiovascular collapse and circulatory failure requiring resuscitation. For the physician, the major point of caution is differentiation between SVT and sinus tachycardia. With SVT, heart rates are usually 230 beats/minute or higher. The P wave axis is usually abnormal (if P waves can be found) and there is little beat-to-beat variation. In sinus tachycardia, there is more rate variation, P waves have a normal axis, and the rate is usually less than 200 beats/minute. Also consider causes for sinus tachycardia such as dehydration, hypoxemia, fever, shock, pneumothorax, cardiac tamponade, or drug induced.

At times, it may also be difficult to differentiate SVT from ventricular tachycardia, particularly if the SVT is associated with aberrant conduction that widens the QRS waves of the ECG. Usually in SVT, the QRS is less than 0.08 seconds. Figure 1.28 outlines the treatment strategy for tachycardia with poor perfusion. If there is no pulse present, the physician should follow asystole management scheme.

Upper Airway Obstruction

If acute upper airway obstruction is not relieved properly and expeditiously, the child is likely to either die or sustain a hypoxemic CNS injury. The emergency department physician, pressured by time to move through a sequence of evaluation and management, knows that inefficiencies or mistakes place the child at increased risk. Unfortunately, this is a circumstance that often generates panic. Panic can be avoided and the child's life saved if calm prevails and an orderly sequence of evaluations and interventions occurs quickly and atraumatically.

Clinical Manifestations

If the clinical process predisposing to upper airway obstruction has not evolved to produce complete airway obstruction, the usual clinical manifestations are related to an increased work of breathing or compromised oxygen exchange. Intercostal retractions qualitatively reflect the increased work of breathing. If the negative intrapleural pressure required to produce gas exchange is increased, retractions will be evident in the intercostal spaces. As the work of breathing continues to increase, retractions will be transmitted beyond the thoracic cage and one may see subcostal or supraclavicular retractions. In the extreme situation, the sternocleidomastoid muscles are outlined by the negative intrapleural pressure transmitted into the neck. Another important clinical marker of airway obstruction is a decrease in breath sounds. Breath sounds should be auscultated both over the trachea and the peripheral lung fields. By following the pattern of inspiratory breath sounds, one can appreciate a gradual diminution during progression of the process of airway obstruction.

Upper airway noises, such as stridor, croupy cough, and “bark,” may be useful signs to identify those at risk for upper airway obstruction. The general appearance of air hunger or of gasping respirations may indicate airway obstruction. In addition, the symptoms of dysphagia, dysarthria, and dyspnea should trigger a thorough evaluation of the airway.

Management

A number of tasks must be accomplished as quickly as possible once the patient has been identified as being at high risk for total airway obstruction. The child should receive supplemental oxygen and have his heart rate and blood pressure monitored. Equipment for managing an obstructed airway should be immediately available (Table 1.5). The hospital's identified airway specialist should be notified of the child's condition and should move as quickly as possible to the emergency department. Beyond the emergency physician, if additional expertise is needed, specialty groups commonly identified as having airway expertise include anesthesiologists, otorhinolaryngologists, nurse anesthetists, general surgeons, respiratory therapists, and cardiothoracic surgeons. It is important to keep in mind that the two important elements for successful management are technical expertise and judgment. These attributes are not necessarily found in the same individual. What may be necessary is a cooperative effort between those with technical expertise and those with clinical experience and judgment.

If obstruction is not imminent, there is usually time to obtain a history and to perform an abbreviated physical examination. In addition, a chest radiograph and lateral neck radiographs may help to define more specifically the pathophysiologic process. These evaluations should occur only if there is no obvious evolution of the airway obstruction and if the natural history of the disease under consideration suggests further diagnostic studies.

If the patient is obtunded, the team must initiate the formal sequence of evaluation and management as described previously. The airway should be opened by flexing the cervical spine on the thoracic spine and tilting the head backward so that the skull is extended on the cervical spine. In addition to this maneuver, the jaw thrust or jaw pull may be used by pushing or pulling the mandibular block of tissue forward off of the posterior pharyngeal wall. At this time, the examiner evaluates the respiratory effort by looking, listening, and feeling for air flow. If the patient is breathing and there is gas exchange, the patient should receive supplemental oxygen and the sequence should progress toward an evaluation of the cardiovascular system. If the patient is not breathing, the oropharynx should be suctioned and an oropharyngeal or nasopharyngeal airway placed in an effort to open the airway. With the airway in place, the physician should reevaluate for gas exchange. If there is now gas exchange, supplemental oxygen should be administered. If there is no gas exchange, the physician should proceed immediately to laryngoscopy.

The purpose of laryngoscopy is twofold: the first priority is placement of an ET tube; second, the laryngoscopy may provide diagnostic information. If a foreign body is evident, the physician should try to intubate before attempting

removal.

If the tracheal tube can be placed, the child should be oxygenated and ventilated. Then the cause of the upper airway obstruction should be evaluated and treated. If the child cannot be intubated, a transcricothyroid membrane catheter should be inserted for delivery of supplemental oxygen (see Appendix D, Section VII). Jet ventilation may be accomplished through the catheter oxygenation system. Once the catheter is in place and oxygen is being administered, it is appropriate to reexamine the upper airway, to look for a cause of the obstruction, and to reattempt placement of an ET tube. If the airway cannot be secured at this time, a surgical team should be mobilized to perform a tracheostomy. Effective needle oxygenation and ventilation may permit transfer of the child to the operating room for tracheostomy. If circumstances do not allow oxygenation and ventilation, a formal tracheostomy may have to be performed in the emergency department.

STABILIZATION AND TRANSPORT

Once resuscitation efforts have achieved cardiorespiratory stability, the patient should be transported to an inpatient special care unit for the critically ill. This transport may be several hundred yards or several hundred miles to an appropriate pediatric hospital. In either circumstance, the patient should be transported with advanced life support technology in place, qualified personnel in attendance, and options for further treatment or intervention immediately available (see Chapter 6 and Chapter 7).

Waddel has reported on the direct and indirect effects of movement within the hospital. Direct effects include patient discomfort, pain, and the physical stimulation of movement. Indirect effects include lack of equipment and facilities and the limitations that occur by being in motion, for example, the ability to provide continuous ventilation while maneuvering the stretcher into a small elevator. All the equipment needed for a complete resuscitation should be available. Table 1.13 lists the equipment that is required. Transport should never be rushed because this may result in direct morbidity to the patient. Pediatric patients are particularly prone to hypothermia and should be warmed to avoid the adverse hemodynamic effects of cold stress. This is particularly important for any young infant who has not established temperature control or for the older child whose CNS pathology has resulted in loss of temperature regulation. Pediatric patients are likely to become physically active once CNS perfusion is adequate. Intravenous lines, ET tubes, and nasogastric tubes must be well secured. Extremities must be restrained. The transport team must include a sufficient number of individuals to fully resuscitation the patient. Responsibility for the patient still rests with the resuscitation team until discharge of that responsibility takes place at the inpatient critical care unit.

1. Airway box—items noted in Table 1.5.
2. Portable suction—compact, battery operated
3. Bag-valve-mask with O_2 reservoir or anesthesia bag
4. Oxygen tank with yoke and flow meter
5. Intravenous fluids
6. Primary and secondary medications
7. ECG monitor defibrillator
8. Infusion pump—battery operated
9. Blood pressure apparatus—battery operated, Doppler or ultrasound
10. Pulse oximeter (optional) and end-tidal CO_2 monitor

Table 1.13. Essential Equipment for Transport in Pediatric Resuscitation

DISCONTINUATION OF LIFE SUPPORT

Termination of life support in the emergency department is usually determined by whether the cardiovascular system can be supported with other than closed-chest massage. If the heart and supporting technology applied to it cannot sustain brain function, resuscitative effort should be discontinued. Obviously, time must be allowed to mobilize and implement for appropriate technology and therefore most unsuccessful resuscitations often go beyond 1 hour. There is growing evidence that if the cardiac muscle is not responsive to the first three doses of epinephrine in the state of adequate oxygenation and ventilation, there is no hope for a successful resuscitation. Additional research will be needed to confirm this as an absolute indicator of when to stop.

Respiratory functions are easily supported mechanically and therefore are not used as markers for continuation or discontinuation of effort.

Brain death is becoming widely accepted as the ultimate determinant of death. It is a clinical diagnosis that should be confirmed by observations over time or by studies documenting absence of cerebral blood flow or cerebral metabolic activity. We have seen infant botulism and postictal depression misdiagnosed as brain death and are therefore extremely cautious of making this diagnosis in the emergency department.

Ultimately, the diagnosis of death and subsequent discontinuation of resuscitative effort is a judgment that should be made by the senior physician who is directly attending the child. A decision not to begin resuscitation is generally not made in the emergency department unless there is a clear plan made with parents and written documentation of the plan transmitted by the child's primary attending physician.

CEREBRAL RESUSCITATION AND OTHER NEW APPROACHES

The term *cerebral resuscitation* refers to an important new focus of resuscitation research. The key concept is to monitor the effects of the hypoxic state on the cerebral nervous system and to discover what can be done to minimize or reverse these effects. Investigators have questioned the ethical appropriateness of “saving a heart” while the resultant brain injury renders the person vegetative. White and others have hypothesized that, in the brain, injury from cardiac arrest occurs during the reperfusion phase. The mechanism for this injury is through ion-dependent lipid peroxidation. White also noted an increase in iron released in the brain for 2 hours following a 15-minute arrest induced in laboratory animals. In the future, cerebral resuscitation may be aimed at reducing lipid peroxidation and iron release during the reperfusion phase following arrest.

Other new approaches have centered on methods of increasing blood flow, so-called new CPR. The techniques have been used to attempt simultaneous chest compression and ventilation to increase ventilatory flow rates and longer systolic compression times. Other efforts have been to use abdominal binding, chest binders, or cardiopulmonary bypass. Mateer et al. have studied the use of interposing abdominal compressions between chest compressions. To date, all these techniques are experimental and none has shown so clear an advantage that use in humans is indicated.

SUMMARY

In most circumstances, resuscitation of the pediatric patient can be approached with a sense of optimism for reversing the process that acutely threatens the child's life. Well-organized and well-qualified personnel can effect a high rate of successful resuscitation. However, organization and qualification require advanced planning, training, and preparation. Inherent in this preparation is the development of personnel disciplined to follow the sequence of evaluation and management for airway, breathing, and circulation. In addition, personnel must be familiar with the nuances of resuscitation peculiar to the age and size ranges of the pediatric population. A complete list of pediatric resuscitation equipment may be found in the Appendix A.

Suggested Readings

Alexander R, Swales H, Pickford A, et al. The laryngeal mask airway and the tracheal route for drug administration. Br J Anaesthesia 1997;78: 220–221.

American Academy of Pediatrics/American College of Emergency Physicians. Advanced Pediatric Life Support. Elk Grove Village, IL: AAP Publication, 1989:1–209.

American Heart Association/American Academy of Pediatrics. In: Chameides L, ed. Pediatric Advanced Life Support. Dallas: AHA Publication, 1997.

American Heart Association/Scientific Statement. Recommended Guidelines for Uniform Reporting of Pediatric Advanced Life Support: The Pediatric Utstein Style. Circulation 1995;92:2006–2020.

Atkins DL, Sirna S, Kieso R, et al. Pediatric defibrillation: importance of paddle size in determining transthoracic impedance. Pediatrics 1988;82:914–918.

Babbs CF, Tacker WA, Paris RL, et al. CPR with simultaneous compression and ventilation at high airway pressure. Crit Care Med 1982;10:501–504.

Barton C, Callaham M. High-dose epinephrine improves the return of spontaneous circulation rates in human victims of cardiac arrest. Ann Emerg Med 1991;20:722–725.

Barton CW, Manning JE. Cardiopulmonary resuscitation. Emerg Med Clin North Am 1995;13:811–829.

Bhende MS, Thompson AE. Evaluation of an end-tidal CO_2 detector during pediatric cardiopulmonary resuscitation. Pediatrics 1995;95: 395–399.

Bhende MS, Thompson AE, Cook DR, et al. Validity of a disposable end-tidal CO_2 detector in verifying endotracheal tube placement in infants and children. Ann Emerg Med 1992;21:142–145.

Bonnin MJ, Pepe PE, Kimball KT, et al. Distinct criteria for termination of resuscitation in the out-of-hospital setting. JAMA 1993;270:1457–1462.

Boxer RA, Gottesfeld I, Singh S, et al. Noninvasive pulse oximetry in children with cyanotic congenital heart disease. Crit Care Med 1987;15:1062–1064.

Brown CG, Martin DR, Pepe PE, et al. A comparison of standard-dose and high-dose epinephrine in cardiac arrest outside the hospital. N Engl J Med 1992;327:1051–1055.

Cairns CB, Niemann JR. Intravenous adenosine in the emergency department management of paroxysmal supraventricular tachycardia. Ann Emerg Med 1991;20:717–721.

Callaham M, Barton CW, Kayser S. Potential complications of high-dose epinephrine therapy in patients resuscitated from cardiac arrest. JAMA 1991;265:1117–1122.

Carpenter TC, Stenmark KR. High-dose epinephrine is not superior to standard-dose epinephrine in pediatric in-hospital cardiopulmonary arrest. Pediatrics 1997;99:403–408.

Chameides L. CPR challenges in pediatrics. Ann Emerg Med 1993;22: 388–392.

Cote CJ, Jobes DR, Schwartz AJ, et al. Two approaches to cannulation of a child's internal jugular vein. Anesthesiology 1979;50:371–383.

Crespo SG, Schoffstall JM, Fush LR, et al. Comparison of two doses of endotracheal epinephrine in a cardiac arrest model. Ann Emerg Med 1991;29:230–234.

Dembo DH. Calcium in advanced life support. Crit Care Med 1981;9:358–359.

Dieckmann RA, Vardis R. High-dose epinephrine in pediatric out-of-hospital cardiopulmonary arrest. Pediatrics 1995;95:901–913.

Eldadah MK, Schwartz PH, Harrison R, et al. Pharmacokinetics of dopamine in infants and children. Crit Care Med 1991;19:1008–1011.

Emergency Cardiac Care Committee and Subcommittees, American Heart Association. Guidelines for cardiopulmonary resuscitation and emergency cardiac care. VI. Pediatric advanced life support. JAMA 1992;268:2262–2275.

Finer NN, Barrington KJ, Al-Fadley F, et al. Limitations of self-inflating resuscitators. Pediatrics 1986;77:417–420.

Fiser DH, Wrape V. Outcome of cardiopulmonary resuscitation in children. Pediatr Emerg Care 1987;3:235–238.

Friesen RM, Duncan P, Tweed WA, et al. Appraisal of pediatric cardiopulmonary resuscitation. Can Med Assoc J 1982;126:1055–1058.

Gillis J, Dickson D, Rieder M, et al. Results of inpatient pediatric resuscitation. Crit Care Med 1986;14:469–471.

Goetting MG. Mastering pediatric cardiopulmonary resuscitation. Pediatr Clin North Am 1994;41:1147–1182.

Goetting MG. Progress in pediatric cardiopulmonary resuscitation. Emerg Med Clin North Am 1995;13:291–319.

Goetting MG, Paradis N. High-dose epinephrine improves outcome from pediatric cardiac arrest. Ann Emerg Med 1991;20:22–26.

Goetting MG, Paradis NA. High dose epinephrine in refractory pediatric cardiac arrest. Crit Care Med 1989;17:1258–1262.

Guyer B, Martin JA, MacDorman MF, et al. Annual summary of vital statistics–1996. Pediatrics 1997;100:905–918.

Hickey RW, Cohen DM, Strausbaugh S, et al. Pediatric patients requiring CPR in the prehospital setting. Ann Emerg Med 1995;25:495–501.

Hodge D. Intraosseous infusions: a review. Pediatr Emerg Care 1985; 1:215–218.

Innes PA, Summers CA, Boyd IM, et al. Audit of paediatric cardiopulmonary resuscitation. Arch Dis Child 1993;68:487–491.

Kanter RK, Zimmerman JJ, Strauss RH, et al. Pediatric emergency intravenous access. Am J Dis Child 1986;140:132–134.

Kaye W, Bircher NG. Access for drug administration during cardiopulmonary resuscitation. Crit Care Med 1988;16:179–182.

Kellerman AL, Hackman BB, Somes G. Predicting the outcome of unsuccessful prehospital advanced cardiac life support. JAMA 1993;270:1433–1436.

Kettrick RG, Ludwig S. Resuscitation in infants and children. Curr Top Emerg Med 1981;1:4.

Krause GS, Joyce KM, Magini NR, et al. Cardiac arrest and resuscitation: brain iron delocalization during reperfusion. Ann Emerg Med 1985;14:1037–1043.

Kuisma M, Suominen P, Korpela R. Paediatric out-of-hospital cardiac arrests: epidemiology and outcome. Resuscitation 1995;30:141–150.

Kulick RM. Pulse oximetry. Pediatr Emerg Care 1987;3:127–130.

Landwirth J. Ethical issues in pediatric and neonatal resuscitation. Ann Emerg Med 1993;22:502–507.

Lavelle JM, Costarino AT Jr, Ludwig S. Recognition and management of the ill child. Compr Ther 1995;21:711–718.

Lee-Terry KW, Templeton JJ, Dougal RM, Downes JJ. Selection of endotracheal tubes in infants and children. Presented to the American Academy of Pediatrics, Annual Meeting, Las Vegas, April, 1980 (Unpublished Data)

Lewis JK, Minter MG, Eshelman SJ, et al. Outcome of pediatric resuscitation. Ann Emerg Med 1983;12:297–299.

Lubitz DS, Seidel JS, Chameides L, et al. A rapid method for estimating weight and resuscitation drug dosages from length in the pediatric age group. Ann Emerg Med 1988;17:576–580.

Ludwig S, Fleisher G. Pediatric cardiopulmonary resuscitation: a review and a proposal. Pediatr Emerg Care 1985;1:40–44.

Ludwig S, Kettrick RG, Parker M. Pediatric cardiopulmonary resuscitation: a review of 130 cases. Clin Pediatr 1984;23:71–76.

Ludwig S, Selbst S. A child-oriented emergency medical services system. Curr Prob Pediatr 1990;20:113–158.

Mateer JR, Stueven HA, Thompson BM, et al. Prehospital IAC-CPR versus standard CPR: Paramedic resuscitation of cardiac arrest. Am J Emerg Med 1985;3:143–146.

McNamara RM, Spivey WH, Sussman C. Pediatric resuscitation without an intravenous line. Am J Emerg Med 1986;4:31–33.

Michaud LJ, Rivara FR, Longstreth WT, et al. Elevated initial blood glucose levels and poor outcome following severe brain injuries in children. J Trauma 1991;31:1356–1362.

Mondolfi AA, Grenier BM, Thompson JE, et al. Comparison of self-inflating bags with anesthesia bags for bag-mask ventilation in the pediatric emergency department. Pediatr Emerg Care 1997;13:312–316.

Nadkarni V, Hazinski MF, Zideman D, et al. Paediatric life support: an advisory statement by the Paediatric Life Support Working Group of the International Liaison Committee on Resuscitation. Resuscitation 1997;34:115–127.

Natanson C, Shelhamer JH, Parrillo JE. Intubation of the trachea in the critical care setting. JAMA 1985;253:1160–1165.

Newman BM, Jewelt TC, Karp MP, et al. Percutaneous central venous catheterization in children: first line choice for venous access. J Pediatr Surg 1986;21:685–688.

Nichols DG, Kehrick RG, Swedlow DB, et al. Factors influencing outcome of cardiopulmonary resuscitation. Pediatr Emerg Care 1986;2:1–5.

O'Marcaigh AS, Koenig WJ, Rosekrans JA, et al. Cessation of unsuccessful pediatric resuscitation: how long is too long? Mayo Clin Proc 1993;68:332–336.

Orlowski JP. Optimum position for external cardiac compression in infants and young children. Ann Emerg Med 1986;15:667–673.

O'Rourke PP. Outcome of children who are apneic and pulseless in the emergency room. Crit Care Med 1986;14:466–468.

Palmi L, Nystrom B, Tunell RB. An evaluation of the efficiency of face masks in the resuscitation of newborn infants. Lancet 1985;1:207–210.

Paradis NA, Martin GB, Rosenberg J. The effect of standard- and high-dose epinephrine on coronary perfusion pressure during prolonged cardiopulmonary resuscitation. JAMA 1991;265:1139–1144.

Plaisance P, Adnet F, Vicaut E, et al. Benefit of active compression-decompression cardiopulmonary resuscitation as a prehospital advanced cardiac life support: a randomized multicenter study. Circulation 1997;95:955–961.

Raffin TA. Withdrawing life support: how is the decision made? JAMA 1995;273:738–739.

Reed AP. Current concepts in airway management for cardiopulmonary resuscitation. Mayo Clin Proc 1995;70:1172–1184.

Ronco R, King W, Donley DK, et al. Outcome and cost at a children's hospital following resuscitation for out-of-hospital cardiopulmonary arrest. Arch Pediatr Adolesc Med 1995;149:21–214.

Ros SP, Fisher EA, Bell TJ. Adenosine in the emergency management of supraventricular tachycardia. Pediatr Emerg Care 1991;7:222–223.

Rosenberg NM. Pediatric cardiopulmonary arrest in the emergency department. Am J Emerg Med 1984;2:497–499.

Sacchetti AD, Linkenheimer R, Lieberman M, et al. Intraosseous drug administration: successful resuscitation from asystole. Pediatr Emerg Care 1989;5:97–98.

Sanders AB. Capnometry in emergency medicine. Ann Emerg Med 1989;18:1287–1289.

Sayah AJ, Peacock WF, Overton DT. End-tidal CO_2 measurement in the detection of esophageal intubation during cardiac arrest. Ann Emerg Med 1990;19:857–860.

Schindler MB, Bohn D, Cox PN, et al. Outcome of out-of-hospital cardiac or respiratory arrest in children. N Engl J Med 1996;335:1473–1479.

Schreiner MS, Kettrick RG, Ludwig S. Pediatric cardiopulmonary resuscitation. Clin Crit Care Med 1986;10:279–303.

Scribano PV, Baker MD, Ludwig S. Factors influencing termination of resuscitative efforts in children: a comparison of pediatric emergency medicine and adult emergency medicine physicians. Pediatr Emerg Care 1997;13:320–324.

Seidel JS. Emergency medical services and the pediatric patient: are the needs being met? Part II. Pediatrics 1986;78:808–812.

Seidel JS, Horinbein M, Yoshiyam AK, et al. Emergency medical services and the pediatric patient: are the needs being met? Part I. Pediatrics 1984;73:769–772.

Selbst SM, Baker MD, Bell LM, et al. Teaching technical skills in pediatrics. Indian J Pediatr 1989;56:35–54.

Sheikh A, Brogan T. Outcome and cost of open- and closed-chest cardiopulmonary resuscitation in pediatric cardiac arrests. Pediatrics 1994;93:392–398.

Singh GK, Yu SM. US childhood mortality, 1950 through 1993: trends and socioeconomic differentials. Am J Public Health 1996;86:505–512.

Slonim AD, Patel KM, Ruttimann UE, et al. Cardiopulmonary resuscitation in pediatric intensive care units. Crit Care Med 1997;25: 1951–1955.

Smith JP, Bodai BI. Guidelines for discontinuing prehospital CPR in the emergency department: a review. Ann Emerg Med 1985;14:1093–1098.

Smith RJ, Keseg DP, Manley LK, et al. Intraosseous infusions by prehospital personnel in critically ill pediatric patients. Ann Emerg Med 1988;17:491–495.

Smyrnios NA, Irwin RS. The jury on femoral vein catheterization is still out. Crit Care Med 1997;25:1943–1946.

Spivey WH. Medical progress. J Pediatr 1987;111:639–643.

Spivey WH, Lathers CM, Mazone DR. Comparison of intraosseous, central, and peripheral routes of sodium bicarbonate administration during CPR in pigs. Ann Emerg Med 1985;14:1135–1140.

Tang W, Weil MH, Schock RB, et al. Phased chest and abdominal compression-decompression: a new option for cardiopulmonary resuscitation. Circulation 1997;95:1335–1340.

Teach SJ, Moore PE, Fleisher GR. Death and resuscitation in the pediatric emergency department. Ann Emerg Med 1995;25:799–803.

Thaler MM, Stobie HC. An improved technique of external cardiac compression in infants and young children. N Engl J Med 1963;269:606–610.

Thompson JE, Bonner B, Lower GM Jr. Pediatric cardiopulmonary arrests in rural populations. Pediatrics 1990;86:302–306.

Torphy DE, Minter MG, Thompson BM. Cardiorespiratory arrest and resuscitation of children. Am J Dis Child 1984;138:1099–1102.

Torres A Jr, Pickert CB, Firestone J, et al. Long-term functional outcome of inpatient pediatric cardiopulmonary resuscitation. Pediatr Emerg Care 1997;13:369–373.

Torres NE, White RD. Current concepts in cardiopulmonary resuscitation. J Cardiothorac Vascul Anes 1997;11:391–407.

Varon J, Fromm RE Jr. Cardiopulmonary resuscitation: new and controversial techniques. Postgraduate Med 1993;93:235–239, 242.

Waddel G. Movement of critically ill patients within hospitals. Br Med J 1975;2:417–419.

Wagner MB, McCabe JB. A comparison of four techniques to establish intraosseous infusion. Pediatr Emerg Care 1988;4:87–91.

Wark H, Overton JH. A paediatric “cardiac arrest” survey. Br J Anaesth 1984;56:1271–1274.

Weil MH, Tang W. Cardiopulmonary resuscitation: a promise as yet largely unfulfilled. Dis Mon 1997;43:429–501.

Weisfeldt ML, Chandra N, Bilik J. Increased intrathoracic pressure—not direct heart compression. Crit Care Med 1981;9:377–378.

White BC, Wiegenstein JG, Winegar CD. Brain ischemic anoxia: mechanisms of injury. JAMA 1984;251:1586–1590.

Zaritsky A. Drug therapy of cardiopulmonary resuscitation in children. Drugs 1989;37:356–374.

Zaritsky A. Pediatric resuscitation pharmacology. Members of the Medications in Pediatric Resuscitation Panel. Ann Emerg Med 1993;22:445–455.

Zaritsky A, Nadkarni V, Getson P, et al. CPR in children. Ann Emerg Med 1987;16:1107–1111.

CHAPTER 2

Neonatal Resuscitation

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EPIDEMIOLOGY

Approximately 4 million infants are born each year in the United States. Although medical personnel attend virtually all births, more than 85% of infants are born in hospitals without neonatal facilities. Despite the fact that more U.S. women are seeking prenatal care in the first trimester of their pregnancies and that there has been a decline in women who receive late or no prenatal care, the United States has one of the highest infant mortality rates of developed countries: 7.2 per 1000 live births in 1996. This rate is markedly affected by birth weight. The infant mortality rate in 1996 was 3.0 per 1000 for infants who weighed 2500 g or more and 65.5 per 1000 for infants who weighed less than 2500 g.

Resuscitative needs also vary greatly by birth weight. Approximately 6% of term newborns will require resuscitation at birth, compared with nearly 80% of infants weighing less than 1500 g. Improvements in prenatal care, infant care, and neonatal resuscitation, as well as the development of the specialty of neonatology and neonatal regional centers, have decreased infant deaths and mortality rates by nearly 45% between 1979 and 1996. The most notable decreases have occurred because of advances in the management of respiratory distress and intrauterine hypoxia and birth asphyxia. Resuscitation of the newborn in the emergency department (ED) is an uncommon yet critical event that can cause a ED team well versed in other resuscitative scenarios to panic. The key to a successful newborn resuscitation for the ED team includes preparedness of staff and equipment and anticipation of high-risk births.

EMERGENCY DEPARTMENT PREPAREDNESS

The best place for the birth of a newborn infant is in the delivery suite. However, because of varying circumstances, infants are born at home, in the prehospital setting, and in the ED. Most neonatal resuscitations in the ED occur without prior notice. Any knowledge that can be obtained before arrival of the laboring mother or recently born infant will aid in the success of the resuscitation. Education of staff, available and functioning equipment, and familiar policies and procedures are critical for preparedness.

Staff

The American Heart Association (AHA) and the American Academy of Pediatrics (AAP) have been leaders in developing guidelines for and offering training in neonatal resuscitation since 1979. Their Pediatric Advanced Life Support (PALS) course offers didactic and skills teaching for neonatal resuscitation that occurs outside of the delivery room. In addition,

resuscitation in the delivery room is addressed in depth in the Neonatal Resuscitation Program (NRP). In concert with the goals of the AHA and the AAP, all personnel responsible for care of newborns should complete courses and maintain their skills in the area of newborn resuscitation. At least one person skilled in newborn resuscitation should attend every delivery. Additional trained personnel must be available for high-risk deliveries, such as most of those occurring in the ED or other locations outside of the delivery room. Ideally, there should be three members on the team that are trained to work together. Identification and training of staff is the first step in preparation for neonatal resuscitation.

Equipment

In addition to a standard obstetric tray, every ED should have a newborn resuscitation kit that is readily accessible, maintained meticulously with other emergency equipment, and rapidly replenished after use. Necessary equipment and medications are listed in Table 2.1. A medication dosing chart by weight and a radiant warmer are invaluable to a neonatal resuscitation.

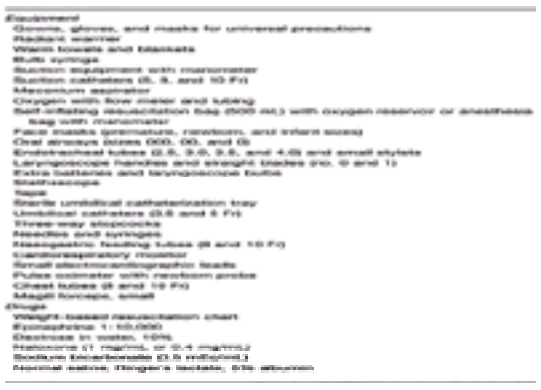

Table 2.1. Neonatal Resuscitation Equipment and Drugs

Policies and Procedures

As soon as the need for neonatal resuscitation becomes evident, a prearranged plan should be activated to organize personnel and assemble equipment. Readily available procedure logs for accessing pediatric and neonatal consultants, as well as neonatal transport teams for transfer to regional centers, are critical. Because neonatal resuscitations in the ED are uncommon, mock codes and scavenger hunts for newborn equipment on a routine basis allow staff to remain familiar with their neonatal resuscitation skills and supplies.

HIGH-RISK BIRTHS

Most births that occur outside of the delivery room have high-risk components such as trauma-induced labor and unexpected or teenage pregnancy. There is usually little time to obtain a complete obstetric history, but a brief period of questioning may reveal pertinent information that will affect a successful newborn resuscitation. Particularly important information includes prematurity, multiple gestation, meconium-stained amniotic fluid, and maternal drug use. The team can then anticipate the need for assisted ventilation, simultaneous resuscitations, tracheal suctioning, and potential naloxone administration, respectively. Table 2.2 lists other risk factors associated with the need for neonatal resuscitation.

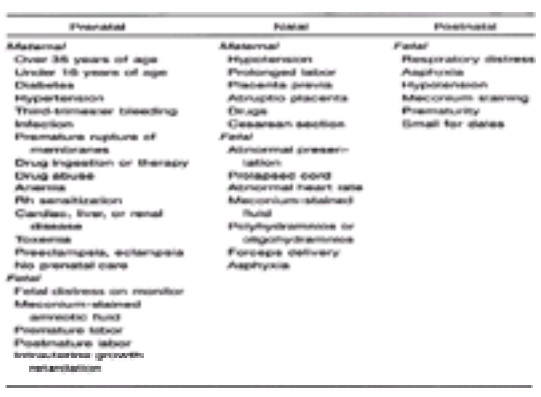

Table 2.2. Neonatal High-Risk Profile

PATHOPHYSIOLOGY

Physiology of Intrauterine Development

The lungs develop over the second and third trimester of pregnancy. Terminal airways develop by approximately 24 weeks' gestation, and the alveoli by 30 to 32 weeks. Surfactant is initially produced by about 23 to 24 weeks; however, sufficient amounts for opening the airways are not present until 34 weeks' gestation. In utero, the lung is filled with amniotic fluid, which is primarily removed by chest compression during vaginal birth. Preterm infants or those born by

cesarean section tend to have more fluid in their lungs. At birth, the key physiologic change is the initiation and maintenance of respiration. Factors such as cold, touch, hypoxia, and hypercarbia help stimulate respiration. However, severe acidosis, hypoxia, maternal drugs, and moderate hypothermia depress this effort.

The heart and circulatory system start developing during the third week of gestation. In utero, the circulation is more like a parallel circuit rather than a series circuit because of the foramen ovale and ductus arteriosus that serve as bypasses. After birth, these structures close physiologically. Severe acidosis, hypoxia, hypovolemia, and hypothermia can impair the closure. Anatomic closure of the bypasses may not occur for 2 to 4 weeks. The fetal heart is also sensitive to hypoglycemia because of the neonate's limited energy stores, and myocardial failure can occur if the infant becomes hypoglycemic.

Changes at Birth

The fetus has two large right-to-left shunts: one from the right atrium to the left atrium through the foramen ovale, and the second from the pulmonary artery to the aorta across the ductus arteriosus. The placenta is the gas-exchange organ, which provides a low-resistance shunt compared with the high resistance of the fetus' peripheral circulation. At birth, two major changes occur that eliminate these shunts: the umbilical cord is clamped and then, respirations are initiated. Expansion of the lungs increases the neonate's Pa_{O_2} and pH, which causes pulmonary vasodilation and a fall in pulmonary vascular resistance. The normal heart rate will vary between 100 and 200 beats/minute initially and then stabilize between 120 and 150 beats/minute.

The normal newborn will begin spontaneous respirations within seconds after birth. The normal rate will be between 35 and 60 breaths/minute. The initial breaths taken by the infant must inflate the lungs and effect a change in vascular pressures so that lung water is absorbed into the pulmonary arterial system and cleared from the lung. This inflation pressure is a powerful mechanism for the release of pulmonary surfactant, which increases compliance of the lung and establishes a functional residual capacity.

The neonate oxidizes free fatty acids released from the brown fat stores for heat production and increases oxygen consumption. The neonate experiences substantial heat loss by all four heat loss mechanisms, especially if he or she is not dried promptly and thoroughly.

Asphyxia

Asphyxia is defined as the failure to provide the cell with oxygen and remove carbon dioxide, resulting in metabolic acidemia. Both circulation and ventilation are essential to avoid asphyxia. There are multiple stimuli at birth to alter the prenatal circulation and initiate respirations. The actual stimuli for initiating respirations are thought to include a rise in $PaCO_2$, interruption of umbilical circulation, and tactile and temperature stimulation.

Neonatal asphyxia can result from multiple factors, as listed in Table 2.3. The initial response to asphyxia will be hyperpnea for 2 to 3 minutes and sinus tachycardia. If there is no significant increase in Pa_{O_2}, respirations will stop for 1 to 1½ minutes (primary apnea). The infant loses muscle tone and becomes mottled, pale, and then cyanotic. The infant may attempt gasping, nonrhythmic respiratory efforts of 6 to 10 times/minute for several minutes, while the heart rate falls below 100 beats/minute. Soon thereafter, the child ceases to gasp (secondary apnea). At this point, ventilatory and circulatory support must be aggressively provided for the newborn to survive. Brain and other organ damage progresses rapidly beyond this point.

Maternal	Fetal
Diabetes	Abnormal presentation
Hypertension	Meconium aspiration
Toxemia	Sepsis
Preeclampsia	Hypovolemia
Eclampsia	Prolapsed cord
Treatment with alcohol, magnesium, β-adrenergic agents, narcotics	Congenital anomalies
Isoimmunization	
Infection	
Abruptio placenta	
Placenta previa	

Table 2.3. Causes of Neonatal Asphyxia

It is important to realize that when one evaluates a neonate in distress or full arrest, the asphyxial event may have begun in utero. It is difficult to document the beginning of the hypoxic period. Indeed, the infant may have passed through both stages of apnea in utero. Thus, there must be aggressive intervention if the infant is to survive. The rule of thumb is that for every minute of secondary apnea, the infant will require 4 minutes of artificial ventilation before rhythmic breathing is reestablished. An apneic infant must be treated as if he or she is in a secondary apneic stage, and resuscitation must begin immediately. If hypoxemia is not treated, there may be further pulmonary vasoconstriction and increased right-to-left shunting through the ductus arteriosus and foramen ovale and a persistence of fetal circulation.

CLINICAL MANIFESTATIONS

Successful resuscitation of a depressed newborn requires accurate assessment of the infant's temperature, respiratory

effort, heart rate, color, and tone. Assessment of these critical parameters occurs simultaneously with management of any detected abnormality in a rapid and timely fashion. The evaluation of the clinical manifestations of a depressed newborn should take place along with resuscitative efforts within the first minute after birth. Complete assessment is performed after the infant is dried and placed in a warm environment, the airway is cleared, and stimulation has been provided.

Temperature

Particular attention must be paid to the thermoregulation of all infants, especially those born in a prehospital setting or in a cool ED. As the patient is dried and placed under a radiant warmer, the temperature should be monitored via the axillary route using electronic thermometers with a disposable tip. Normal axillary temperatures fall between 36.5 and 37.4°C. Rectal temperatures are reserved for infants whose core temperature may be in question. Recovery from acidosis is delayed by hypothermia. In addition, hypothermia increases metabolic needs and produces hypoxia, hypercarbia, metabolic acidosis, and hypoglycemia. Thus, efforts to maintain a normal body temperature are crucial to a successful resuscitation.

Respiratory Effort

Most newborns will begin to breathe effectively in response to mild stimulation. The infant should be assessed for respiratory rate (between 35 and 60 breaths/minute is normal). Adequacy of respirations is noted by evaluating chest rise, auscultating good air movement, and confirming a heart rate above 100 beats/minute with improving color of the infant. Observation of tachypnea, retractions, or grunting warrants close evaluation and management. A gasping, cyanotic, or unresponsive infant requires immediate respiratory support with oxygenation and ventilation (see Management section following).

Heart Rate

The newborn's heart rate is an excellent objective measurement of the success of the resuscitation and should be monitored closely after assessment of respiratory effort. The heart rate may be determined by one of many ways: 1) palpation of the pulse at the base of the umbilical cord, 2) auscultation of heart tones with a stethoscope, 3) palpation of the femoral or brachial pulse, or 4) placement of a cardiac monitor. Auscultation of the apical heart rate is often difficult in a noisy environment, and the electrodes of a cardiac monitor may be difficult to place while vernix covers the newborn's body. The normal infant's heart rate is greater than 100 beats/minute at birth. The average awake infant's heart rate is between 120 and 150 beats/minute shortly after birth. Variations in heart rate commonly occur with hypoxia, hypovolemia, hypothermia, and maternal drug use. Trends in heart rate are followed closely during resuscitation and postresuscitation stabilization.

The average mean arterial pressure of term infants in the first 12 hours of life is between 50 and 55 mm Hg.

Color

As respirations begin and pulmonary vascular pressures fall, the newborn rapidly becomes pink. Acrocyanosis, or persistent cyanosis of the distal extremities, may persist for several hours after birth. Acrocyanosis is not a reflection of inadequate oxygenation, but it may indicate hypothermia if persistent. Pallor may be a sign of decreased cardiac output, anemia, hypovolemia, hypothermia, or acidosis. Its cause should be investigated and corrected promptly. Central cyanosis that has not resolved with administration of oxygen and ventilation within the first minute of life must be emergently evaluated for heart disease, sepsis, diaphragmatic hernia, other congenital anomalies, or other causes.

Apgar Score

The Apgar score is a useful guide to evaluate the newborn at specific intervals after birth. Five objective signs—heart rate, respirations, muscle tone, grimace, and color—are assessed 1 minute and 5 minutes after birth. Each sign receives a score between 0 and 2, and the points are then totaled for the final score (Table 2.4). If the 5-minute Apgar score is less than 7, additional scores are obtained every 5 minutes until the infant is 20 minutes old. The Apgar score has been used as an indicator of responsiveness to resuscitative efforts. The score at 5 minutes and beyond is more predictive of survival and neurologic status. Although experienced physicians have developed these guidelines, they have not undergone rigorous clinical trials. Thus, if resuscitative efforts are needed for a newborn infant, they should be started immediately and not be delayed while the Apgar score is obtained.

	Score		
Sign	0	1	2
Heart rate	Absent	<100 beats/min	≥100 beats/min
Respirations	Absent	Slow, irregular	Good, crying
Muscle tone	Limp	Some flexion	Active motion
Reflex irritability	None	Grimace	Cough, sneeze, cry
Color	Blue or pale	Pink body, blue hands and feet	Completely pink

Table 2.4. Apgar Score

MANAGEMENT

The initial steps of neonatal resuscitation include prevention of heat loss, stimulation, clearing of the airway, initiation of respirations, and evaluation of circulation. When possible, all resuscitation equipment should be ready for use, the radiant warmer on, and a team with preassigned roles assembled. Figure 2.1 is a flow diagram of neonatal resuscitation.

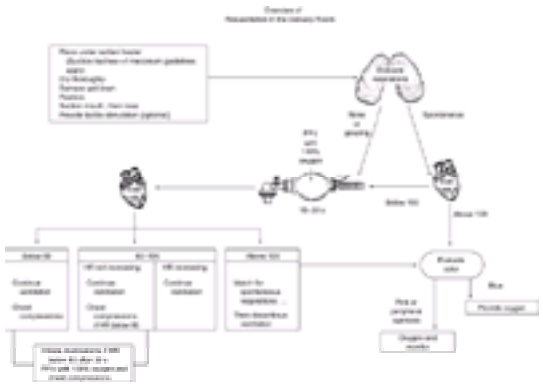

FIGURE 2.1. Overview of resuscitation in the delivery room.

When and if to begin resuscitative efforts on a newborn is fraught with emotion and difficult to objectively study. Studies have shown that between 40 and 50% of term newborns who were stillborn survived. Approximately two-thirds of these infants had a normal neurologic outcome. It thus seems obvious that resuscitative efforts should be performed on any term infant. A difficult decision is when to stop resuscitation. One predictor is the Apgar score. Survival is extremely unlikely if the 10-minute Apgar score remains zero. There are multiple ethical issues regarding initiation of resuscitation of the very low–birth weight infant. However, with surfactant therapy and improved management of these infants, outcomes have improved over time and the controversy of resuscitation remains. At the stressful time of an emergency delivery, if there is any question of viability, it is probably best to initiate resuscitative efforts.

Thermoregulation

The initial step of drying the infant to minimize heat loss is extremely important, and further resuscitation is continued after warming has begun. As previously stated, recovery from acidosis is delayed by hypothermia, and hypothermia is a special problem for the infant born outside of the hospital. Thus, simply resuscitating the baby in a warm environment, under a prewarmed radiant warmer (Fig. 2.2) while drying the amniotic fluid from the infant and removing wet linens from contact with the skin will markedly decrease heat loss. These maneuvers will maximize the infant's chance at recovery. Alternative methods of warming infants, particularly while awaiting a radiant warmer in the case of an unexpected delivery, include warm blankets and towels. Placing the infant naked against the mother's body and covering both mother and infant with blankets may also warm the stable infant.

FIGURE 2.2. Radiant warmer.

Suctioning

Many newborns have excessive secretions, including amniotic fluid, cervical mucus, and meconium, which may obstruct their airways. (Meconium is a special situation discussed in the following section.) These secretions can generally be removed by placing the infant on his or her side and gently suctioning the buccal pouch with a bulb syringe. Mechanical suction with an 8- or 10-Fr suction catheter may also be used. To avoid soft tissue injury, negative pressure from mechanical suctioning should not exceed –100 mm Hg. Deep suctioning of the oropharynx in a newborn is likely to cause vagally mediated bradycardia and/or apnea. Excessive suctioning may also contribute to atelectasis. Most clear fluid is resorbed by the lungs into the arterial system. Consequently, suctioning should be gentle and limited to 5 seconds per pass. Adequate time must be allowed between necessary suctioning attempts for spontaneous or assisted ventilation.

Stimulation

Most newborns will begin effective breathing during routine drying and suctioning. Other methods of safe stimulation include flicking the heels and rubbing the back of the newborn infant. More vigorous methods of stimulation are unnecessary. If after 10 to 15 seconds of stimulation, effective respirations have not been established, positive-pressure ventilation (PPV) is initiated.

Airway and Breathing

Oxygen Administration and Airway Positioning

Most infants require only warming, drying, stimulation, and suctioning after birth for a smooth transition to their extrauterine environment. If a newborn is exhibiting signs and symptoms of airway obstruction after routine suctioning, the airway should be repositioned. This maneuver is often accomplished by placing a towel or blanket beneath the shoulders and upper back of the supine infant. By elevating the shoulders and upper back approximately 1 inch, the airway is slightly extended into a neutral position, compensating for the infant's relatively large occiput. Avoid flexion or hyperextension of the newborn's neck, which is likely to exacerbate airway obstruction.

An infant who exhibits central cyanosis, yet is making adequate, spontaneous respirations and has a heart rate above 100 beats/minute, needs supplemental oxygen. Oxygen is delivered at 100% and a flow rate of at least 5 L/minute by blow-by through tubing, a face mask attached to an anesthesia bag, an appropriately sized simple mask, or an oxygen hood. Ideally, oxygen should be warmed and humidified. Although this may not always be possible initially in an emergency setting, efforts to warm and humidify oxygen delivered to a newborn should be made as soon as possible.

Bag-Valve-Mask Ventilation

Adequate expansion of the lung is often the only and most important measure needed for successful resuscitation of the newborn. The fluid-filled lungs must be inflated with air. Adequate inflation stimulates surfactant secretion and also allows some gas trapping during exhalation to create a functional residual capacity. Although this is best done by negative pressure generated by a vigorous term infant with a strong chest wall, some infants require PPV to initiate lung expansion. Indications for PPV are listed in Table 2.5.

Apnea or gasping respirations
Heart rate <100 beats/min
Persistent central cyanosis despite administration of 100% oxygen

Table 2.5. Indications for Positive-Pressure Ventilation

Lung expansion is best achieved with a well-fitted face mask, which covers the infant's nose and mouth but does not place pressure on the eyes. A cushioned rim on the face mask allows the best possible seal. A relatively high inflation pressure, between 25 and 40 cm H_2O, delivered slowly over several seconds is necessary for the infant's first breath. Subsequent ventilations typically require less pressure and are best judged by good chest wall rise and breath sounds. If effective ventilation does not result, the airway should be repositioned and suctioning of the oropharynx considered. An assisted ventilatory rate of 40 to 60 breaths/minute will provide effective oxygenation and ventilation. Typically 100% oxygen is delivered via PPV. However, some authors advocate resuscitation with room air because of concerns about the generation of free radicals from high concentrations of oxygen, which may exacerbate brain injury. Although current findings do not justify changes in guidelines for resuscitation, further work in this arena may have implications for neonatal resuscitation in the future.

PPV may be delivered by a self-inflating bag or an anesthesia bag. Although self-inflating bags do not require a gas source to operate, they must be used with an oxygen source and a reservoir to deliver high concentrations of oxygen. They are straightforward and easy to use, but several caveats must be kept in mind. First, relatively small volumes of air (approximately 6 to 8 mL/kg) are delivered to newborns during PPV. A 450-mL self-inflating bag rather than the larger bags should be used to avoid complications from barotrauma such as a pneumothorax. In addition, many self-inflating bags have a pressure-limiting pop-off valve set at 30 to 45 cm H_2O. In some circumstances, when an infant requires higher initial inflation pressures, the bag may not allow the resuscitator to deliver enough pressure to the newborn for an adequate first breath. Unless the valve is occluded, effective inflation may be prevented.

Anesthesia bags require air flow into them as well as a good mask seal to inflate. Consequently, the resuscitator must be facile at positioning the airway and mask, controlling the flow valves, and monitoring the manometer, which is needed to monitor peak ventilatory pressures delivered to the infant. Benefits of the anesthesia bag include the ability to deliver a wide range of peak inspiratory pressures, positive end-expiratory pressure, and high concentrations of oxygen compared with the self-inflating bag. Proper use requires training and practice.

Recently, investigators have successfully used a size 1 laryngeal mask airway to resuscitate 20 newborns who required PPV. This method of airway management warrants further investigation.

If bag-valve-mask ventilation is prolonged or results in gastric distension, an orogastric tube should be placed to decompress the stomach so that further effective ventilation is not inhibited. The infant should be reevaluated after 30 seconds of PPV for spontaneous respirations and heart rate. If the infant has begun breathing and the heart rate is above 100 beats/minute, PPV may be slowly discontinued. If respirations are inadequate or the heart rate remains less than 100 beats/minute, assisted ventilation must be continued and endotracheal (ET) intubation must be considered.

Endotracheal Intubation

Most resuscitative efforts succeed with bag-valve-mask ventilation alone. Indications for ET intubation are listed in Table 2.6. Once the decision to intubate the trachea has been made, supplies from the newborn resuscitation tray are organized. Sizes of airway equipment can be determined by birth weight (Table 2.7). ET tube size can also be estimated by gestational age:

$$\text{ET tube size in mm} = \frac{\text{Gestational age in weeks}}{10}$$

Ineffective bag-valve-mask ventilation
Prolonged need for positive-pressure ventilation
Suctioning of meconium
Administration of resuscitation medications

Table 2.6. Indications for Endotracheal Intubation

Weight (g)	Endotracheal Tube Size (mm)	Suction Catheter (Fr)	Oral Airway	Laryngoscope Straight Blade
<1000	2.5	5	000	0
1000-1250	2.5, 3.0	5, 6	000	0
1250-2500	3.0	6	00	0, 1
2500-3000	3.0, 3.5	6, 8	0	1
>3000	3.0, 3.5, 4.0	8	0	1

Table 2.7. Selection of Airway Equipment by Weight

Thus, a 35-week premature infant would require a 3.5-mm ET tube.

ET intubation is typically performed via the orotracheal route during direct laryngoscopy. Laryngoscopy in the newborn is challenging because of the infant's large tongue and secretions, which may obscure airway landmarks. Hancock et al. advocate finger intubation of the trachea in newborns. They successfully and quickly used this method to intubate 37 infants and had no complications. The technique requires some practice, as does laryngoscopic intubation.

Successful ET intubation also requires proper tube positioning. Most neonatal ET tubes have a black vocal cord line near the tip. When this guide is placed at the level of the vocal cords, the tip of the tube is likely to be positioned properly in the trachea. Another estimate for the insertion distance of the ET tube is:

$$\text{Total centimeters at gum line} = 6 + \text{Weight of the infant in kilograms}$$

Proper positioning of the ET tube must be confirmed by auscultation of equal breath sounds in both axillae; good, symmetric chest wall movement; improvement of the infant's cardiorespiratory status; and detection of exhaled carbon dioxide. Once positioning is clinically verified, the ET tube must be securely taped in place, and positioning may then be confirmed with a radiograph as indicated.

Circulation

Chest Compressions

Chest compressions are rarely needed during neonatal resuscitation. In 1993, Jain reported that 0.03% of newborns delivered required chest compressions (in 1995, Perlman demonstrated a need in 0.12%). Bradycardia and asystole in the newborn are virtually always a result of respiratory failure, hypoxemia, and tissue acidosis. Consequently, oxygenation and ventilation are critical to successful resuscitation even of the infant's circulation. Indications for chest compressions, which are always performed simultaneously with PPV with 100% oxygen, are listed in Table 2.8.

Heart rate <60 beats/min
Heart rate 60-80 beats/min and not rapidly increasing despite positive-pressure ventilation with 100% oxygen for 30 seconds

Table 2.8. Indications for Chest Compressions

Current recommendations state that 3 chest compressions are followed by a brief pause for 1 ventilation. Thus, in 1 minute, the newborn should receive 90 chest compressions and 30 ventilations. This technique, when compared with previous recommendations of 120 compressions and 40 to 60 simultaneous respirations per minute, allows for optimal lung expansion by not compressing the chest during PPV. The most important aspect of reversing neonatal asphyxia, good oxygenation, and ventilation is maximized.

Two techniques of performing chest compressions in the neonate or young infant are recommended. The preferred method involves placing the thumbs on the middle third of the sternum, encircling the chest and supporting the back with the fingers (Fig. 2.3). Ultimately, the thumbs should be placed side by side just below the nipple line. However, if the neonate is very small or if the resuscitator is large, the thumbs may need to be superimposed. Pressure must be placed on the sternum and not the adjacent ribs. In the event that the resuscitator's hands are too small to encircle the newborn's chest, or encircling the chest obstructs other resuscitative efforts such as umbilical line placement, then the two-finger technique may be used. This method entails placing the ring and middle fingers on the sternum just below the nipple line for chest compressions.

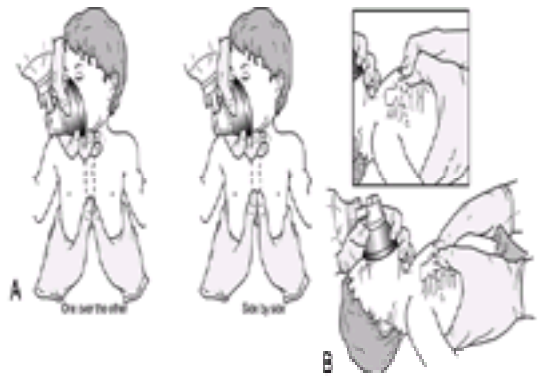

FIGURE 2.3. A. Thumb method of chest compressions. Infant receiving chest compressions with thumb 1 fingerbreadth below the nipple line and hands encircling chest. **B.** Hand position for chest encirclement technique for external chest compressions in neonates. Thumbs are side by side over the midsternum. In the small newborn, thumbs may need to be superimposed (inset). Gloves should be worn during resuscitation.

With either method of chest compression, the resuscitator should compress the chest approximately ½ to ¾ of an inch in a smooth fashion. The compression and relaxation phases should be equal in duration, and the fingers or thumbs should not be lifted off of the chest at any time. The spontaneous heart rate should be checked periodically during the resuscitation, and chest compressions discontinued when the heart rate exceeds 80 beats/minute.

Vascular Access

A newborn requires vascular access for administration of medications or volume expansion. Bradycardia or asystole unresponsive to effective oxygenation, ventilation, and chest compressions warrant pharmacologic therapy. Infants exhibiting signs of poor perfusion, particularly those with risk for hypovolemia such as fetal hemorrhage or maternal hypotension from placental abruption, require volume expansion.

Several methods of vascular access may be used in the newborn. The umbilical vein is often considered a preferred site for vascular access during neonatal resuscitation because it is easily located and cannulated. See Section VII, Umbilical

Vein Catheterization for methods. A skilled resuscitator may elect to cannulate the umbilical artery to obtain arterial blood gases as well as to monitor arterial pressures in a significantly ill infant. See Section VII, Umbilical Artery Catheterization. Vascular access may also be obtained by placing peripheral catheters in the extremities or scalp (see Section VII, Intraosseous Infusion). In a newborn resuscitation scenario, peripheral venous access may be difficult. In the event that fluids and medications are required and other methods of vascular access have failed, intraosseous lines may be used. Although experience with other infants is extensive in this arena, experience in the neonate is limited. A 20- or 22-gauge spinal needle may replace the 16- or 18-gauge larger intraosseous needles; however, the procedure for line placement in the proximal tibia is the same as for older children (see Appendix 3.8). Finally, the ET tube may be used for administration of epinephrine and naloxone hydrochloride when vascular access has not yet been established.

Medications and Volume Expanders

Epinephrine

Epinephrine is the most commonly needed medication for neonatal resuscitation. Because asystole and bradycardia are usually the result of respiratory failure and tissue acidosis, epinephrine therapy is indicated when the newborn's heart rate remains less than 80 beats/minute despite effective ventilation with 100% oxygen and chest compressions for approximately 30 seconds. Epinephrine works because of its a-adrenergic effects. Swine models have demonstrated that a-vasoconstriction in infants increases the diastolic and mean arterial pressures and thus increases the perfusion pressure to the coronary arteries, enhancing oxygen delivery to the heart. The b-adrenergic effects of increased myocardial contractility and stimulation of spontaneous contractions appear less important.

The dose of epinephrine therapy in neonates is 0.01 to 0.03 mg/kg of a 1:10,000 concentration, or 0.1 to 0.3 mL/kg (Table 2.9). It may be administered via an umbilical catheter, a peripheral IV, an intraosseous line, or the ET tube. The dose should be repeated every 3 to 5 minutes as needed throughout the resuscitation. The safety and efficacy of high-dose epinephrine (0.1 to 0.2 mg/kg) has not been studied in neonates. A concern that large doses of epinephrine may lead to prolonged hypertension and subsequent intracranial hemorrhage in neonates has precluded changing dosing recommendations. In addition, the AHA continues to recommend that the dose of endotracheally administered epinephrine remain at 0.01 to 0.03 mg/kg. This is because of the findings by Lindemann in 1984 that 10 infants unresponsive to PPV and chest compressions were successfully resuscitated using regularly recommended doses of epinephrine. If the neonate has no vascular access and fails to respond to PPV with 100% oxygen, and a standard dose of epinephrine by the ET route, administration of a larger epinephrine dose (0.1 mg/kg) by the ET route may be considered.

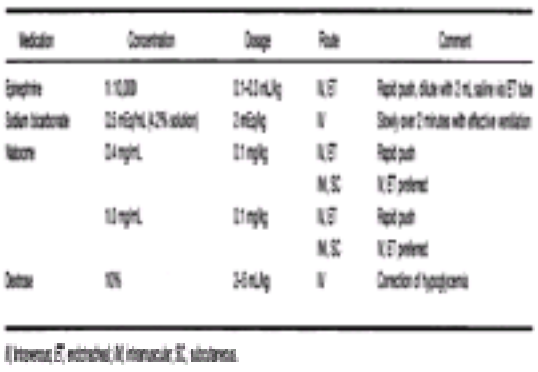

Table 2.9. Medications for Neonatal Resuscitation

Sodium Bicarbonate

Bicarbonate therapy is indicated in neonatal resuscitation, after establishment of adequate ventilation, for documented metabolic acidosis or presumed metabolic acidosis when other resuscitative measures have failed. The dose is 1 to 2 mEq/kg administered intravenously and slowly over 2 minutes to decrease adverse effects associated with its hypertonicity. For the same reason, only the 0.5 mEq/kg (4.2%) solution should be administered to neonates. If only the 1 mEq/kg solution is available, it should be diluted 1:1 with sterile water before intravenous delivery.

Recommendations for bicarbonate therapy have changed recently. In animal studies performed over the last 40 years, acidosis has been shown to reduce cardiac contractility, blood pressure, and heart rate responses to catecholamines. Subsequently, sodium bicarbonate therapy was recommended before administration of epinephrine. Correction of metabolic acidosis is very dependent on carbon dioxide removal by pulmonary gas exchange. This in turn is reliant on minute ventilation and pulmonary blood flow. Recently, investigators have discovered that in a resuscitation scenario, both minute ventilation and pulmonary blood flow are decreased. As a result, hypercarbia ensues and exacerbates intracellular acidosis. Therefore, bicarbonate should be administered only to the well-ventilated infant with a documented metabolic acidosis.

Naloxone Hydrochloride

Naloxone is a narcotic antagonist that reverses respiratory depression induced by narcotics. Naloxone is indicated for infants displaying signs of respiratory depression whose mothers have received narcotics within the 4 hours before delivery. Prompt and effective oxygenation and ventilation must be provided before administration of naloxone. The current dosing recommendation for naloxone is 0.1 mg/kg, which may be given as 0.1 mL/kg of the 1 mg/mL concentration or 0.25 mg/mL of the 0.4 mg/mL concentration (Table 2.9). This increased dosage from previously

published statements has been found more effective in opiate reversal. Naloxone is best administered via the intravenous, intraosseous, or ET routes. Sporadic and delayed absorption may occur if the medication is given intramuscularly or subcutaneously, particularly in an infant with poor perfusion. Furthermore, the resuscitator must remember that repetitive doses of naloxone may be required because the duration of action of narcotics may exceed that of naloxone.

Atropine

Atropine is a parasympatholytic drug that reduces vagal tone and accelerates sinus or atrial pacemakers and atrioventricular conduction. Because vagal stimulation does not cause bradycardia in neonatal resuscitation, atropine is not indicated. Furthermore, many investigators believe that the bradycardic vagally mediated response to hypoxia is a valuable reflex to guide resuscitative efforts and should not be pharmacologically abolished by atropine.

The usual dose of atropine is 0.02 mg/kg with a minimum dose of 0.1 mg and a maximum dose of 2 mg. Because most newborns weigh less than 5 kg, their dose would require the 0.1 mg minimum. If smaller doses are given, paradoxical bradycardia and slowed atrioventricular conduction will likely occur. In conclusion, the efficacy of atropine in newborn resuscitation is unproven and anecdotal and could have deleterious consequences.

Volume Expanders

Volume expanders are indicated for the treatment of hypovolemia. Both historical and physical examination findings suggest the need for volume expansion. Historical factors include fetal hemorrhage from an avulsed cord or trauma, or maternal hypotension from placenta previa, placental abruption, or trauma. Umbilical cord prolapse may cause hypovolemia in the newborn. Physical examination findings include pallor that persists despite oxygenation, weak peripheral pulses with a good heart rate, and a poor response to resuscitation, including effective ventilation.

Volume expanders (Table 2.10) are given intravenously in 10 mL/kg aliquots; after each infusion, the infant is reassessed for improvements in perfusion, blood pressure, and oxygenation. It is appropriate to begin with a bolus of normal saline or Ringer's lactate and proceed to albumin or packed red blood cells based on the infant's clinical response. Volume replacement should be given over 10 minutes to decrease the risk of intracranial hemorrhage from delicate vascular beds.

Fluid	Dosage	Route
5% albumin	10 mL/kg	IV
Normal saline	10 mL/kg	IV
Ringer's lactate	10 mL/kg	IV
Packed red blood cells	10 mL/kg	IV

IV, Intravenous.

Table 2.10. Volume Expanders for Neonatal Resuscitation

Post-resuscitation Stabilization

After appropriate resuscitative efforts, continuous monitoring and anticipation of complications must occur until the patient is safely transported to a neonatal facility. Priority must be given to thermoregulation by providing the infant with a warm environment and repetitively monitoring the temperature. Measures of effective oxygenation and ventilation are assessed. Pulse oximetry and arterial blood gases are performed. ET tubes are securely taped and a chest radiograph is ordered to confirm tube and venous access placement. Vascular access is secured, and correction of metabolic acidosis and hypovolemia is continued.

If mechanical ventilation is required while waiting transport to the neonatal facility, a pressure ventilator is used. Peak pressures are determined by clinical evaluation of adequate chest wall rise and blood gas analyses. A good starting point for peak pressure is that pressure needed for good chest wall rise and breath sounds during resuscitation as shown on the manometer. In general, this is between 15 and 30 cm H_2O. The physician should try to use the lowest pressure necessary for good clinical and laboratory response. Excessive positive pressure will decrease venous return to the heart, decrease cardiac output, and cause injury to lung tissue.

SPECIAL SITUATIONS

Meconium

Meconium staining of the amniotic fluid complicates between 10 and 20% of all pregnancies. The risk of meconium at delivery increases to nearly 30% in infant's born after 42 weeks' gestation. Approximately 2 to 5% of infants born with meconium in the amniotic fluid will experience some degree of aspiration syndrome, ranging from mild tachypnea to very severe pneumonitis with persistent pulmonary hypertension (Fig. 2.4). The management of an infant born through meconium differs from that previously discussed for other depressed infants. Efforts to remove meconium from the

oropharynx and trachea must precede other interventions because of the risk of its aspiration into the lungs with the infant's first breath.

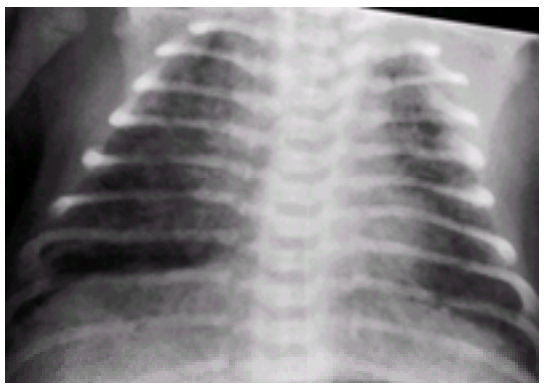

FIGURE 2.4. Meconium aspiration radiograph.

When meconium staining is detected during delivery, a sequence of events to prevent aspiration must occur (Fig. 2.5). As soon as the infant's head is delivered, suctioning of the mouth, nose, and posterior pharynx must occur with a 12- or 14-Fr suction catheter or bulb syringe at the perineum. Suctioning must be performed before delivery of the shoulders and thorax to decrease the risk of aspiration. After delivery, the infant is placed in a warm environment in a slight Trendelenburg position, and before other usual resuscitative efforts, meconium suctioning is completed. First, the oropharynx is visualized with a laryngoscope, and any remaining meconium is removed with a suction catheter. Next, the trachea is intubated, and suctioning of the lower airway occurs. Because the ET tube itself is the largest diameter item placed in the trachea, it is the most effective means of suctioning viscous meconium. Thus, a meconium aspirator directly attached between the ET tube and mechanical suction is the preferred method of removing meconium from the lower airway. Negative pressure is applied by occluding the opening on the side of the aspirator with a finger. Mechanical suctioning should not exceed –100 mm Hg. It may be necessary to repeat intubation and suctioning with another ET tube until the aspirated material is clear. Upon completion of tracheal suctioning, orogastric suctioning is performed to empty meconium from the newborn's stomach, which could potentially be aspirated later.

FIGURE 2.5. Diagram of management of infant born with meconium-stained amniotic fluid.

Several controversies exist regarding the management of meconium-stained amniotic fluid. Although all sources agree that thick, particulate meconium necessitates ET suctioning, management of thin meconium in a vigorous infant remains without consensus. Many authors, including the AHA and AAP, advocate ET suctioning for all infants with meconium-stained fluid. Their recommendations are supported by the finding that meconium aspiration syndrome necessitating prolonged periods of mechanical ventilation occur in infants with thin meconium as well as those vigorous infants with Apgar scores greater than 8. In addition, several studies have demonstrated no adverse sequelae of intubation. However, many authors contend that infants born through thin meconium warrant visualization of the cords with a laryngoscope and ET suctioning only if meconium is present in the larynx and the infant is depressed. As yet, no large prospective studies have investigated the benefits or complications of either a selective or nonselective approach to the management of the infant with thin meconium-stained amniotic fluid.

Prematurity

Prematurity greatly increases the likelihood of needing newborn resuscitation. Early involvement of neonatologists and neonatal centers adept in the management of low–birth weight infants is crucial to improve outcome. Only 15% of hospitals have specialized neonatal units. Hospitals without neonatal units need easily available guidelines and established relationships for accessing neonatal consultation and transport. Several factors have added importance in the resuscitation of the preterm infant. These include greater risk for heat loss, greater mechanical ventilation needs, and greater risk of intraventricular hemorrhage.

Premature infants are at greatest risk for heat loss because of their higher ratio of body surface area to body mass. Premature infants require strictest attention to maintenance of normal body temperature.

Premature infants are more likely to develop respiratory distress than term infants are. As a result, assisted ventilation

must be provided effectively but gently. ET intubation is usually necessary for surfactant administration and transport to a neonatal facility. Too much ventilatory pressure may result in barotrauma to the lungs and decreased cardiac output as a result of decreased venous return. Good clinical judgment should be used by watching for adequate chest wall rise and listening for good breath sounds. Then, the physician should use the lowest pressure necessary to achieve these clinical end points, which can be confirmed by blood gas analysis. Hyperoxia may lead to complications such as retinopathy of prematurity in low–birth weight infants. Once the infant is stabilized after initial resuscitative care, the fraction of inspired oxygen can be decreased while monitoring pulse oximetry.

The germinal matrix of the preterm infant's brain is vulnerable to bleeding. Factors contributing to subsequent intracranial hemorrhage include excessive pressure or osmolality delivered to an already maximally dilated vascular bed. Subsequently, in premature infants, hyperosmolar solutions such as 25% dextrose or 8.4% sodium bicarbonate should be avoided. Volume expanders, dextrose, and sodium bicarbonate solutions, when indicated, should be administered slowly to minimize injury to these vascular beds.

Pneumothorax

Pneumothorax is a potentially lethal problem in the neonate because it can rapidly progress to a tension pneumothorax and thereby decrease cardiac output. It is often the result of PPV, positive end-expiratory pressure, or resuscitation.

Pneumothorax is also more common in premature infants with surfactant deficiency and in infants with meconium aspiration. Signs and symptoms include grunting respirations; intercostal, sternal, and substernal retractions; elevated respiratory rate; and tachycardia followed by bradycardia and hypotension. The physical examination findings may include decreased breath sounds and distant heart tones. However, it often may not be possible to diagnose or localize a pneumothorax by auscultation. Transillumination by a high-intensity light in a dark room will reveal increased light transmission on the side of the pneumothorax.

If significant respiratory distress is present and pneumothorax is suspected, rapid decompression may be achieved with a large syringe, 20-gauge needle or catheter over needle, and three-way stopcock. The chest is cleansed with antiseptic solution, and the needle is advanced at the fourth intercostal space in the anterior axillary line or the second interspace in the midclavicular line. This will relieve the tension and decompress the pleural space. Subsequently, a chest tube (8 Fr) may be placed using a standard technique (see Section VII, Insertion of a Chest Tube). If the infant is stable, an expedient portable anteroposterior (AP) chest radiograph may be taken to confirm the diagnosis.

Diaphragmatic Hernia

Diaphragmatic hernia is a true neonatal emergency and may be suspected by tachypnea, asymmetric chest wall motion, and a scaphoid abdomen. The diagnosis is confirmed by a chest radiograph showing bowel gas within the thorax (Fig. 2.6). The patient should be given oxygen and a nasogastric tube placed to decompress the stomach. Intubation and PPV are often necessary. The infant must be rapidly evaluated by a pediatric surgeon after ventilation is stabilized and venous access is achieved.

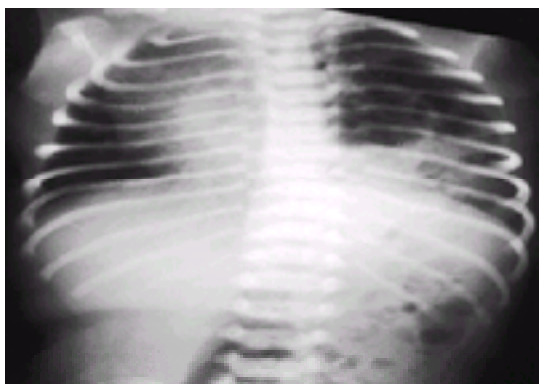

FIGURE 2.6. Left diaphragmatic hernia.

Omphalocele/Gastroschisis

Omphalocele and gastroschisis are defects of the umbilical ring that allow herniation of the abdominal contents outside of the abdominal wall. An omphalocele is covered by a thin layer of amnion that may be intact or broken. The abdominal contents are free-floating in the amniotic fluid in gastroschisis (Fig. 2.7). Cardiovascular malformations are commonly associated with omphalocele. The infant must be kept dry and warm, and the eviscerated bowel covered by warm saline-soaked gauze and placed in a plastic bag. If a sac covers the omphalocele and the sac is intact, it should be covered with saline-soaked gauze. A nasogastric tube must be placed and oxygen and IV fluids given. The infant may be hypovolemic from peritoneal fluid loss. The physician should maintain good peripheral perfusion and a urine output of 1 to 2 mL/kg per hour. The infant must be evaluated promptly by a pediatric surgeon who can repair these defects.

FIGURE 2.7. Gastroschisis.

Spina Bifida

Spina bifida (meningocele, meningomyelocele, and lipomeningocele) involves a wide array of defects. It can range from the least significant form (spina bifida occulta, nonfusion of vertebral laminar arches) to the severe form with meninges and neural tissue protruding, with poorly organized cord tissue exposed to the surface. Neurologic deficit ranges from none to severe impairment and associated hydrocephalus. The child should receive proper supportive care, oxygen and fluid (as needed), sterile moist dressings to the exposed sac or tissues, and prompt referral to a pediatric neurosurgeon.

Suggested Readings

Adamsons K, Gandy GM, et al. The influence of thermal factors upon oxygen consumption of the newborn human infant. J Pediatr 1965;66:45.

Apgar V. A proposal for a new method of evaluation of the newborn infant. Anesth Analg 1953;32:260.

American Academy of Pediatrics Committee on Drugs. Emergency drug doses for infants and children and naloxone use in newborns: clarification. Pediatrics 1989;83:803.

American Heart Association. Standard and guidelines for cardiopulmonary resuscitation (CPR) and emergency care (ECC). JAMA 1992;268:2276–2281.

Berkowitz D, Gervais B, et al. Epinephrine dosage affects cerebral and myocardial blood flow in an infant swine model of cardiopulmonary resuscitation. Anesthesiology 1991;73:1041–1050.

Bloom RS, Cropley C, eds. Textbook of Neonatal Resuscitation. Dallas: American Heart Association and American Academy of Pediatrics, 1987.

Boon AW, Milner AD, et al. Lung expansion, tidal volume, and formation of the functional residual capacity during resuscitation of asphyxiated neonates. J Pediatr 1979;85:1031.

Burchfield DJ. Medication use in neonatal resuscitation: epinephrine, sodium bicarbonate. Neonatal Pharmacol Q 1993;2:2.

Chameides L, Hazinski MF, eds. Newborn Resuscitation. Pediatric Advanced Life Support. Dallas: American Heart Association, 1997.

Cordero L, Hon EH. Neonatal bradycardia following nasopharyngeal stimulation. J Pediatr 1971;78:441.

Cunningham AS, Lawson EE, et al. Tracheal suction and meconium: a proposed standard of care. J Pediatr 1990;116:153.

David R. Closed chest cardiac massage in the newborn infant. Pediatrics 1988;81:552–554.

Goetting MG, Paradis NA. High-dose epinephrine improves outcome from pediatric cardiac arrest. Ann Emerg Med 1991;20:22–26.

Guyer B, Martin JA, et al. Annual summary of vital statistics 1996. Pediatrics 1997;100:905–918.

Hancock PJ, Peterson G. Finger intubation of the trachea in newborns. Pediatrics 1992;89:325.

Handal KA, Schauben JL, et al. Naloxone. Ann Emerg Med 1983;12:438–445.

Hoskyns EW, Milner AD, Hopkin IE. A simple method of face mask resuscitation at birth. Arch Dis Child 1987;62:376–378.

Jain L, Ferre C, et al. Cardiopulmonary resuscitation of apparently stillborn infants: survival and long-term outcome. J Pediatr 1991;118:778–782.

Kauffman RE, Banner W, et al. Naloxone dosage and route of administration for infants and children: addendum to emergency drug doses for infants and children. Pediatrics 1990;86:484.

Kelsall AW. Resuscitation with intraosseous lines in neonatal units. Arch Dis Child 1993;68:324–325.

Lantos JD, Miles SH, Silverstein MD, et al. Survival after cardiopulmonary resuscitation in babies of very low birth weight. Is CPR futile therapy? N Engl J Med 1988;318:91–95.

Leuthner SR, Janes RD, et al. Cardiopulmonary resuscitation of the newborn. Pediatr Clin North Am 1994;41:893.

Milner AD. Resuscitation of the newborn. Arch Dis Child 1991;66:66–69.

Moore KL. The Developing Human. Philadelphia: WB Saunders, 1977.

Perlman JM, Risser R. Cardiopulmonary resuscitation in the delivery room: associated clinical events. Arch Pediatr Adolesc Med 1995;149:20.

Peterson SJ, Byrne PJ, et al. Neonatal resuscitation using the laryngeal mask airway. Anesthesiology 1994;80:1248.

Sangstad OD, Rootwelt T, Allen O. Resuscitation of asphyxiated newborn infants with room air or oxygen: an international controlled trial, The

Resair 2 study. Pediatrics 1998;102:e 1.

Sinkin RA, Davis JM. Cardiopulmonary resuscitation of the newborn. Pediatr Rev 1990;12:136–141.

Taeusch HW, Ballard RA, eds. Avery's Diseases of the Newborn. 7th ed. Philadelphia: WB Saunders, 1998.

Tarnow-Mordi WO. Room air or oxygen for asphyxiated babies? Lancet 1998;352:341.

Todres ID, Rogers MC. Methods of external cardiac massage in the newborn infant. J Pediatr 1975;86:781.

Upton CJ, Milner AD. Endotracheal resuscitation of neonates using a rebreathing bag. Arch Dis Child 1991;66:39–42.

Wiswell TE, Tuggle JM, et al. Meconium aspiration syndrome: have we made a difference? Pediatrics 1990;85:715–721.

CHAPTER 3

Shock

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All physicians who care for ill children will be faced with managing the clinical syndrome of shock. Many common childhood illnesses, such as trauma, gastroenteritis, infection, and accidental drug ingestions, can lead to shock. Ultimately, without timely medical intervention, the child in shock will follow a common pathway to multiorgan system failure and death. Early recognition and appropriate therapy are vital if we hope to reduce the morbidity and mortality associated with this serious syndrome.

The first section of this chapter is devoted to early recognition, which demands a clear understanding of the definition, pathophysiology, and clinical manifestations of shock. Next, the etiologic types of shock, including hypovolemic, cardiogenic, distributive (septic), dissociative, and obstructive shock, are discussed.

Finally, the appropriate therapy and prevention of shock are discussed. Exciting developments are occurring within this area. Advances in molecular biology and immunology have led to a better understanding of the biochemical mediators involved in initiating and maintaining shock. Treatment modalities that antagonize or prevent the inflammatory cascade that lead to shock, multiorgan system failure, and death are now being studied. Combined with aggressive supportive and microbial therapies, these experimental immunotherapies may further reduce the morbidity and mortality associated with shock.

DETERMINANTS OF CARDIAC OUTPUT AND THE DEFINITION OF SHOCK

Circulation in the Child

Normal circulatory function is maintained by the complex interplay between the central pump (heart) and blood flow at the regional level, all done with the sole purpose of delivering oxygen and nutrients to the tissues.

The cardiac output is calculated by multiplying the stroke volume (volume of blood ejected by the left ventricle) by the heart rate (ejection cycles per minute). The stroke volume depends on the filling volume of the ventricle (preload), myocardial contractility (Starling's curve), and the resistance against which the heart is pumping blood into the systemic vasculature (afterload).

Heart rate is controlled through the vagus nerve and endogenous catecholamine release. Hypertension and severe hypoxemia can lead to increases in vagal tone and bradycardia. In times of “flight, fright, or stress,” endogenous catecholamine release increases adrenergic tone with an increased heart rate. In the infant, who has relatively less myocardial contractility, in times of metabolic need, increase in cardiac output depends on an increasing heart rate rather than on an increase in stroke volume. This is also the reason bradycardia is poorly tolerated in this age group; cardiac output falls quickly because there is little ability to compensate with an increase in stroke volume. Conversely, faster heart rates are best tolerated in infants, in whom ventricular filling time is less critical in contributing to stroke volume and, ultimately, to cardiac output.

Definition of Shock

An understanding of normal physiology allows us to define *shock* as an acute syndrome that occurs because of cardiovascular dysfunction and the inability of the circulatory system to provide adequate oxygen and nutrients to meet the metabolic demands of vital organs. Note that this definition recognizes that shock can and does exist without hypotension.

PATHOPHYSIOLOGY

Microcirculatory Dysfunction

The clinical manifestations of shock can be directly related to the abnormalities seen on the tissue, cellular, and biochemical levels. Microcirculatory dysfunction, common to all etiologic types of shock, is characterized by maldistribution of capillary blood flow. Local sympathetic, vasoconstrictor nerve activity and circulatory vasoactive substances (Table 3.1) cause smooth muscle contraction in the precapillary sphincters and arterioles. As shock continues, mechanical obstruction of capillary beds occurs by blockage with cellular debris. Normally, polymorphonuclear leukocytes undergo extensive deformation as they squeeze through the capillaries. Hydrostatic pressure within the capillary makes this possible. However, hydrostatic pressures fall by 30 to 40% during shock states. As a result, capillary beds are blocked and endothelial damage occurs. Subsequent complement activation causes still further aggregation of platelets and granulocytes. During septic shock, exposure to endotoxin directly damages vascular endothelium. Once damaged, endothelial cells can generate procoagulant activity, which may explain the mechanism by which fibrin is deposited in the microcirculation. Superoxide radicals, lysosomal metabolites, and cytokines produced by macrophages and neutrophils for bacterial killing can result in further tissue damage, especially to endothelium, adding to the vicious cycle of damage to the microcirculation.

Mediator	Stimulus	Major Source	Major Actions
Norepinephrine	Hypovolemia	Sympathetic nervous system	Vasoconstriction
	Head trauma	Adrenal medullae	β_1, β_2 stimulation
Epinephrine	Hypovolemia	Adrenal medullae	Vasoconstriction
	Hypercapnea		α_1, β_1 stimulation
Angiotensin II	Hypovolemia	Kidneys, brain, blood	Vasoconstriction
Arachidonic acid metabolites			
Leukotrienes	Tumor necrosis factor	Macrophages	Capillary permeability
	Bacterial antigens		Vasoconstriction, release of myocardial hydrolases
Thromboxane A_2	Hypoxia	Platelets	Vasoconstriction, platelet aggregation
Prostaglandins $F_{2\alpha}$	Hypoxia	Platelets	Vasoconstriction
		Vascular smooth muscle	
Prostaglandins I_2	Hypoxia	Healthy vascular endothelium	Vasodilator counterbalance thromboxane A_2
Myocardial depressant factor	Ischemia	Pancreas	Direct negative inotropic effects
	Tissue damage		
Opiates (β-endorphins)	Hypoxia	Pituitary	Decreased myocardial contractility
			Decreased sympathetic tone hypotension

Table 3.1. Endogenous (Host-Derived) Vasoactive Mediators in Shock

Tissue Ischemia

Tissue ischemia is also basic to all forms of shock. The consequences of poor tissue perfusion sustain the cascade of events that occur during shock. When there is a lack of oxygen, energy production at the cellular level becomes inefficient, producing only 2 moles of adenosine triphosphate (ATP) per mole of glucose, instead of the normal 38 moles of ATP produced by aerobic metabolism.

In addition, anaerobic metabolism depletes glycogen stores with an accumulation of lactate and associated acidosis. The decreasing energy and acidosis lead to an efflux of potassium and an influx of sodium and calcium with an obligate influx of water into the cell. Cellular swelling and further cellular dysfunction occur, which is seen clinically as edema.

Release of Biochemical Mediators

Biochemical mediators play an important role in the development and continuation of all types of shock. These vasoactive and inflammatory mediators are endogenous (host-derived) products primarily from cells of nervous system and hematopoietic origin. Although in septic shock these mediators are stimulated after exposure to microbial products (e.g., endotoxin) and play a primary role in initiating shock, in hypovolemic and cardiogenic shock, they are released secondarily in response to ischemic cellular injury as just described.

VASOACTIVE MEDIATORS

The vasoactive mediators exert their effect primarily by induction of severe vasoconstriction and vasospasm, induction of platelet aggregation and thrombus formation, increased capillary permeability, and redistribution of blood flow away from vital tissues (Table 3.1).

INFLAMMATORY MEDIATORS

In the past, it was believed that invasive microbial agents were directly responsible for the cellular damage and microcirculatory dysfunction seen in septic shock. However, within the last decade, it has become clear that endogenous inflammation mediators are the real culprits in the pathogenesis of septic shock and that lethal tissue injury occurs when

production of these mediators escalates out of control.

Septic shock starts with exposure to microbial products. Perhaps the most potent stimulator of the inflammatory cascade is the outer cell membrane of Gram-negative bacteria, a lipopolysaccharide coat (LPS), also called endotoxin. Once in the bloodstream, the LPS attaches to a plasma protein called LPS-binding protein, or LBP. This complex (LPS-LBP) binds to the CD14 receptor on the surface of the monocyte/macrophage, which leads to stimulation of tumor necrosis factor (TNF) and interleukin-1 (IL-1) and ultimately to the entire cascade of inflammatory mediators.

As a group, these newly described protein mediators are called cytokines (Table 3.2). TNF plays the pivotal role in triggering the production of not only other cytokines but also other inflammatory mediators. TNF is one known endogenous factor that is capable of inducing a broad range of vasoactive and inflammatory mediators. Because of this, treatment of shock with anti-TNF antibodies has been attempted with mixed results (see “ Initial Therapy ,” following). TNF in physiologic amounts has beneficial effects in tissues and promotes wound healing, tissue remodeling, and neovascularization. In pathogenic amounts, TNF and other inflammatory mediators (Table 3.2) cause severe septic shock in animal models and act primarily by inducing fever, increasing the white blood cell counts, inducing production of procoagulant and cell adhesion molecules by endothelial cells, causing aggregates of hematopoietic cells, and increasing vascular permeability.

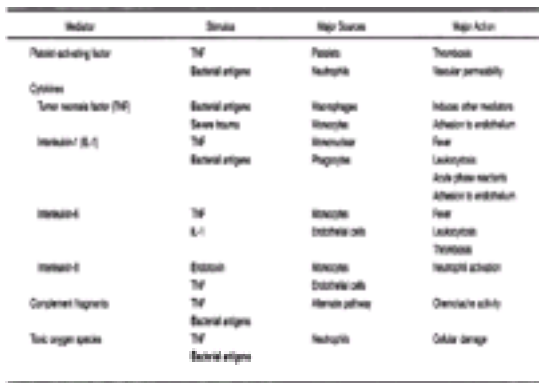


Table 3.2. Endogenous (Host-Derived) Inflammatory Mediators in Shock

NITRIC OXIDE

In 1990, several groups noted that circulatory shock appears to be intimately associated with an increased production of nitric oxide by a variety of different cells. After exposure to TNF-a and IL-1 a variety of cells, including, macrophages, vascular endothelium, vascular smooth muscle, hepatocytes, and cardiac myocytes, are induced to increase nitric oxide production. In pathologic amounts, nitric oxide causes vasodilation, vascular hyporesponsiveness, and hypotension. Although nitric oxide has beneficial effects that make it important in host defense against infection, in excessive amounts, nitric oxide leads to hypotension, worsening the shock state. Studies are ongoing to determine whether drugs that inhibit nitric oxide production may be useful in controlling shock.

COMPLEMENT ACTIVATION

The complement system is activated by circulating bacteria and bacterial products. The low-molecular-weight peptides that are released as a result induce both vasoactive and inflammatory effects. Vasoactive effects are seen with C3 and C5 fragments, which promote the release of histamine and other vasoactive mediators, which produces increased permeability and vasodilation. Complement fragments also stimulate an inflammatory response by promoting the activation and aggregation of platelets and granulocytes.

MYOCARDIAL DEPRESSANT FACTOR

Myocardial depression occurs in all types of shock. Recent investigation suggests that myocardial depression may occur as a result of mediators that act directly on myocardial tissue. Discovered in 1970, a small peptide called myocardial depressant factor is produced when the pancreas is ischemic and hypoperfused. Myocardial depressant factor has been shown to have negative inotropic effects in isolated heart muscle and causes constriction of the splanchnic vascular bed. In 1985, Parker, using isolated heart muscle preparation, found altered inotropic responsiveness within 1 to 2 hours of endotoxin treatment, demonstrating in vitro what is apparent in patients with shock syndrome.

Intrinsic myocardial depression either primarily as in septic shock or secondarily as in hypovolemic or cardiogenic shock adds to other circulatory derangements that have been discussed. Understanding this intrinsic or direct cardiac depression is important in designing treatment strategies.

CLINICAL MANIFESTATIONS

Early or Compensated Shock

Regardless of the etiology, shock begins when there is absolute or functional hypovolemia. Absolute hypovolemia exists in cases of severe emesis and diarrhea, trauma with blood loss, peritonitis, and “third-spacing” of fluids or increased capillary permeability, as in sepsis. Functional hypovolemia exists when vascular capacity increases, as in septic shock,

spinal cord injury, anaphylaxis, and barbiturate overdose.

The signs of early shock include tachycardia, mild tachypnea, slightly delayed capillary refill (more than 2 to 3 seconds), orthostatic changes in blood pressure or pulse, and mild irritability. These earliest symptoms result from an effort to compensate for shock and increase cardiac output and maintain perfusion of vital organs (brain, heart, kidneys). Unexplained tachycardia without other signs may be one of the earliest signs of shock. Tachycardia occurs to compensate for a diminished stroke volume (Fig. 3.1). Delayed capillary refill occurs as increases in sympathetic tone by endogenous catecholamines cause peripheral vasoconstriction. In some cases of early septic shock, the skin may be warm and dry without a decrease in capillary refill, reflecting cutaneous vasodilation in a state of increased cardiac output and increased venous capacitance—so-called warm distributive shock. As systemic vascular resistance falls, cardiac output must increase to maintain normal arterial pressure. Often, in this form of distributive (septic) shock, the pulse will be bounding and the pulse pressure widened.

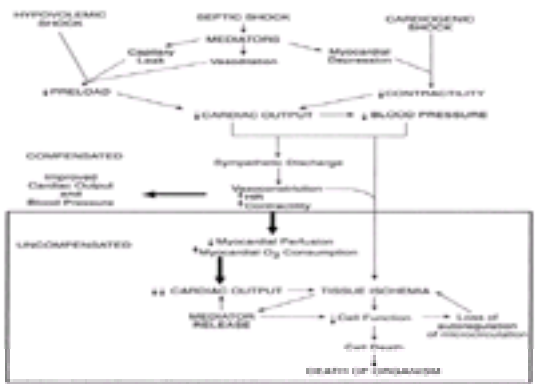


FIGURE 3.1. Sequence of pathophysiologic events in clinical shock states. (Reprinted with permission from Witte MK, Hill JH, Blumer JL: Shock in the pediatric patient. Adv Pediatr 34:139–174, 1987.)

Late or Uncompensated Shock

As shock continues, these early compensatory mechanisms are not enough to meet the metabolic demands of the tissue, and uncompensated shock follows (Fig. 3.1). In uncompensated shock, the effects of cellular ischemia with the associated release of vasoactive and inflammatory mediators begin to effect the microcirculation, and the child shows signs of brain, kidney, and cardiovascular compromise.

Tachycardia and tachypnea continue. Tachypnea becomes more severe because an increasing acidosis elicits a compensatory increase in the minute ventilation, resulting in a fall in Pa CO_2 and a compensatory respiratory alkalosis. The skin may be mottled or pale and extremities cool as vasoconstriction and diminished blood flow to the skin occur. Capillary refill becomes markedly delayed (more than 4 seconds). Hypotension is noted. Decreased cardiac output and vasoconstriction cause renal perfusion, and oliguria is noted. The gastrointestinal tract is also underperfused and may become ischemic. Under these conditions, decreased motility, distension, release of vasoactive and inflammatory mediators, and fluid accumulation may occur. In patients with septic shock, fever (greater than 38.3°C rectally) or hypothermia (less than 35.6°C rectally) may occur. As perfusion of the brain occurs, irritability progresses to agitation, confusion, hallucinations, alternating periods of agitation and stupor, and finally coma. The multiorgan dysfunction secondary to ongoing shock and exaggerated inflammatory responses has been termed systemic inflammatory response syndrome (SIRS).

The effects of a dysfunctional microcirculation, tissue ischemia, and release of vasoactive and inflammatory mediators obviously affect all tissues, including the pulmonary tissues and vasculature. Damage to the capillary endothelium in the lung allows fluid to fill the interstitium of the intra-alveolar septum. If the shock syndrome progresses, fluid accumulation will eventually lead to fluid leakage into the alveolar spaces, which prevents adequate gas exchange. As the damage to the lungs continues, the child demonstrates dyspnea, tachypnea, cyanosis refractory to oxygen therapy, decreased lung compliance, and diffuse alveolar infiltrates. These signs, when grouped together, have been termed the adult respiratory distress syndrome (ARDS), which, despite the name, has been seen in infants as early as 2 weeks after birth. For example, Carcillo found that 11 (32%) of 34 children with septic shock develop ARDS.

TYPES OF SHOCK

Hypovolemia

Hypovolemia (decreased circulating blood volume) is the most common cause of shock in children. The most common cause of hypovolemic shock occurs from water losses associated with diarrhea and vomiting (see Chapter 16). The World Health Organization estimates that in developing countries, 5 to 10 million diarrhea-associated infant deaths occur annually, primarily because of hypovolemic shock, secondary to the vomiting and diarrhea that occurs with a variety of infectious agents, such as rotaviruses. Other causes of hypovolemic shock include blood losses (trauma, gastrointestinal, intracranial hemorrhage), plasma losses (burns, hypoproteinemia, peritonitis), and water losses (glycosuric diuresis, sunstroke).

Distributive Shock

Distributive shock occurs primarily because of vasodilation and pooling of blood in the peripheral vasculature. Causes

include anaphylaxis, central nervous system (CNS) or spinal injuries, drug ingestions, and most commonly in children, sepsis. The primary derangements in septic shock results from exposure to microbial components (e.g., endotoxin, teichoic acid, viral proteins), which trigger the cascade of inflammatory and vascular mediators described. The bacterial etiology of septic shock (and meningitis) is listed in Table 3.3. *Haemophilus influenzae* type b used to be the most common bacteria associated with septic shock in infants and children but has been virtually eliminated through vaccination.

Organism
Streptococcus pneumoniae
Neisseria meningitidis
Group B *Streptococcus*
Listeria monocytogenes
Haemophilus influenzae type b
Gram-negative bacilli[b]
Staphylococcus aureus
Pseudomonas aeruginosa
Salmonella enteritidis

[a]Listed in order of most to least frequently isolated from blood or cerebrospinal fluid. Based on data from Schuchat A, et al. *N Engl J Med* 1997;337:970–976.
[b]Includes *Escherichia coli* and *Enterobacter* species.

Table 3.3. Bacterial Etiology of Invasive Disease in Infants and Children[a]

In 1990, Jacobs et al. retrospectively analyzed more than 2000 admissions to their pediatric intensive care unit over 3 years. They found that 27% (564 cases) of admissions met criteria of sepsis syndrome, which included 1) clinical manifestations of sepsis, 2) fever (greater than 38.3°C) or hypothermia (less than 35.6°C), 3) tachycardia, 4) tachypnea, and 5) signs of inadequate tissue perfusion (e.g., decreased capillary refill, hypoxemia, oliguria, acidosis, altered mental status). Inotropic support was required to maintain an adequate blood pressure in 268 of 564 patients that met this criteria. However, an etiology for septic shock was found in only 143 (25%) of cases. Meningitis was found in half of these children (71 of 143), and mortality was 10% (14 of 143). In some cases of septic shock, superantigenic bacterial toxins are responsible. Toxins such as staphylococcal toxic shock syndrome toxin-1 (TSS-1) and streptococcal exotoxin-A (SPEA) are suspected to cause profound hypotension, leading to inflammation and multiorgan failure. Both of these superantigens have been shown to stimulate monocyte/macrophage production of TNF-a, IL-1b, and IL-6.

Cardiogenic Shock

Cardiogenic shock can usually be distinguished from other forms of shock because of associated signs of congestive heart failure, including rales auscultated throughout the lungs, a gallop cardiac rhythm, enlarged liver, and jugular venous distension.

Regardless of the etiology, cardiogenic shock leads to decreased cardiac output, in most cases as a result of a decrease in myocardial contractility. As we have seen, direct myocardial damage occurs in all types of shock as a late manifestation. Other common etiologies of cardiogenic shock in children include viral myocarditis, arrhythmia, drug ingestions, postoperative complications of cardiac surgery, metabolic derangements (hypoglycemia), and congenital heart disease. Occasionally, congenital heart disease is diagnosed in an infant, usually within the first 3 months of life, when the infant presents to the emergency department in congestive heart failure and shock. These infants invariably have congenital heart abnormalities, such as truncus arteriosus, transposition of the great vessels, or left hypoplastic heart syndrome, that depend on flow through the ductus arteriosum to maintain adequate oxygen delivery. The closure of the ductus precipitates congestive heart failure and eventually cardiogenic shock.

The management of obstructive shock, which is caused by mechanical obstructions to ventricular outflow and occurs with pericardial tamponade or tension pneumothorax (see Chapter 100 and Section VII), and dissociative shock, which occurs secondary to carbon monoxide poisoning (see Chapter 86) or methemoglobinemia, are discussed elsewhere.

TREATMENT

Initial Therapy

To determine proper therapy, we should recall the definition and pathophysiology of shock. *Shock* is defined as an acute syndrome that occurs because of cardiovascular dysfunction and the inability of the circulatory system to provide adequate oxygen and nutrients to meet the metabolic demands of vital organs. Therefore, initial therapy in the emergency department can be applied universally, regardless of the etiology of shock, and is directed to reverse or halt further tissue injury. To underscore this, in 1989, Carcillo compared hemodynamic and oxygen use in children with either cardiogenic shock or septic shock. These data suggested that there was little difference physiologically, and therefore, initial treatment should be similar.

As noted previously, the basic defects are in shock hypovolemia, microcirculatory dysfunction, tissue ischemia, and cardiovascular dysfunction. Each of these defects becomes more severe the longer the shock state exists, so prompt and aggressive treatment is mandatory. The etiology of shock can be determined as therapy begins.

With this pathophysiology in mind, the first steps of therapy are to 1) establish an adequate airway; 2) determine whether breathing is adequate; 3) provide oxygen at 100% Fi O_2; 4) establish vascular access and obtain laboratory samples; and 5) provide aggressive fluid resuscitation, beginning with 20 mL/kg of crystalloid 0.9% sodium chloride or Ringer's lactate

given intravenously as rapidly as possible (minutes). Reassessment after each therapeutic maneuver is vital (Fig. 3.2). After the initial therapy, the following questions should be addressed: 1) Is tracheal intubation needed? 2) Should additional intravenous therapy be given? If so, blood, crystalloid, or colloid? 3) Are positive inotropic drugs needed? If so, which one initially? 4) What is the urine output? 5) What other drugs are needed (antibiotics)? 6) Should arrangements for admission to the intensive care unit be initiated?

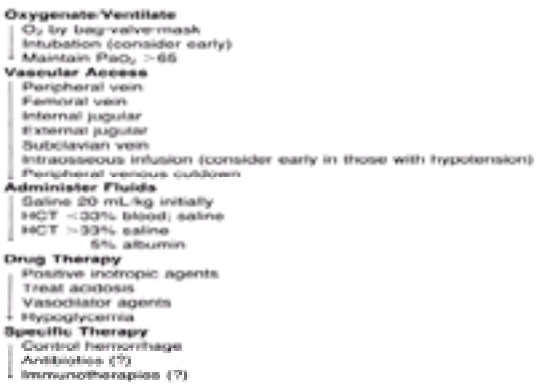


FIGURE 3.2. Management of shock—overview.

Simultaneously, a history should be obtained while the initial treatment is started. If possible, another physician can obtain a history from the caretakers. Questions pertaining to trauma, fever, diarrhea, vomiting, medication, allergies, heart disease, and seizures should be addressed.

Decision and Monitoring in the Emergency Department

Oxygenation

Oxygen delivery to the tissues remains our primary focus in children with shock. While the airway and ventilatory effort is assessed, 100% oxygen should be provided via a bag-valve-mask apparatus. Assisted bag-valve-mask may be indicated. If there is any question that the airway is obstructed or that ventilatory effort is inadequate, the insertion of an artificial airway is indicated. We suggest the orotracheal intubation route initially. Measurements of the Pa O_2 by an arterial blood sample or pulse oximetry should be done throughout the decision-making process. The goal is to maintain the arterial oxygen tension above 65 mm Hg; therefore, 100% oxygen should be continued until that is achieved.

Vascular Access

Vascular access is vital in treatment. If possible, a large-bore intravenous catheter should be inserted in a peripheral vein. However, in many instances, the peripheral extremities will be cool because of vasoconstriction and no vein is found. Central vein venous placement by the Seldinger technique (see Section VII, Procedure 3.2) is the next step. Use of the femoral vein is preferred in infants and younger children. In older children and adolescents, cannulation of the internal jugular, external jugular, and subclavian veins can also be considered. If there is any delay in accomplishing prompt placement of a central venous catheter, an intraosseous line should be placed.

In children less than 5 years old, needle placement into the marrow space of the medial portion of the proximal tibia, angulated away from the growth plate and 1 to 2 inches below the tibial tuberosity, is indicated (see Section VII, Procedure 3.8). In older children (more than 5 years old) and adults, needle placement 1 to 2 inches above the medial maleolus is indicated. If central line placement is delayed, intraosseous fluid replacement is an excellent interim step in infants and children who require fluid resuscitation. In 1991, Velasco et al. found this technique to be useful in resuscitation in a hemorrhagic shock model. Others have commented on the safety and ease of placement. Intravenous fluid, blood products, bicarbonate, and catecholamines are among the therapies successfully given using this technique and are comparable in effect to the central or peripheral intravenous routes.

The last resort in establishing venous access would be for a venous cutdown (see Section VII, Procedure 3.1). Simultaneous with attempts at vascular access, venous blood samples can be obtained for complete blood count, platelets, prothrombin and partial thromboplastin times, electrolytes, blood urea nitrogen, creatinine, glucose, and blood culture (if indicated). An arterial blood sample should also be obtained.

Fluid Administration

After venous access is established, 20 mL/kg of 0.9% normal saline or Ringer's lactate is infused as rapidly as possible. Then reassessment should occur. The decision to give additional intravenous fluids can be based on arterial pressures, heart rate, and oxygenation. If blood pressure is normal, additional fluids will depend on urine output, heart rate, capillary refill, and mental status. If the child remains hypotensive after the initial fluid challenge, an additional 20 mL/kg should be infused and, if possible, titrated against central venous or right atrial pressures because they correlate better with intravascular volume than does systemic arterial pressure. If there is a delay in transfer to the intensive care area, central venous pressure monitoring can be accomplished in the emergency department. Once in the intensive care setting, placement of a balloon-tipped, flow-directed pulmonary artery catheter may be needed to assess more accurately the filling pressures of the heart.

If there is delay in transfer to the intensive care area, fluid management may be directed by right heart filling pressures

(central venous pressures). This can be accomplished by using a modification of Weil's “4 and 2 rule” (Fig. 3.3). Central venous pressure (CVP) is observed for 10 minutes. If that pressure is less than 6 mm Hg, 4 mL/kg is infused over 10 minutes, discontinuing the infusion if CVP rises at any time more than 4 mm Hg. If, after the infusion, CVP has risen by less than 4 mm Hg but more than 2 mm Hg, the patient should be observed for 10 minutes. If CVP remains above 2 mm Hg of the starting value, the patient should be monitored without administering additional fluid. If CVP declines to within 2 mm Hg of the initial value, again 4 mL/kg should be infused over 10 minutes. We repeat these maneuvers until the systemic arterial pressure reaches a normal value, the patient manifests other signs of restored circulation integrity, or the “4 and 2 rule” is violated. If the initial CVP lies between 6 and 10 mm Hg, we administer 2 mL/kg over the 10-minute period and look for the same CVP changes. If the initial CVP exceeds 10 mm Hg, we infuse 1 mL/kg fluid over 10 minutes and again observe the CVP change.

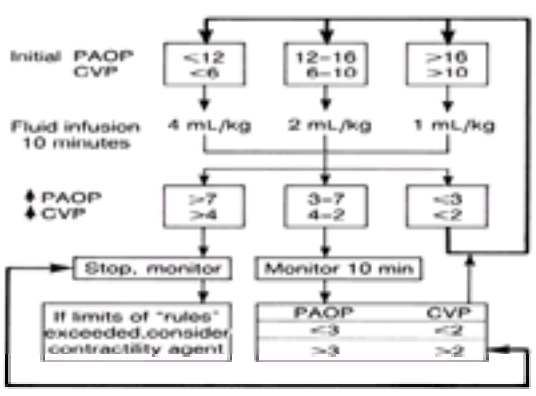


FIGURE 3.3. “7-3, 4-2 Rules” of Weil. *CVP,* central venous pressure; *PAOP,* pulmonary artery occluded pressure. (Adapted from Weil MH. J Am Med Assoc 1969;207:337–340.)

Two points should be emphasized. First, we examine the change in CVP measurement in response to the fluid infusion, rather than look for an absolute value. Second, because the veins are very distensible vessels and act as a reservoir for fluid, a threefold increase in the volume of fluid contained in the venous system may be necessary before we see changes in CVP. When the pulmonary artery occluded pressure (PAOP) is used, 7 and 3 mm Hg should be substituted for 4 and 2 mm Hg CVP measurements (Fig. 3.3).

If this type of management is not possible in the emergency department or if transfer to the intensive care is imminent, changes in vital signs and perfusion can be used to guide fluid management. Despite the fact that we do not base fluid management on an absolute amount and are guided by the child's clinical condition, we can give some ballpark figures for initial fluid resuscitation. In 1991, Carcillo et al. studied all children with septic shock who presented to the emergency department over a 6-year period and who had a pulmonary artery catheter inserted within 6 hours of presentation. Interestingly, fluid resuscitation in excess of 40 mL/kg in the first hour improved survival and was not associated with an increased risk of either cardiogenic pulmonary edema or ARDS, compared with children who received smaller amounts of fluid. Therefore, in severe shock, fluid resuscitation, if indicated, up to 60 mL/kg or approximately 50% of the circulation blood volume may be given within the initial phase of therapy. The important point to remember is that, in most cases of shock, not enough fluid is given and the child remains in relative hypovolemic shock. Monitoring such signs as heart rate, capillary refill, mental status, and urine output (at least 1 mL/kg/hour) is helpful in determining the amount of fluids needed in the initial phases of resuscitation. Monitoring of cardiac output is essential for children with cerebral damage of those in cardiogenic shock, in which case the need for adequate fluid resuscitation must be balanced with concerns over cerebral edema or cardiac disease, respectively.

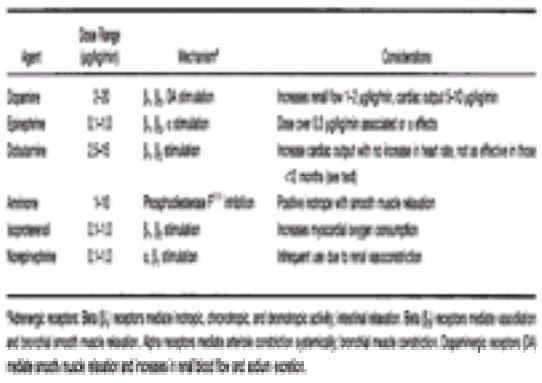
Choice of Fluids and Blood Products

The initial choice of fluid should be 0.9% saline or Ringer's lactate given as described earlier, beginning with 20 mL/kg over minutes. Packed red blood cells should be given at 10 mL/kg over 1 to 2 hours to maintain a hematocrit of 33%. Children with cyanotic heart disease, or neonates, may require higher hematocrit percentages to ensure adequate tissue delivery.

If the hematocrit is over 30% or if blood is not available, 5% albumin in 0.9% sodium chloride can be used in combination with crystalloid fluids. Initially, albumin can be given in 10 mL/kg doses. When colloids (albumin) are used, appropriate intravascular monitoring should be considered when possible to guard against circulatory overload.

Improving Myocardial Function

Catecholamines (adrenergic agents) are the drugs of choice for improving myocardial contractility in patients with shock because of their very short half-life (2 to 3 minutes) and potency (Table 3.4). A brief review of adrenergic receptor physiology is important if we are to have a rational approach to their use.



Agent	Dose Range (µg/kg/min)	Mechanism[a]	Considerations
Dopamine	2–20	β_1, β_2, DA stimulation	Increases renal flow 1–2 µg/kg/min; cardiac output 5–10 µg/kg/min
Epinephrine	0.1–1.0	β_1, β_2, α stimulation	Dose over 0.3 µg/kg/min associated w α effects
Dobutamine	2.5–10	β_1, β_2 stimulation	Increase cardiac output with no increase in heart rate, not as effective in those <12 months (see text)
Amrinone	1–10	Phosphodiesterase F^{111} inhibition	Positive inotrope with smooth muscle relaxation
Isoproterenol	0.1–1.0	β_1, β_2 stimulation	Increases myocardial oxygen consumption
Norepinephrine	0.1–1.0	α, β_1 stimulation	Infrequent use due to renal vasoconstriction

[a]Adrenergic receptors: Beta (β_1) receptors mediate inotropic, chronotropic, and dromotropic activity; intestinal relaxation. Beta (β_2) receptors mediate vasodilation and bronchial smooth muscle relaxation. Alpha receptors mediate arteriole constriction systemically; bronchial muscle constriction. Dopaminergic receptors (DA) mediate smooth muscle relaxation and increases in renal blood flow and sodium excretion.

Table 3.4. Positive Inotropic Agents

There are at least three broad populations of adrenergic receptors, termed alpha (a), beta (b), and dopaminergic (DA) receptors. Although all have been subdivided further, in general, b $_1$-receptors mediate inotropic (contractility), chronotropic (rate), and dromotropic (increased conduction velocity) activity. b $_2$-Receptors mediate vasodilation and bronchial smooth muscle relaxation. a-Receptors mediate arteriole constriction systemically and bronchial muscle constriction. Dopaminergic receptors, termed DA_1 and DA_2 mediate smooth muscle relaxation and increase renal blood flow and sodium excretion.

Catecholamines may stimulate some adrenergic receptors more strongly than others, providing some rationale for selection. In general, the mechanism of action for most positive inotropic agents seems to be an increased concentration of or sensitivity to intracellular calcium during systole. If the desired effect is not achieved with one agent, combinations of several agents together may be necessary. It is important to note that there may be decreased responsiveness to adrenergic stimulation in patients with congenital heart disease, after heart transplantation, or in those with bronchopulmonary dysplasia.

Currently, dopamine is the first choice to improve cardiac function and improve splanchnic and renal circulation if the patient is relatively stable but remains hypotensive after initial fluid resuscitation. At low dosages (2 µg/kg/minute), dopamine increases renal blood flow up to 50% and sodium excretion up to 100%. Cardiac output is increased with dosages of 5 to 10 µg/kg/minute. Improvement in perfusion as measured by increased urine output, blood pressure, and warming of the extremities can be seen early. More accurate measurements of cardiac index, arterial mixed venous difference, or left ventricular stroke work can be used to titrate the dosage in the intensive care unit.

In some cases of profound septic shock and hypotension, epinephrine should be considered initially. A low dosage (less than 0.2 µg/kg/minute) of epinephrine stimulates both b $_1$ cardiac effects and b $_2$ peripheral vascular effects, which results in an increase in skeletal muscle blood flow and a decrease in diastolic blood pressure. Dosages higher than 0.3 µg/kg/minute are associated with increased a-adrenergic effects and increases in blood pressure. If the child is unresponsive to dopamine, epinephrine may be useful in maintaining blood pressure and cardiac output.

When epinephrine is being used, one may need to consider vasodilator therapy. Nutrient flow may be improved, left ventricular stroke work enhanced, and myocardial oxygen consumption decreased by lowering impedance to left ventricular ejections. A short-acting vasodilating drug such as sodium nitroprusside, beginning with 0.1 µg/kg/minute, is preferred. The infusion can be increased until evidence of decreased peripheral vascular resistance exists or until the generally accepted safe dosage of between 8 and 10 µg/kg/minute is reached. Toxicity results from the accumulation of cyanide, which should be monitored. The use of this therapy in the emergency department is rarely needed, although in cases in which transfer to the intensive care unit is delayed, it may be indicated.

Dobutamine should be considered initially in patients with cardiogenic shock because it is a very selective stimulant of b 1 receptors. In patients with cardiogenic shock, it tends to increase cardiac output without increasing the heart rate. Starting dosages should be 2 to 5 µg/kg/minute. However, Perkins found that infants with cardiogenic or septic shock who were less than 12 months of age derived little benefit from dobutamine, demonstrating insignificant increases in cardiac index and stroke index. If dobutamine fails, epinephrine should be used.

Finally, amrinone represents a class of inotropic agents distinct from the catecholamines. Although the mechanism of action is not fully understood, data favor inhibition of phosphodiesterase F ^{111}. A direct relaxant effect on vascular and vasodilation that results in smooth muscle causes afterload, and preload reduction contributes to the improved hemodynamic state. Amrinone facilitates atrioventricular conduction, relaxes smooth muscle, and dilates coronary arteries. At this time, there is seldom an indication to start this agent in the emergency department.

Acid-Base Abnormalities

It is clear that unless perfusion and ventilation are adequate, infusion of sodium bicarbonate rarely maintains arterial pH. Therefore, bicarbonate should be considered as a temporary and immediate therapy to acutely alter arterial pH so that myocardial performance will be optimized. Ultimately, improved blood flow will result in a decrease in acid products of anaerobic metabolism and only then will pH concentration remain normal. We calculate the dose of sodium bicarbonate according to the following formula: bicarbonate administered in milliequivalents (mEq) equals the body weight (kg) times base excess (BE) times 0.6. In the presence of acute hypercapnea, one-half of the calculated dose is administered and pH remeasured. If correction is not achieved, the remainder is infused. The suggested rate of administration should not exceed 2 mEq/minute.

Steroids

Bone, in a prospective, randomized, double-blind, placebo-controlled study of adults, concluded that high-dose corticosteroids provided no benefit in the treatment of septic shock. Patients in this study who exhibited serum creatinine levels above 2 mg/dL and received steroids experienced a significantly higher mortality rate than the placebo group. Furthermore, patients who received methylprednisolone died as a result of secondary infection more often than their placebo-treated counterparts. Currently, there is no good evidence to support the use of steroids in shock syndrome.

Disseminated Intravascular Coagulation

As we have seen, microcirculatory dysfunction, tissue ischemia, and cardiovascular dysfunction, regardless of etiology, leads to shock and consumption of coagulation factors and platelets. This consumption is characterized by thrombocytopenia, an increase in fibrin split products, a decrease in fibrinogen, and abnormally prolonged prothrombin time and partial thromboplastin times (see Chapter 36). Management includes platelet transfusions (if indicated for bleeding or platelet counts less than 50,000/mm^3) with infusion 0.2 unit/kg. Fresh frozen plasma, 10 mL/kg intravenously, may be given for prolonged prothrombin and partial thromboplastin times.

Antibiotics

Antibiotics are given presumptively in most cases of severe shock when the etiology is unclear. Antibiotics are chosen based on age and suspected bacterial pathogens. If the child is less than 4 weeks of age and there is no suspicion or evidence of meningitis, ampicillin and gentamicin is one effective combination. From 4 to 12 weeks of age, ampicillin and cefotaxime are favored by many pediatric infectious diseases specialists when meningitis is ruled out.

In some cases, the infant or child is too unstable to tolerate a lumbar puncture. Presumptive antibiotics should not be delayed in these cases. After resuscitation, the lumbar puncture can safely be performed and the cerebrospinal fluid (CSF) profile will still indicate a bacterial etiology. Latex agglutination tests can be obtained on urine plus CSF if the CSF culture and blood culture are negative. Presumptive antibiotics in children with shock and meningitis or when meningitis cannot be ruled out would include Vancomycin and cefotaxime for the possibility of a strain of *Streptococcus pneumoniae* that is resistant to penicillin or cefalosporins.

Immunotherapies

Over the last several years, the identification of the mediators of septic shock has lead to a better understanding of how these mediators cause and contribute to the pathophysiology of shock. Coincidentally, the use of monoclonal antibody techniques has allowed for the production of large quantities of antibody that are free from human infection and are of known isotype and epitope specificity. This new knowledge has led to the development of a number of investigational therapies aimed at different components of the inflammatory cascade. These therapies can be divided into three broad categories (Table 3.5): agents aimed at blocking the effects of circulating microbial products, agents that block cytokines, and finally agents that reduce or prevent nitric oxide production.

Category	Product and Mechanism of Action
Antiendotoxin	
E5	Murine IgM monoclonal antibody; binds to lipid A of endotoxin
HA-1A	Human IgM monoclonal antibody; binds to lipid A of endotoxin
Soluble CD14	Blocks binding of endotoxin to macrophage
Anti-CD14 antibody	Blocks binding of endotoxin to macrophage
Anticytokine	
Anti-TNF antibody	Blocks inflammatory cascade
TNF soluble receptor	Binds TNF, inhibits inflammatory cascade
IL-1 receptor antagonist	Blocks IL-1 binding
Nitric Oxide (NO) Inhibitors	
L-NMA	L-arginine analog; inhibits NO synthase and decreases NO production
L-NAA	Same as above

*The safety and efficacy studies of these agents in humans and animals have been mixed. In some cases, use of the above products have resulted in an increased mortality as compared to controls. Further research is needed.
TNF, tumor necrosis factor; IL-1, interleukin-1.

Table 3.5. Experimental Therapies[a] for the Treatment of Septis Shock (Selected)

Although initially there was much optimism that these products would reduce mortality by blunting the detrimental effects of inflammatory mediators, subsequent animal and human studies have revealed mixed results. In fact, some of these studies have been associated with an increased mortality.

The use of these agents needs further study in both animals and humans to first establish safety then efficacy. If proven effective, we can almost certainly expect to use these agents in the emergency department. Therefore, emergency medicine physicians will need to understand the indications and risks of these agents as new data are made available for review.

Laboratory Indications of Improvement in Shock

The initial phase of treatment for shock are directed to improve oxygen delivery to the tissues by ensuring adequate ventilation, correcting hypovolemia, and improving cardiac function.

The success of the initial resuscitation is usually reflected in signs of improved perfusion (skin, kidneys, brain, heart rate)

and indications of normal CVP if invasive monitoring is required. However, other laboratory parameters may be useful, if monitored sequentially, in determining if the shock syndrome is persisting despite early clinical improvement. Sequential measurements of serum lactate levels, and expired CO_2 gas (end-tidal CO_2) may indicate that continued aggressive resuscitation is needed despite signs of improvement.

In 1983, Vincent et al. found that a 5% reduction in serum lactate in the first hour was a good prognostic sign for patients presenting in circulatory shock. Lactate should fall over time as perfusion and oxygen delivery to the tissue improves. High serum lactate levels (approximately 4 mm/L) or serum lactate levels that continue to rise despite therapy are indications of severe shock. Aggressive therapeutic maneuvers to reverse the shock should be redoubled in this setting.

In addition, end-tidal CO_2 is a noninvasive laboratory value that may indicate continued hypovolemia. In hypovolemia and poor perfusion of the peripheral tissues and pulmonary tissues, CO_2 is not excreted in the lungs. Subsequently, there is a reduction of expired CO_2 gas, which is measured as a decreased end-tidal CO_2. As perfusion improves, end-tidal CO_2 increases. Investigators hope to correlate whether sequential end-tidal CO_2 measurements can be used to titrate the amount of intravenous fluid needed during resuscitation of the patient in shock. These measurements are noninvasive and may be a valuable adjunct in accessing improvements in perfusion in children with severe hypovolemia in the emergency department.

Suggested Readings

Bone RC. A critical evaluation of new agents for the treatment of sepsis. JAMA 1991;266:1686–1691.

Bone RC, Fisher CJ, Clemmer TP, et al. A controlled clinical trial of high-dose methylprednisolone in the treatment of severe sepsis and septic shock. N Engl J Med 1987;317:653–658.

Carcillo JA, Pollack MM, Ruttimann UE, Fields AI. Sequential physiologic interactions in pediatric cardiogenic and septic shock. Crit Care Med 1989;17:12–27.

Carcillo JA, Davis AL, Zaritsky A. Role of early fluid resuscitation in pediatric septic shock. JAMA 1991;266:1242–1245.

Crone RK. Acute circulatory failure in children. Pediatr Clin North Am 1980;27(3):525–538.

Falk JL, Rackow EC, Leavy J, et al. Delayed lactate clearance in patients surviving circulatory shock. Acute Care 1985;11:212–215.

Falk JL, Rackow EC, Weil MH. End-tidal carbon dioxide concentration during cardiopulmonary resuscitation. N Engl J Med 1988;318:607.

Glauser MP. The inflammatory cytokines: new developments in the pathophysiology and treatment of septic shock. Drugs 1996;52(suppl):9–17.

Greenman RL, Schein RMH, Martin MA, et al. A controlled clinical trial of E5 murine monoclonal IgM antibody to endotoxin in the treatment of gram-negative sepsis. JAMA 1991;226:1097–1102.

Jacobs RF, Tabor DR. The immunology of sepsis and meningitis—cytokine biology. Scand J Infect Dis 1990;73:7–15.

Jacobs RF, Sowell MK, Moss M, Fiser DH. Septic shock in children: bacterial etiologies and temporal relationships. Pediatr Infect Dis J 1990;9: 196–200.

Lefer AM. Role of a myocardial depressant factor in the pathogenesis of circulatory shock. Fed Proc 1970;29:1836–1847.

Mink RB, Pollack MM. Effect of blood transfusion on oxygen consumption in pediatric septic shock. Crit Care Med 1990;18:1087–1091.

Natanson C, Hoffman WD, Suffredini AF, et al. Selected treatment strategies for septic shock based on proposed mechanisms of pathogenesis. Ann Internal Med 1994;120:771–783.

Orlowski JP, Porembka DT, Gallagher JM, et al. Comparison study of intraosseous, central intravenous, and peripheral intravenous infusions of emergency drugs. Am J Dis Child 1990;144:112–117.

Parker JL, Adams HR. Development of myocardial dysfunction in endotoxin shock. Am J Physiol 1985;248:H818–H826.

Perkin RM, Levin DL. Shock in the pediatric patient. Part I. Pediatr 1982;101:163–169.

Perkin RM, Levin DL. Shock in the pediatric patient. Part II. J Pediatr 1982;101:319–332.

Perkin RM, Levin DL, Reedy J, et al. Hemodynamic effects of dobutamine in children with shock. Crit Care Med 1981;9:171.

Royall JA, Levin DL. Adult respiratory distress syndrome in pediatric patients. 1. Clinical aspects, pathophysiology, pathology, and mechanisms of lung injury. J Pediatr 1988;112:169–179.

Saez-Lloren X, Ramilo O, Mustafa MM, et al. Molecular pathophysiology of bacterial meningitis: current concepts and therapeutic implications. J Pediatr 1990;116:671–684.

Schuchat A, Robinson K, Wenger JD, et al. Bacterial meningitis in the United States in 1995. N Engl J Med 1997;337:970–976.

Spivey WH. Intraosseous infusions. J Pediatr 1987;111:639–643.

Sprung CL, Schultz DR, Marcial E, et al. Complement activation in septic shock patients. Crit Care Med 1986;14:525–528.

Tracey KJ. Tumor necrosis factor (cachectin) in the biology of septic shock syndrome. Circ Shock 1991;35(2):123–128.

Velasco AL, Delgado-Paredes C, Templeton J, et al. Intraosseous infusion of fluids in the initial management of hypovolemic shock in young subjects. J Pediatr Surg 1991;26:4–8.

Vincent JL, DuFaye P, Berre J, et al. Serial lactate determinations during circulatory shock. Crit Care Med 1983;11:449–451.

Virgilio RW, Rice CL, Smith DE, et al. Crystalloid vs. colloid resuscitation: is one better? Surgery 1979;85:129–139.

Weisel RD, Vito L, Dennis RC, et al. Myocardial depression during sepsis. Am J Surg 1977;133:512–521.

Witte MK, Hill JH, Blumer JL. Shock in the pediatric patient. Adv Pediatr 1987;34:139–174.

Wolfe JE, Rabinowitz LG. Streptococcal toxic shock like syndrome. Arch Dermatol 1995;131:73–77.

Wolfe T, Dasta JF. Use of nitric oxide synthase inhibitors as a novel treatment for septic shock. Ann Pharmacother 1995;29:36–46.

Word BM, Klein JO. Current therapy of bacterial sepsis and meningitis in infants and children: a poll of directors of programs in pediatric infectious diseases. Pediatr Infect Dis J 1988;7:267–270.

Zaritsky A, Chernow B. Use of catecholamines in pediatrics. J Pediatr 1984;105:341–350.

Ziegler EJ, Fisher CJ, Sprung CL, et al. Treatment of gram-negative bacteremia and septic shock with HA-1A human monoclonal antibody against endotoxin. N Engl J Med 1991;324:429–436.

CHAPTER 4

Sedation and Analgesia

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Pain is an unpleasant sensory and emotional experience usually defined in terms of tissue damage, which is signaled by some form of visible or audible behavior. Acute pain serves a useful function and is necessary for survival; it alerts us to avoid certain painful stimuli, and it warns us that some body tissues may be damaged. It often helps the emergency physician make a correct diagnosis. For example, a fracture might be diagnosed by localizing pain with palpation of an extremity. Appendicitis might be diagnosed by finding tenderness in the right lower abdominal quadrant. Chronic pain, however, serves no useful function; thus, a child's pain must be relieved as soon as possible.

One of the most common reasons for seeking care in an emergency department (ED) is to relieve pain and suffering. Injuries and painful medical conditions are common among children, and ED physicians are expected to manage this pain appropriately. Unfortunately, pain control is not often addressed satisfactorily. In 1990, Selbst and Clark found that in the ED setting, 60% of adult patients received adequate analgesia for painful conditions, but only 28% of pediatric patients received adequate analgesia for similar painful conditions. Emergency physicians and pediatricians were equally unlikely to give analgesics to children. There have been great advances in recognition, assessment, and management of pain in children since then. The care of infants and children who undergo painful procedures in the ED has improved considerably. However, two recent studies showed that children still often fail to receive analgesia for painful conditions. Inadequate dosing of medications for children upon discharge from the ED is a significant problem.

Many theories try to explain why pediatric pain is not successfully managed in the ED. Physicians expect babies to cry, so this nonspecific response to pain often is tolerated instead of controlled. Moreover, because young children and infants cannot describe or localize their pain, it is often ignored or presumed not to exist. Adult patients who clearly indicate that they are in pain generally get a direct response from a physician, whereas a young child who is crying or whimpering may not. In addition, some ED physicians avoid giving adequate analgesics to children because they fear it will lead to drug addiction. This is fear unfounded, however, because narcotic addiction is extremely rare when medications are used appropriately to manage acute pain. Hypotension and respiratory depression are other feared consequences of narcotic use with children, and although these fears may be legitimate, respiratory depression and hypotension are unlikely to occur if proper protocols are adhered to. These unlikely occurrences should be manageable in the ED, and they should not inhibit the attempt to control pain. Furthermore, it is likely that ED physicians often are forced to concentrate on other aspects of resuscitation and care before managing pain. Plans for pain control, therefore, may be forgotten because of other priorities. Also, in some cases, pain is ignored because it is inconvenient to wait for analgesics to take effect. Thus, some physicians may convince a young child that a painful procedure or repositioning of an extremity will hurt only for a minute. Brute force (instead of medication) is then used, and more pain is inflicted on an already uncomfortable child. Table 4.1 summarizes some reasons for inadequate pain control with children.

Inability of young children to talk
Misconception that infants cannot feel pain
Misconception that children will not remember pain
Misconception that children will get addicted to narcotics
Fear of respiratory depression and hypotension
Unfamiliarity with analgesics and dosages
Other conditions taking priority

Table 4.1. Possible Reasons for Inadequate Pain Control in the Emergency Department

Pain control may not be properly addressed with children because many physicians lack knowledge about the management of such pain. Some textbooks and residency programs do not emphasize the management of acute pain. Until recently, pain control has been poorly studied in children. Children are often unable to cooperate or provide good verbal descriptions of pain, so it may be difficult to assess or quantitate pain or to measure success in treating it. Great progress has been made lately in the assessment of pain in children. Self-report pain scales are the best indicators of pain and are the gold standard for assessing pain in children. Pain measured with visual analog scales and "thermometers" have been validated for children. A visual analog scale is used with children older than 4 years of age. This scale has some limitations but seems useful regardless of the child's education, background, or gender. It consists of a 10-cm horizontal line with endpoints marked as "no pain" (0 score) to "worst possible pain" (10 score). Children can indicate the level of their pain by marking the line.

There are modified visual analog scales, such as color scales for use with younger children. Children score their pain on these scales by interpreting the pain as red, black, or purple. Also, children as young as 3 or 4 years old can use a face scale (e.g., the Oucher Scale), which has six pictures of a young boy's face, each displaying a different expression of pain, placed along a thermometer scale of increasing pain levels from 0 to 100.

Behavioral pain scales and physiologic pain scales (measuring heart rate, blood pressure, respiration, palm sweating, and transcutaneous oxygen) provide information about general distress levels in infants and young children. The Children's Hospital of Eastern Ontario Pain Scale (CHEOPS) is a behavioral scale that measures the child's face and verbal behaviors, as well as crying and movement. The scale has been found to be valid for assessing pain in younger children and infants who cannot use self-report scales. Research in the area of pain management for children has increased dramatically in recent years, and this should allow us improve our abilities to control pediatric pain in the future.

Emergency physicians must understand that pain is an individual experience and that many factors contribute to the degree of pain that a child feels for any given condition. For example, the age and cognitive development of a child are important. Children of all ages can experience pain. It is believed that even newborns feel pain, and many react to a painful stimulus (e.g., circumcision) with wriggling motions and crying. Younger children have lower pain thresholds than older children, and they do not tolerate painful procedures as well. For example, they often exaggerate the size and power of needles. Older children may be better able to understand the need for a painful procedure; they are usually less anxious and better able to tolerate this pain. However, an older child may have a better understanding about the significance of an injury or an illness that could cause depression, anxiety, and more pain. Similarly, parental response (anxiety or reassuring calm) may affect a child's perception of pain. Other psychological factors such as the child's emotional state or personality traits may create more or less anxiety, and this also can alter the degree of pain.

Some children seem to have a hypersensitivity to pain, whereas others tolerate it well. The child's gender or cultural background is likely to influence the pain experience, but this is not well studied in children. A study of Israeli children on a kibbutz found they did not scream or cry when they received inoculations. This was in contrast to 26% of urban Israeli children who expressed their pain quite emphatically with inoculations. It was thought that the kibbutz philosophy that "doctors are friends" accounted for the different responses of the two groups.

The context of the situation also plays a role, because children who are punched in play may not complain of pain. Yet, the same impact could elicit a significant response if it was meant as an attack or as punishment. A child's past experience with painful stimuli also is meaningful. A recent study showed inadequate analgesia for one painful procedure may diminish the effect of adequate analgesia in subsequent procedures. Of course, the painful stimulus itself is important, and a stimulus that causes a great deal of tissue damage may hurt more than one that causes minor injury. Figure 4.1 summarizes the components of the pain experience.

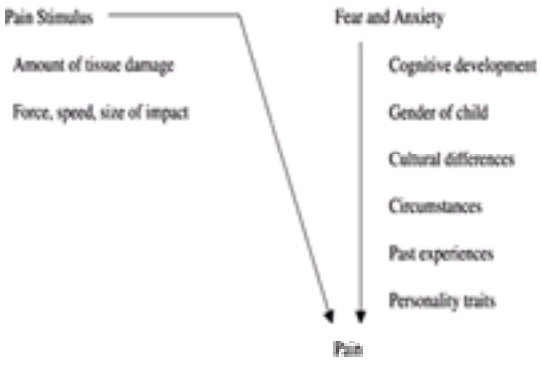

FIGURE 4.1. Components of pain. (Adapted from Schecter NL. Pain and pain control in children. Curr Prob Pediatr 1985;15:4–67.)

PAIN MANAGEMENT

Realistically, pain in the pediatric patient who presents to the ED is never eliminated completely. Efforts must be made, however, to relieve this pain as much as possible. Several means are available for managing pain in children, and various medications and techniques can be used, depending on the type of pain that requires treatment.

Minor Pain

Conditions such as headache, myalgia, chest pain, pharyngitis, otitis media, arthralgia, sunburn, strains, and sprains often produce minor pain in children. For treatment of this mild pain, aspirin, acetaminophen, ibuprofen (nonnarcotics), and codeine (narcotic) are excellent oral analgesic medications. *Aspirin* is one of the oldest analgesic medications, but its use has recently declined. It has the advantage of being inexpensive, and it has anti-inflammatory effects. Sustained high dosages are needed, however, for the anti-inflammatory effect. Thus, in most instances, aspirin is not much better than non–anti-inflammatory drugs. Aspirin may be given every 4 hours at a dosage of 10 to 15 mg/kg per dose. Higher dosages increase only the anti-inflammatory effect—not the analgesic effect. Buffered aspirin may be tolerated better and absorbed faster, but there is no evidence to show that it acts more rapidly or lasts longer than the nonbuffered variety. Likewise, enteric-coated aspirin is better tolerated but has variable absorption.

There are some definite disadvantages to using aspirin for pain management. It has many side effects, particularly gastrointestinal irritation in some patients, which can lead to nausea and vomiting. Aspirin also inhibits platelet function, which can lead to bleeding with overdosage, and it may cause reversible liver toxicity and central nervous system (CNS) problems (tinnitus, dizziness). Moreover, it may induce bronchospasm in asthmatic patients. Reye syndrome has been associated with the use of aspirin for varicella and flulike illness, but not for control of pain from trauma.

Acetaminophen acts centrally on nonopioid receptors in the brain to inhibit prostaglandin synthetase. Acetaminophen is more expensive than aspirin but is probably a better choice for pain associated with minor trauma or otitis media because it is tolerated better and it comes in liquid form, making it easier to give to young children. In some studies, acetaminophen has been shown to be a less potent analgesic than aspirin, but most claim it is equipotent. One study showed that 1000 mg of acetaminophen equals to 60 mg of codeine for postpartum pain. In addition, acetaminophen does not cause bleeding and is less likely than aspirin to cause bronchospasm in asthmatics. It is dosed at 15 mg/kg per dose every 4 hours and takes effect in 20 to 40 minutes, with a peak effect in 2 hours. High dosages are usually well tolerated, but therapy should not exceed 4 to 6 g/day. Acetaminophen has no anti-inflammatory effects, and therapeutic doses rarely are associated with side effects; overdose, however, can cause liver toxicity.

Nonsteroidal anti-inflammatory drugs (NSAIDs) (e.g., ibuprofen) are excellent choices for treating minor pain such as headache, dysmenorrhea, or musculoskeletal injuries. NSAIDs are thought to be more potent drugs than aspirin, and they have a longer half-life. Like aspirin, NSAIDs act on the peripheral nervous system to block formation of prostaglandins. The dosage is 8 to 10 mg/kg given every 6 hours. The recommended dosage for ibuprofen in older children is 400 mg every 4 to 6 hours for mild to moderate pain. Ibuprofen is available in liquid form, making it suitable for use in very young children. Ibuprofen is nonaddictive and does not cause respiratory or cardiac depression. They may cause gastrointestinal bleeding, but this risk is small. Like aspirin, these agents also cause renal and hepatic dysfunction and should be used with caution in children with renal or hepatic disease. Unlike aspirin, however, their effect on platelets (acetylation) is reversible. They can cause sodium retention, and they have cross-reactivity with aspirin, so they should not be used with patients who have aspirin sensitivity.

Finally, *codeine* is a narcotic analgesic that can be given orally (even to young children) to control minor pain. Codeine usually is given orally because it maintains two-thirds of its effectiveness in oral form compared with parenteral use. It is thought to be more potent than aspirin but less potent than meperidine; thus, it is valuable for moderate pain (dental abscess, severe otitis media, or stomatitis) and has a low addiction potential. The dosage is 0.8 to 1.5 mg/kg per dose every 4 to 6 hours. Codeine can be changed into a liquid form for use in young children, and it can be combined with acetaminophen or aspirin to produce an even greater analgesic effect than when either is used alone. Codeine can cause respiratory depression, but this occurs very rarely. It has no renal or hepatic toxicity and does not alter platelet function, but it can cause the same gastrointestinal side effects (e.g., nausea and vomiting) as noted for other narcotics.

The benefits of using codeine and the other analgesics described here for mild pain far outweigh the few side effects involved with their use in young children. Table 4.2 summarizes the advantages and disadvantages of these analgesics for treating minor pain in children.

Analgesic	Advantages	Disadvantages
Acetaminophen	Well tolerated, safe	Liver toxicity if overdosed
Aspirin	Inexpensive	Gastrointestinal irritation
Ibuprofen	Long duration of action	Gastrointestinal irritation
Codeine	Potent opioid analgesia	Nausea, constipation

Table 4.2. Analgesics for Mild Pain

Moderate and Severe Pain

Opioid medications are extremely important for treating patients in the ED with moderate to severe pain (burns, fractures, sickle cell vaso-occlusive crises). Most opioids can cause important adverse effects (respiratory depression and hypotension), but these often are dose related and may be reversed with naloxone, making opioids safe in the ED setting. Because of pharmacokinetic differences in young infants that may predispose them to respiratory depression, these drugs should be used with caution in infants less than 6 months of age who are not ventilated. Such infants should receive one-fourth to one-third of the initial calculated dosage recommended for older children, and they should be closely monitored. Opioid analgesics can be given by various routes. In general, the intramuscular route should be avoided because the injection itself is painful, it causes delayed drug absorption, and the dosage of drug given cannot be titrated. The intravenous route is more advantageous because titration is possible, although some pain is involved in starting the intravenous line. Some physicians choose to deliver the opioids with a patient-controlled analgesia (PCA) device that allows the patient to self-administer the drug at a safe dosage as it is needed. This allows patients to have some control over their own pain while relieving the ED nurse of time needed to administer the drugs.

For severe pain from a significant burn, sickle cell crisis, fracture, or other injury, *morphine* is an excellent choice. The usual dosage of morphine is 0.1 to 0.15 mg/kg per dose given intravenously over a few minutes. The maximum dose is 10 mg for opioid naive subjects, which is repeated every 3 or 4 hours. The higher dosage, and a dosing interval of every 1 to 2 hours, is suggested for those who get narcotics often (e.g., those with sickle cell disease or cancer) because they may have some tolerance to the drug. If needed, a subsequent dose is reduced to 0.05 mg/kg if the patient is moderately sedated. Morphine can also be given as a continuous infusion at 0.07 to 0.10 mg/kg per hour, and it can be titrated, if needed (increase 25% every 3 hours). When given intravenously, its effect is almost immediate, with the peak effect occurring in 20 minutes. Morphine can cause pooling of blood by decreasing peripheral vascular resistance, which may result in hypotension. However, this is a concern only in the patient with a severe injury who may be hypovolemic. Certainly, the fluid status of an injured child requires careful attention from the ED staff, but morphine should not be withheld after intravenous fluids have corrected volume depletion. If the child is awake, alert, and screaming in pain, morphine can be given safely as long as the patient is monitored carefully.

Meperidine is another opioid agent that can be used to treat moderate to severe pain. It can be given intravenously or intramuscularly at a dose of 0.5 to 1 mg/kg, given every 2 to 3 hours. The maximum recommended dose for opioid-naive patients is 125 mg. Meperidine is more effective than morphine when given orally, and it reaches peak effectiveness more quickly than morphine when given by the intramuscular route (about 90 minutes). Otherwise, it has no significant advantages over morphine, but there can be problems with it use. For example, it may cause nervousness, tremors, disorientation, and even seizures when used intravenously. Morphine therefore may be a better opioid for severe pain unless a patient indicates that meperidine works best.

Fentanyl is a synthetic opioid that should be given slowly at a dose of 2 to 3 µg/kg intravenously over 3 to 5 minutes. It has a rapid onset of action (a few minutes) and a short duration of action (30 to 40 minutes), which makes it useful in the ED. It is an excellent agent for severe pain from fractures. Fentanyl also has several other advantages. It is a relatively safe drug and rarely causes hypotension, making it an excellent choice for injured children in severe pain. A study of adult patients found that the drug caused hypotension in only 0.4% of patients, and alcohol intoxication may have been responsible for some cases of hypotension. It is thought that hypotension is unusual because histamine is not released when the drug is given. Respiratory depression can occur within minutes of fentanyl administration, but this is reported in only 0.7% of adult patients, some of whom were intoxicated with alcohol. There is a greater risk of respiratory depression when coadministered with other sedatives and in infants younger than 3 months of age. Apnea occurs even less often, and this may be related to the rate of infusion of fentanyl rather than the dosage. Although these adverse effects are serious, they can be reversed with naloxone and reduced if the dosage guidelines are followed and the drug is given slowly. Individualized dosing titrated to effect may reduce these side effects. Also, equipment and personnel who can manage an obstructed airway should be nearby when fentanyl is used.

An uncommon event caused by fentanyl is neuromuscular blockade, with severe thoracic and abdominal muscle rigidity. However, this is expected only when high dosages (greater than 15 µg/kg) are used and especially when fentanyl is administered rapidly. Most often, this side effect is reversible with naloxone, but succinylcholine may be required. Despite the problems already noted, fentanyl remains a valuable analgesic that should be used liberally in the ED when a child has severe pain.

Ketorolac tromethamine is a parenteral nonsteroidal anti-inflammatory drug that has been used to treat moderate to severe pain. This drug is relatively new, and few data are available regarding its use with children. However, studies with adult patients show intramuscular ketorolac is comparable to narcotic agents and oral ibuprofen for treatment of

musculoskeletal pain, headaches, sickle cell crises, and orthopedic injuries with less sedation and fewer side effects. It does not cause respiratory depression or nausea or vomiting. Intravenous ketorolac has been found to be as effective as morphine for postoperative pain in children with fewer side effects. It may have an opioid sparing effect, but this is not well proven. The drug is not yet approved by the Food and Drug Administration for intravenous use, although studies have demonstrated this route to be safe and efficacious in adults and children. Many pediatric centers use ketorolac with a loading dose of 1 mg/kg intravenously to a maximum of 60 mg followed by 0.5 mg/kg IV (maximum 30 mg) every 6 hours.

Other Agents

Another semisynthetic agent used occasionally with children in the ED is hydromorphone (Dilaudid), which can substitute for morphine and codeine. This is often given orally, rectally, or parenterally, and the analgesic effects last 4 to 5 hours. Also, hydrocodone (Hycodan) is an oral analgesic that is more potent than codeine with less associated nausea and vomiting. Likewise, oxycodone is 10 times more potent than codeine and about equal to the strength of morphine. Like codeine, it retains about 50% of its efficacy when given orally. It is often combined with aspirin (Percodan) or acetaminophen (Percocet, Tylox). The agents described next are not used in the ED for children. Propoxyphene (Darvon) has been shown to be inferior to aspirin, and in some studies, it was no better than placebo. It has significant addiction potential. Pentazocine (Talwin) is a narcotic agonist–antagonist that is better than placebo but not much better than aspirin. It is expensive and causes dizziness and hallucinations, as well as sedation and gastrointestinal discomfort. Methadone is an excellent analgesic with actions similar to those of morphine. It is well absorbed from the gastrointestinal tract and reaches peak concentration 4 hours after ingestion. This is a useful drug for treating chronic pain because it has few gastrointestinal side effects, less sedation, and less euphoria. Heroin is not available in the United States for pain control.

Table 4.3 summarizes the advantages and disadvantages of several analgesics used to treat moderate to severe pain.

Analgesic	Advantages	Disadvantages
Morphine	Rapic onset, potent analgesia	Respiratory depression, hypotension
Meperidine	Potent analgesia	Respiratory depression, seizure
Fentanyl	Potent analgesia, less hypctensicn	Respiratory depression, spnea
Ketorolac Tromethamine	Nonnarcotic	Not well studied in children

Table 4.3. Analgesics for Severe Pain

PROCEDURAL PAIN/CONSCIOUS SEDATION

Children often undergo painful procedures in the ED, including fracture reduction, laceration repair, incision and drainage, burn debridement, gynecologic examination, and foreign body removal. The pain involved with these procedures can be alleviated largely with the appropriate use of analgesics. Because a child's anxiety may further exacerbate the feeling of pain, it is often useful to give the child a combination of an anxiolytic with an analgesic medication before a painful procedure is performed.

The narcotics mentioned earlier commonly are given intravenously just before and during a painful procedure. For example, Fentanyl has been used for safe repair of complicated facial lacerations that might otherwise have required general anesthesia. It should be noted that fentanyl causes an unusual tendency for children to reach up and scratch their faces. If fentanyl is used for repair of lacerations, restraints may be needed to prevent a child from contaminating a sterile field. Fentanyl can also be given in the form of a lollipop. Oral transmucosal fentanyl citrate is a raspberry flavored fentanyl impregnated lozenge that allows oral transmucosal absorption, resulting in a narcotized child within 15 to 30 minutes. This is advantageous because it does not require a needle and is nonthreatening to young children. A disadvantage of delivering the drug in this form is that vomiting is a common side effect, occurring in up to 45% of children who received the agent before laceration repair. Oxygen desaturation is also common.

Some emergency physicians combine opioids with phenothiazines. Morphine and hydroxyzine, combined in an intramuscular injection, are used by some physicians. For years, the combination of meperidine 2 mg/kg, promethazine 1 mg/kg, and chlorpromazine 1 mg/kg has been used for sedation and analgesia (“lytic cocktail,” or “DPT”). Because this combination can produce deep and prolonged sedation with respiratory depression, its use is undesirable in the ED. It also requires a painful intramuscular injection and can result in dystonic reactions. Therefore, the use of the lytic cocktail, or DPT, is not recommended.

Sedatives

Sedatives should be used for painless procedures or in combination with analgesics when pain is anticipated.

Diazepam has been used as a sedative in doses of 0.1 to 0.2 mg/kg intravenously or orally. It can also be administered rectally, but the intramuscular route is discouraged because of pain and poor absorption. One retrospective study of

children undergoing orthopedic procedures found the combination of fentanyl and diazepam to be safe.

Midazolam has become more popular and may be given at a dose of 0.05 to 0.2 mg/kg slowly, intravenously every 15 minutes as needed. Intranasal midazolam (dose of 0.2 to 0.4 mg/kg) and oral midazolam (dose of 0.2 to 0.5 mg/kg) have also been used with narcotic analgesics before painful procedures are performed. The intranasal route is irritating to the nasal mucosa and may not be tolerated well by the pediatric patient. Midazolam may be preferable over diazepam because it has a shorter onset of action (about 5 minutes) and a more rapid recovery (about 1 to 4 hours). The combination of a benzodiazepine and a narcotic analgesic produces amnesia, sedation, and muscle relaxation, which is preferred for orthopedic procedures and laceration repair. The likelihood of respiratory depression increases when a sedative is added to a narcotic, so proper precautions must be taken to protect the airway.

Propofol is an intravenous anesthetic used for sedation during short procedures, gynecologic examinations, and cardioversion. Propofol does not have analgesic properties of its own. This agent has been used rarely in the ED, but one report found it to be effective when used in combination with fentanyl for fracture reduction, abscess drainage, and chest tube placement in 20 adult patients. There were few side effects in that study, and propofol is advantageous because it has a rapid onset of action, a rapid recovery phase, and amnesic properties. It is a potent sedative, associated with significant respiratory depression, so great caution should be exercised with its use.

Pentobarbital is another sedative that is useful for pediatric procedures. Pentobarbital has no analgesic properties of its own and may increase pain perception, so it is not ideal for painful procedures. It has greatest use for nonpainful procedures such as sedation of children for imaging studies. Narcotics can also be combined with pentobarbital 4 to 5 mg/kg, but this may result in prolonged sedation, making the combination less convenient to use in a busy ED. Sedation occurs rapidly after intravenous administration, and the effect dissipates in 15 to 20 minutes.

Chloral hydrate is a pure sedative hypnotic. It has no analgesic properties but may also be useful for painless procedures such as imaging studies. Unfortunately, the sedation effect is variable and sometimes long lasting, making it less desirable for use in the ED. The peak effect may take 40 to 60 minutes or longer to accomplish. It is usually given orally in a dose of 20 to 75 mg/kg. Vomiting and paradoxical hyperactivity may occur with its use, but it is a relatively safe drug. Cardiac arrhythmia is a rare event associated with large doses, and respiratory depression may be a risk if the patient has obstructive sleep apnea.

In one study of 95 children who received 25 to 30 mg of chloral hydrate orally, there were no adverse cardiovascular, respiratory, or gastrointestinal effects. All sedated children recovered promptly in less than 60 minutes.

Nitrous Oxide

Nitrous oxide gas has been used for analgesia in an emergency setting since 1969. A few studies on children have shown that it is an effective agent for reducing pain when performing painful procedures, and it can be delivered painlessly.

Nitrous oxide has a short duration of action and a sedative, dissociative effect on the patient. The patient may complain of pain but does not seem to experience it (dissociative effect) and does not remember it later (amnesic effect). Patients who receive this gas feel as if they are floating, drowsy, or euphoric. It takes about 4 minutes to induce these effects with a nitrous oxide–oxygen mixture and about 4 to 5 minutes for them to wear off. When used in a 50/50 mixture with oxygen, nitrous oxide provides analgesia, not anesthesia. That is, the patient remains awake during a procedure and is able to follow instructions. These properties make it useful in the emergency setting.

The nitrous oxide–oxygen mixture has been used on children as young as 16 months of age, but it is certainly more difficult to use in young, uncooperative children. It may be best for children 8 years and older. (Masks for administering the gas can be flavored to help gain acceptance with young patients.) It has been shown to be useful to treat lacerations, burns, or Colle's fractures or to reduce a dislocated radial head injury. It has been used in adults to drain abscesses and pilonidal cysts, for which local anesthesia is difficult to administer. It has been proved useful for managing a child whose penile foreskin got caught in his zipper.

When used properly in a mixture with more than 20% oxygen, nitrous oxide does not produce serious side effects. Nitrous oxide affects the cerebral cortex (not the brainstem), so circulatory and respiratory depression does not occur and there is little relaxation of skeletal muscle. The gas is nonallergenic and not flammable or explosive, but it must be used with oxygen, and it will support combustion if a fire develops. It must be used with a fail-safe system that shuts off the flow of nitrous oxide when oxygen flow stops. The child can hold the mask on his or her face so that it will fall off if the child becomes unresponsive. Nitrous oxide also can be given with a nasal mask, enabling the patient to breathe through the mouth during the procedure. These precautions will avoid possible anoxia, which could be catastrophic. Some patients have experienced vomiting with this mixture of nitrous oxide, but this usually is an inconvenience rather than a danger because the patients maintain their cough and gag reflexes. In one study, 7% of patients fell asleep with the gas, but they all could be easily aroused. Prolonged intermittent dizziness has been reported after nitrous oxide–oxygen administration, but bone marrow suppression, testicular dysfunction, and liver and CNS disorders are seen only with chronic exposure to the gas.

Still there are some limits to the use of nitrous oxide. Personnel demands are great because staff who can manage an obstructed airway in an unconscious patient should be present. Nitrous oxide is not recommended for children who are already sedated, unconscious, or intoxicated or who have head or chest injuries. In addition, the equipment required costs several thousand dollars and must be inspected daily to ensure safety. Also, a scavenger device is needed to eliminate nitrous oxide from the ED so that workers and other patients do not experience the effects of the gas. A simple device can be attached to the machine to accomplish this.

Finally, not all patients report complete pain relief with nitrous oxide. Some studies have shown it to be helpful in 80 to

95% of patients. There was marked pain relief in 29% of patients and partial pain relief in another 61%. Table 4.4 reviews the advantages and disadvantages of nitrous oxide–oxygen analgesia.

Advantages	Disadvantages
Has rapid onset, short duration of action	Fail-safe system required
Causes sedation, dissociation, and amnesia	Expensive equipment
Is useful when local anesthesia is impractical	Scavenger device needed
May be used for young children	Not all patients benefit
Is safe when mixed with oxygen	More personnel required

Table 4.4. Nitrous Oxide–Oxygen Analgesia

Ketamine

Ketamine hydrochloride is another anesthetic agent used when performing wound debridement, foreign body removal, laceration repair, and other painful procedures. It can provide intense analgesia at subanesthetic doses. It causes dissociative amnesia and a trancelike state in which the child can follow commands but cannot respond verbally. The drug can be given intramuscularly (at a dosage of 3 to 4 mg/kg), and it also can be given intravenously (at a dosage of 0.5 to 2 mg/kg). This can be followed by an infusion of 0.01 to 0.2 mg/kg per minute if anesthesia is needed for more than 20 minutes. Ketamine has the advantage of rapid onset of action (1 minute if given intravenously, 5 to 10 minutes if given intramuscularly). It does not cause prolonged sedation and a single intravenous dose has an effect for about 15 minutes. With repeated doses or continuous infusion, ketamine's effect depends on its elimination half-life and is considerably prolonged (1 to 2 hours). Ketamine can also be given orally (10 mg/kg), in which case sedation occurs in 30 to 45 minutes with duration of effect about 2 hours.

Ketamine does have disadvantages, however, in addition to the pain of injection. Rarely, ketamine causes unusual, unpleasant sensations and dreams with subsequent flashbacks, but benzodiazepines may decrease this reaction if given with ketamine. In a recent prospective study, no children experienced nightmares. Ketamine also can cause increased production of saliva in 11%, and vomiting upon awakening in about 6% of those who receive this medication. Excess salivation can be effectively prevented by prior administration of atropine (0.01 mg/kg) or glycopyrrolate (0.005 mg/kg). Ketamine does not generally impair pharyngeal or laryngeal function or protective airway reflexes such as cough or swallowing. Thus, the risk of airway compromise is less with ketamine than with some other agents. However, a rare adverse effect is laryngospasm, so precautions should be taken to manage the airway if this develops. Neonates and those with upper respiratory infection are more likely to have laryngospasm, and they should not receive ketamine. Ketamine also can elevate blood pressure, intracranial pressure, and pulmonary artery pressure, so patients with head trauma, CNS malformations, and cardiovascular or respiratory disease should not receive this drug.

Local Anesthetic Agents

Lidocaine

Lidocaine is an excellent local anesthetic that has been used frequently in the ED for wound repair, foreign body removal, insertion of intravenous infusion lines or lumbar puncture needles, drainage of abscesses, and arterial puncture. Lidocaine has been shown to reduce pain of lumbar puncture in neonates without decreasing the success of the procedure.

Lidocaine usually is administered as a 1% solution (10 mg/mL) at a dosage of 3 to 5 mg/kg. A 0.5% solution is used for infiltration when large volumes are needed or in smaller patients when it is desirable to limit the total number of milligrams per kilograms given. When vasoconstriction is desired for suturing, lidocaine can be used in combination with epinephrine, at a dosage of (lidocaine) 7 mg/kg. Lidocaine should not be combined with epinephrine for use in areas supplied by end arteries such as the digits, penis, or pinna of the ear. Lidocaine is advantageous because it provides excellent local anesthesia and takes effect quickly (within a few minutes). The effect lasts long enough to complete most procedures (about 1½ to 2 hours). It is a safe drug, as few people have a true allergy to it. Serious toxicity, such as seizures and cardiac arrest, can occur, but only when large amounts are injected inadvertently or when the drug is injected directly into a blood vessel.

The major disadvantage of using lidocaine as a local anesthetic is that a painful injection is required for administration. This pain can be reduced, however, if a long, small (27- or 30-gauge) needle is used to produce a “fanning” effect of the anesthetic. When injecting deep into tissue, a larger needle is needed to aspirate blood so that inadvertent injection into a vessel is avoided. Otherwise, the small needle is recommended, and only a small amount of lidocaine should be injected to avoid tissue distortion. Some physicians recommend using a syringe with a thumb ring during infiltration for better control. A recent study shows that warming the lidocaine, by storing the medication and syringes in fluid warmed to 98.6°F, seems to reduce the pain of infiltration. It may also hurt less to inject the lidocaine into the damaged tissue inside the wound instead of in the intact skin. The needle should be pulled out to the tip and the injection given again at 90-degree angles to minimize the number of punctures. Subsequent injections to extend the area of anesthesia should be given through anesthetized tissue when possible. It is helpful to rub the skin near the site of injection first because this reduces pain by stimulating other nerve endings according to the gate theory of pain. The skin should be pulled tightly,

and only the smallest amount of lidocaine needed should be injected to avoid wound distension.

It may also help to use buffered lidocaine, which is more alkaline and perhaps less painful when administered. Some physicians recommend mixing 10 parts lidocaine and 1 part sodium bicarbonate before injection to minimize burning when injecting the anesthetic. A recent study showed that rate of lidocaine injection has a greater impact on perceived pain of administration than does buffering. It is best to inject lidocaine slowly, perhaps over 30 seconds. This may cause less rapid distension of local tissue and activation of fewer nerve endings. In all cases, a few minutes should be allowed for the anesthesia to take effect. Table 4.5 summarizes some hints to reduce the pain of lidocaine infiltration.

1. Do not allow child to see needles involved in preparing lidocaine.
2. Warm and buffer lidocaine with sodium bicarbonate.
3. Use a long, small needle for infiltration.
4. Rub skin around injection site before infiltration.
5. Infiltrate through devitalized tissue or anesthetized areas.
6. Inject slowly, only what is needed.
7. Wait for anesthetic effect.

Table 4.5. Hints to Reduce Pain of Lidocaine Infiltration

Other Injectable Local Anesthetics

Bupivicaine 0.25% is similar to lidocaine but may have a longer duration of action and may help reduce pain for 6 hours after a wound is repaired. Diphenhydramine, 0.5% and 1%, has been used and studied as a local anesthetic in adult patients. However, it seems to cause more pain on infiltration than does lidocaine and may cause tissue necrosis. In addition, a recent study showed that saline with 0.9% benzyl alcohol additive (the bacteriostatic compound in multidose vials of physiologic saline solution) provides anesthesia when injected into the skin of children just before placement of an intravenous line. With injection of 1 mL, anesthesia lasts about 2 minutes and there is minimal pain of injection and no side effects.

Topical Anesthetic Agents

LET

Wound repair can often be done painlessly with the use of a topical anesthetic such as LET. LET is a solution of 4% lidocaine, 0.1% epinephrine, and 0.5% tetracaine. It can be made in gel form with hydroxyethyl cellulose. LET has been used successfully and safely for repair of uncomplicated facial and scalp lacerations in children. The major advantage to using LET for suturing is that the anesthetic can be applied painlessly, without the use of a needle. This should reduce the fear and anxiety involved in wound repair and may help the suturing go more smoothly. Even in the small number of children who have inadequate anesthesia from LET, the application of this topical anesthetic will reduce the pain of subsequent administration of lidocaine by injection. The gel can be applied directly to the wound for 15 to 20 minutes, or the solution can be “painted” on to the wound with a cotton-tipped swab, or a saturated cotton ball can be applied to the wound and held in place manually or with tape. Subsequently, the surrounding skin will be well blanched, indicating adequate local anesthesia. Like lidocaine, LET should not be applied to body parts where vasoconstriction is contraindicated.

Table 4.6 suggests a protocol for the use of LET for wound repair.

1. Apply LET gel directly to the wound.
2. Apply LET for 10–20 minutes.
3. Alternatively apply 3–5 mL of LET solution for 3-cm wound.
4. Use cotton-tipped swab to paint solution onto wound.
5. Drip some LET into wound directly.
6. Tape cotton saturated with LET onto wound.
7. Avoid eyes, digits, and mucous membranes.

Table 4.6. Suggested Protocol for Use of LET for Wound Repair

TAC

In many centers, LET has replaced tetracaine, adrenaline, cocaine compound (TAC) as the preferred topical anesthetic

for wound repair because it is much less costly and has reduced toxicity. TAC is a clear solution of 2% tetracaine 25 mL, 1:1000 epinephrine 50 mL, and cocaine 11.8 g. This is diluted with normal saline to make a volume of 240 mL, which is divided into 3 or 5 mL aliquots for a 3-cm wound. An additional 3 ml of TAC can be used for each additional 3 cm of laceration. TAC has been shown to be as effective as lidocaine for most facial and scalp wounds, but perhaps less effective for wounds on the trunk or extremities. The maximum safe dose of TAC has not been established, but some recommend using no more than 0.09 mL/kg. TAC should be applied directly to the wound just as described for LET. The major problem associated with TAC is the risk of cocaine toxicity. This can lead to seizures and death, so TAC should never be used near mucous membranes where rapid absorption can occur. Death of a child was reported after TAC was absorbed through nasal mucous membranes. The solution can also cause corneal abrasions, so TAC should not be used near the eye.

EMLA Cream

Some physicians have used topical EMLA cream, which is a eutectic mixture of lidocaine and prilocaine with an occlusive dressing to supply local anesthesia. EMLA has been found helpful in relieving pain associated with intravenous catheter placement, lumbar puncture, and venipuncture in children. This cream may also be useful for draining perirectal abscesses or paronychias, draining arthrocentesis, or accessing subcutaneous drug reservoirs. The EMLA cream must be applied directly to the skin for 60 minutes before it is effective, so it is not practical for many situations in the ED. (Lidocaine does not penetrate well through intact skin.) If preparing for intravenous line placement, one should prepare multiple sites in case the first attempt is unsuccessful. Also, young children may become agitated, perspire, and have difficulty keeping the anesthetic on the skin. This could then mandate additional nursing time for a procedure. The cream works best for procedures where the child is unlikely to view the procedure needle and become agitated (e.g., lumbar puncture, bone marrow aspiration).

Despite these disadvantages, EMLA cream has been gaining popularity in the ED setting when time permits. In one recent study, EMLA was compared with TAC in a controlled, blinded fashion during wound repair in children. It was found to be superior to TAC in that patients treated with EMLA required less supplemental anesthesia than those who received TAC.

Other Topical Anesthetics

Smith et al. studied new combinations of anesthetics, including bupivacaine with norepinephrine (bupivanor), Etidocaine with norepinephrine (etidonor), Prilocaine with norepinephrine (mepivanor), and prilocaine with norepinephrine (prilonor). Bupivanor proved as effective as TAC in relieving pain upon infiltration of lidocaine for laceration repair. This offers the advantage of not risking the toxicity of TAC; however, none of the combinations were compared with LET.

Nerve Block

Lidocaine can be used for peripheral nerve block if the physician has appropriate knowledge of anatomy and the nerve supply to the wound is superficial. The skin at the nerve site should be anesthetized, and then lidocaine (5 mg/kg) should be infiltrated more deeply into the nerve in the same manner as would be used for local anesthesia. During this infiltration, the physician should aspirate to ensure that a blood vessel has not been penetrated accidentally.

Some EDs use lidocaine for regional nerve block. The preferred technique for fracture reduction at some institutions is a “mini-Bier” block. With this technique, a double pneumatic tourniquet or two blood pressure cuffs are placed above the elbow and intravenous lines are started in the child's upper extremities. One of these lines is for administration of lidocaine, and one is for other medications, if needed. Diazepam, midazolam, and thiopental should be available in case seizures result from lidocaine infiltration, and the child should be attached to a cardiac monitor during the procedure. The affected limb is elevated for exsanguination as the upper cuff is inflated to occlude the arterial blood supply. Then lidocaine (without epinephrine) is infused into the affected extremity at a dosage of 1.5 mg/kg (maximum 100 mg). This should be given slowly, and tourniquet pressures must be maintained so that the drug does not escape under the tourniquet. Now that the injured extremity has local anesthetic in an isolated compartment, the lower cuff, which should be wrapped around an anesthetized area, can be inflated. The upper cuff can be deflated. This lower cuff can be deflated very slowly after the procedure but not less than 15 minutes after the lidocaine infusion was given. The child should be observed in the ED for at least 1 hour. There are risks to this procedure, including seizures, coma, confusion, and cardiac arrest if the child were to receive accidentally a massive amount of lidocaine. Some physicians, therefore, prefer to perform the mini-Bier block in the operating room, where circumstances can be better controlled.

NONPHARMACOLOGIC METHODS FOR PAIN CONTROL

Hypnosis

Hypnosis has been used to treat pain in children for several years, and it has been successful in children as young as 2 years of age. Some physicians believe that the technique better serves children than adults because children have vivid imaginations and can more easily intertwine fantasy and reality. Also, it is thought that patients who present to the ED are already in a hypersuggestible state and thus may be more receptive to hypnosis.

This technique involves creating visual images for the child in an effort to reduce pain. For example, one could ask the child to think of the funniest movie he or she has ever seen and to imagine the pain getting less intense with each laugh. Or, the child could be asked to imagine the pain as a color that is fading away and is painted over with the child's favorite color.

Hypnosis is of value for chronic pain syndromes such as migraine headaches or long-term illnesses. It has also been used to treat children with sickle cell disease and painful crises. Such children have been taught self-hypnosis to reduce

the frequency and intensity of painful crises, but some required a few outpatient sessions before it became effective.

Hypnosis has also been recommended for the acute management of burns, fractures, and other injuries. In fact, it is thought to be more useful for those entities than when emotional problems are triggers for pain. However, hypnosis may again be less practical than analgesics in this setting in the ED. It may have a role in conjunction with analgesics, but hypnosis cannot completely replace the need for medications in the acute situation. The use of hypnosis for many painful procedures in a busy emergency department has not been well studied.

Distraction and Relaxation Techniques

Distracting a child (e.g., with counting or squeezing a parent's hand) during a painful procedure may help reduce pain. One study showed that asking children to “blow out” air as if they were blowing bubbles was helpful in reducing pain from needle sticks for immunizations. Singing a song or telling a story to a child is also helpful. Children become very involved when listening to their favorite story, and parents can help with this. Paintings on the walls or ceiling of a procedure room may also distract a young child. However, distracting a child may have short-lived success because of a child's limited attention span, and it is less effective than involving the child in a detailed imaginary story. One study showed that allowing adult patients to listen to musical audio tapes during suture repair provided a safe, inexpensive, and effective adjunct for pain management. It is also possible that such tapes or television would provide helpful distractions to older children undergoing painful procedures.

Restraint and Reassurance

Properly restraining a child for a painful procedure can be useful in reducing pain. Although this does not reduce fear or anxiety (in fact, it may heighten them), it allows the physician to perform the task better. This indirectly reduces pain because fewer attempts may be necessary to accomplish the task. One should never attempt a painful procedure on a moving subject! The need for restraint should be explained to the parents (who should not be involved in the actual process). Instead, the child might be wrapped in hospital sheets or papoose boards with Velcro straps, with the parents attempting to calm the child afterward. There are only rare complications with this restraint, including minor bruising, edema, or transient vascular compromise. More important, children need gentle reassurance and carefully chosen words during a procedure to reduce fear and pain. One should keep in mind that young children understand more than they say. Casual teasing, condescension, or talking about the child while excluding him or her should be avoided. Choices should not be offered if they do not exist. The child should be warned honestly about the possible pain, but it is best to allow for the possibility that it may not hurt as much as he or she expects. Table 4.7 summarizes possible means of reducing pain from procedures. One should not allow long delays between the explanation and the actual procedure because the anticipation may be the most distressing part of the procedure. One study showed that an empathic (age-appropriate) explanation of an upcoming needle stick reduced crying among patients compared with a group of children who received impersonal instructions. Fassler showed that allowing a child to read about a procedure and then allowing role-playing and discussion was helpful in reducing pulse rates and other physiologic and behavioral responses to pain. Such explanations and role-playing are a time-consuming processes, and this is not always possible in a busy pediatric ED. The value of empathy and reassurance, however, cannot be emphasized enough. Well-prepared parents can assist their child and reduce anxiety and pain associated with painful procedures.

Agent	Advantages	Disadvantages
Narcotics	Easy administration, may be titrated	Respiratory depression
Nitrous oxide	Painless administration, rapid onset, short duration, dissociative state	Requires nurse personnel, not always effective
Ketamine	Rapid onset, dissociative state	Hypersalivation, bad dreams (rare), laryngospasm (rare)
IST	Painless administration, low toxicity	Delays procedure slightly
TAC	Painless administration	Seizures, death if ingested
Lidocaine	Rapid onset, long duration, safe drug	Painful injection
Hypnosis	No side effects	Time-consuming, not always effective
Restraint	Rarely dangerous, increases success of procedure	May increase child's fear

Table 4.7. Summary of Some Choices Useful for Painful Procedures

During discharge from the ED, it is helpful to reassure the patient, to be honest, and to listen to the child. Perhaps writing a prescription, even for an over-the-counter analgesic, will help give a message that you are concerned about the child's pain. It is important to encourage the family to contact the ED or a private physician if pain persists.

CONSCIOUS SEDATION PROTOCOLS

Because of the potential for respiratory depression with the sedative and analgesic agents discussed previously, it is imperative that EDs develop protocols for their use. Conscious sedation generally refers to the state where the patient has a minimally depressed level of consciousness and can maintain protective airway reflexes and follow commands while responding to physical stimuli. Deep sedation refers to the state where the patient has a more depressed level of consciousness, may not have protective airway reflexes, and is difficult to arouse. Several organizations have prepared guidelines for conscious sedation in children, but the specifics of the ED protocol should be modified at each individual institution. The American Academy of Pediatrics recommends that if children are to have conscious sedation, there should be “emergency equipment” available for children of all ages and sizes. This should include a suction device and a positive-pressure oxygen delivery system capable of delivering at least 90% Fi O_2. Personnel who are trained in pediatric life support should be available, and at least one additional support personnel should be on hand to monitor the patient.

Patients should have continuous monitoring of oxygen saturation and heart rate with (at least) intermittent monitoring of blood pressure and respiration during and after the procedure. Pulse oximetry is essential because of the proven difficulty in recognizing hypoxemia even by experienced personnel.

If a patient is expected to have deep sedation, the requirements just discussed remain. In addition, an intravenous line should be established before sedation. Furthermore, there should be frequent monitoring of the child's blood pressure, cardiac rhythm, and temperature. Additional equipment recommendations include a nearby defibrillator and medications that might be needed for resuscitation. Reversal agents such as naloxone and flumazenil are also useful. Finally, there should be a third person trained in pediatric resuscitation assisting the patient.

Continuous pulse oximetry and monitoring of heart rate should continue until the patient has met discharge criteria. Before discharge, any child who has received conscious sedation should be awake enough to sit and speak without assistance and preferably able to ambulate. Younger children should be able to perform age-appropriate functions. The child should also have adequate hydration status, documentation of stable cardiovascular function, and an adequate airway.

SUMMARY

It is common to encounter children in pain in the pediatric ED. It is often impossible to avoid inflicting pain on some children in this setting. Therefore, the proper management of this pain is essential. This management should be accomplished with various narcotic and nonnarcotic analgesics, as well as with local and topical anesthetics. Other agents, such as nitrous oxide, ketamine, and other techniques such as hypnosis and distraction, play a more limited role in pain management. Gentle restraint and reassurance are of paramount importance.

Suggested Readings

Altman RS, Smith-Coggins R, Ampel LL. Local anesthetics. Ann Emerg Med 1985;14:1209–1217.

Amadio P. Peripherally acting analgesics. Am J Med 1989;77:17–23.

American Academy of Pediatrics. Guidelines for monitoring and management of pediatric patients during and after sedation for diagnostic and therapeutic procedures. Pediatrics 1992;89:1110–1115.

Anderson AB, Colecchi C, Baronoski R, et al. Local anesthesia in pediatric patients: topical TAC vs lidocaine. Ann Emerg Med 1990;19:519–522.

Arts SE, Abu-Saad HH, Champion GD, et al. Age related response to lidocaine-prilocaine (EMLA) emulsion and effect of music distraction on the pain of intravenous cannulation. Pediatrics 1994;93:797–801.

Bartfield JM, Gennis P, Barbera J, et al. Buffered vs. plain lidocaine as a local anesthetic for simple laceration repair. Ann Emerg Med 1990;19:1387–1389.

Bartfield JM, Sokaris SJ, Raccio-Robak N. Local anesthesia for lacerations: pain of infiltration inside vs. outside the wound. Acad Emerg Med 1998;5:100–104.

Berde CB. Toxicity of local anesthetics in infants and children. J Pediatr 1993;122:S14.

Billmire DA, Neale HW, Gregory RO. Use of fentanyl in the outpatient treatment of pediatric facial trauma. J Trauma 1985;25:1080–1085.

Binder LS, Leake LA. Chloral hydrate for emergent pediatric procedural sedation: a new look at an old drug. Am J Emerg Med 1991;9:530–534.

Birmingham PK. Recent advances in acute pain management. Curr Probl Pediatr 1995;25:99–112.

Brogan GX, Giarruso E, Hollander JE, et al. Comparison of plain, warmed, and buffered lidocaine for anesthesia of traumatic wounds. Ann Emerg Med 1995;26:121–125.

Callahan JM. Pharmacologic agents. In: Dieckman R, Fiser D, Selbst SM, eds. Illustrated Textbook of Pediatric Emergency and Critical Care Procedures. St Louis: Mosby, 1997:53–67.

Carraccio C, Feinberg P, Sinclair Hart L, et al. Lidocaine for lumbar punctures: a help not a hindrance. Arch Pediatr Adolesc Med 1996;150:1044–1046.

Christoph RA, Buchanan L, Begalla K, et al. Pain reduction in local anesthetic administration through pH buffering. Ann Emerg Med 1988;17:117–120.

Chudnofsky CR, Wright SW, Dronen SC, et al. The safety of fentanyl use in the emergency department. Ann Emerg Med 1989;18:635–639.

Clark S, Radford M. Topical anesthesia for venipuncture. Arch Dis Child 1986;61:1132–1134.

Cohall AT. Self medication: a new approach to pain. Contemp Pediatr 1989;6:68–108.

Cole TB, Sprinkle RH, Smith SJ, et al. Intravenous narcotic therapy for children with severe sickle cell pain crises. Am J Dis Child 1986;140:1255–1259.

Cote CJ. Sedation for the pediatric patient: a review. Pediatr Clin North Am 1994;41:31–58.

Connors K, Terndrup TE. Nasal versus oral midazolam for sedation of anxious children undergoing laceration repair. Ann Emerg Med 1994;24:1074–1079.

Dachs RJ, Innes GM. Intravenous ketamine sedation of pediatric patients in the emergency department. Ann Emerg Med 1997;19:146–150.

Dailey RH. Fatality secondary to misuse of the TAC solution. Ann Emerg Med 1988;17:159–160.

Dronen SC. Complications of TAC. Ann Emerg Med 1983;12:333.

Dula DJ, Skiendzielewski JJ, Snover SW. The scavenger device for nitrous oxide administration. Ann Emerg Med 1983;12:759–762.

Ernst AA, Marvaez-Valls E, Mall G, et al. 1% lidocaine versus 0.5% diphenhydramine for local anesthesia in minor laceration repair. Ann Emerg Med 1994;23:1328–1332.

Ernst AA, Marvez-Valls E, Nick TG, et al. Lidocaine adrenaline tetracaine gel versus tetracaine adrenaline cocaine gel for topical anesthesia in linear scalp and facial lacerations in children aged 5 to 17 years. Pediatrics 1995;95:255–258.

Ernst AA, Marvez-Valls E, Nick TG, Wahle M. Comparison trial of four injectable anesthetics for laceration repair. Acad Emerg Med 1996; 3:228–233.

Farrell RG, Swanson SL, Walter JK. Safe and effective IV regional anesthesia for use in the emergency department. Ann Emerg Med 1985;14:239–240.

Fassler D, Wallace N. Children's fear of needles. Clin Pediatr 1982;21:59–60.

Fatovich DM, Jacobs IG. A randomized controlled trial of oral midazolam and buffered lidocaine for suturing lacerations in children (the SLIC trial). Ann Emerg Med 1995;24:209–214.

Fernald C, Corry J. Empathic vs. directive preparation of children for needles. Child Health Care 1981;10:44–47.

Fitzmaurice LS, Wasserman GS, Knapp JF, et al. TAC use and absorption of cocaine in a pediatric emergency department. Ann Emerg Med 1990;19:515–518.

French GM, Painter EC, Coury DL. Blowing away shot pain: a technique for pain management during immunization. Pediatrics 1994;93:384–388.

Friedland LR, Kulick RM. Emergency department analgesic use in pediatric trauma victims with fractures. Ann Emerg Med 1994;23:203–207.

Friedland LR, Pancioli AM, Duncan KM. Pediatric emergency department analgesic practice. Pediatr Emerg Care 1997;13:103–106.

Froese N, Costarino AT. Sedation and pain relief: other agents. In: Dieckman R, Fiser D, Selbst SM, eds. Illustrated Textbook of Pediatric Emergency and Critical Care Procedures. St Louis: Mosby, 1997:68–71.

Gamis AS, Knapp JF, Glenski JA. Nitrous oxide analgesia in a pediatric emergency department. Ann Emerg Med 1989;18:177–181.

Green SM, Nakamura R, Johnson NE. Ketamine sedation for pediatric procedures: part I. A prospective series. Ann Emerg Med 1990;19:1024–1032.

Green Sm, Rothrock SG, Gorchynski J. Validation of diphenhydramine as a dermal local anesthetic. Ann Emerg Med 1994;23:1284–1289.

Griffin GC, Campbell VD, Jones R. Nitrous oxide-oxygen sedation for minor surgery, experience in a pediatric setting. JAMA 1981;245:2411–2413.

Hagenbarth MA, Altieri MF, Hawk WH, et al. Comparison of topical tetracaine adrenaline, and cocaine anesthesia with lidocaine infiltration for repair of lacerations in children. Ann Emerg Med 1990;19:63–67.

Halperin DL, Koren G, Attias D. Topical skin anesthesia for venous subcutaneous drug reservoir and lumbar punctures in children. Pediatrics 1989;84:281–284.

Hennes HM, Wagner V, Bonadio WA, et al. The effect of oral midazolam on anxiety of preschool children during laceration repair. Ann Emerg Med 1990;19:1006–1009.

HO K, Spence J, Murphy M. Review of pain-measurement tools. Ann Emerg Med 1996;27:427–432.

International Association for the Study of Pain, Subcommittee on Taxonomy. Pain terms: a list of definitions and notes on usage. Pain 1979;6:249–252.

Jay SM, Ozolins M, Elliot CH, et al. Assessment of children's distress during painful medical procedures. Health Psychiatry 1983;2:133–147.

Klein EJ, Shugerman RP, Leigh-Taylor K, et al. Buffered lidocaine: analgesia for intravenous line placement for children. Pediatrics 1995;95:709–712.

Koenig KL, Hodgson L, Kozak R, et al. Ketorolac vs. meperidine for the management of pain in the emergency department. Acad Emerg Med 1994;1:544–549.

Koren G. Use of eutectic mixture of local anesthetics in young children for procedure related pain. J Pediatr 1993;122:S30.

Krauss BS, Shannon M, Damian FJ, et al. Guidelines for Pediatric Sedation. Dallas: American College of Emergency Physicians, 1995.

Lesko SM, Mitchell AA. As assessment of the safety of pediatric ibuprofen: a practitioner-based randomized clinical trial. JAMA 1995;273:929–933.

Lewis N. The needle is like an animal. Child Today 1978;7:18–21.

Liebelt EL. Reducing pain during procedures. Curr Opin Pediatr 1996; 8:426–441.

Litman RS. Recent trends in the management of pain during medical procedures in children. Pediatr Ann 1995;24:158–163.

Lovejoy FH. Aspirin and acetaminophen: a comparative view of their antipyretic and analgesic activity. Pediatrics 1978;62:904–909.

Mader TJ, Playe JJ, Garb JL. Reducing the pain of local anesthetic infiltration: warming and buffering have a synergistic effect. Ann Emerg Med 1994;23:550–554.

McGrath PA. Pain in the pediatric patient: practical aspects of assessment. Pediatr Ann 1995;24:126–138.

Means LJ. Pain relief for children: new concepts, new methods. Contemp Pediatr 1994;11:70–93.

Menegazzi JJ, Paris PM, Kersteen CH, et al. A randomized, controlled trial of the use of music during laceration repair. Ann Emerg Med 1991;20:348–350.

Olness K. Hypnosis in pediatric practice. Curr Prob Pediatr 1981;12:1–47.

Olney BW, Lugg PC, Turner PL, et al. Outpatient treatment of upper extremity injuries in childhood using intravenous regional anesthesia. J Pediatr Orthop 1988;8:576–579.

Petrack EM, Christpher NC, Kriwinsky J. Pain management in the emergency department: patterns of analgesic utilization. Pediatrics 1997;99:711–714.

Pierce MC, Fuchs S. Evaluation of ketorolac in children with forearm fractures. Acad Emerg Med 1997;4:22–26.

Pohlgeers AP, Friedland LR, Keegan-Jones L. Combination fentanyl and diazepam for pediatric conscious sedation. Acad Emerg Med 1995;2: 879–883.

Pryor GS, Kilpatrick WR, Opp DR. Local anesthesia in minor lacerations, topical TAC versus lidocaine infiltration. Ann Emerg Med 1980;9:568–571.

Sacchetti A, Schafermeyer R, Gerardi M, et al. Pediatric analgesia and sedation. Ann Emerg Med 1994;23:237–250.

Scarfone RJ, Jasani M, Gracely EJ. Pain of local anesthetics: rate of administration and buffering. Ann Emerg Med 1998;31:36–40.

Schecter NL. Pain and pain control in children. Curr Prob Pediatr 1985;15:5–67.

Schutzman SA, Liebelt E, Wisk M, Burg J. Comparison of transmucosal fentanyl citrate and intramuscular meperidine, promethazine and chlorpromazine for conscious sedation of children undergoing laceration repair. Ann Emerg Med 1996;28:385–390.

Schilling CG, Bank DE, Borchert BA, et al. Tetracaine, epinephrine (adrenaline) and cocaine (TAC) versus lidocaine, epinephrine and tetracaine (LET) for anesthesia of lacerations in children. Ann Emerg Med 1995;25:203–208.

Selbst SM. Analgesia in children: why is it underused in emergency departments? Drug Safety 1992;7:8–13.

Selbst SM. Managing pain in the pediatric emergency department. Pediatr Emerg Care 1989;5:56–63.

Selbst SM. Pain management in the emergency department. In: Schechter NL, Berder CB, Yaster M, eds. Pain in Infants, Children and Adolescents. Baltimore: Williams & Wilkins, 1993:505–518.

Selbst SM, Clark M. Analgesic use in the emergency department. Ann Emerg Med 1990;19:1010–1013.

Shapiro A. Behavior of kibbutz and urban children receiving an injection. Psychophysiology 1975;12:79–82.

Smith GA, Strausbaugh SD, Harbeck-Weber C, et al. Comparison of topical anesthetics without cocaine to tetracaine-adrenaline-cocaine and lidocaine infiltration during repair of lacerations. Pediatr 1996;97:301–307.

Stewart RD. Nitrous oxide sedation/anesthesia in emergency medicine. Ann Emerg Med 1985;14:139–148.

Sutters KA, Levine JD, Dibble S, et al. Analgesic efficacy and safety of single-dose intramuscular ketorolac for postoperative pain management in children following tonsillectomy. Pain 1995;61:145–153.

Terndrup TE, Cantor RM, Madden CM. Intramuscular meperidine, promethazine, and chlorpromazine: analysis of use and complications in 487 pediatric emergency department patients. Ann Emerg Med 1989;18: 528–533.

Terndrup TE, Dire DJ, Madden CM, et al. A prospective analysis of intramuscular meperidine, promethazine, and chlorpromazine in pediatric emergency department patients. Ann Emerg Med 1991;20:31–35.

Tessler M, Ward J, Sareda M. Developing an instrument for eliciting children's description of pain. Percept Mot Skills 1983;56:315–321.

Theroux MC, West DW, Corddry DH, et al. Efficacy of intranasal midazolam in facilitating suturing of lacerations in preschool children in the emergency department. Pediatrics 1993;91:624–627.

Tobias JD, Phipps S, Smith B, Mulhern RR. Oral ketamine pre-medication to alleviate the distress of invasive procedures in pediatric oncology patients. Pediatrics 1992;90:537–541.

Torturro MA, Paris PM, Seaburg DC. Intramuscular ketorolac versus oral ibuprofen in acute musculoskeletal pain. Ann Emerg Med 1995;26: 117–120.

Unruh A, McGraph P, Cunningham SJ, et al. Children's drawings of their pain. Pain 1983;17:388–392.

Weinberger M. Analgesic sensitivity in children with asthma. Pediatrics 1978;62:910–915.

Weisman SJ, Bernstein B, Schechter NL. Consequences of inadequate analgesic during painful procedures in children. Arch Pediatr Adolesc Med 1998;152:147–149.

Wright SW, Norris RL, Mitchell TR. Ketorolac for sickle cell vasoocclusive crises pain in the emergency department: lack of a narcotic-sparing effect. Ann Emerg Med 1992;21:925–928.

Yaster M, Nichols DG, Deshpand JK, et al. Midazolam fentanyl intravenous sedation in children: case report of respiratory arrest. Pediatrics 1990;86:463–466.

Yaster M, Tobin JR, Fisher GA, et al. Local anesthetics in the management of acute pain in children. J Pediatr 1994;124:165.

Zeltzer L, Dash J, Holland JP. Hypnotically induced pain control in sickle cell anemia. Pediatrics 1979;64:533–536.

Zeltzer L, Lebaron S. Hypnosis and non-hypnotic techniques for reduction of pain and anxiety during painful procedures in children and adolescents with cancer. J Pediatr 1982;101:1032–1035.

Zempsky WT, Karasic RB. EMLA versus TAC for topical anesthesia of extremity wounds in children. Ann Emerg Med 1997;30: 163–166.

CHAPTER 5

Emergency Airway Management—Rapid Sequence Intubation

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Management of the airway is a critical initial step in the stabilization of patients who present to the emergency department (ED) with a life-threatening emergency. Tracheal intubation often is the most reliable means of maintaining airway control. Indications for tracheal intubation include cardiopulmonary arrest, apnea, respiratory insufficiency, actual or potential airway obstruction, respiratory depression, severe burns, severe multiple trauma, severe head injury, increased intracranial pressure (ICP), a depressed sensorium, and a loss of the normal protective airway reflexes.

Airway management is most optimal when the ED staff is trained to recognize the need for intubation and to accomplish this while minimizing complications. The equipment necessary for tracheal intubation should be readily available at the bedside for immediate access and use during the management of any critically ill patient in the ED (Table 5.1).

Pulse oximeter
End-tidal CO_2 monitor or detector
Electrocardiogram monitor
Uncuffed endotracheal tubes, sizes 2.5 to 6.0
Cuffed endotracheal tubes, sizes 6.0 to 8.5
Endotracheal tube stylets
Laryngoscopes (straight blade sizes 0 to 3, curved blade sizes 2 to 4)
Oral airways
Oxygen masks, preferably a nonrebreather
Ventilation masks in all sizes for bag-valve-mask ventilation
Large and small self-inflating ventilation bag with oxygen reservoir tail and positive end-expiratory pressure (PEEP) valve attachment
Laryngeal mask airways (LMA) in all sizes
Oxygen source
Suctioning source
Large-bore stiff suction tips
Flexible suction catheters
Nasogastric tubes
Tracheostomy tubes
Tracheostomy surgical instrument set
12- and 14-gauge needle catheters for needle cricothyrotomy
Percutaneous transtracheal ventilation setup

Table 5.1. Equipment Needed for Rapid Sequence Intubation

Tracheal intubation can be difficult because of seizures, agitation, combativeness, and inadequate muscle relaxation, resulting in poor airway visualization. Laryngoscopy and intubation result in ICP elevation, pain, bradycardia, and a higher risk of gastric regurgitation and hypoxemia. Patients who arrive in the ED often have full stomachs.

Although intubation often can be accomplished under these circumstances, the conditions are optimized and the adverse effects are minimized when a patient is intubated using rapid sequence intubation (RSI)—a rapid induction of general anesthesia that induces unconsciousness and muscle relaxation. The purpose of RSI is to rapidly render a patient unconscious and paralyzed so that intubation can be facilitated. Emergency intubations done under a fully relaxed state are usually easier to perform and have fewer adverse effects, such as pain and ICP elevation.

Pharmacologic paralysis makes it impossible to perform a neurologic examination on the patient and eliminates all respiratory effort by the patient. Would it be preferable to sedate a patient without paralysis for intubation because of these considerations? Although this was a common practice in years past, intubation using sedation alone has a higher complication rate than RSI does, making RSI the preferred technique.

RSI is not necessarily indicated in cardiac arrest. In cardiac arrest, for example, intubation without RSI would generally be preferable unless cardiopulmonary resuscitation (CPR), brain perfusion, muscle tone, and/or some degree of consciousness were maintained, in which case RSI may be of benefit. Understanding the principles of RSI is the best

means of determining when it is indicated.

A typical RSI consists of providing atropine to block vagal stimulation, a sedative to induce unconsciousness, and a muscle relaxant to induce paralysis. This typical sequence can become rather complicated when more considerations are added. It should be noted that the drugs for pediatric RSI have been recommended in their most basic forms as 1) atropine, 2) sedative, and 3) muscle relaxant. This chapter addresses the following controversies surrounding RSI: sedative selection, muscle relaxant selection, priming or defasciculation, and adjunctive medications such as lidocaine and fentanyl.

RAPID SEQUENCE INTUBATION SEDATIVES

The most common sedatives used in RSI include thiopental, midazolam (and other benzodiazepines), ketamine, and etomidate. Narcotic analgesics such as fentanyl can also be used, but this is less common. The use of propofol has increased in frequency, but its use is mostly for brief sedation for procedures, rather than for RSI. Each of these drugs has beneficial and detrimental properties that must be understood to select the best sedative for RSI in the patient at hand. The advantages and disadvantages of each drug are summarized in Table 5.2.

Drug	Onset	Duration	Cerebrovascular Effect	Cardiovascular Effect	Bronchial Effect	Other Considerations
Thiopental	Rapid	Brief	Good	Significant depression	Bronchospasm	
Midazolam	Less rapid	Brief	Modest	Neutral	Neutral	Titration recommended; not available in PRC
Ketamine	Rapid	Brief	Adverse	Stimulatory	Bronchodilatory	Psychic reactions and excessive airway secretions
Etomidate	Rapid	Brief	Good	Neutral	Neutral	Myoclonus, cortisol suppression
Fentanyl	Less rapid	Brief	Modest	Neutral	Neutral	Succinylcholine activity and chest wall rigidity
Propofol	Rapid	Brief	Good	Significant depression	Neutral	Less experience with agent in ED PRS

Table 5.2. Significant Properties of Rapid Sequence Intubation Sedatives

Thiopental (an ultra-short-acting barbiturate) was initially one of the most commonly used sedatives for RSI. Its advantages are reliable and rapid onset (10 to 20 seconds), short duration, and a cerebral protective effect accomplished by reducing ICP, cerebral metabolism, and oxygen demand. Its main disadvantages are vasodilation and myocardial depression, which may result in hypotension. Thiopental should be avoided or used in lower dosages in hypotensive or hypovolemic patients. These effects can be minimized by slowing the rate of injection. Thiopental causes respiratory depression and may result in coughing, laryngospasm, and bronchospasm. Thiopental is contraindicated in porphyria and status asthmaticus.

Midazolam (and other benzodiazepines such as diazepam) has a slower onset than thiopental does. It is more commonly used for conscious sedation or as an adjunctive agent in general anesthesia. Midazolam is capable of anesthesia induction at higher dosages. Cardiovascular and respiratory depression occur less often than with barbiturates. Lack of recall or anterograde amnesia results from benzodiazepines used for anesthesia. Benzodiazepines should not be used in patients with glaucoma. Midazolam has many properties that suggest its use for RSI in the ED. Many reports in the literature have advocated its use despite the lack of data to document its efficacy and safety. The dosing range is suggested at 0.1 to 0.3 mg/kg. The lower dosage is probably insufficient to reliably induce unconsciousness. Because these drugs also result in amnesia, studies may never be able to retrospectively assess the degree of unconsciousness attained during RSI.

Although attractive, benzodiazepines have a slower onset than thiopental and have an excessively wide dosing range during which titration is recommended. In RSI, however, titration is undesirable because a rapid induction of unconsciousness and paralysis is the goal, and paralysis makes it impossible to assess consciousness. The use of midazolam in RSI has grown in popularity, largely because it can be used in nearly all clinical situations. Despite this, there appear to be drugs that have advantages over benzodiazepines in specific clinical situations.

Ketamine produces rapid sedation, amnesia, and analgesia. It is described as a dissociative agent that induces a trancelike state in which the patient is unaware, but not necessarily asleep. In combination with a paralyzing agent as in RSI, this difference is not noticeable. Ketamine results in sympathetic stimulation and an increase in systemic blood pressure (BP); however, reduced doses or no sedative are still recommended in potentially hypovolemic patients. Adverse effects, which include ICP elevation, intraocular pressure elevation, hallucinations, excessive airway secretions, and laryngospasm, limit its use to ED patients who have hypotension, hypovolemia, or status asthmaticus. Ketamine increases airway secretions, so routine atropine premedication is recommended. Ketamine has a bronchodilating effect in addition to its sympathetic effect, making it useful for RSI of patients with severe bronchospasm that requires intubation. Ketamine is contraindicated in patients with hypertension, head injury, psychiatric problems, glaucoma, or an open globe injury. The psychic actions of ketamine have severely limited its use. These reactions usually disappear upon awakening but can be recurrent. The frequency of psychic disturbances with ketamine (5 to 30%) are less common in children. Other reports have shown no significant adverse psychological effects compared with other sedatives. The use of lorazepam during ketamine anesthesia has been recommended to reduce the likelihood of adverse psychological effects.

Etomidate has more recently been advocated as an RSI sedative. Although most RSI sedatives have advantages in certain clinical situations and disadvantages in others, etomidate has advantages in a broader range of RSI patients. It

shares some advantages of thiopental—rapid and reliable onset of unconsciousness, ICP reduction, and reduction of cerebral metabolic demand. Etomidate has minimal cardiovascular depression compared with thiopental. Etomidate appears to have superior cerebroprotective properties because it lowers ICP and cerebral metabolism with better preservation of cerebral perfusion pressure compared with thiopental. Etomidate's major disadvantage is its association with myoclonus resembling seizures. The frequency of this ranges from 10 to 80% in various reports. The etiology of this is not well defined. Etomidate has some anticonvulsant properties, but it also appears to stimulate seizures in others. Etomidate suppresses glucocorticoid and mineralocorticoid levels. This effect is clinically significant in long-term administration of etomidate. Single use as in RSI results in measurable decreases in corticosteroid levels, but this is not clinically significant unless etomidate is used over a long period. Replacement corticosteroids may be considered to offset this problem. Although unclear, etomidate should not be used in patients with partial seizures and adrenal insufficiency. Despite its broad applicability for RSI, etomidate has not been used frequently for RSI. It has only recently been recommended for RSI in the ED; thus, many ED physicians are not familiar with it. There are some disadvantages of using etomidate, and its ultimate use pattern in the future has yet to be determined.

Fentanyl is a short-acting narcotic analgesic that results in rapid analgesia and unconsciousness at higher dosages. Adverse effects associated with fentanyl are less than those of morphine. The doses of narcotics required to produce complete anesthesia are much higher than the doses required for analgesia alone and may vary extensively. Chest wall rigidity may occur with rapid injection of fentanyl, but this is reversible with a muscle relaxant or with naloxone. Fentanyl use has been associated with seizurelike activity. Fentanyl is used most often in cardiovascular surgery in combination with other anesthetics. Although it has some properties useful for RSI done in the ED, literature sources to support fentanyl use for RSI in the ED are lacking. Fentanyl should not be used with monoamine oxidase (MAO) inhibitors.

Propofol use in the ED is in its infancy. Largely used by anesthesiologists for short-term sedation and general anesthesia, propofol is gaining wider acceptance as an agent for short-term sedation for procedures in the intensive care unit (ICU) and ED. It can be used as a sedative in RSI, but its degree of experience here is limited and it does not have substantial advantages over other sedatives. Propofol shares many features with thiopental. Propofol decreases ICP and cerebral metabolism. Propofol's onset is rapid and brief, but it has significant cardiovascular depression. Although propofol can be used in instances when thiopental could be used, there is more experience with thiopental.

Although myocardial depression is most pronounced with thiopental, all sedatives cause some degree of cardiovascular depression, especially in hypotensive or hypovolemic patients. Because no sedative is entirely free of cardiovascular depression in the hypovolemic or hypotensive patient, such patients should receive reduced dosages or no sedative at all, depending on their cardiovascular status.

SEDATIVE SELECTION

Sedative selection remains one of the most controversial aspects of RSI. The addition of etomidate as a sedative for RSI has extended the sedative selection options. Etomidate appears to have the broadest applicability, but each agent should be well understood so that individual practitioners can make the best decisions about which agent would be most optimal in each clinical situation. No sedative is universally superior. The clinical situation determines the optimal sedative selection. Table 5.3 summarizes sedative selections in different clinical categories.

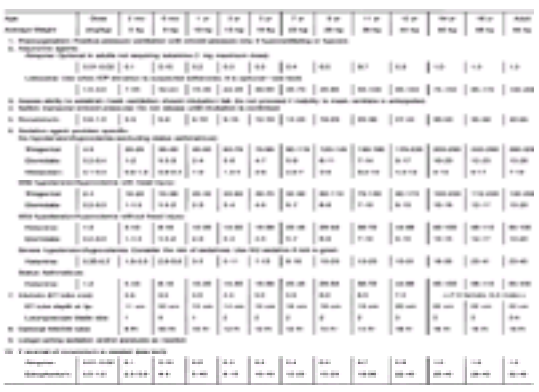

Table 5.3. Rapid Sequence Intubation Drugs and Doses (in milligrams)

Thiopental can be used for all patients except those with hypotension, hypovolemia, status asthmaticus, or porphyria. In potentially hypovolemic patients with suspected head injuries, midazolam or low-dose thiopental may be used in conjunction with volume resuscitation. Midazolam has the major disadvantage of ideally requiring titration, which is not feasible in RSI. Midazolam's dosing range is broad because its sedative effect is highly variable. Despite these major disadvantages, midazolam appears to be a popular choice in RSI; however, this popularity is difficult to justify because other agents appear to be superior.

Perhaps etomidate would be superior to thiopental in all these indications, but there is concern that patients with head injuries may be at risk for partial seizures. When thiopental can be used, propofol can be used as well, but its experience level does not approach that of thiopental.

In patients with suspected head injuries who have severe hypovolemia or hypotension, thiopental should be avoided. Any sedation agent may compromise cardiovascular function in such a state. There is not enough information in the literature to establish a clear consensus about which sedative would be the most optimal in this situation. A low-dose sedative or no sedative would be among the best options. Although etomidate appears to have many desirable characteristics, there

is concern that patients with head injuries may be at risk for partial seizures.

In a patient without head injuries who has hypotension/hypovolemia, ketamine or etomidate may produce the least adverse effects. Atropine should be used with ketamine. No sedative or a low dosage of sedative (ketamine, etomidate, or any acceptable sedative) is recommended in severe hypotension/hypovolemia to reduce the likelihood of cardiovascular collapse in patients with maximal endogenous sympathetic stimulation.

Ketamine or benzodiazepines have been recommended for sedating patients with status asthmaticus who require intubation; however, ketamine is associated with a bronchodilatory effect while diazepam is not. Etomidate can be used here, but ketamine has superior characteristics for severe status asthmaticus.

In patients with severe cardiovascular compromise or unconsciousness, a sedative may be considered optional to prevent further cardiovascular compromise.

MUSCLE RELAXANTS

Muscle relaxants result in total muscle paralysis, yet the patient may be fully conscious.

Succinylcholine is a depolarizing muscle relaxant with a rapid onset (30 to 60 seconds) and short duration (3 to 12 minutes). Even though it has been the most common muscle relaxant used in RSI, it has numerous disadvantages. Although not contraindicated in head injuries, it causes elevated ICP. Intraocular and intragastric pressure elevations may occur. Muscle fasciculations that may result in muscle pain, rhabdomyolysis, and myoglobinuria also may occur. These are more severe in muscular patients and can be prevented by a defasciculating dose of vecuronium before succinylcholine (this is not necessary in children under 5 years of age). Atropine premedication can prevent the bradycardia and excessive bronchial secretions associated with succinylcholine. Other less preventable adverse effects include negative inotropic and chronotropic effects, an association with malignant hyperthermia, hyperkalemia, hypertension, and arrhythmias. Succinylcholine is contraindicated in patients with glaucoma, penetrating eye injuries, significant neuromuscular disease, history or family history of malignant hyperthermia, and pseudocholinesterase deficiency. At 3 to 60 days after trauma or burns (in other words, day 3 or later following trauma or burns), succinylcholine results in an increased frequency of the risks described here and should not be used. Patients with severe burns or large crush injuries may already be acutely hyperkalemic; thus, the decision to use succinylcholine should consider this as well.

Of the nondepolarizing muscle relaxants, rocuronium currently has the fastest onset and shortest duration. Vecuronium is still commonly used because it was the most popular agent before rocuronium. Both agents have minimal cardiovascular effects. Onset times for rocuronium and vecuronium are 30 to 90 seconds and 90 to 120 seconds, respectively. Duration for both drugs is 25 to 60 minutes. Rocuronium comes in a premixed vial ready to use, whereas vecuronium comes in a powder that must be reconstituted. This factor gives rocuronium another advantage when the time to intubation may be prolonged because of medication preparation.

Other nondepolarizing muscle relaxants include pancuronium and atracurium. Pancuronium has a slower onset and more cardiovascular side effects. Atracurium has an onset time similar to that of vecuronium; however, it is associated with histamine release and cardiovascular side effects. Rocuronium is currently the nondepolarizing muscle relaxant of choice, but there is less experience with this drug compared to vecuronium. Other nondepolarizing agents should be eliminated from consideration.

MUSCLE RELAXANT SELECTION

In comparing rocuronium and succinylcholine, rocuronium has fewer adverse effects, whereas succinylcholine has a shorter duration. The onset times are similar. Some view rocuronium as preferable because it is safer. The contrary view is that because of its shorter duration (which allows restoration of spontaneous ventilation within 3 to 12 minutes compared with 25 to 60 minutes for rocuronium), succinylcholine is better if intubation fails. Rocuronium can be reversed pharmacologically with edrophonium and other similar agents; however, this is not clinically routinely useful because reversal cannot be achieved immediately. Reversal must wait for some degree of spontaneous recovery to occur, which happens later than the duration of succinylcholine.

In most patients, neuromuscular blockade facilitates both intubation and bag-valve-mask ventilation. For patients with risk factors that suggest a difficult intubation, succinylcholine may be preferable. For others, rocuronium may be preferable. Whether the shorter duration of succinylcholine justifies its greater risk of adverse effects is essentially a personal judgment that individual clinicians must make.

Defasciculation and Priming

Defasciculation refers only to the use of succinylcholine, which may cause fasciculations, muscle pain, rhabdomyolysis, and myoglobinuria. This effect is most pronounced in muscular individuals. Fasciculations may increase muscle tone and increase the risk of gastric regurgitation during RSI. To prevent fasciculations, “defasciculation” is recommended, where one-tenth the paralyzing dose of a nondepolarizing muscle relaxant (e.g., vecuronium 0.01 mg/kg) is administered 1 to 3 minutes before succinylcholine administration. This “defasciculating” dose of vecuronium will prevent fasciculations caused by succinylcholine. Defasciculation is most beneficial in muscular individuals. Defasciculation is not necessary in children 5 years of age or younger. Note that this defasciculating step delays the time to intubation and adds complexity to RSI.

Priming in RSI refers to nondepolarizing muscle relaxants only. Its purpose is to shorten the onset time of nondepolarizing muscle relaxants. A priming dose is one-tenth the paralyzing dose of a nondepolarizing muscle relaxant.

Using vecuronium as an example, a priming dose of 0.01 mg/kg is administered. Five minutes should elapse for the "priming" to take effect. The paralyzing dose of 0.1 mg/kg is now administered. The full paralyzing onset time of vecuronium is about 100 seconds without priming, 50 seconds with priming. Unfortunately, priming adds an additional 5-minute delay to intubation while saving 50 seconds in accelerating the onset of the full dose of vecuronium. Because the onset time of rocuronium is considerably faster, the advantage of priming is minimal. Although some experts still recommend priming, it appears to have little benefit in the ED when immediate intubation is required.

Defasciculation and priming are often confused because they both require one-tenth of the paralyzing dose of a nondepolarizing muscle relaxant. However, the two principles are different even though they have similar characteristics in that they both are optional and they both delay the time to intubation and they both add complexity to the drug administrations in RSI.

ADJUNCTIVE AGENTS

The RSI sequence shown in Table 5.3 considers the use of atropine and lidocaine. Atropine use is considered routine in children to prevent bradycardia, but it is optional in adults, unless ketamine is used as a sedative, in which case, atropine is recommended in adults as well.

Lidocaine is more controversial. It has been shown to reduce ICP and airway reactivity under certain conditions when given 2 minutes before intubation. If ICP elevation is suspected, a cerebroprotective sedative (thiopental or etomidate) is generally preferred in RSI. Lidocaine is cerebroprotective in isolation, but it is unclear whether lidocaine results in additional benefit when added to a cerebroprotective RSI regimen that includes thiopental or etomidate. Despite this controversy, most practicing academic centers and consensus reports recommend the use of intravenous lidocaine if ICP elevation is suspected. In addition to intravenous lidocaine, topical lidocaine has been recommended to blunt the adverse reaction to tracheal intubation. This adds considerable complexity to the laryngoscopy procedure, especially in patients in whom neck immobilization is critical and/or airway visualization may be less than optimal. The recommendation that lidocaine be used in intubating asthmatics stems from its beneficial effect in attenuating bronchospasm. If one truly believes that lidocaine has such a benefit, then if possible, it should be administered long before the patient requires intubation, as opposed to administering it during RSI.

Opiate analgesics such as fentanyl and morphine have been advocated as adjunctive agents in RSI to further reduce the adverse effects of intubation. Ketamine has analgesic properties; thus, coadministration of analgesics is unnecessary with ketamine. Sedatives such as benzodiazepines, etomidate, and thiopental have little or no analgesic properties. The coadministration of analgesics have been recommended to address this. When considering that the patient is fully unconscious when reliable sedatives such as etomidate and thiopental are used, the additional benefit of analgesics to reduce the amount of "pain" felt by an unconscious patient becomes small. Narcotic analgesics have adverse reactions and the additional risk that these pose may not justify their routine use. Benzodiazepines must ideally be titrated to assess the degree of sedation; thus, the degree of sedation with these agents is less reliable in RSI, in which case, coadministration of analgesics may be more beneficial.

If RSI is to accomplish its goal of rapid tracheal intubation, the addition of adjunctive agents should be critically considered because each additional agent adds time and complexity to RSI. This factor is often not considered when discussing the benefit of an individual adjunctive agent in isolation. Therefore, the RSI protocol described in Table 5.3 includes atropine for pediatric patients and intravenous lidocaine as an optional adjunctive agent for head trauma. Other adjunctive agents may be considered, but they have not been included in the table.

RAPID SEQUENCE INTUBATION PROTOCOL

After patient assessment, immediate stabilization, and intravenous access, patients should be assessed for any contraindications to RSI or its agents. The major contraindication to RSI is a likelihood that intubation or ventilation might not be possible, as in cases of limited cervical mobility, a receding mandible, limited jaw opening, major facial or laryngeal trauma, upper airway obstruction, or distorted facial or airway anatomy.

To simplify RSI, a table such as Table 5.3 should be adapted and modified to your preferences. This table should be taped to the wall in the critical care area of your ED. This table is not a substitute for thoroughly understanding the characteristics of each agent and the critical thinking necessary to select the agents. What follows explains the protocol given in Table 5.3:

1. Preoxygenation (by spontaneously inspiring or mask-ventilating 100% oxygen for 2 to 5 minutes) results in an oxygen reserve. Positive-pressure mask ventilation may inflate the stomach and increase the likelihood of gastric regurgitation; thus, positive pressure in conjunction with cricoid pressure (see step 4) should be done only if needed to oxygenate and ventilate the patient adequately. Hyperoxygenation is impossible in some patients. Pulse oximetry and an electrocardiogram monitor should be considered mandatory for patients undergoing RSI. If a self-inflating bag is used, oxygen is *not* delivered to the mask unless the bag is squeezed. Thus, if the patient is spontaneously breathing, a mask attached to a self-inflating bag should *not* be used; instead, a standard oxygen mask, such as a nonrebreather, should be used.
2. Atropine premedication is routinely administered in children. It prevents bradycardia and reduces oral secretions. This is considered optional for adults unless ketamine is used, in which case atropine is recommended. Lidocaine lowers ICP and suppresses the cough reflex. It may be beneficial to patients with ICP elevations. When used in conjunction with other ICP-lowering agents, however, its additional benefit is unclear. Lidocaine is generally recommended for patients with suspected ICP elevation.
3. Before proceeding, it should be ascertained that a good mask seal and an open airway can be maintained. In most instances, muscle relaxation facilitates mask ventilation and intubation. If an inability to intubate and/or mask ventilate is suspected, RSI should not proceed until additional assistance can be obtained.
4. The Sellick maneuver (application of pressure on the cricoid ring sufficient to occlude the esophageal lumen

without compressing the airway lumen or moving the cervical spine) reduces the likelihood of passive gastric regurgitation and aspiration. It should be maintained until tracheal intubation is confirmed. This also reduces the likelihood of gastric distension resulting from mask ventilation and therefore should be done before any positive-pressure mask ventilation unless this results in gagging.

5. After appropriate selection, a muscle relaxant and a sedative should be administered simultaneously or in rapid sequence. Administering the muscle relaxant first allows the sedative to be administered gradually while waiting for the full onset of the muscle relaxant. Some experts prefer the reverse sequence. For muscle relaxants, Table 5.3 lists only rocuronium. Succinylcholine or vecuronium may be substituted here. Optional priming (applies to nondepolarizing agents only) and defasciculation (applies to succinylcholine only) are not included in the sequence described in Table 5.3.
6. Intubation can take place when there is full relaxation of the airway muscles, usually 45 seconds after rocuronium administration (90 seconds after vecuronium).
7. Once intubation is completed, proper endotracheal (ET) tube placement should be confirmed by auscultation, end-tidal CO_2 detection, and the maintenance of oxygenation monitored by pulse oximetry.
8. Gastric evacuation can be done with a nasogastric or orogastric tube at this time.
9. Longer-acting sedatives and nondepolarizing muscle relaxants should be administered to maintain unconsciousness and paralysis as needed.
10. If reversal of rocuronium is necessary, edrophonium together with atropine can be administered to accelerate recovery; however, some degree of spontaneous recovery must be present for reversal to occur.

Nasal Intubation Compared with Oral Intubation in the Trauma Patient

Trauma victims who arrive in the ED have suspected cervical spine (C-spine) injuries in addition to other injuries. When it is not possible to rule out a C-spine injury before RSI, the head and neck should be immobilized during intubation.

Unless contraindicated, emergency intubation of pediatric patients should always be done orally. In spontaneously breathing patients, older literature sources have recommended blind nasal tracheal intubation, whereas newer recommendations prefer oral tracheal intubation using RSI. If the need for intubation is emergent, nasal tracheal intubation may not be as reliable as oral tracheal intubation. Nasal tracheal intubation is noxious, and it may cause the conscious patient to gag or become agitated, resulting in more neck movement, an increase in ICP, and possible vomiting. Nasal tracheal intubation is more difficult in children. Epistaxis, sinusitis, and cribriform fracture complications are other concerns with nasal tracheal intubation. Studies have not been able to show that nasal tracheal intubation results in less C-spine movement than oral tracheal intubation.

There is concern that laryngoscopy during oral tracheal intubation may displace a C-spine fracture. When using RSI, however, laryngoscopy manipulation and neck movement are minimized under these more ideal conditions. The concern that the loss of cervical muscle tone on an unstable C-spine will reduce its splinting effect and increase its instability has not been supported by evidence.

In a critical situation, intubation is best carried out by the means with which the clinician is most familiar for the given clinical condition. Oral tracheal intubation using RSI appears to be the best means of securing an airway for most clinicians.

Cervical Spine Immobilization During Tracheal Intubation

The terms that describe C-spine immobilization are ambiguous. Traction has been used synonymously with immobilization, although “traction” generally indicates a pulling action with an undefined degree of force. Manual cervical immobilization implies that hands are somehow used to immobilize the neck. Cervical immobilization implies the use of a stiff collar and other devices to immobilize the neck. Philadelphia collars used with manual stabilization do not provide any additional C-spine stability compared with manual stabilization alone. Axial (in-line) traction has been shown to worsen C-spine stability in patients. One preferred method is to remove the anterior portion of the cervical collar with an assistant immobilizing the head and neck once the patient is rendered unconscious and paralyzed. This enables the jaw to open wider, providing better visualization during laryngoscopy without sacrificing cervical immobilization.

ALTERNATIVE INTUBATION AND AIRWAY TECHNIQUES

Because the experience level of most ED physicians is greatest with oral tracheal intubation, it is unwise to deviate from this in managing a critically ill child who requires intubation. Alternative procedures should be reserved for instances in which conventional airway techniques prove unsuccessful.

Flexible fiberoptic scopes, lighted stylets to guide nasal tracheal intubation, retrograde intubations, and surgical airways all require high skill and experience levels to be done optimally. These procedures have a minimal degree of documented experience in children. Directly visualizing the airway through a fiberoptic scope is appealing; however, it requires extensive practice, and it may be especially difficult in critical intubations or in agitated patients. Intubation aided by bronchoscopy, lighted stylets, and the retrograde wire technique are not recommended in ED RSI because of the lack of spontaneous breathing during RSI and the time required for these procedures.

The Combitube (a registered trademark of Sheridan Catheter Corporation, Argyle, NY) is a double- lumen airway that is blindly inserted through the mouth. One lumen exits through the distal end of the Combitube. The other lumen exits through multiple side-holes proximal to the distal end. An inflatable (distal) balloon separates these two (the distal end-hole and the more proximal side-holes). Because the Combitube is inserted blindly, it will enter either the trachea or the esophagus. If it enters the trachea, the distal balloon is inflated and the distal end-hole lumen is used to ventilate the patient just as if this were a conventional ET tube. If the Combitube enters the esophagus, the inflation of the distal balloon occludes the esophagus and the lumen ending in the more proximal side-holes is used to ventilate the patient. The esophageal position of this tube is similar to an esophageal obturator airway. Use of the Combitube requires

familiarity with its function and method of insertion. It has been demonstrated to be effective in providing an airway during resuscitation, but failures occur as well. Widespread experience with the Combitube in pediatric patients is lacking.

The laryngeal mask airway (LMA) is a newer airway device. The LMA is not disposable and must be sterilized between uses. LMAs come in several different sizes, and their use in pediatric patients has been demonstrated. Experience with LMAs is growing. The correct insertion and placement position of the LMA is critical. The LMA is inserted blindly, taking about 15 to 20 seconds. LMA insertion methods are best taught using video or hands-on instruction. In-depth understanding of the LMA and previous hands-on experience are required to consider it as an airway management option. The LMA does not prevent aspiration.

Surgical airways may be considered. Complications, including incorrect tube placement, subcutaneous emphysema, pneumomediastinum, pneumothorax, bleeding, tracheal stenosis, subglottic stenosis, arterial injury, blood aspiration, and persistent tracheocutaneous fistulae, are more common when the procedure is performed on an emergency basis in children. Cricothyrotomy is faster and easier to perform than tracheostomy and also has a lower complication rate. However, in small children, the cricothyroid membrane is not readily palpable, and it may be too small for an airway. It is not recommended in children less than 10 years of age. Electrocautery devices should be avoided during these procedures because the presence of high-flow oxygen can result in spontaneous combustion.

Because surgical airways are difficult to perform in children, needle cricothyrotomy may be beneficial for children who cannot be ventilated by any other route, although the complications are similar to those of surgical airways and the experience level with this procedure in children is minimal as well. An over-the-needle 12- or 14-gauge intravenous catheter is directed inferiorly through the cricothyroid membrane, the needle is removed, and the catheter is left in place. It is vital to have a ventilation device preassembled and ready to use before such an emergency because ventilation through a transtracheal catheter requires a special setup. Many recommendations for transtracheal ventilation have appeared in the literature. Special transtracheal airway kits are available commercially. Two examples are shown in Figure 5.1. These use a wall outlet or tank oxygen pressure directly into the transtracheal catheter. It is the most optimal means to deliver an adequate tidal volume through the small catheter. By occluding the T-tube or the side-hole, oxygen is forced through the catheter at high pressure. Chest movement should be used as a visible indicator of adequate ventilation. Exhalation occurs passively through the larynx and not through the catheter. If exhalation is obstructed as well, transtracheal ventilation is contraindicated. The catheter must be held securely because a kink or movement of the catheter would compromise this fragile airway. Another common recommendation is to attach a ventilation bag to the transtracheal catheter through an ET tube connector (about size 3.0). Delivering an adequate tidal volume by squeezing the bag, however, is nearly impossible with this method.

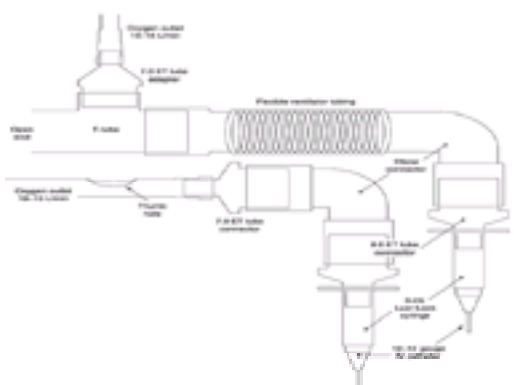

FIGURE 5.1. Transtracheal ventilation setup. (Adapted from Yamamoto LG. Rapid sequence anesthesia induction and advanced airway management in pediatric patients. Emerg Med Clin North Am 1991;9:611–638, copyright by WB Saunders, with permission from author and publisher.)

Transtracheal ventilation is only a temporizing measure, and a more definitive airway should be established as soon as possible. It is important to have ED staff members familiarize themselves with the transtracheal ventilation setup. By attaching a balloon or glove to the transtracheal catheter, air flow can be visualized during practice sessions.

AVOIDING PROBLEMS

Being prepared by becoming aware of the following problems commonly encountered during RSI can improve the success of the procedure greatly.

1. Oxygen delivery devices must be well-understood by the staff. New nurses and residents often do not know that oxygen is *not* delivered to the mask attached to a self-inflating bag unless the bag is squeezed. Thus, putting such a mask in front of a patient who is spontaneously breathing will deliver only 21% oxygen (room air), and the patient will fail to hyperoxygenate. For spontaneously breathing patients, oxygen should be delivered using a standard oxygen mask such as nonrebreather. A mask attached to a closed circuit bag (anesthesia-type bag, Rusch bag) with 100% oxygen can also be used. For patients who require positive-pressure mask ventilation, a self-inflating bag with an oxygen reservoir tail is satisfactory.
2. Drug preparation is time-consuming. Thiopental comes in kits that require familiarization before use. Other drugs are less troublesome.
3. Sedatives and muscle relaxants may precipitate if administered together. The line between the two medications should be cleared. When RSI begins, it is better to have the IV running at a high rate to more effectively clear the line to prevent precipitation. The IV rate should be turned back to baseline when RSI drug administration is completed. It may be more optimal to have two IVs running so that the drugs can be administered in rapid sequence

in two different lines.

4. Paralysis is not a substitute for sedation. Conscious patients remain conscious during paralysis (a frightening experience). It may be safer, however, to avoid sedatives in hemodynamically compromised or unconscious patients.
5. Intubation using a sedative without paralysis has a higher complication rate than RSI.
6. Incorrect weight estimates can result in underdosing or overdosing of drugs and can cause significant delays in the onset of RSI.
7. Avoiding unrecognized extubation and esophageal intubation is critical. Frequent clinical reevaluation, end-tidal CO_2 ($ETCO_2$) monitoring, and pulse oximetry are highly recommended because they often can prevent this serious complication. End-tidal CO_2 monitoring (capnometry) is preferable to $ETCO_2$ color detection because it permits quantitative PCO_2 monitoring, which is useful in patients who are difficult to ventilate or in those who have head injuries and require hyperventilation. $ETCO_2$ color detectors work only for a short period. After 15 minutes or so, moisture renders them nonfunctional; thus, these should not be used for long transports unless they are frequently replaced.
8. Bronchial mainstem intubation can occur easily in small children who have a small degree of ET tube movement.
9. RSI protocols should be reviewed with the ED staff at periodic in-services. A common problem occurs when the person who applies the Sellick maneuver releases it prematurely to tend to a seemingly more important task. This person should be dedicated to this maneuver alone and must not release it until intubation is confirmed.
10. Suction devices may malfunction; therefore, there always should be a back-up available. Standard Yankauer tips may be clogged by food particles, so newer stiff suction tips with larger openings may be more useful.
11. Be prepared for transtracheal ventilation. This may be lifesaving in a patient who cannot be intubated or mask ventilated.

MULTIPLE TRAUMA

A disciplined airway, breathing, circulation (ABC) approach should take priority in the management of multiple trauma victims. Deformities and open wounds must not distract team members from these priorities. The ABC assessment should be made quickly, followed by a brief neurologic assessment (*D* for disability) that specifically checks for signs of ICP elevation.

Airway management must include precautions for a possible C-spine injury. (C-spine immobilization and nasal versus oral tracheal intubation are discussed earlier in this chapter.) Available information suggests that, in most instances, children with multiple trauma who require intubation are best intubated orally using RSI. Succinylcholine may worsen hyperkalemia in patients with severe crush injuries or severe burns. In children with severe craniofacial or airway injuries, oral tracheal intubation may prove to be difficult. In such cases, a surgical airway may be indicated. This can be done emergently in the ED, or it may be done as a standby procedure if oral tracheal intubation fails. (The complications of this procedure are discussed earlier in this chapter.) The child's airway status may worsen during the procedure because of bleeding, agitation, or additional airway trauma. Needle cricothyrotomy with transtracheal ventilation should be available as a standby procedure if further airway difficulties occur.

Breathing is best assessed clinically by auscultation and by checking the patient's color. Arterial blood gas monitoring is useful to measure ventilation, oxygenation, and any occult metabolic acidosis objectively. Pulse oximetry is useful to monitor oxygenation continuously and noninvasively. It is also an indirect monitor of perfusion because it will show a shorter perfusion bar or waveform with pulsation in slightly marginal perfusion states and it will fail to register any signal in poor perfusion states. In patients who require intubation, colorimetric $ETCO_2$ detectors are an inexpensive way of confirming tracheal intubation; however, they are unable to measure $ETCO_2$ reliably. Continuous capnometry is preferable because it permits an ongoing estimate of $PaCO_2$ in most instances. As an exception, $ETCO_2$ measurements often are erroneous in extremely poor perfusion states (during cardiopulmonary resuscitation) because they depend on pulmonary perfusion as well. Maintaining PCO_2 in the 30–35 mm Hg range is beneficial for patients with head injuries because it lowers ICP. Capnometry prevents excessive hyperventilation and reduces the need for frequent blood gas analysis.

Oxygenation may be compromised by pulmonary injuries such as pneumothorax, hemothorax, chest wall injuries, pulmonary contusions, or aspiration. In addition to these, ventilation may be compromised by airway trauma and central nervous system (CNS) depression. Oxygen administration should be considered routine for all multiple trauma victims. Intubation is indicated in patients who have ventilatory compromise, a potential for airway compromise, a head injury that requires hyperventilation, moderate or severe shock, and hypoxemia despite supplemental oxygen. Positive-pressure ventilation may worsen a pneumothorax if a chest tube is not in place.

Circulation should be assessed by using multiple clinical parameters. Early mild shock should be treated aggressively to reverse any progression toward late shock and hypotension, which is associated with a poorer outcome. Acute symptoms and signs of early shock are subtle and often underestimated. These signs and symptoms include agitation, restlessness, lethargy, pallor, delayed capillary refill, coolness of the feet, metabolic acidosis, a short perfusion bar/wave on the pulse oximeter, and difficulty in picking up a pulse oximetry signal. Tachycardia and hypotension are indicators of late, severe shock. Paradoxical bradycardia has been noted in shock with hypotension; therefore, the absence of tachycardia cannot be used to rule out shock. Dismissing a low BP as “normal for a child” because of the absence of tachycardia is not valid.

Sedatives may have significant adverse effects on BP because of myocardial depression or vasodilation. In hypovolemic or hypotensive children, sedative doses should be reduced or avoided, depending on the clinical situation.

The initial treatment of shock related to hypovolemia consists of frequent clinical reassessment and fluid restoration with volume-expanding crystalloids such as normal saline or lactated Ringer's solution. The usual initial bolus should be 20 mL/kg. This is followed by reassessment of clinical shock parameters. This process should be repeated if evidence of shock persists until fluid volume is restored. Red blood cells should be transfused if excessive hemorrhage is sustained.

Large volume fluid resuscitation can result in complications because of hypothermia. When large fluid volumes are anticipated or the patient is very small, crystalloid solutions should be warmed gently and blood products should be infused through a blood warmer.

HEAD TRAUMA

The same priorities should be followed with head trauma as with multiple trauma. Children with head injuries who have depressed sensoriums may be hypoxic, hypovolemic, hypotensive, or acidotic. Although intracranial hemorrhage alone cannot account for significant hypovolemia that results in shock in an older child or adult, this is possible in an infant.

Unconscious patients should be intubated using RSI and should be hyperventilated. Patients responsive only to painful stimuli may need to be intubated as well. Some patients with lesser degrees of sensorium depression also may need intubation using RSI and hyperventilation, depending on the degree of head injury and the rate of deterioration. Patients at risk of ICP elevation should be given thiopental or etomidate in addition to lidocaine pretreatment as part of RSI unless hypovolemia or hypotension exist, in which case the dose should be reduced or eliminated, depending on the clinical circumstances. Thiopental and etomidate lower ICP and cerebral metabolic oxygen demand.

In patients with evidence of ICP elevation, cerebral dehydration may reduce ICP and prevent impending herniation. This can be done using diuretics (furosemide 1 mg/kg) or osmotic diuretics (mannitol 0.25 to 1.0 mg/kg).

Patients with head injuries may develop posttraumatic seizures related to cerebral contusions, cerebral edema, or intracranial hemorrhage. Under RSI, these seizures are not visible because of pharmacologic paralysis. It is prudent in most instances to give a loading dose of phenytoin (10 to 20 mg/kg) after intubation is confirmed to treat prophylactically any undiagnosed posttraumatic seizure focus.

BURNS

Children with severe burns represent a special form of multiple trauma. Burn patients may also sustain blunt trauma, and ABCs are still the priority. Early intubation using RSI is advocated for patients at risk of airway injury because airway edema is expected to worsen rapidly. These cases include children with evidence of soot in sputum or vomitus, burns of the face, singed nasal hairs, lip burns, wheezing, stridor, or severe burns. The possibility of carbon monoxide poisoning should be considered. It may be preferable to avoid succinylcholine in patients with severe burns for fear of worsening hyperkalemia.

Pulmonary compromise may result from smoke inhalation, burn injury, bronchial edema, bronchospasm, blunt trauma, or adult respiratory distress syndrome (ARDS). Initial chest radiographs may fail to show some of these injuries.

Children with severe burns have significant hypovolemia because of external fluid losses. Lactated Ringer's or normal saline boluses of 20 mL/kg should be used to immediately correct hypovolemia. Guidelines for fluid replacement based on body surface area (BSA) of the burns provide a means to estimate fluid requirements. The Parkland formula recommends 4 mL/kg (3 mL/kg if BSA is less than 30%) of lactated Ringer's solution for each BSA percent burn, required over the next 24 hours in addition to maintenance fluids. These guidelines require careful monitoring of urine output, gastric fluid output, and the subtle parameters that may identify early shock. Fluid boluses may be required to reverse any trend toward shock to restore circulatory volume immediately. Burns exude high-sodium and high-protein fluids, resulting in subsequent hyponatremia and hypoproteinemia. Syndrome of inappropriate secretion of ADH may develop in patients with cerebral injury, pulmonary injury, or prolonged bed rest. These factors should be considered when determining the initial fluid management.

Hypothermia caused by body exposure and fluid administration must be anticipated and prevented. The use of radiant warmers and the warming of IV solutions can prevent this complication.

Circumferential burns around an extremity may result in additional swelling and venous and arterial occlusion. Infarction of the extremity distal to this injury can be prevented by early recognition of this fact and surgical release of the constriction. Similarly, circumferential burns about the chest may result in chest wall constriction and respiratory difficulties.

STATUS EPILEPTICUS

In patients presenting to the ED with prolonged seizures, the standard approach of ABC support and immediate administration of benzodiazepines is generally initiated. Loading with IV phenytoin and/or phenobarbital may also be considered if seizures continue. This process of administering anticonvulsants and waiting to assess its effect occurs in 5- to 15-minute cycles. If seizures fail to respond to several doses of anticonvulsants, the child could be continuously seizing for an additional 30 to 60 minutes.

Prolonged seizures result in hypoxia and respiratory acidosis due to poor ventilation. The brain is simultaneously hypermetabolic with greater oxygen demand. RSI using thiopental effectively reverses this process. Skeletal muscle activity stops. Oxygenation and ventilation are restored. The brain may still be hypermetabolic because it may still be epileptogenic, but at least the patient is no longer hypoxic and acidotic. Thiopental has potent cerebroprotective and anticonvulsant activity. Simultaneous administration of high dosages of benzodiazepines, phenytoin, and phenobarbital can potently reduce the seizure potential of the brain while maintaining oxygenation and ventilation. The major adverse effect of most anticonvulsants is respiratory depression. Following RSI, this is no longer a concern and maximum doses of anticonvulsants can be administered to provide maximum anticonvulsant activity.

In refractory status epilepticus, the duration of seizures, hypoxia and acidosis is likely to contribute to cerebral injury. At

some point in the management of status epilepticus (e.g., 60 minutes), the failure of conventional anticonvulsants should be recognized and RSI followed by maximum anticonvulsant administration should be considered. Because potentially injurious seizure durations of 40 minutes or more can occur in the management of such patients, it has been my preference to initiate RSI earlier rather than later. The concern that seizures can no longer be visibly appreciated after RSI should be tempered by the clinical benefits of RSI as described. EEG monitoring in the ED is often recommended, but it is not feasible in most hospitals.

AGITATED PATIENTS WHO REQUIRE PROCEDURES OR TRANSPORT

Agitated or combative patients with head injuries or possible intracranial lesions that require computed tomography (CT) scanning cannot be scanned in such a condition. Sedation alone can be considered for such patients. In patients who fail to respond to standard sedation measures or in patients who may benefit from tracheal intubation, RSI provides an effective means of immediately securing airway control, breathing, and movement so that imaging can be completed with minimal trauma to the patient. Patients who require transport to another facility and who are agitated and hard to control may be difficult to manage during transport. After the clinical situation has been assessed, RSI may be indicated if its benefits outweigh its risks. Patients are more difficult to monitor during procedures and transports. The immediate detection of unrecognized extubation or hypoxemia is crucial. Portable pulse oximeters and $ETCO_2$ monitors can monitor oxygenation and confirm intubation continuously.

Suggested Readings

Atherton GL, Johnson JC. Ability of paramedics to use the Combitube™ in prehospital cardiac arrest. Ann Emerg Med 1993;22(8):1263–1268.

Bergen JM, Smith DC. A review of etomidate for rapid sequence intubation in the emergency department. J Emerg Med 1997;15(2):221–230.

Criswell JC, Parr MJA. Emergency airway management in patients with cervical spine injuries. Anaesthesia 1994;49:900–903.

Gerardi MJ, Sacchetti AD, Cantor RM, et al. Rapid-sequence intubation of the pediatric patient. Ann Emerg Med 1996;28(1):55–74.

Gnauck K, Lungo JB, Scalzo A, et al. Emergency intubation of the pediatric medical patient: use of anesthetic agents in the emergency department. Ann Emerg Med 1994;23(6):1242–1247.

Ivy ME, Cohn SM. Addressing the myths of cervical spine injury management. Am J Emerg Med 1997;15(6):591–595.

Lev R, Rosen P. Prophylactic lidocaine use preintubation: a review. J Emerg Med 1994;12(4):499–506.

Ruchs-Buder T, Tassonyi E. Intubating conditions and time course of rocuronium-induced neuromuscular block in children. Br J Anaesth 1996;77:335–338.

Scannell G, Waxman K, Tominaga G, et al. Orotracheal intubation in trauma patients with cervical spine fractures. Arch Surg 1993;128(8):903–905.

Scheulen JJ, Munster AM. The Parkland formula in patients with burns and inhalation injury. J Trauma 1982;22:869–871.

Silverman BK, Haller A, Johnston C, Luten R, Singer J. APLS: The Pediatric Emergency Medicine Course. American Academy of Pediatrics and American College of Emergency Physicians, 1993.

Stanwood PL. The laryngeal mask airway and the emergency airway. J AANA 1997;65(4):364–370.

Staudinger T, Brugger S, Watschinger B, et al. Emergency intubation with the Combitube™: comparison with the endotracheal airway. Ann Emerg Med 1993:22(10):1573–1575.

Tobias JD. The laryngeal mask airway: a review for the emergency physician. Pediatr Emerg Care 1996;12(5):370–373.

Tryba M, Zorn A, Thole H, Zenz M. Rapid-sequence orotracheal intubation with rocuronium: a randomized double-blind comparison with suxamethonium—preliminary communication. Eur J Anaesth 1994;11(suppl 9):44–48.

Walls RM. Lidocaine and rapid sequence intubation. Ann Emerg Med 1995;27(4):528–529 (Letter).

Walls RM. Rapid-sequence intubation comes of age. Ann Emerg Med 1996;28(1):79–81 (Editorial).

Yamamoto LG. Rapid sequence anesthesia induction and advanced airway management in pediatric patients. Emerg Med Clin North Am 1991;9(3):611–638.

Yamamoto LG, Kim GK, Britten AG. Rapid sequence anesthesia induction for emergency intubation. Pediatr Emerg Care 1990;6(3):200–213.

Zemenick RB. No support for intravenous lidocaine airway reflex suppression during rapid sequence intubation. Ann Emerg Med 1995;26(5):660 (Letter).

CHAPTER 6

Prehospital Care

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Fortunately, childhood is a period of life that generally is free of critical illnesses and injuries. In large populations, however, such cases are commonly seen. Injuries (both intentional and unintentional), acute medical diseases, and acute exacerbations of chronic disease afflict the pediatric population. Although prevention is the ultimate “treatment” for many of these disorders, it is inevitable that a substantial number of emergencies will continue to affect children in all geographic locations, emphasizing the need for appropriate pediatric prehospital care services.

In response to the need for such services, governmental efforts have been initiated to help states improve their emergency medical services for children (EMS-C). Eloquently summarized by Ludwig and Selbst, EMS-C is an all-encompassing, multidisciplinary system that includes parents, primary care providers, prehospital care providers and transport systems, community hospital and tertiary care referral center emergency departments, and pediatric inpatient units, including critical care facilities. Ideally, the elements of this system should be linked by effective communication and transportation systems and governed by well-established policies and procedures. The provision of prehospital emergency services, although a single link in this chain, is a critical component. This chapter familiarizes the reader with the structure of and services provided by these systems and their inherent limitations. There is great need for physician involvement in many aspects of prehospital care; we hope to stimulate involvement through better understanding of these services.

HISTORY OF EMERGENCY MEDICAL SERVICES AND PREHOSPITAL CARE

The concept of EMS was born centuries ago in the battlefield care of the sick and wounded. The roots of modern ambulance services reach back at least as far as the Napoleonic wars, when covered carts were used to carry casualties to rear dressing stations. Subsequent world conflicts have resulted in rapid advancements in medical care. The development of prehospital care as a specialty, however, is a more recent development. Arguably, this can be traced to the formation of formal EMS systems, which took place in the early 1970s.

In 1973, the critical components of EMS systems were outlined in the Emergency Medical Services Act (PL-93-154), which was designed to develop regional EMS systems. This act outlined 15 components that an EMS system had to address to receive federal funding. Notably absent from this list was the issue of medical control (i.e., physician involvement). Subsequently, Roush and McDowell have suggested that effective EMS systems should include the following elements: medical direction, prehospital transport agencies, interfacility transport agencies, dispatch, communications, protocols (for triage, treatment, transport, and transfer), receiving facilities, specialty care units, training, financing, auditing and quality assurance, public information and education, mutual aid, and disaster plans. Optimal function of this system requires uniformity of purpose and communication between all components, including prehospital care. The multiplicity of goals and functional independence of many of these components occasionally interferes with the service provided. Clearly, administrative streamlining and improved organization of EMS services are major challenges that lie ahead.

Although prehospital care and EMS systems have existed for more than 25 years, only recently has attention been focused on issues related to pediatrics. The development of pediatric emergency medicine as a specialty has highlighted the need for a greater emphasis on improvement of EMS systems for children. Pediatric emergency medicine specialists are increasing not only in numbers but also in sophistication, with the latter reflected by the improved level of medical care offered and the depth of research produced.

Many contributions have been made by pediatric researchers in terms of prehospital care issues that affect pediatric patients. These studies have helped elucidate the strengths and weaknesses of prehospital care systems' pediatric services. Studies by Seidel et al. have demonstrated the lack of readiness many prehospital care systems possess to manage childhood emergencies adequately. On the other hand, Scribano et al. have demonstrated overuse of both invasive and noninvasive interventions by prehospital personnel when transporting children. Research efforts such as these reveal the deficiencies in pediatric prehospital practice and the need for further evaluation of these issues.

Epidemiologic studies also have helped clarify the pediatric needs of EMS systems. Tsai et al. found those at the pediatric age extremes to be the principal users of prehospital services: teenagers for trauma and infants and preschoolers for illness (primarily seizures, ingestions, and respiratory diseases). Yamamoto et al. showed that the handicapped were also more likely to use an ambulance; however, ambulance personnel felt less prepared to handle those patients. In a survey of how pediatric practitioners used EMS services in Pennsylvania, Baker and Ludwig found that infants with serious illness were more likely to be transported by EMS services than were adolescents. The combination of these findings and the fact that most childhood medical arrests occur in children under four years of age highlights the need of EMS systems to provide age-appropriate equipment and personnel properly trained in pediatric emergency care. For the youngest children, these services often have been found to be lacking.

Even when appropriate EMS systems are available, they might be significantly underused. In the study by Baker and Ludwig, although 93% of practitioners surveyed had some form of professional transport services available, 54% most often chose to transport ill children to tertiary care centers in family automobiles, unaccompanied by advanced life support personnel or equipment. This was true even in the case of suspected epiglottitis. It is clear that further education is needed for EMS providers and EMS users alike regarding pediatric issues. However, education should be provided that has been properly studied and demonstrated to be effective. Although several studies have been conducted to evaluate the effectiveness of classroom training in increasing the knowledge and confidence of prehospital personnel in the management of acutely ill and injured children, few, if any, have included outcome measures of actual performance.

EMS-related research, although plentiful, suffers from general lack of rigor. Callaham has noted that of the 5842 publications on prehospital EMS that appeared in Medline from 1985 to 1997, only 54 were randomized controlled trials. Of the 54, only 7 randomized controlled trials showing a positive outcome of the intervention were uncontradicted. Only one of those examined a major outcome such as survival, and only one compared the intervention with a placebo and could therefore evaluate the efficacy of EMS itself. This paucity of scientific scrutiny of EMS highlights the need for future research focusing on EMS practice. In the meantime, we must resist the temptation to quickly add new technologies, procedures, and protocols to prehospital care without ensuring that these modalities have proven efficacy. There is a real concern that in an effort to aid one patient, many others will suffer from unnecessary intervention. Furthermore, additional interventions must be balanced against the need for rapid transport to the hospital described classically as scoop and swoop versus stay and play.

SERVICES AND GOVERNANCE OF PREHOSPITAL CARE SYSTEMS

The intentions of most prehospital care services are to provide immediate medical care to ill or injured persons who are outside of the hospital setting and to transport these patients to appropriate medical facilities for physician evaluation and management. Although prehospital care and other EMS components have improved significantly since the early 1970s, the capabilities of these components vary on a regional basis. In all 50 states, EMS legislation exists, providing a statutory basis for statewide systems. Under these laws, state agencies can set forth regulations that carry the weight of the law. Because EMS systems often are regionally based, however, municipal agencies ultimately govern the extent of services that can be provided by prehospital care personnel. Furthermore, many jurisdictions place restrictions on procedures that prehospital care providers may perform on children.

All states have identified lead agencies that provide coordination of EMS activities and purpose within the state. In most states, the lead agency is headed by an EMS director who reports to the state department of health. Often, state-level advisory councils exist to aid in the development of protocols and minimum standards of operation.

States often are divided into EMS regions, at which level prehospital care becomes operational and local government, hospitals, and ambulance services interact with each other. At this level, regional advisory councils exist as well. Physicians should be encouraged to become involved in these regional committees as advocates of the pediatric needs within their systems. The American Academy of Pediatrics (AAP) prehospital care committee of the section on emergency medicine has prepared an informative resource manual that addresses pediatric prehospital care issues. Agencies, educational programs, and other available avenues of physician involvement are outlined in this manual.

COMPONENTS OF PREHOSPITAL CARE SYSTEMS

Prehospital care is a multicomponent system that involves various personnel and equipment, both of which have undergone remarkable changes over the past 20 years. To understand the extent of the services provided by prehospital care systems, it is essential that physicians who interact with these systems understand the training and capabilities of prehospital personnel and the equipment available to them.

Prehospital Personnel

Prehospital personnel have not always been trained specially to provide the care that their patients require. It was not until 1964 that the first reports of attempts to train fire and police personnel in basic cardiopulmonary resuscitation (CPR) appeared in the literature. Since then, several classifications of prehospital care providers have emerged, each with different levels of training and varying degrees of capabilities. Three general categories of prehospital personnel exist (Table 6.1): first responders, basic life support (BLS) providers, and advanced life support (ALS) providers. Training standards and requirements for certification exist for all of these groups. Federal standards often are superseded by more precise state or local training prerequisites for practice. Many jurisdictions either offer to or require of the providers various supplemental training modules. As a result, many intermediate levels of providers have evolved, with variable capabilities.

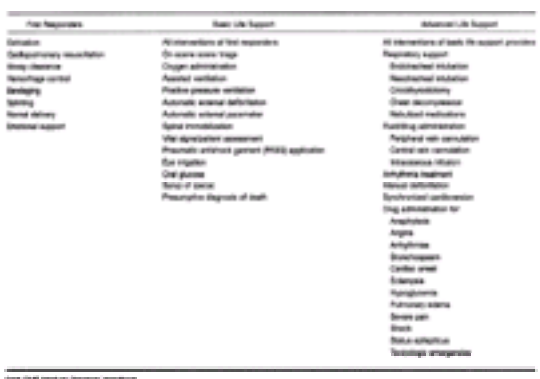


Table 6.1. Categories of Prehospital Care Providers and Their Capabilities

First Responders

The true definition of a first responder is the person who happens on the scene first to provide patient aid. In the context of classification of capabilities, however, *first responder* is a generic term that refers to a large group of persons who have limited but significant lifesaving capabilities. A national first responder course with a standardized 40-hour curriculum has been developed. Eisenberg et al. have demonstrated that properly trained community-based responders can shorten the time to CPR significantly and can enhance resuscitation outcome.

Although the exact capabilities of first responders might vary according to local standards, most are trained to help clear an obstructed airway, to provide lifesaving hemorrhage control and limb-preserving splinting, and to administer BLS while awaiting the arrival of ALS personnel. Spinal immobilization usually is beyond the capabilities of most first responders. First responders generally do not provide patient transport in ambulances; however, in some rural systems, first responders might replace emergency medical technicians (EMTs) as initial ambulance personnel.

Basic Life Support Providers

BLS providers are trained to give all the services of first responders, as well as to evaluate the severity of a patient's condition, provide other simple interventions, and transport patients. In different locations, alternative titles for BLS providers might exist; however, many refer to these personnel as EMTs.

The capabilities of EMTs exceed those of first responders. EMTs are capable of patient assessment, spinal immobilization, ventilatory assistance (exclusive of intubation), and application of pneumatic antishock garment (PASG). In addition to first responder recognition of pulselessness, apnea, upper airway obstruction, and extremity deformity, BLS providers are trained also to recognize respiratory distress, altered mental status, shock, mechanisms of injury, and obvious death. Initial patient triage and routing to receiving medical centers is another crucial responsibility of BLS providers.

For first responders, standardized EMT courses exist that provide 100 hours or more of didactic and clinical instruction. A basic curriculum, which often is expanded further, has been developed by the U.S. Department of Transportation. Occasionally, additional training modules are available to persons who have achieved such training. Satisfactory completion of this supplemental instruction might result in advancement and designation to an intermediate level of provider status. For example, many areas now offer EMT training in defibrillation. Once certified in this technique, the provider often is called an EMT-D.

Advanced Life Support Providers

More advanced than BLS providers are those who are trained to provide ALS. As previously stated, BLS providers often can advance their skills and levels of expertise in modular training formats. Certification exists for progressive skill levels in vascular access, respiratory support, and drug administration. Many intermediate levels have been created between BLS and ALS providers.

Just as BLS providers often are referred to as EMTs, so are ALS providers often called paramedics. The most intensively trained paramedics have more than 400 hours of training and are capable of administering advanced resuscitation in the field. Their capabilities include advanced general diagnostic skills, recognition of arrhythmias as well as drug and electrical conversion of arrhythmias, and advanced airway management (including endotracheal intubation). In many locations throughout the United States, field ALS providers have significantly decreased mortality following cardiac

arrest.

Intermediate Levels of Providers

Numerous intermediate levels of providers have been mentioned other than the basic three groups already described. Many such provider classifications developed in response to specific needs of local jurisdictions, regions, or states. For example, some localities might have found that patient outcome could improve by adding training in peripheral venous access to the basic EMT curricula. As a result, this and many other training permutations have been developed.

Many negative aspects of these hybrid programs have been realized. Intermediate-level designees often focus their efforts on learning interventions that (independently) are of little value or are easily forgotten because of infrequent use rather than concentrating on perfecting basic clinical skills. It is incumbent on the local and state medical directors to review existing and new interventions constantly and to make thoughtful decisions regarding additional training requirements of intermediate-level providers.

Equipment and Modes of Transport

Ground Transport

Napoleon's chief surgeons are credited with providing the concept from which modern ambulances developed. From these original covered carts to the most sophisticated mobile intensive care unit of the 1990s, many changes have occurred. Before the 1970s, most changes concerned the types of vehicles used as ambulances. Up until that time, ambulances were little more than taxis, sparsely equipped and staffed, at best, by first-aid-capable personnel. In 1969 and 1973, the National Academy of Science and the U.S. Department of Transportation published documents that generally defined the purpose of an ambulance and its contents. In 1974, the Department of Transportation and General Services Administration published another document (KKK Standards) that provided engineering specifications that incorporated the general recommendations of the ambulance design criteria. Although KKK standards have been amended, they remain the specifications by which federal agencies purchase ambulances. In 1983, the National Highway Transportation Safety Administration published minimum standards for ambulances not purchased by federal agencies but using federal highway safety funds. According to these guidelines, ambulances must comply with the essential equipment list established by the American College of Surgeons, have space for two litter patients, have 60 inches of head room for EMTs, be manned by two basic-level EMTs, and have adequate exterior lighting. No mention was made of capabilities for ALS or communication with medical command. As a result, many states have established more comprehensive standards.

Three types of ground transport vehicles generally are available today. Type I is a conventional truck cab chassis on which a modular transferable ambulance body is mounted. Type II is a standard van, the body of which is converted to conform with ambulance standards. Type III is a specialty van with a unitized cab and body or with a cab-over-engine chassis to which a modular transferable ambulance body is mounted.

Ambulances also are categorized according to their equipment and staffing capabilities. BLS units should be equipped to conform at least to the list of “Essential Equipment for Ambulances” published in 1983 by the American College of Surgeons (Table 6.2). Included in this list are ventilation and airway equipment, immobilization devices, bandages, pneumatic antishock garments, two-way communication equipment, obstetric kits, extrication equipment, and other miscellaneous items.

Table 6.2. Required Equipment and Supplies for Ambulances Providing Basic Life Support

In 1988, the American College of Emergency Physicians (ACEP) published a position paper that detailed staffing and equipment appropriate for ALS units (Table 6.3, Table 6.4 and Table 6.5). In addition to most of the equipment contained in the BLS list, these ALS units are recommended to carry intubation and vascular access equipment, a portable battery-operated monitor-defibrillator, and a variety of ALS medications.

Equipment	Burn pack—standard pack; towels or gel burn sheet acceptable
Oxygen tank with tubing, with humidified source for long transport times	Thermal absorbent blanket
Oral airways—sizes 0–5	Equipment sizing tape or equipment/age/weight chart
Nasopharyngeal airways with lubricant—12–30 Fr or equivalent sizes in mm	Heat source for ambulance
Self-inflating bags with oxygen reservoir—250-, 500-, 1000-mL bags	Supplies
Oxygen masks—infant, child, and adult	Adhesive tape
Nasal cannulae—infant, child, and adult	Alcohol sponges
Masks for bag-valve-mask—infant, child, and adult sizes 1–3	Arm boards—various sizes
Stethoscope	Povidone-iodine prep pads
Blood pressure cuffs—infant, child, and adult	Elastic bandages
Portable suction unit	Extra batteries and bulbs for equipment needs
Suction catheters (flexible and rigid)—6–14 Fr	Flashlight, bulb, and batteries
Backboard for spinal immobilization—short and long board neck immobilization	Gauze rolls
Towel rolls, blanket rolls, or equivalent	Gauze sponges
Rigid cervical collar (for children more than 2 years of age—infant, child, small, medium, and adult)	Protective eyewear, gloves, and masks
Femur splint—designed for pediatric patients	Scissors
	Tincture of benzoin
	Tongue blades

Table 6.3. Basic Life Support: Minimum Pediatric Equipment and Suppliers

Equipment	3-way stopcocks or adapter that allows administration of additional fluids or medications
BLS minimum equipment and supplies, plus	Syringes—various sizes
Monitoring	Blood sample tubes
Transport monitor—battery-operated with 3 or 4 lead wires	Intravenous tubing, Buretrol
Defibrillator with ECG and 8-cm paddles (or paddle adapter) and pads capable to dial down to appropriate watt-sec for pediatric patients (when replacing current equipment, new equipment should have settings below 20 watt-sec)	Tuberculin syringes
Monitoring electrodes	Intravenous Solutions
Equipment and drug dosage tape or age weight chart	Normal saline or lactated Ringer's
Glucose detection strips	D5W (diluent)
Airway	Sodium chloride—bacteriostatic, for injection
Laryngoscope handle with extra batteries and bulbs	Water—bacteriostatic, for injection
Laryngoscope blades—straight and/or curved, 0, 1, 2, 3	Medications/Concentrations
Stylettes—pediatric sizes	Atropine sulfate—0.1 mg/mL
Endotracheal tubes	Bicarbonate, sodium—4.2% (1.0 mEq/mL)
Uncuffed range 2.5–6.0	Diazepam or anxiolytic choice—5 mg/mL
Cuffed range 5.5–8.0	Epinephrine—1:1000 (1 mg/mL)
Magill forceps (Pediatric)	Epinephrine—1:10,000 (0.1 mg/mL)
Lubrication (water soluble)	Lidocaine hydrochloride (20—10 mg/mL)
Nasogastric tube—sizes 8–18 Fr	Naloxone hydrochloride (adult)—1.0 mg/mL
Vascular Access	Pain medication per medical control
Intravenous catheter of choice—16–22 gauge	D50 (dextrose 50 + water and diluent)
Intraosseous needles of choice	Inhaled β-adrenergic agent of choice
Tourniquet/rubber bands	Activated charcoal

Table 6.4. Advanced Life Support: Minimum Pediatric Equipment and Medications

Basic Life Support (BLS) for Newborn
Oxygen cylinder
Stethoscope
Bulb syringe
Portable suction
Suction catheters—5–10 Fr range
Resuscitation bag—(750 mL (250 or 500 mL)
Face mask (infant)—premature and newborn sizes
Gauze
Sterile scissors
Thermal absorbent blanket and head cover
Cord clamps
Appropriate heat source for ambulance compartment
Advanced Life Support (ALS) for Newborn
BLS equipment for newborn as listed above, plus:
Endotracheal tube uncuffed—3.0–4.0 mm
Endotracheal tube stylet—6 Fr
Laryngoscope—straight blades 0 and 1
Infusion set, microdrip unit

Table 6.5. Basic and Advanced Life Support: Minimum Resuscitation Equipment and Supplies for the Newborn

Pediatric Equipment

It has only been recently that the prehospital care needs of the pediatric patient have been addressed formally. When formulating equipment lists for pediatric prehospital emergencies, it is tempting to be all-inclusive. This desire should be tempered by the limitations of budget and carrying space. Because of the tremendous variability in pediatric-appropriate equipment carried by EMS agencies, an attempt was made to establish minimum standards for pediatric equipment (Table 6.6). The equipment listed centers mostly around airway, circulation, and immobilization needs. More technically sophisticated equipment can be added if patient needs and provider expertise warrant.

Equipment	Quantity
Airway[b]	
Suction catheter (for tracheostomy care)	
6.0 Fr	2
8.0 Fr	2
Ventilation	
Face mask	
Newborn	1
Infant	1
Child	1
Bag-valve-mask with pop-off valve	
500–750 mL	1
Oxygen delivery mask—partial rebreathing	
Infant	1
Child	1
Spinal immobilization	
Short board	1
Sand bag (5–10 lb)	4
Circulation[c]	
Cardiac monitor (with pediatric paddles approximately 20 cm²)	1
Blood pressure cuff	
Infant (6–8 cm)	1
Child (8–10 cm)	1
IV catheter (over the needle)	
22-gauge	[illegible]
24-gauge	[illegible]
Miscellaneous	
Short-arm board	2
Long-arm board	2
Bulb suction	1
Cord clamp	2
Sterile scissors	1

[a] Luten R, Foltin G, eds. Pediatric Resources for Prehospital Care. American Academy of Pediatrics, 1989:42–46.
[b] Consider oral/pharyngeal airways, equipment for endotracheal intubation.
[c] Consider intraosseous needle, infant and toddler [illegible]; IV catheter recommendations assume larger size catheters (20 gauge and greater) are available.

Table 6.6. Minimal Standard for Pediatric Equipment, 1989 (ALS Units)[a]

Ground Vehicle Staffing

Although vehicle staffing configurations vary by locality, the most common configuration consists of two persons trained at least to the level of EMT-ambulance. The ACEP position paper of 1988 outlines the minimum capabilities of providers who staff ALS units. Such personnel should have BLS as well as ALS skills and should be able to recognize, assess, and initially manage medical and trauma emergencies alike. In an ACEP (1989) publication that discussed the principles of EMS systems, Roush estimates that nine EMTs are needed to provide 24-hour staffing of one EMS vehicle and also suggests that five of them should be paramedic level if the service offers ALS.

In many geographic areas, the medical needs of the population served demand specialty transport services. Such services can be either ground-based or air-based. Examples of ground specialty transport units include mobile coronary care units, neonatal transport units, high-risk obstetrics units, and mobile intensive care units. For each of these, specialized equipment and staffing are incorporated.

Air Transport

Air medical transport had been envisioned by many immediately after the first successful flight of the Wright brothers in 1903. Again, however, the demands of war made air medical transportation a reality. The Korean and Vietnam wars provided tremendous experience with this prehospital service. Reduced mortality of battlefield casualties was attributable largely to the efficiency of air medical transport.

The gradual introduction of air medical transport into civilian prehospital care began in the 1960s. During that time, a National Academy of Sciences National Research Council document recommended the initiation of pilot programs to evaluate ground and air ambulance services in sparsely populated areas. During the 1970s and 1980s, air medical transport developed into a common part of EMS services.

Regardless of their degree of involvement, physicians should be aware of the air medical transport services in their region. Two basic types of air transport exist: helicopter and fixed-wing. Helicopters (rotor craft) are used more commonly in densely populated areas. Their unique capabilities of rapid, direct scene response give these craft a distinct advantage over fixed-wing craft. Helicopter services have allowed for the provision of ALS services to larger rural areas incapable of sustaining independent ALS units and have provided access to tertiary care centers for patients in regions without such centers. The second type of craft, fixed-wing, has the advantage of being smoother, quieter, faster, and more spacious. These craft are limited, however, by their need for runway facilities, and they are incapable of scene access.

Air medical transport is a specialized service and is an adjunct to and not a substitute for ground transport. Proper provision of air medical services requires specialized equipment and staffing, both on the ground and in the air. Physicians and transport personnel knowledgeable in flight physiology and experienced in working within the confines of cramped cabins are essential components of these systems. Air transport services carry higher safety risks and higher operational costs than their ground counterparts.

ACCESS TO EMERGENCY MEDICAL SERVICES FOR CHILDREN

For the EMS-C system to provide rapid prehospital care to an acutely ill child, the system must be accessed by a person who recognizes that the child needs emergency treatment. Children rarely access the system themselves; this decision usually is made by a parent, a bystander, or a primary care physician. Because it is a parent who is most often with the child when an emergency happens, it is imperative that the parent be able to recognize a child with an emergency illness or injury. Although most parents do recognize signs and symptoms of obvious life-threatening illness (e.g., seizures, respiratory distress), other emergencies with more subtle symptoms (e.g., dehydration, toxic ingestions) may be missed. However, parents may place undo emphasis on a relatively common symptom, such as fever, and activate the EMS-C system to transport a well-appearing child with fever rather than call the child's physician. If a child is involved in a severe accident when the parent is not present or when the parent is also involved in the accident, the responsibility of accessing the EMS-C system is with a bystander. Proper education directed toward recognizing emergencies and knowing how and when to access the system must be directed not just at parents and caretakers, then, but to all members of the community.

In the event that the parent calls or visits the primary care physician concerning the child's illness, the physician must then direct emergency care. The physician should be able to perform a rapid assessment of the child's condition and, if needed, initiate BLS. Depending on the nature of the illness or the injury, the physician may direct the transport of the child to an emergency facility (either by the parents or by activation of the EMS-C system). It is important that the physician take responsibility for the child's transport and, in some cases (e.g., acute epiglottitis), accompany the child by the safest, most expedient fashion.

MEDICAL COMMAND

Medical command is the entity primarily responsible for the overall medical supervision of the emergency medical system in a community. In general, medical command is composed of a group of base stations—facilities integrated into the paramedic program—with paramedics assigned to that facility as their operational base. Most base stations are hospitals or specialized fire department stations. A medical control facility is a hospital that provides direct medical control for advanced prehospital treatment and is responsible for designation of a medical director, a nurse liaison, and an administrative liaison officer to support a paramedic program. Medical control can be achieved by off-line direction of paramedics and other field personnel through training, provision and monitoring of protocols, and data collection and evaluation and by on-line medical consultation by base station physicians with emergency medical technicians and paramedics. This control is exercised on three distinct levels: 1) the EMS director, who has an off-line, systemwide

administrative role; 2) the base station hospital medical director, who has an off-line regional administrative role; and 3) the base station hospital emergency physician, who has an on-line physician supervisor role directing the care of individual cases in the field.

The EMS director can be an individual person or a group of representatives from areas hospitals, which comprise a Physician Advisory Board. This advisory board has the advantage of representing a consensus of opinion that may be more acceptable to system participants and local politicians. Furthermore, the supervisory work can be distributed among several physicians. However, the bureaucracy often associated with an advisory board may result in a delay in implementation of important policies.

Medical control systems for EMS in the United States are diverse, predominantly because of the various circumstances under which each system operates. Local geographic and medical constraints, financial restrictions, and experience of available personnel affect the extent of the regional system. Nevertheless, any system design for prehospital care should include the basic components described in the following sections.

Off-Line Medical Control

A fundamental requirement of an EMS system is that an off-line medical director be responsible for the planning, implementation, operation, and monitoring of the system. Essential components of the system include development of guidelines for prehospital care; establishment of protocols for patient treatment, triage, and transfer; development of minimum standards for participating facilities; and development of standards for certification of field personnel. In EMS systems with more than one base station, it also is necessary to have an off-line medical director of a regional base station who coordinates the prehospital care in the assigned region. This regional director is the medical liaison with other receiving hospitals in the region.

On-Line Medical Control

The management of individual cases by field personnel is the responsibility of the emergency physician on duty at the centralized medical control facility or at the regional base station. After patient assessment at the scene, field personnel follow operational protocols and then contact the on-line medical control physician for voice authorization of certain ALS procedures. This system allows immediate medical consultation and adjustment of treatment protocols for each case. To be most effective, the on-line physician must be experienced in efficient evaluation of the patient's emergent medical needs. Often, on-line medical command is associated with longer on-scene time and an infrequent physician-directed deviation from the written treatment protocols. Clearly, effective medical control rests on a close and trusting relationship between medical control physicians and field personnel.

An important component of on-line medical command is clear communication between the field personnel and the base station. Typical radio systems allow only one person to speak at a time, which may delay communication. Landline telephones allow both parties to speak at the same time and thereby, enhance the transfer of information. Furthermore, telephones have more clarity than radio communications. Clearer communication has been associated with shorter call duration. In the future, we can anticipate conversion of voice, data, and other communications to digital or other more modern technologies. These changes will make computerization universal and lead to more effective processing of information.

MECHANICS OF THE SYSTEM

The essence of an efficient EMS-C system depends on quick and reliable communication systems (Fig. 6.1). If the parent is unable to access the system, treatment will be delayed. Regardless of how good the receiving hospital services are, if the system cannot get the child to the hospital in a timely fashion, outcome is affected. Most regions with centralized emergency aid have a communications center that receives all calls and coordinates emergency response. This center may be located in a central city agency, such as the police or fire headquarters, or it may be located in a hospital facility. The goal of the communication center is to receive all distress calls, identify the type of emergency need, and route immediate BLS (and if necessary ALS) personnel to the scene. Although the mechanics of the EMS-C response vary from region to region, many components are common to the prehospital phase.

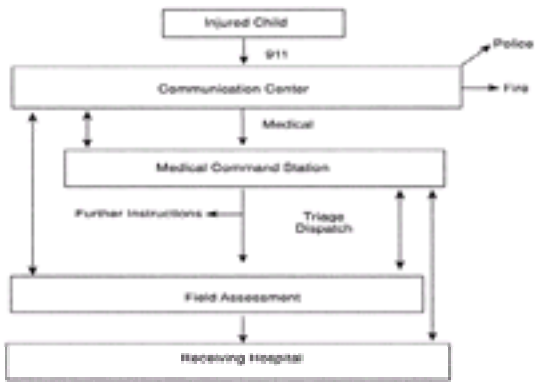


FIGURE 6.1. EMS-C communication system diagram.

Initiation of Emergency Medical Services for Children

Although the parent can use many methods to initiate an EMS response, the telephone is the easiest and most common. A major difficulty in accessing the system occurs when the parent is not sure who should be called—the primary care physician, the police, the fire department, or a nearby hospital. Faced with this dilemma, the parent often calls the operator at the telephone company, which delays the EMS response because the operator then must transfer the call into the regional emergency system. Furthermore, telephone operators are not trained in appropriate triage of calls, leaving room for errors in judgment that may affect patient outcome.

In an attempt to institute a single, coordinated telephone access system, most regions have adopted one easy-to-remember telephone number (911) to access a response for any emergency. In fact, some areas are using an enhanced 911 system that will automatically provide the dispatcher with the caller's name and telephone number. For this to be effective, residents in the region must be taught how to then access EMS. In addition, care must be taken to emphasize the fact that “911 must be saved for the real thing.” Depending on the local resources for accessing the health system, residents may become accustomed to using “911” for transport of nonemergent medical problems that may lead to unnecessary delays in response to other cases of life-threatening illness.

Receiving

In a developed, centralized EMS system, the distress call is made directly to a communication center. A trained operator at the communication center then routes the call to an operator at the police, fire, or medical center. These specialized operators then gather essential information concerning the nature of the emergency and relay the location of the child and the triage complaint to the EMS system for dispatch of a first responder. This first interaction of the parent and the communication system perhaps is the most crucial. In a matter of seconds, the medical operator must calm the caller, extract the complaint, make the triage decision, and obtain the address. While the EMS unit is being dispatched, the operator continues to talk with the caller. At this point, the operator usually uses computerized algorithms to provide the caller with additional basic lifesaving instructions.

Dispatch

When the distress call is received, a first responder is sent to the scene immediately. The first responder is the closest personnel (police, fire, EMT, or paramedic) with BLS training. Based on the severity of the complaint, additional responders (usually paramedics with ALS training) will be dispatched. In the event of a medical disaster, multiple response teams, including police, fire, and medical personnel, are activated.

Field Treatment

The first responder begins BLS techniques until the arrival of more experienced medical personnel. When the paramedics arrive, ALS is begun using established protocols and, if available, on-line medical command. The communication center and the medical command center then decide on the closest, most appropriate hospital for treatment. The hospital selected is based on various factors, including bed availability and critical care, trauma, or burn management capabilities. A severely injured child may be transported to a pediatric level 1 trauma center, a level 1 trauma center, or a community hospital, depending on the relative time available to transport the child and the extent of injuries.

Completion of Prehospital Service

Once the child is en route to the receiving hospital, medical command then notifies the receiving hospital of the transport. Based on the nature of the child's illness or injury, the receiving hospital then can begin to assemble personnel and equipment for prompt treatment. On arrival, essential information concerning the child's condition and the field treatment is transferred to the accepting physician.

TYPE OF TRANSPORT USED

The decision about whether to use ground or air transportation depends on the child's condition, the type and location of emergency facilities, and the resources in the community. In general, ground transportation is more readily available and safer if the appropriate emergency facility is within 20 to 30 minutes from the scene. Air transport is more valuable in rough terrain (helicopter) or when there is a long travel distance (fixed-wing). In the first instance, the first responder notifies the regional emergency air transport system (which may be hospital- or police-based) directly, and the helicopter then is dispatched to the scene. For long-distance transport, patients are taken to airports by ground transport vehicles and then are transferred to fixed-wing craft. In no other transport system is effective and efficient communications between prehospital providers and emergency department physicians more important than in air transport. It also must be emphasized that air transport is not without risk, especially in inclement weather, and should be used only when absolutely necessary. It is not intended for routine or nonemergent transport. When choosing between alternative forms of transport, safety factors should always be balanced against the urgency of transport and other special needs of the patient.

EMERGENCY MEDICAL SERVICE ORGANIZATIONS

Many organizations at the local, state, and national levels are available to a wide range of EMS personnel. These organizations serve as forums for discussing, teaching, and implementing policies used to promote the specific needs of EMS. A list of national organizations is shown in Table 6.7.

American College of Emergency Physicians (ACEP) Dallas, TX
American Trauma Society Upper Marlboro, MD
Association of Air Medical Services (AAMS) Columbia, MO
Emergency Medical Foundation Washington, DC
National Association of Emergency Medical Technicians Kansas City, MO
National Association of EMS Physicians Pittsburgh, PA
National Association of First Responders Orange Beach, AL
National Association of State EMS Directors Lexington, KY
National Emergency Number Association (NENA) Columbus, OH
National Registry of Emergency Medical Technicians Columbus, OH

Table 6.7. National EMS Organizations

MEDICAL-LEGAL ISSUES

Prehospital care providers are legally responsible for their actions or lack of actions. Malpractice suits increase annually, affecting all components of the health care system. More claims originate from urban areas than from rural areas. Because they act in an official capacity, for which they are paid, prehospital care providers generally are not protected by Good Samaritan laws.

Prehospital care providers practice in a precarious setting. They commonly attend to children in cramped, poorly lighted quarters, encounter large crowds or hostile family members, and lack equipment appropriate for pediatric patients. Despite these and other obstacles, prehospital care providers often must make important decisions about the management and transport of critically ill children with rapidly changing conditions. Because of inadequate pediatric training or experience, some prehospital care providers might not be prepared well enough to provide such management. Most lawsuits that involve prehospital care providers result from transport of patients to inappropriate facilities, deviation from standardized procedures, slow response, or failure to transport patients when indicated. Although prevention of all legal actions is unlikely, prehospital care providers can minimize their risks by attending to the three Ds: duties, details, and documentation. When there is doubt, prehospital care providers should consult with the on-line medical control physician.

Duties to Provide Care

All health care providers must understand their duties to provide care. Questions often arise concerning issues of consent. Most states allow for the treatment of minors without parental consent when an emergency exists. In general, any condition that threatens “life and health” or “life and limb” is considered an emergency and can and should be treated immediately. Patients with impaired cognitive function (i.e., irrational, unconscious, or retarded) also should be treated. It is best to err on the side of treatment.

If parents are present and refuse care for their children, they should be asked to sign a (witnessed) form releasing the prehospital care provider from responsibility. Regardless of religious beliefs or parental desires, the child must be treated, however, if there is a life-threatening emergency or possible child abuse. Treatment also should be rendered if the parents are incapable of understanding the risks of refusing care. Off-duty prehospital care providers in uniform still have some responsibility to provide emergency care.

Prehospital care providers are not required to provide care if they would be put at risk for personal injury. Some states also permit prehospital care providers not to administer CPR after being shown written physician orders directing such care.

Details of Standards of Care

Prehospital care providers must always provide care in accordance with local standards. Many municipalities have established written protocols that help streamline decision making. Communication with the command center is crucial; inquiries should be made when doubt exists.

Care should be taken to provide proper immobilization, when indicated. Lawsuits have resulted from improper immobilization. Furthermore, to protect themselves, prehospital care providers should insist that the receiving physician examine the traumatized patient before removal of immobilization equipment. Although treatment of the patient is the first priority, care should be given to preserve evidence if trauma resulted from a criminal act. Clothing and other items (e.g., poison containers) should be brought with the child.

Local regulations also must be obeyed. Laws that detail lines of authority among police, fire, and prehospital care units often exist. Likewise, local driving and speed laws govern ambulance road behavior.

Documentation

All actions by prehospital care providers should be documented. These documents become part of the child's record and may be used as evidence. Therefore, the person who provides documentation should strive for completeness. Each patient should have an individual report that contains vital signs, physical findings, medications and fluids given, procedures performed, position of the patient during transport, and communications (including failed) with medical command. Although these records are confidential, prehospital care providers are required to report suspected child

abuse, gunshot wounds, and illegal drug use.

SAMPLE PATIENT MANAGEMENT ALGORITHMS

Prehospital patient care often is required for seriously ill or injured children. For this reason and because pediatric skills often are suboptimal because of infrequent use, it is wise to develop standing protocols for various medical and surgical emergencies. Logically, such protocols should be developed regionally or locally so that they can incorporate the facilities and services available. Protocols always should reflect the principles of optimal medical and surgical management.

The chapter appendix (pp. 94–101) contains sample algorithms developed by the Philadelphia Emergency Medical Services Council, under the direction of Steven J. Davidson, MD. These protocols were created by the emergency medicine staff of The Children's Hospital of Philadelphia in 1990, and they are updated periodically.

APPENDIX
SAMPLE PATIENT MANAGEMENT ALGORITHMS

ASYSTOLE

Confirm presence of asystole in two lead positions.

[1] When given IV/IO, epinephrine should be repeated every 5 minutes. If IV/IO is unsuccessful, epinephrine 0.01 mg/kg may be administered via endotracheal tube.

[2] Intubation is preferable if it can be accomplished simultaneously with other techniques.

[3] Atropine administration may be repeated once in 5 minutes. If IV is unsuccessful, atropine 0.02 mg/kg (minimum dose = 0.1 mg) may be administered via endotracheal tube. **Maximum dose = 2 mg.**

[4] Sodium bicarbonate administered after 10 minutes of down/arrest time. For children less than 1 year of age, dilute this drug 1:1 with sterile water.

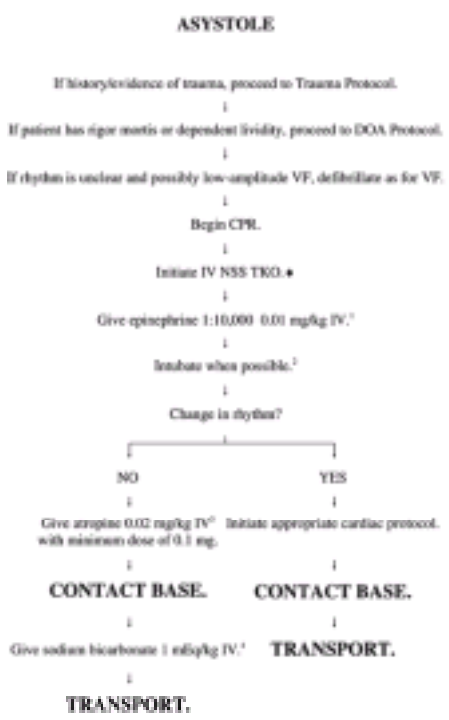
[u] Place an IO line if unable to obtain IV access. Once established, the IO line replaces the IV line as the primary route of administration for fluid and medications.

Time on scene for “cardiac code” generally should not exceed 30 minutes including contact with Medical Command physician.

Fiftieth Percentile Weights by Age

Age (yr)	Average Weight(kg) Boy	Girl
0.5	8	7
1	10	9.5
1.5	11	11
2	12	12
3	14	14
4	17	16
5	19	18
6	21	20
7	23	22
8	25	25
9	28	28
10	32	32
11	36	37
12	40	41
13	45	46
14	50	50
15	55	54
16	60	56

NOTE: If child's weight is **greater than** 50 kg, enter the Adult Protocol.



BRADYCARDIA

The rate at which a child is bradycardic depends on his or her age. The ECG is characterized by a slow rate, in addition to alteration of the P:QRS ratio and P-R interval.

[5] In a bradycardic child, therapy is reserved for the patient who is symptomatic as manifested by these symptoms that identify the presence of decreased blood flow to end organs.

[u] Place an IO line if unable to obtain IV access. Once established, the IO line replaces the IV line as the primary route of administration for fluid and medications.

Fiftieth Percentile Weights by Age Average Weight(kg)

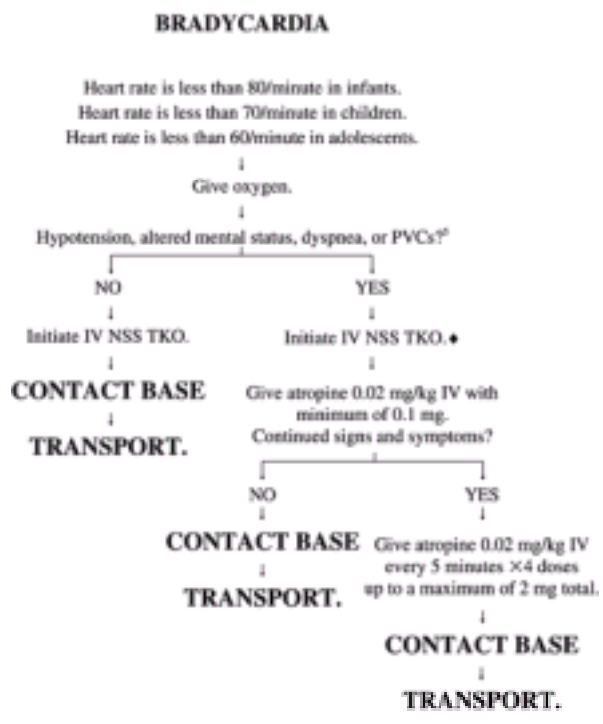
Age(yr)	Boy	Girl
0.5	8	7
1	10	9.5
1.5	11	11
2	12	12
3	14	14
4	17	16
5	19	18
6	21	20
7	23	22
8	25	25
9	28	28
10	32	32
11	36	37
12	40	41
13	45	46
14	50	50
15	55	54
16	60	56

NOTE: If child's weight is **greater than** 50 kg, enter the Adult Protocol.

AVERAGE PEDIATRIC VITAL SIGNS

Temperature	Pulse	Blood Pressure+	Respirations
36-37°C	Newborn: 140-160	80/50	Infant: 40
	1 year: 120	82/54	Preschool: 30
	2 year: 110	84/56	School-age: 20
	4-6 year: 100	90/60-110/76	
	8-10 year: 90	116/78	
	12 year: 80	120/80	

+ Hypotension implies a systolic blood pressure 10 mm Hg less than the average value for age; when in doubt; check with Medical Command physician.



APNEA/INADEQUATE RESPIRATIONS

Inadequate respirations are indicated by anxiety, restlessness, poor or unequal chest expansion, cyanosis, extremely shallow respirations, respiratory rate as listed below, and abnormal or absent breath sounds.

[6] Endotracheal (ET) intubation is the preferred method of airway maintenance. Refer to the table below for pediatric endotracheal tube size guidelines. EOA is contraindicated in children less than 16 years of age.

[7] Consider intubation of the right mainstem bronchus. If necessary, remove initial ET tube. Hyperventilate for 1 minute, then reattempt intubation.

[8] If ET intubation is unsuccessful after three attempts, use manual methods to maintain airway and ventilations. Immediately transport to closest appropriate hospital.

Rates of assisted ventilations for:

- Infants: 20–25 breaths/minute
- Children: 16–20 breaths/minute
- Adolescents: 12–15 breaths/minute

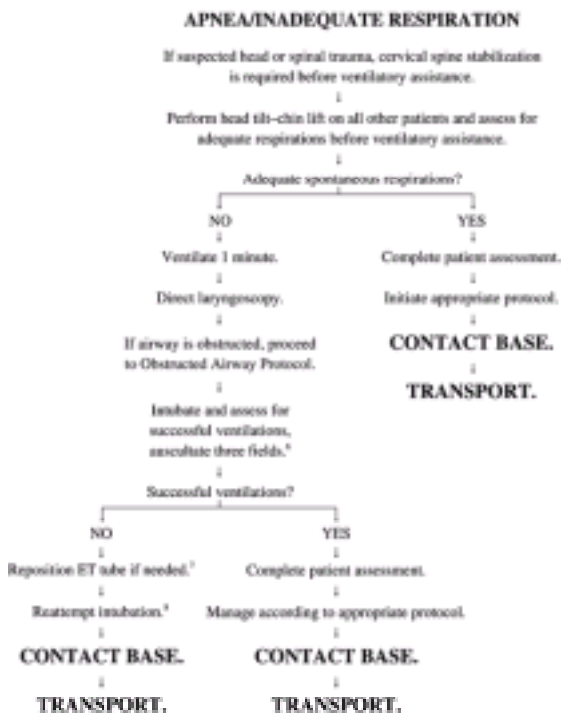
Pediatric ET Tube Size Guidelines

Age	ET Size(mm)
Newborn	3.0
6 months	3.5
1 year	4.0
2 years	4.5
4 years	5.0
6 years	5.5
8 years	6.0
10 years	6.5
12 years	7.0

$$\text{ET size} = \frac{16 + \text{age (yr)}}{4}$$

Laryngoscope Blade Size

Age	Laryngoscope Blade Size
Premature	0 Straight
Term-1 year	1 Straight
1-1½ year	1½ Straight
1½-12 years	2 Straight/curved
13+ years	3 Curved



OBSTRUCTED AIRWAY

If acute obstruction of the airway is due to systemic allergic reactions, proceed to Hypersensitivity Protocol. If maxillofacial trauma is present, proceed to Trauma Protocol.

[9] For children <1 year of age, put head down and use back blows/chest thrusts. For children >1 year of age, use Heimlich maneuver.

[10] ET intubation is the preferred method of airway maintenance after removal of a foreign body with Magill forceps. Refer to the table below for pediatric ET intubation tube size guidelines.

If ET intubation is unsuccessful, use manual methods to maintain airway and ventilate with immediate transport to closest hospital. EOA is **NOT** to be used in patients presenting with obstructed airway.

Successful ventilation is indicated by:

- Bilateral chest expansion
- Adequate tidal volume
- Adequate lung sounds

[11] Remove initial ET tube. Hyperventilate for 1 minute, then reattempt intubation.

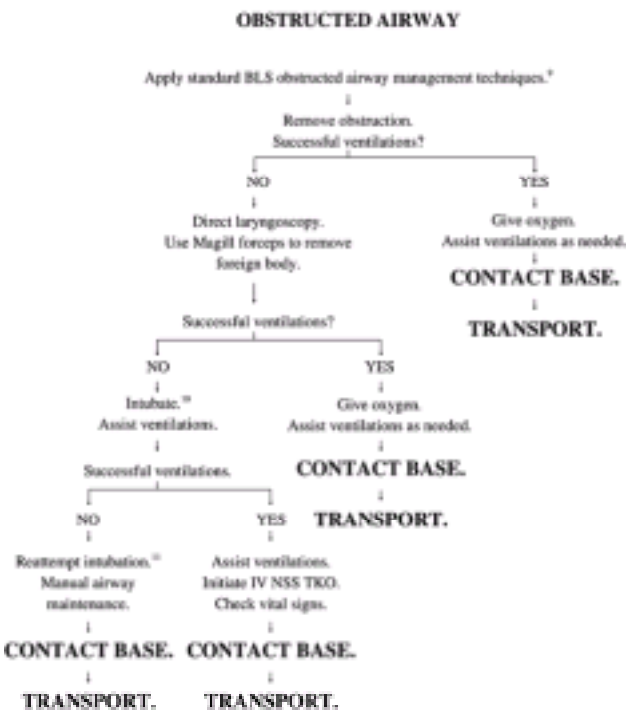
Pediatric ET Tube Size Guidelines

Age	ET Size(mm)
Newborn	3.0
6 months	3.5
1 year	4.0
2 years	4.5
4 years	5.0
6 years	5.5
8 years	6.0
10 years	6.5
12 years	7.0

$$\text{ET size} = \frac{16 + \text{age (yr)}}{4}$$

Laryngoscope Blade Size

Age	Laryngoscope Blade Size
Premature	0 Straight
Term-1 year	1 Straight
1-1½ year	1½ Straight
1½-12 years	2 Straight/curved
13+ years	3 Curved



Suggested Readings

American College of Emergency Physicians. Prehospital advanced life support skills, medications, and equipment. Ann Emerg Med 1988;17:1109–1111.

Baker MD, Ludwig S. Pediatric transport and the private practitioner. Pediatrics 1991;88:691–695.

Callaham M. Quantifying the scanty science of prehospital emergency care. Ann Emerg Med 1997;30:785–790.

Cobb LA, Alverez H, Copas MK. A rapid response system for out of hospital cardiac arrests. Med Clin North Am 1976;60:283–290.

Crampton RS, Aldrich RF, Gascho JA, et al. Reduction of prehospital, ambulance and community death rates by the community-wide emergency cardiac care system. Am J Med 1975;58:151–163.

Eisenberg M, Bergner L, Hallstrom A. Paramedic programs and out of hospital arrest. I. Factors associated with successful resuscitation. Am J Public Health 1979;69:30–38.

American College of Surgery Bulletin. Essential equipment for ambulances. ACS Bull 1983;68:36–38.

Hergenroeder P, Berk W. Levels of prehospital providers. In: Kuehl AE, ed. National Association of EMS Physicians: EMS Medical Directors' Handbook. St Louis: Mosby, 1989:39–48.

Ludwig S, Selbst S. A child-oriented emergency medical services system. Curr Probl Pediatr 1990;20:115–158.

Luten R, Foltin G, eds. Pediatric Resources for Prehospital Care. Elk Grove Village, IL: American Academy of Pediatrics, 1989.

Roush WR, McDowell RM: Emergency medical services systems. In: Roush WR, ed. Principles of EMS Systems. Dallas: American College of Emergency Physicians Press, 1989:11.

Scribano PV, Baker MD, Holmes J, et al. Utilization of prehospital interventions for the pediatric patient in an urban emergency medical service system (Abstract). Arch Pediatr Adolesc Med 1996;150(suppl):36.

Seidel JS. Emergency medical services and the pediatric patient: are the needs being met? II. Training and equipping emergency medical services providers for pediatric emergencies. Pediatrics 1986;78: 808–812.

Selbst SM. Medical/legal issues in prehospital pediatric emergency care. Pediatr Emerg Care 1988;4:276–278.

Sherman MA. Mobile intensive care units: an evaluation of effectiveness. JAMA 1979;241:1899–1902.

Smith GA, Thompson JD, Shields BJ, et al. Evaluation of a model for improving emergency medical and trauma services for children in rural areas. Ann Emerg Med 1997;29:504–510.

Stewart RD: Historical perspective. In: Roush WR, ed. Principles of EMS Systems. Dallas: American College of Emergency Physicians Press, 1989:5–8.

Storer DL, Wilcox DE. Ground and air medical services specialty transport. In: Roush WR, ed. Principles of EMS Systems. Dallas: American College of Emergency Physicians Press, 1989:61–76.

Suler JM, Montes MF, Egol AB, et al. The ten year malpractice experience of a large urban EMS system. Ann Emerg Med 1985;14:982–985.

Tsai A, Kallsen G. Epidemiology of prehospital care. Ann Emerg Med 1987;16:284–292.

Weigand JR. Prehospital ground transportation: system structure and requirements. In: Roush WR, ed. Principles of EMS Systems. Dallas: American College of Emergency Physicians Press, 1989:51–60.

Yamamoto LG, Wiebe RA, Maiava DM, et al. A one year series of pediatric prehospital care. Pediatr Emerg Care 1991;7:206–214.

CHAPTER 7

Transport Medicine

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Pediatric interfacility transport involves patient transfer from one medical location to another. Patient status can vary from relatively stable to critically ill. The disease processes involved are detailed throughout this text and include inpatients with progressive or unresolved problems and the entire spectrum of neonatal illness. Although the ABC (airway, breathing, circulation) approach to care remains the integral focus during interfacility transport, it is essential to understand the disease process and expected progression of the disease during the transport period.

Interfacility transport can originate from or be directed to hospitals, emergency care centers, physician offices, clinics, or other medical care facilities. There may not be a clear distinction between interfacility and prehospital transport in terms of equipment, process, and in some instances, personnel. Many interfacility transport teams routinely transport patients from the prehospital care environment, and emergency medical services (EMS) may be involved in interfacility transport. The differences between prehospital and interfacility transport teams often revolve around extrication issues, personnel education, and experience (see Chapter 6).

Although transport teams care for patients with disease processes similar to those seen in emergency department (ED) and critical care units, the delivery of care can differ. The transport environment offers many opportunities for problems if care is not managed appropriately. Although issues of transport team organization can be found in other texts, we briefly review important concepts of interfacility transport here.

Interfacility transport begins with the recognition of a need or a desire for medical care not available at the patient's current location. Reasons for transport include the need for advanced levels of care or specialized care or services, a person's preference for a particular caretaker, the desire to obtain a second opinion, insurance issues, and parent or provider frustrations. However, as outlined in the Consolidated Omnibus Budget Reconciliation Act of 1985 (COBRA) and the federal Emergency Medical Treatment and Active Labor Act (EMTALA) regulations, interfacility transport cannot be used as a method to avoid initial assessment, stabilization, or intervention, especially with regard to a patient's ability to pay.

To prepare for interfacility transport, a transport system must have access to specialized equipment, trained personnel, and appropriate licenses (Table 7.1 A, Table 7.1 B, Table 7.1 C and Table 7.1 D). The transport system is responsible for ensuring the safety of the patient, team, and family members during the transport as well as guaranteeing that the patient is not cared for in a less medically sophisticated environment. The transport system should have an identifiable medical director who is responsible for ensuring adequate training and education as well as continuing assessment of the transport personnel and process. The medical director is the person ultimately responsible for ensuring a safe, reliable transport system.

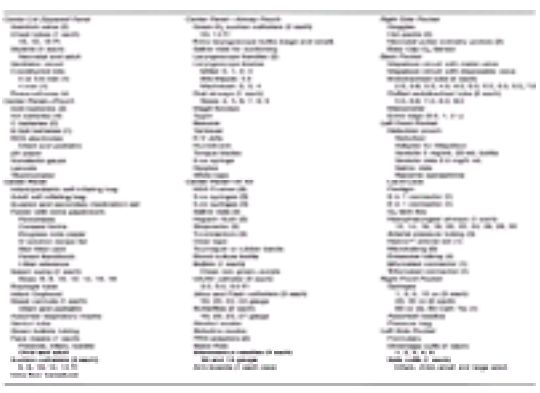

Table 7.1. ***An Example of Equipment and Pharmaceutical Supplies Useful for Pediatric Critical Care Transport. Because Transport Teams Must Be Self-Sufficient During the Transfer Process, Care Should Be Taken to Ensure Adequate and Appropriate Supplies. Specifics May Vary As Per Team Function and Intended Patient Population.***
A. Contents of a "Universal" Pediatric Critical Care Transport Equipment Bag

Cardiorespiratory Monitors	Infusion Pumps
Noninvasive	Single and/or multichannel
Invasive	Blood Sample Measuring Devices
Ventilators	Point of care testing
Pressure	Glucometer
Volume	Portable Suction
Capnography	Portable Oxygen
Pulse Oximeter	Cellular Phone(s)
Defibrillator with Pacing Capability	

Table 7.1.B. Additional Transport Equipment to Be Considered When Equipping a Pediatric Critical Care Transport Team

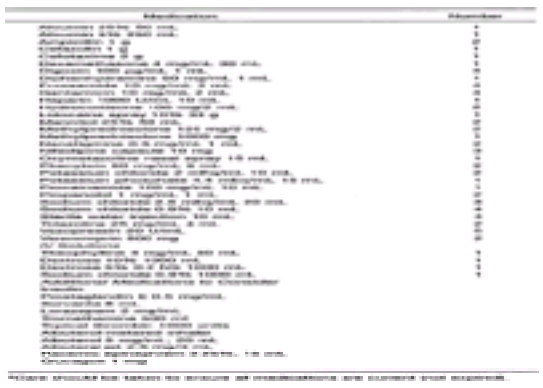

Table 7.1.C. A Standard Pediatric/Neonatal Critical Care Transport Medication Supply[a]

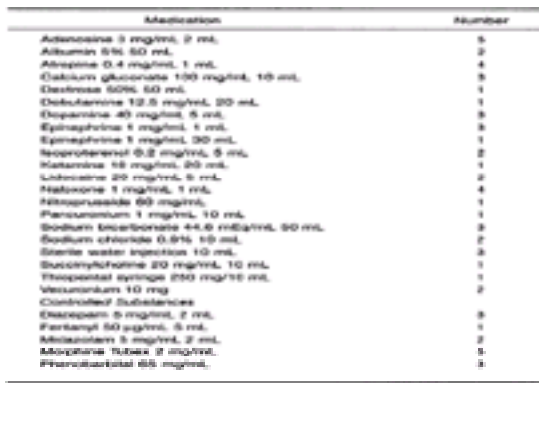

Table 7.1.D. A Rapidly Accessible "STAT" Medication Supply Should Be Included

Significant preparation is required to be an efficient user of transport services. The users of a transport system (the referral hospitals and physicians) must ensure that the transport services meet the standards required for the transfer of their patients. The referral physician must avoid the mind-set of getting the patient out of the ED as quickly as possible without first ensuring transport safety and medical integrity.

Transport medicine is a recognized section within the American Academy of Pediatrics (AAP). Many other groups, such as the Air Medical Physicians Association (AMPA), are dedicated to ensuring optimal care for transported patients. These organizations offer continuing education and are conduits for information regarding transport medicine. The AAP published the "Guidelines for Air and Ground Transport of Neonatal and Pediatric Patients" in 1993, and an updated revision is scheduled for publication in 1999.

TRANSPORT CONSIDERATIONS

Familiarity with the transport environment is necessary for optimal transport care, and the ability to understand the environment and troubleshoot as necessary is critical for the successful transport of a patient. Therefore, configuring a transport team in response to an acute request for patient transport, unless there has been adequate preparation and

training, should be avoided. Although the medical care of the transport patient is similar to that provided in an ED or an intensive care unit (ICU), the potential for errors during the transport process is significant. Issues to consider include ensuring proper equipment and medication supplies; intervening in a cramped, moving environment; safely securing the patient to the stretcher and the stretcher to the transport vehicle; and recognizing and managing the loss of inverter power. In addition, oxygen delivery and suction—as well as the issue of motion sickness—can be difficult for the non–transport-oriented participant. Noise, vibration, and temperature can also be formidable problems for the patient and provider if not anticipated and planned for.

In the transport process, one must be prepared for all types of patients and complications. When the transport team arrives, the patient's status may be significantly different than that initially described. This can be the result of a change in the patient's condition, incomplete assessment by the referring physician or transport team, or inadequate information flow. The ability to correctly assess severity of illness or injury before transport, from both the referring as well as receiving physician perspectives, helps facilitate appropriate triage, mode of transport, and personnel configuration decisions. The transport team may also be asked to transfer a different patient if a more critical patient has presented to the same referring institution. Inadequate numbers or types of personnel, equipment, or medications can render the transport team less effective in these situations.

The medical capabilities of the transport systems are important to assess. All transport teams do not have equivalent levels of pediatric skills. Transport services can vary from specialized pediatric teams, such as those supplied by tertiary care pediatric hospitals, to generalized transport services. A generalized transport service accepts all ages and types of patients; unfortunately, there are no universal standards or regulations regarding the level of experience or expertise in pediatrics required to transport pediatric patients. Although pediatric patients can be efficiently and safely transported by different types of transport systems, the referring physician is responsible for assessing each program for medical sophistication and safety. Pediatric diseases and processes are different from those in adults, and one should not assume that a general transport service has adequate experience in pediatrics to offer the appropriate level of care. Although patient transport by a general team may be quickly accomplished, the process may be classified as getting the patient to pediatric care quickly as opposed to rapidly bringing pediatric care to the patient. For many patients, this is an academic distinction. For example, the stable trauma patient, the child with a clearly defined medical process, or the patient needing referral for a nonprogressive, non–life-threatening issue may be adequately transported by a transport team without extensive pediatric experience. It is imperative, however, that general critical care skills be available for most of these transports. Consultation with a pediatric expert should also be included. When the differential diagnosis needs to be explored during the transport process or when the patient's condition is rapidly changing, an experienced pediatric team is usually preferred. In geographic locations where specialized pediatric transport is unavailable, involvement of a pediatrician on the referring and/or receiving end is important. When possible, a patient should not be referred from a nonpediatric provider to a pediatric institution via a transport team inexperienced in pediatric disease process and management. Ideally, advanced pediatric care should begin the moment the transport team is contacted. Pediatric medical or surgical advice, as well as adequate instruction before arrival of the transport team, can be lifesaving for that particular patient.

Technical skill capability in general transport teams does not necessarily translate into technical skill competence in the treatment of children. This is perhaps most evident in children needing advanced airway intervention. A child's anterior larynx and the recommendation to avoid nasal intubation in children can preclude successful airway intervention by personnel who are not sufficiently educated or experienced in managing the pediatric airway. This discussion is not meant to cause one to avoid nonpediatric transport services but only to ensure that skills necessary for adequate assessment and care of the patient during the transport process are optimal and that the cognitive components, if necessary, be augmented by an outside pediatric specialist.

When a decision is made to transport a pediatric patient, there is often a discussion of the appropriate mode of transport. Nonmedical transports include a parent's automobile or a taxicab. Problems with these choices include no assurance of direct transport to the receiving facility, inability to ensure patient safety, and the lack of available medical care during the transfer. Even the accompaniment of a physician or nurse does not markedly improve the ability for medical intervention in these nonmedical vehicles. A basic life support (BLS) ambulance offers direct transportation to the receiving institution but does not offer much in the way of pediatric expertise or intervention capability. Physician or nurse accompaniment in a BLS ambulance increases potential level of medical care; however, the BLS environment is limited with regard to basic personnel, pediatric equipment, and medications. Advanced life support (ALS) transport offers more sophisticated resuscitation abilities but often provides minimal pediatric expertise. Physician accompaniment may increase the level of available medical care within the ALS environment but may be limited by the particular service's use of protocols and off-line medical direction. Critical care transport services can increase the medical sophistication of ALS providers by ensuring the presence of a critical care transport nurse. Pediatric critical care transport systems include critical care participants (nurses and physicians) with significant pediatric experience and expertise.

When transport services are needed, the modes to be considered include ground ambulance, rotor-wing aircraft (helicopter), and fixed-wing aircraft (jet or prop plane) (Fig. 7.1, Fig. 7.2 and Fig. 7.3). Often, the decision is easy. For example, the available transport system may be ground only, or inclement weather may prevent air transport. Alternatively, the distance may be so great or traffic issues significant enough that ground ambulance transport is impractical. Several important issues should be considered when mode of transport is discussed. The first is personnel availability. If the transport modes require or offer different personnel configurations, the mode with the most appropriate personnel should be strongly considered. Ideally, a pediatric transport service with the capability for air and ground transport is available so that comparative personnel issues are not the major deciding factors. A choice between speed of transport and appropriate medical personnel can be difficult, although they are not necessarily mutually exclusive. Often, the referral hospitals or physicians want to have a patient taken to the receiving hospital as quickly as possible. They are sometimes willing to accept a transport team with little pediatric sophistication based solely on speed. Occasionally, this may be appropriate, but caution needs to be exercised to ensure that the transport system is capable of handling issues that can occur during the transport process. The referral physicians must be proactive in the mode of transport decision. This includes being aware of the transport systems that are available and evaluating those systems before using them. In areas without tertiary care pediatric transport options, local pediatric providers and the receiving pediatric physicians

should be available to provide expertise to the general transport services to help bridge the gap between the general (primarily adult) and pediatric provider.

FIGURE 7.1. A, B. Pediatric interfacility ambulance environment. Examples of patients being transported within the ambulance environment. Note relative limitations of space and patient access. (Figures used with permission. © The Children's Hospital of Philadelphia, Philadelphia, PA.)

FIGURE 7.2. A-D. Air medical transport environment. Examples of design of a medical helicopter (“rotor wing”). Note relative space and patient access limitations. (**A–C.** used with permission, Hahnemann University Hospital, University MedEvac, Philadelphia, PA; **D** used with permission. © The Children's Hospital of Philadelphia, Philadelphia, PA.)

FIGURE 7.3. Fixed-wing transport. Both jet and piston (propeller) aircraft are used.

The disease process must also be considered in mode of transport decisions. The patient with developing petechiae, fever, and hypotension should not be transported several hours by ground if a quicker method of transport is available. On the other hand, a short air transport for a stable patient may not be an appropriate use of resources or be in the patient's or team's best interest. One must be cognizant of the many issues surrounding the mode of transport choices. These choices should be individualized for each patient. Although appropriate medical care should not be withheld for financial reasons, a cost comparison of air and ground transports is often useful, especially if done before the acute transport. This may be an important factor in the decision process, however, if the referral or receiving physician or hospital is responsible for guaranteeing the cost of the transport. In general, rotor-wing (helicopter) transport costs two to three times as much as a ground transport for local transfers. However, the cost is often offset by the savings in time. A helicopter, which can travel directly to and land at the patient's location, is much quicker than an ambulance, which must take a more circuitous route. If the helicopter cannot land directly at the referring or receiving center, however, the time savings by air transport may be less significant. In that situation, in addition to the decreased time savings, the patient may be placed at greater risk with the multiple transfers from referral center to ambulance to helicopter to ambulance to receiving hospital. The riskiest time for the patient is often during transfer from stretcher to stretcher or vehicle to vehicle (Fig. 7.4). These transfers increase the opportunities for dislodgement of endotracheal tubes, central venous catheters, chest tubes, and other lifesaving equipment.

FIGURE 7.4. A–C. Transfer of patient during transport process. Patient transfer between vehicles or stretchers can be risky to the patient. Tube, line, oxygen, or medication disconnection or disruption, as well as shifts in immobilization, must be avoided. (**A** and **C** used with permission. © The Children's Hospital of Philadelphia, Philadelphia, PA; **B** used with permission, Hahnemann University Hospital, University MedEvac, Philadelphia, PA.)

PERSONNEL ISSUES

Many types of providers can function effectively as part of a pediatric transport team. Nurses, respiratory therapists, emergency medical technicians, paramedics, and physicians serve on various transport teams. The choice of personnel depends on several factors, but the most important are the team's primary mission and the resources available for training and skill maintenance. In general, the personnel chosen for the transport team should have experience in the care of critically ill infants and/or children. The transport environment is not the appropriate place to learn basic pediatric critical care skills.

The primary mission of the team must be kept in mind when selecting personnel and planning training. For example, a team devoted to neonatal transport should consider team members with experience in the care of critically ill neonates, whereas teams that perform transports from nonhospital locations may want to employ personnel with prehospital care experience. These team members are likely to adapt more easily to the demands of their new roles. Teams that have multiple missions, such as those that transport both neonates and older children, should attempt to recruit team members from varied backgrounds. By necessity, such teams have to devote considerable time to the medical cross-training of staff members. However, having team members from varied backgrounds offers the potential for those members to assist in the training process. Regardless of the medical background of the transport participants, education and experience in the transport environment is imperative.

Transport team capabilities and types of personnel vary significantly depending on the transport system. However, pediatric critical care transport teams, the ideal interfacility transport configuration for children, often have several specific types of providers. At the heart of most pediatric critical care transport teams are highly trained pediatric nurses. These nurses usually have significant critical care or emergency medicine experience before becoming members of the transport service. They have often had their technical and cognitive skills enhanced by formal or informal specialized training as described in the following. Such training may allow them to be classified as advanced care practitioners. Depending on the sophistication of the transport system, training opportunities, skills and assessment, and medical licensure issues, transport nurses often provide advanced management for these children. This can include diagnosis and assessment skills as well as interventions (e.g., advanced airway management, central venous access, resuscitation). In addition to their cognitive and technical skills, transport nurses have become experts in the environment in which they practice. The transport nurse should be intimately aware of all operating systems within the transport environment as well as safety procedures for the patient and the transport team. The medical skills of pediatric transport nurses can often be complemented by the addition of an attending, fellow, or resident physician; respiratory therapist; nonpediatric transport nurse; or paramedic. Team compositions vary greatly in different systems, and no single team configuration is preferred. The ideal team composition is one that addresses the acute and projected needs of a particular patient and that has the flexibility to be amended when necessary.

In addition to the personnel already described, transport teams usually include drivers or pilots who may have no role in patient care or who may assist the other personnel. Communications specialists may be employed as part of the team's call receipt and dispatch process. Finally, all transport teams should have a clearly identified medical director or directors. The functions of the communications specialists and the medical directors are discussed elsewhere in this chapter.

Other important educational considerations are the resources of both time and money available to devote to training team members. If training time is limited, the transport team must consider hiring staff who are already well trained. For instance, a neonatal team could hire neonatal nurse practitioners. However, such well-trained staff usually demand higher wages, making them ultimately more expensive than a training program. Many teams do not have the luxury of hiring nurse practitioners or other highly skilled personnel; therefore, teams have to devote significant time and resources to training team members. The amount of time necessary varies with the team's mission and its customary personnel composition. A team with a well-defined scope of practice, like neonatal transport, should employ experienced neonatal nurses. In this circumstance, only those skills that are new to the team members are taught. Likewise, if a team usually includes a physician, the other team members may not need to learn advanced skills such as tracheal intubation. Teams that do not routinely include a physician must ensure that their members are competent in all procedural and management skills that may be required during transport. Such extensive training usually includes a didactic component, a skills segment, and rotations through various clinical care areas. If the group is large enough, the didactic component may be a series of lectures. However, this type of experience is difficult to arrange for one or two new team members.

Alternatives to formal lectures include videotaped or audiotaped lectures or a modular self-study curriculum.

Unfortunately, skill acquisition is only the beginning. Rarely used skills are quickly forgotten, so a process for skill maintenance must be established. Furthermore, as in all areas of medical practice, the knowledge base in transport medicine is constantly changing, making continuing education vital. Skill retention and continuing education are best accomplished using a three-part process. The first component is renewal of basic procedural and cognitive skills. Such retraining may include rotations through the operating room to practice airway techniques; dry or animal laboratory experiences for interosseous infusion, cricothyroidotomy, thoracostomy tube placement, and other important but rarely needed procedures; and mock codes to practice resuscitation.

The second component is formal continuing education through regularly scheduled programs, including lectures, journal clubs, and presentations of particularly unusual or difficult patients. In addition, such forums may be used to learn about new medical equipment, communication devices, and vehicle issues.

The final component of an effective education program is quality assurance. Routine, periodic case reviews should take place by the transport service in conjunction with other medical experts. A formal morbidity and mortality conference may be included as a part of such a program. In addition, topics such as response times and parent satisfaction may be discussed. The focus of these sessions should be on the process of patient transport. Determining and assigning blame for less-than-optimal outcomes is important only when the staff member involved was clearly negligent. It is far more important to focus on ways in which the team's practices may be changed to improve performance and minimize risk of similar events in the future.

COMMUNICATION

A key component of any transport team is effective communication. Referring physicians must be able to contact the transport team quickly and easily, and teams in the field must be able to communicate with the receiving facility. The ability to communicate with a command physician is particularly important for teams using resident physicians or nonphysician practitioners because they may need on-line medical direction. Ideally, a single point of contact (e.g., a dispatch center) should be established to help ensure that no calls are missed and that all communications are properly documented (Fig. 7.5).

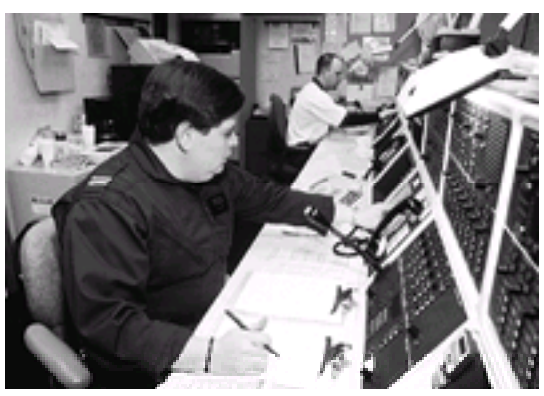

FIGURE 7.5. Transport communication center. A dedicated transport communication center and personnel are invaluable in coordinating all aspects of pediatric transport. The system need not be as elaborate as demonstrated, but should include dedicated phone lines, radio access, personnel notification capability systems, and personnel. (Used with permission, Hahnemann University Hospital, University MedEvac, Philadelphia, PA.)

Communication with the transport team begins with an initial call from the referring hospital. This initial contact is best managed by the use of a protocol or template, which helps ensure that all of the necessary patient and logistical information is properly received by the transport team (Fig. 7.6). During the initial call or soon thereafter, the referring provider may request advice regarding the medical management of the patient. Alternatively, such advice may be offered by the receiving physician. For critically ill infants and children, medical advice via telephone may be needed intermittently from the time of initial contact until the transport team arrives at the referring facility. For these reasons, it is preferable to have transport requests initiated and received by senior physicians who can ask for and offer advice directly. The more people between the source of the information and the final recipient, the greater potential for significant changes or omissions that may be vital to the patient. We also recommend that nurse-to-nurse conversation to evaluate the patient from the nursing perspective be an expected part of the transport process. Together, these two avenues of information flow offer the greatest potential for complete awareness of all aspects of the patient's disease process and current medical condition.

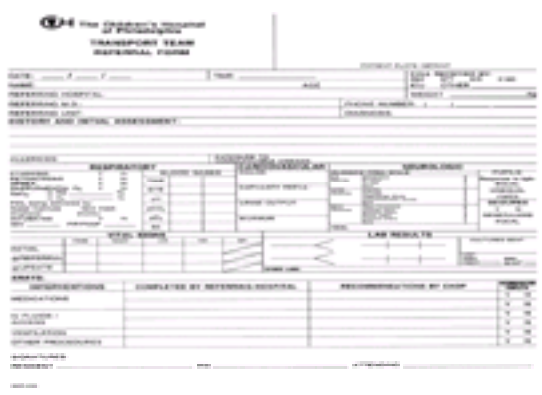

FIGURE 7.6. Transport referral form. A standardized form for recording transport referral information is important. This form should be readily accessible to those who receive the referral. Copies can also be distributed to referral centers to help streamline the process. The forms should, at least, be in duplicate to allow for an official medical record copy (which stays with the command physician during the transport process to document transport progress) and one to accompany the transport team which eventually resides in the patient's transport record.

After the transport team has arrived at the referring facility and performed a preliminary evaluation of the patient, they often need to communicate with one or more people at the receiving hospital. These calls can involve patient review and logistical issues such as patient disposition, scheduling of studies, and need for consultants. Such calls are best facilitated by a communication center.

En route to the receiving hospital, it may be necessary for the transport team to contact the medical command physician either for advice or because the patient's medical condition has changed. Reliable communications are especially important at this point in the transport. The team should be equipped with redundant systems to ensure that a reliable means of communication is always available. These should include cellular technology, long-range alpha beepers, and land radio communications systems. Newer technologies, including telemedicine and global positioning system (GPS), may improve communication and logistics capabilities. The transport command physician should be immediately accessible to the transport team by telephone or designated beeper.

After arrival at the receiving hospital, the transport team is responsible for ensuring an efficient and informative transition of care to the inpatient physician and nursing team. Adequate communication and information flow must take place to fully inform the inpatient team of the patient's disease process and care to date. Complete documentation, written in a clear, concise fashion, is mandatory (Fig. 7.7). Anything less than a complete transfer of information from referral physician to transport team to receiving physicians is a disservice to the patient and a source of potential liability.

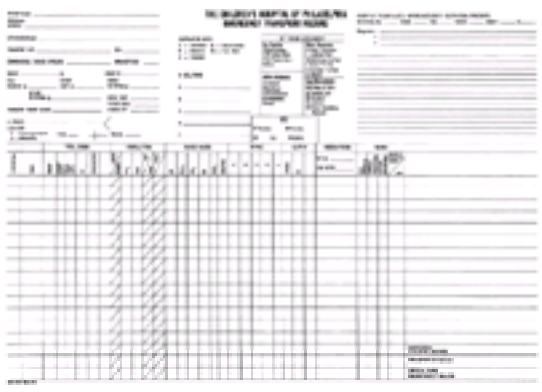

FIGURE 7.7. A. Transport record form (flow sheet). The intratransport period must be documented in an efficient fashion. It must document general medical information as well as progression and response to specific interventions in a timed, sequential fashion. This should also be at least in duplicate to allow for a medical record copy as well as one that remains with the transport system. (Figure **7.7 A**, Figure **7.7 B.1**, Figure **7.7 B.2**, Figure **7.7 C.1**, Figure **7.7 C.2**, Figure **7.7 D**, Figure **7.7 E** used with permission. © The Children's Hospital of Philadelphia, Philadelphia, PA.)

FIGURE 7.7. B.1. Drug infusion charts, as well as medication dosage charts based on weight, may be useful for the transport team.

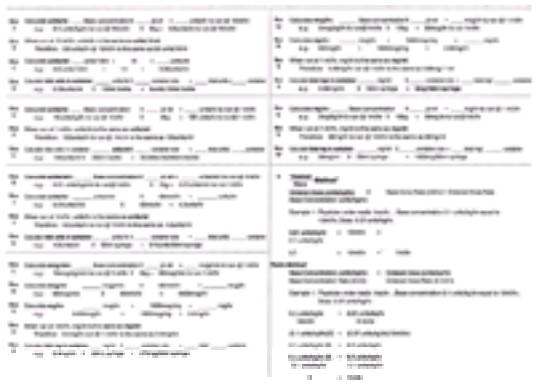

FIGURE 7.7. B.2.

FIGURE 7.7. C.1. Written consent for transport is mandatory.

FIGURE 7.7. C.2.

FIGURE 7.7. D. A written release for medical records will help with transmission of information at time of discharge to referring physicians as well as the primary care physician.

FIGURE 7.7. E. A standardized form can be distributed to referral centers to allow demographic information to be collected prior to the arrival of the transport team. Identification of all principals involved in the patient's care can help future flow of information regarding the patient's diagnosis and outcome.

It is important to remember that transport communications have other medicolegal ramifications. The most important of these involve the giving of medical advice and the assumption of legal responsibility for patient management. When giving or receiving management advice by telephone, both parties should remember that the transport command physician is unable to see or examine the child in question. Therefore, his or her advice will often be somewhat general. The receiving physician must do his or her best to offer clear and complete information, especially when specific information is requested. Suggestions for care must be clearly and completely communicated. For example, if the transport command physician believes that a fluid bolus is needed, that advice should include type and amount of fluid and speed of infusion to avoid any misinterpretation of advice or an inadvertent mistake in one of those parameters. All advice should be documented in writing or by tape recording. The referring physician is under no obligation to accept the advice of the receiving physician, but he or she would be prudent to give it serious consideration. If the referring physician is unable or unwilling to perform suggested interventions because of personnel issues, equipment limitations, or other reasons, this should be discussed with the receiving physician. Likewise, results from interventions or marked changes in the patient's condition during the referral process should be communicated to the receiving physician and transport service. Clear, precise, efficient, and honest communication is imperative for the patient to receive the most appropriate care.

Communication with the patient and family is also important during the transport process. Straightforward communications about disease process and expectations can help prepare a family to accept the consequence of the illness or injury. Lack of communication or reluctance to give bad news can cause a family to expect different outcomes than they should and may pave the way for anger and resentment.

Written communication of all data regarding the patient's care is imperative. Patient summaries, copies of all medical paperwork, laboratory values, and radiographs should be available for the transport team on their arrival. A transport referral checklist may be useful to help streamline the process (Fig. 7.8). Adequate preparation of these documents before arrival of the transport team can greatly improve the efficiency of the transport process and the transition of care. Availability of the referring personnel at the time of transport can also make the process more efficient for all concerned. Telephone and fax numbers and the addresses of the referring and primary physicians should be available to the transport team so that follow-up information may be easily conveyed. Likewise, the referring and primary physicians should be given contact numbers for the transport team and its medical director.

FIGURE 7.8. Referral checklist. Transport referral checklists can decrease the transition time at the referring hospital by allowing the referral team to anticipate the logistics of the pre-transport process and accomplish many tasks prior to the arrival of the transport team. (Used with permission. © The Children's Hospital of Philadelphia, Philadelphia, PA.)

Finally, the family should receive preprinted directions to the receiving facility. Information about parking, mass transit, and the visiting policies of the receiving unit should also be provided. Allowing a family member to accompany the transport team may be useful for both the patient and family.

PATIENT AND TEAM SAFETY

Safety is a key consideration for the transport team. The team should do everything possible to provide for the safety of all involved in the transport process. This includes more than providing pediatric medical expertise for the patient during the transport. It starts with vehicle selection, driver or pilot capabilities, and ongoing licensure requirements. The transport medical director is responsible for continually assessing the capabilities of the particular modes of transport as well as the personnel involved with those functions. This goes beyond licensure issues. Active inspections and evaluations as well as continuous quality improvement (CQI) issues are important. Unsafe vehicles or personnel must be attended to or removed from service. Safety of the transport personnel must also be a priority. Avoiding the use of rotor-wing transport in bad weather is a good example of a safety decision in the transport environment. Fewer air medical transport accidents occur now than in the past. Part of this improvement results from pilots being isolated from patient care information. Instead of being informed that a critical child might die without their intervention, pilots now often make “go” or “no go” decisions based solely on weather and equipment issues. If an appropriate “no go” decision is made, this should not be countermanded by medical or administrative personnel. If a “no go” decision is made based on

weather considerations, another mode of transport or other patient care options must be considered. Competition between transport programs or aeromedical providers can be a safety hazard. In their desire to gain a competitive advantage, one or all of the programs (or specific personnel) may be willing to bend weather and safety rules. It is the policy of our systems that if one air service has denied a transport for weather-related issues, another air service is not contacted unless it is located in a separate environment that may change the weather issues. There is no excuse for risking or losing the lives of several caretakers for any one particular transport.

HOW TO EVALUATE A TRANSPORT TEAM

When assimilating information about pediatric transport options, the following recommendations are offered. First, an attempt to find the transport team with the most pediatric expertise available should be made. Ideally, this team should have the capabilities for both air and ground transport. An assessment of transport personnel should be made through discussions with the medical director or other representative of the transport system. Ideally, the transport team members are dedicated transport personnel who have significant experience in both pediatrics and the transport environment. In assessment of transport programs, one should look for a team that has the capability to vary personnel composition depending on the specific needs of the patient. One should not assume that a more sophisticated medical degree is synonymous with optimal care in the transport environment. The addition of a physician to a transport system does not necessarily suggest a better or more capable system. The system's capabilities depend on the skills of the transport team members as well as the skills and experience of the physician. The use of inexperienced physicians or junior residents who are not familiar with the transport environment may actually decrease the level of delivered care. The inexperienced transport physician may be an unequal partner in the transport process and one who may actually make the transport less effective. For example, the transport nurse may be required to perform most of the patient care and management functions as well as monitor the logistics of the environment. If the transport nurse also needs to tend to an inexperienced provider's personal medical needs during the transport (e.g., motion sickness), this detracts from direct patient care. The addition of a senior pediatric resident, fellow, or physician can be invaluable, however, if that person has transport experience and the patient requires the additional cognitive or technical skills the physician may offer.

The transport system should be easily contacted through an identifiable transport referral number that is answered by a transport communication specialist. One should be able to relay information efficiently and receive suggestions from a transport command physician regarding medical care. It is more efficient to directly contact the transport system regarding patient referral than to call a receiving unit or a particular physician. In an efficient system, a call to the transport communication center should allow the referring personnel to speak to the appropriate receiving physician in a timely fashion. This physician should be clearly identified as the single spokesman for the receiving institution. A single transport command physician allows the process to be streamlined for both the referring and receiving personnel. Transport direction will be from one source, with the use of consultants as necessary. The referring physician does not have to offer transfer information repeatedly, allowing him or her to direct attention to patient care issues. Calling a communication center should also enable the transport team to participate in the conversation as well as simultaneously prepare themselves for the transport. Pretransport preparations can be significant and time-consuming, and early notification can be invaluable. Calls to personnel, ambulances or helicopters, hospital admissions' office, other referral personnel (nurses), and arrangement of tests or procedures at the referral center can be undertaken while medical information is being relayed to the receiving physician. For the nonacute patient, insurance precertification can also be accomplished more efficiently by those who routinely address that issue. The simultaneous completion of these steps can lead to a quicker and more efficient transport. An estimated time of arrival should be given and updated as necessary, recognizing that transport resources may be limited and there may be a necessity to triage patients due to severity of illness.

In addition to initial medical advice, one should expect follow-up from the transport provider regarding the patient. The referring physicians and other caretakers should expect follow-up communication from the transport service regarding care of the patient during transport and within the receiving hospital, including location of the patient and his or her caretakers. There should be an easily identifiable path to the medical director or program manager for issues that develop during or around the transport process. One should expect concerns to be addressed in a timely and satisfactory fashion. In addition, the transport service should be willing to visit the referral physician and location to review specific transport issues. Because the referring physician should have a clearly defined path to the transport service regarding transport issues, the transport team should also have an opportunity to discuss problems with the referral physicians and facilities. These issues could involve timing of referral, preparation for transport, and diagnostic issues. Outreach education by the critical care transport service can be invaluable not only for the physicians but also for nurses and other personnel involved in patient transport preparation.

LEGAL ISSUES

Patient transport is governed by a variety of federal, state, and local statutes. Transport teams and their members may be sued for malpractice under traditional tort law. Therefore, it is important that all members of the transport team understand applicable regulations and avoid unnecessary medicolegal risk.

Of all regulations, the EMTALA has the greatest impact on the management of patient transport. This act places clear duties on both the referring and receiving hospitals. The referring clinicians must do everything possible to stabilize the patient's medical condition before transport and may not transfer a patient against his or her will unless the facility cannot provide the appropriate level of care. Furthermore, the referring physician must obtain informed consent for transfer and, as a part of this process, must advise the patient or the parents of a minor about the risks and benefits associated with transfer. Under EMTALA rules, these discussions should not include the financial ramifications of the decision. This aspect of the law is particularly important when the patient is being transferred solely because of a managed care contract. It seems only fair that parents know that a refusal of transfer may leave them responsible for a hospital bill. However, under EMTALA rules, such information, however well intentioned, may be construed as financial coercion to accept transfer. Instead, the patient should be told to contact the representatives of his or her insurer or the hospital financial personnel to discuss these issues. The referring hospital is also responsible for selecting an appropriate means

of transport. Obviously, the more critical the need for medical care and expertise, the more sophisticated the means of transport. This is an important point for the referral physician to remember. The desire to transfer a child to a more appropriate medical facility as soon as possible is understandable, but if the method chosen places the child in a medical environment that does not offer at least the level of care at the referral center, that center and physician will be liable for any untoward effects that can be construed as having occurred because of the choice of transport. Finally, the EMTALA requires the receiving hospital to accept the patient in transfer if the appropriate type and level of care are available. The ability of the patient to pay for medical care cannot be considered by either the referring or the receiving facility. In addition to the EMTALA, there are often local regulations that govern transport services. For example, some cities have laws designating certain agencies as official providers of prehospital services. Such laws must be considered when offering transport services.

Traditional tort law also applies to the transport team. Most of these issues are no different than those encountered in other health care venues. However, one potential source of medicolegal risk is unique to the transport team. As stated earlier, the transport team gradually becomes more and more involved in the care of the patient. At first, this involvement is limited to giving advice and management suggestions. It is the referring physician's responsibility to carry out these suggestions as he or she deems appropriate. At this stage in the transport process, transport personnel should try to gain the clearest possible picture of the patient's condition so that the most appropriate suggestions may be given. Furthermore, advice may be best prefaced with general phrases such as “Most patients with this condition” or “We often manage this problem by doing.” Finally, the transport team should clearly document any advice given in writing or by tape recording in case disagreements regarding what advice was given arise later.

The next stage of involvement occurs when the transport team arrives at the referral facility and begins to care for the patient, often along with one or more members of the referring hospital's staff. At this point, the greatest medicolegal risks are conflicts over management and difficulties in determining who gave or carried out medical orders. When management conflicts arise, the medical command physician should be contacted, and he or she should resolve these conflicts by speaking directly with the referring physician. The medical record should clearly reflect who gave and who carried out each order.

Finally, the transport team assumes total responsibility for the care of the patient when they leave the referral center. The team should be convinced that the patient is as stable as possible before leaving. If an unstable patient is transported, the team must document why it was in the patient's best interest to undertake transport at that time.

In some cases, transfer agreements exist between hospitals. Typically, such agreements stipulate that the receiving hospital will accept all transfers from the referring hospital. In the past, transfer agreements served to decrease the time needed to accept the patient by eliminating or shortening the approval process. More recently, transfer agreements have become less important for two reasons. First, the EMTALA places a duty on the receiving hospital to accept the patient as long as there is an appropriate bed location available. Second, for patients who are not critically ill, managed care organizations often stipulate certain facilities. In such cases, the transfer agreement exists between the managed care organization and the receiving hospital. However, the referring hospital must still meet obligations to the patient under the EMTALA.

Finally, if the transport team operates under specific guidelines or protocols, these may become the focus of legal action. Therefore, it is imperative that all guidelines represent the current standard of care. Furthermore, guidelines should be designed to ensure that providers do not exceed their scope of practice as defined by state and local regulations. Periodic review of existing protocols and guidelines is warranted. New guidelines should be developed in conjunction with recognized authorities and should be reviewed by risk management specialists before implementation.

STABILIZATION FOR TRANSPORT

The patient care philosophy of most interfacility transport teams stands in contrast to that of prehospital care systems. EMS providers are usually bringing a patient from an environment without medical care (e.g., home or accident scene) to a hospital. In many of these cases, the patient is better served to have the minimum stabilization necessary at the scene followed by rapid transport to an appropriate hospital, with further intervention being performed en route or on arrival. On the other hand, the transport team is most often taking a patient from a hospital, often an ED or another monitored setting, to a monitored bed within a more sophisticated care center. The transport team, therefore, is responsible for maintaining an appropriate level of care between the two centers. Ideally, the transport team should provide the level of care that the patient will have at the receiving facility. At a minimum, the transport team must maintain the patient's present level of care and must do this under difficult circumstances. Stabilization before transport is the key to this process.

Initial preparation for transport often begins when the referral caregivers recognize that the patient requires care beyond the capabilities of their center. Appropriate advice and suggestions from transport personnel or the receiving physician may allow much of the necessary preparation for transport to be accomplished before the team arrives.

When the transport team arrives, they should review the medical history, including all therapeutic maneuvers and interventions performed at the referring hospital. An immediate and thorough physical examination is mandatory. During the pretransport process, endotracheal tubes, chest tubes, intravenous and intra-arterial catheters, and other indwelling devices should be checked for proper placement and stability. When doubt exists, devices should be replaced or better secured.

After this initial assessment, the transport team, in concert with the medical command physician, should decide which, if any, further medical interventions are required before leaving the referring center. Such interventions are most appropriate when they may have a direct impact on patient outcome. For example, the child who may have meningitis should definitely receive antibiotics before transport, but a lumbar puncture may be deferred until he or she arrives at the receiving hospital. The appropriateness of interventions will, to some degree, be dictated by the distance to the receiving

hospital. For example, a child with a circumferential burn of an extremity may require a fasciotomy to prevent vascular compromise. If the receiving hospital is 15 minutes away, this procedure can be accomplished there. However, if the receiving hospital is 2 hours away, it may be better to have the procedure performed before departing from the referring center.

After the patient is optimally prepared for transport, he or she must then be moved from the referral facilities bed to the transport stretcher and then to the vehicle. Such movements represent great risk to the patient. If an intravenous (IV) catheter or an endotracheal tube is going to be displaced, it will likely be while the patient is being moved. This fact has several implications. First, patients should be subjected to the fewest transfers necessary to get them from the referring hospital to the bed they will occupy at the receiving hospital. Movement from the transport stretcher to a holding bed in the ED is often unnecessary and can be avoided with advanced planning. Second, extra vigilance should be used during patient transfer. Personnel should be assigned to secure lines and tubes, and the movement should be coordinated by a team leader. Precautions such as planned, temporary disconnection of the ventilator from the endotracheal tube may need to be considered during these moves. Finally, the patient should be reassessed immediately after each movement. The team must be ensured that the airway is stable and that potentially lifesaving tubes and lines have not become dislodged.

Monitoring is imperative during the transport process. Observation and palpation may be hindered by patient position relative to the caretaker within the vehicle. This may be especially evident in a small transport helicopter. Auscultation may also be impaired in a noisy transport environment. The air transport environment, especially in a rotor-wing or turbo prop aircraft, may be 50% louder than a comparable ground transport. Therefore, more reliance is placed on sophisticated monitoring tools, including cardiorespiratory, pulse oximetry, capnography, and gas delivery monitors with audible and visual alarms, as well as point-of-care laboratory testing. The transport team must ensure stability of the ABCs, and these monitoring tools help them accomplish this effectively.

ALTITUDE PHYSIOLOGY AND THE AIR MEDICAL ENVIRONMENT

When pediatric patients are transported by helicopter or fixed-wing aircraft, one must be cognitive of issues regarding altitude physiology. An increase in altitude brings with it a decrease in ambient oxygen as well as the potential for an increase in the size of air spaces. For most patients, however, these should be not major issues. For patients with severe hypoxia at sea level, diving injuries, or large, enclosed pockets of air, however, air transport can be dangerous.

Two gas laws are most important in the transport process. Boyle's law states that with a constant temperature, the volume of a gas varies inversely with the pressure ($P_1V_1 = P_2V_2$) (Fig. 7.9). As altitude increases, barometric pressure decreases; therefore, the volume of the gas increases. Dalton's law (the law of partial pressure) says that the partial pressure of a gas mixture is the sum of all the partial pressures of the gas within the mixture ($PT = P_1 + P_2 + P_3 . . .$) (Fig. 7.10). For example, the total pressure of air is 1. The partial pressure of nitrogen is 0.78, oxygen is 0.21, and other gases is 0.01. The partial pressure of oxygen will always be 21%. At higher altitudes, air becomes less dense and the partial pressure of oxygen, while still 21%, offers diminished oxygen availability.

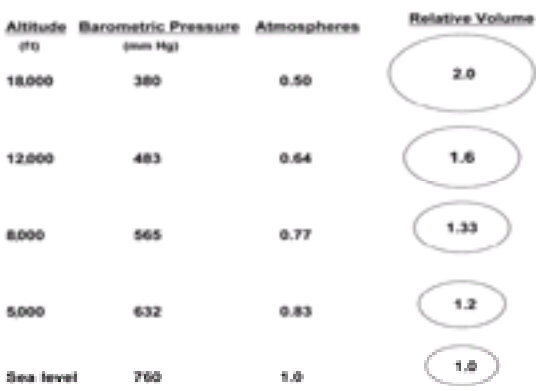

FIGURE 7.9. Boyle's Law ($P_1V_1 = P_2V_2$ or $P_1/P_2 = V_1V_2$). As altitude increases, barometric pressure decreases and volume of gas increases. The diagram illustrates enclosed gas expansion at specific altitudes. “Atmospheres” is comparison to the amount of pressure exerted by an overlying one square inch air column. At sea level, this equals 14.7 pounds per square inch (psi) and one-half that amount (7.35 psi) at 18,000 feet. (Used with permission. Woodward GA, Vernon DD. Pediatric and Neonatal Transport Medicine. Jaimovich DG and Vidyasagar D (eds). Philadelphia: Hanley & Belfus, Inc. page 40, 1995.)

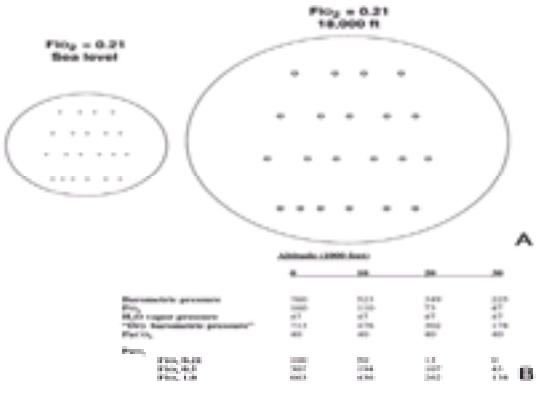

FIGURE 7.10. A. Dalton's Law (law of partial pressure): ($P_T = P_1 + P_2 + P_3...$). The total pressure of a gas is the sum of its component gases. The diagram illustrates that the percentages of air components (oxygen illustrated and represents

21% of air) at different altitudes does not change, although air is less dense at a higher altitude. **B.** The effects of altitude and decreased barometric pressure on oxygen availability. (Used with permission. Woodward GA, Vernon DD. Pediatric and Neonatal Transport Medicine. Jaimovich DG and Vidyasagar D (eds). Philadelphia: Hanley & Belfus, Inc. page 41, 1995.)

These issues can be important during the air medical transport. Entrapped air, if not vented, can be painful (middle ear sinus, teeth, bowel), annoying (flatus, belching), and dangerous (pneumothorax). Use of tight-fitting earplugs in flight can cause an artificial air pocket that may trigger the same problems. More significant air space issues include simple pneumothorax and pneumocephalus, which can become symptomatic at high altitudes. Patients with bowel obstructions may have increased gas volume leading to vomiting and potential aspiration. Air in military antishock trousers (MAST) may vary with altitude as will the air in pressure splints and blood pressure cuffs. Air in endotracheal tube cuffs and Foley catheters may also be affected and might need to be adjusted during flight. Patients with an air embolism from a diving injury or other cause are especially prone to gas volume–related issues during air transport. Hypoxia can be a major issue as well. This can usually be overcome with addition of 100% Fi O_2--unless the patient is already hypoxic at lower altitudes on 100% oxygen. Positive end-expiratory pressure is usually not effective in augmenting the hypoxemia that occurs secondary to an increase in altitude.

Most air transports, however, do not reach an altitude that will greatly influence the patient's care. Helicopter transport routinely occurs at 1000 feet or less above ground level, although this can be greatly altered in mountainous regions where traversing high-altitude peaks may be necessary. Fixed-wing transport aircraft are usually pressurized, meaning that they can simulate the atmospheric pressure of a lower environment. Air pressure in these aircraft is often set at a level of 5000 to 8000 feet, which can, however, still lead to a significant increase in gas volume. Opportunities for achieving higher ambient pressure (lower altitude pressure) may be available depending on the limitations of the particular aircraft. If an aircraft cannot be pressurized to a higher pressure, one can consider flying at a lower altitude. Tradeoffs include increased turbulence, speed restrictions, and fuel issues.

Other issues to be considered in air transport include vibration, turbulence, noise, humidity, temperature, air sickness, exhaust fumes, specific aircraft dangers, and gravitational forces. Helipad availability, landing zones, and other issues of aircraft accessibility must also be determined. Although these logistical issues are ultimately the responsibility of the air transport service and are usually predetermined for the interfacility transport, any additional information should be offered by the referral center personnel. One must be especially careful around running (hot) aircraft. The helicopter's tail rotor and plane's propellers are often invisible when turning but capable of inflicting severe damage upon those who venture too close. Loose clothing, medical equipment, and stretcher pads can become entangled in a helicopter's rotors, causing severe damage to the vehicle and to the passengers of the transport. In general, no one should approach an aircraft other than crew and specifically trained personnel.

SPECIFIC TRANSPORT ISSUES

Airway Management

Airway skills are of paramount importance for transport. There are several general principles of airway management for patients being transported that differ somewhat from those of patients in other clinical settings. First, when a patient being transported has the potential to have airway instability, elective intubation before transport should be considered. In an ED or ICU, similar patients might warrant observation; however, the potential difficulties with intubation in a transport vehicle make elective intubation before transport the more prudent option. This is not to say that the transport environment precludes successful airway intervention, only that the logistics and personnel available in a hospital setting may be more conducive to an efficient, successful procedure. This must be kept in mind when disagreements about airway management are encountered at the time of transfer. It is helpful for the transport personnel to recognize the referral physician's potential reluctance to perform an invasive procedure on a relatively stable patient, and the referral personnel should recognize the potential limitations of the transport environment. Inclusion of the medical command physician may be helpful if a stalemate regarding intervention is reached between the referring physician and the transport personnel. Addressing conflicting airway intervention issues by leaving the ED and “electively” intubating the patient in the ambulance is not an acceptable option. Second, transport teams must ensure that all artificial airways are well secured before transport. The team must be nearly fanatical about this issue. Loss of an airway is to be avoided at all costs. Third, as previously stated, the airway should be checked after each transfer of the patient from one stretcher or vehicle to another. Finally, intubated patients should have continuous end-tidal CO_2, as well as pulse oximetry and cardiorespiratory monitoring, during transport. These technologies allow rapid detection of displaced or ineffective tracheal tubes and are particularly important in aeromedical transport, where other methods of assessment, such as auscultation, are difficult to use.

Even under the best of circumstances, a tracheal tube may be lost during transport. When this occurs, the team members should first remember that their objective is to oxygenate and ventilate the patient. If this can be safely accomplished with a bag-valve-mask device, this type of ventilation should be used initially and perhaps continued for the remainder of the transport process. Reintubation, if necessary, should be performed by the most experienced team member. Each transport team should have at least one member skilled in airway management. Airway skills should not be limited to bag and mask ventilation and tracheal intubation. Transport team members should be prepared to perform needle or surgical cricothyroidotomy and/or retrograde intubation if necessary.

Status Epilepticus

The management of status epilepticus during transport differs little from its management in an ED. The initial approach is to address the ABCs. For most children, this involves provision of supplemental oxygen and appropriate positioning to

ensure a patent airway. Once these issues have been addressed, the team must treat the seizure itself. This activity is divided into two components that should be accomplished simultaneously. The team should attempt to identify immediately treatable causes for the seizures while concurrently preparing to administer anticonvulsant medications. Monitoring oxygen saturation and administering supplemental oxygen will identify and eliminate hypoxemia as a possible cause. Hypoglycemia may be identified by a simple finger-stick glucose. Newer modalities (e.g., handheld multichannel testing devices) can identify hyponatremia and other electrolyte disorders. If any of these causes are identified, appropriate treatment can be initiated.

In many cases, the patient will require anticonvulsant medications; the initial drug of choice is usually a benzodiazepine. In an ED, the usual agent chosen is lorazepam. However, lorazepam should be refrigerated, making it problematic for transport services. Alternatives to lorazepam include diazepam and midazolam. When possible, anticonvulsants should be given intravenously; however, if the patient does not have an IV line, he or she can be given rectal medication. Alternatively, midazolam and fosphenytoin may be given intramuscularly. Anticonvulsants may, of course, be given via the interosseous route. Finally, as a last resort, benzodiazepines may be given via a tracheal tube. If the initial dose of a benzodiazepine fails to stop the seizure within 10 minutes, the same dose may be repeated up to two more times. As more doses are given, the risk of respiratory depression increases, and the transport team should be prepared to support respiration.

If the patient continues to seize after a reasonable number of doses of a benzodiazepine, an alternative agent should be administered. Phenytoin, fosphenytoin, and phenobarbital are the second-line agents most often chosen. Phenytoin will not adversely affect the child's respiratory status and is an inexpensive and effective anticonvulsant. Unfortunately, phenytoin can be administered no faster than 1 mg/kg per minute without risk of bradycardia and is very toxic to tissues. If a child's IV line infiltrates while he or she is receiving phenytoin, a severe skin injury is likely to result. Like phenytoin, fosphenytoin will not contribute to respiratory depression. Fosphenytoin is not toxic to tissues and may be administered much faster than phenytoin. As noted, fosphenytoin may be safely given intramuscularly. It is, however, more expensive than phenytoin.

Phenobarbital will act synergistically with the benzodiazepines to cause severe respiratory depression. It also has cardiovascular depressant effects, making it a poor choice for children with hypovolemia, septic shock, and other forms of circulatory impairment. The use of phenobarbital will also likely depress the patient's mental status during the postictal period, making acute neurologic assessment more difficult. If this agent is chosen, the transport team should ensure that the patient has a large-bore IV line capable of supporting the circulation. Furthermore, elective intubation should be considered before initiating patient transport.

When both the first- and second-line agents have failed, the patient should undergo elective tracheal intubation and be placed on a continuous infusion of midazolam or pentobarbital. If these measures fail to control the seizures or cannot be initiated, the child should be given a nondepolarizing neuromuscular blocking agent and adequate sedation during transport. The purpose of neuromuscular blockade is protection of the patient and the team during transport. Neuromuscular blockade ensures adequate ventilation of the patient and prevents the patient from inadvertently harming himself or herself or a team member. It is particularly dangerous to attempt to transport a seizing patient by helicopter unless he or she is intubated, sedated, and paralyzed.

Active Cardiac Arrest

The reasons for transport should always be fully explored. Transport to avoid stopping a resuscitation when there is not an expected change in therapy is inappropriate. Transport teams may have different philosophies about response to calls for patients receiving active resuscitation. Many will not mobilize until the patient has been stabilized. Others will mobilize to offer assistance if needed or in anticipation of impending stabilization but will not transport the child unless he or she is stabilized. Still other teams may accept the patient for transport during resuscitation if there is a modality available at the receiving hospital that might make a difference, such as extracorporeal membrane oxygenation (ECMO). It is important to recognize that transfer of a patient with ongoing cardiopulmonary resuscitation (CPR) can be dangerous for the transport team because they must remain unsecured in the vehicle. It can also be difficult for families, who may expect that transfer suggests a positive resolution of the medical crisis, when it is clear to the medical personnel that the outcome will be poor. In general, transport of the patient with active CPR should be arranged—but not undertaken—until adequate stabilization has been achieved.

Shock

Transport of the hypotensive or hypovolemic patient must be anticipated. Care is similar to that performed in an ED, although the supplies and vascular access may be limited. Ensuring and securing initial and perhaps secondary IV access routes before transport is important. Almost all transported patients should have at least one patent IV line. Those with reliance on those lines should have backup lines in place.

Shock management is fundamentally the same in transport as it is in the hospital. Adequate volume status should be ensured and augmented with isotonic fluids, colloid, or blood products as indicated. Vasoactive drugs should available if required. Assessment should use all components of evaluation, including mental status, capillary refill, skin color, urine output, acid-base status, and vital signs. The transport team is responsible for assimilating and communicating all the care to date, so it is helpful to have a running total of all fluids and medications administered when the transport team arrives. This will help ensure adequate patient evaluation and continuation of appropriate care.

Trauma

Transport of the severely injured trauma patient is similar to the transport of the medical patients. If an urgent or potentially surgical lesion is present that cannot be adequately managed at the referral site, rapid transport is important. Adequate assessment and management of the patient during transport is imperative, and vigilance to the ABCs, including

cervical spine immobilization, is crucial. In general, clearing of the cervical spine should not be attempted by the transport team, but by the receiving service. Appropriate cervical immobilization of the patient should be maintained throughout the transfer process. Blood products should be available for the transport team if they might be needed during the transport. Inclusion of all patient radiographs and studies can help ensure a more efficient evaluation at the receiving center. Notification of the receiving hospital and service regarding transport of a trauma patient with a potential surgical lesion is important to allow for preparations to be made. A rapid transport with simultaneous preparation of an operating room can lead to improved outcomes for these patients.

Sedation

Many children will require sedation or analgesia during transport. These children can be can be divided into three general categories of patients. The first are the children who required analgesia or sedation at the referral hospital and most likely need to continue sedation medications during and after the transport. The postoperative patient, the child with sickle cell disease, and the patient with a femur fracture are examples. The second category involves children whose disease process and sedation requirements are evolving. This category might include the child who is intubated just before or during transport, the trauma patient who has deteriorated and requires insertion of a chest tube, or the patient whose mental status has improved enough to allow recognition of chest tube–related pain. The third category includes children for whom the transport process itself may sufficiently increase pain or anxiety enough to warrant additional analgesia or sedation. This group might include the child with a long-bone fracture or peritonitis who, while comfortable in a quiet, stationary hospital bed, experiences significant pain during the transport. Each time a patient is moved (e.g., hospital to ambulance, ambulance to helicopter, helicopter to plane) additional stress or pain can occur. In addition, the stress of a loud transport environment with accompanying vibration and temperature fluctuations can lead to reactive pathophysiologic changes. If necessary, the level of sedation and analgesia can be titrated in response to these environmental issues.

The stress of leaving family members and the concept of abandonment are potential issues for the child who needs interfacility transport. The transport team should be cognizant of nonpharmacologic pain and anxiety management options, including calming and distraction, positioning and immobilization, and play therapy. A parent, depending on his or her own level of anxiety, may be helpful in these situations. The parent may be of assistance with immobilization, calming, and distraction. The family should also be informed about transport and hospital expectations so that they can be of assistance in preparing the child for the ambulance journey as well as any procedures or diagnostic tests that will be performed at the receiving institution. Accompaniment of a family member during the transport can help the patient adjust to the new environment(s) and, perhaps, lessen the need for pharmacologic intervention.

Appropriate-sized airway equipment and resuscitation medications should be available for any patient receiving sedation or analgesia during transport. These medications should include reversal agents such as naloxone and flumazenil. Naloxone is a benign medication that has rapid and dramatic effects when administered to reverse the respiratory depressant actions of opiate overdose. The effects of naloxone, however, last only a short time, and the drug may need to be repeated during the transport. Titration of naloxone via infusion may be necessary for patients who are in severe pain yet experiencing respiratory compromise or loss of protective airway reflexes. This can be accomplished by placing 2 to 4 mg/kg of naloxone into 100 mL of saline and titrating to reverse the untoward effects while maintaining some pain relief. Naloxone can be useful for the patient who has received a large dose of fentanyl and is experiencing chest wall rigidity.

Flumazenil has been used to reverse adverse effects of therapeutic benzodiazepine administration. The recommended dose is 0.01 mg/kg, with a maximum initial dose of 0.2 mg. The drug may be repeated (0.005 mg/kg) every 1 minute to a total maximum dose of 1 mg. Because seizures are a potential side effect of flumazenil, it should not be used in patients who have ingested multiple medications or who have a propensity for seizures.

Suggested Readings

Aoki BY, McCloskey K. Evaluation, Stabilization, and Transport of the Critically Ill Child. St Louis: Mosby Year-Book, 1992.

Beyer AJ, Land G, Zaritsky A. Nonphysician transport of intubated pediatric patients: a system evaluation. Crit Care Med 1992;20:961–966.

Bhende MS, Karr VA, Wiltsie DC, Orr RA. Evaluation of a portable infrared end-tidal carbon dioxide monitor during pediatric interhospital transport. Pediatrics 1995;95:875–878.

Black RE, Mayer T, Walker ML, et al. Air transport of pediatric emergency cases. N Engl J Med 1992;307:1465–1468.

Boyko SM. Interfacility transfer guidelines: an easy reference to help hospitals decide on appropriate vehicles and staffing for transfers. J Emerg Nursing 1994;20:18–23.

Brink LW, Neuman B, Wynn J. Transport of the critically ill patient with upper airway obstruction. Crit Care Clin 1992;8:633–647.

Britto J, Nadel S, Maconochie I, et al. Morbidity and severity of illness during interhospital transfer: impact of a specialized paediatric retrieval team. Br Med J 1995;311:836–839.

Bruhn JD, Williams KA, Aghababian R. True costs of air medical vs. ground ambulance systems. Air Medical J 1993;August:262–268.

Burney RE, Hubert D, Passini IL, Maio R. Variation in air medical outcomes by crew composition: a two-year follow-up. Ann Emerg Med 1995;25:187–192.

Day S, McCloskey K, Orr R, et al. Pediatric interhospital critical care transport: Consensus of a national leadership conference. Pediatrics 1991;88:696–704.

Day SE. Intra-transport stabilization and management of the pediatric patient. Pediatr Clin North Am 1993;40:263–274.

Day SE, Chapman RA. Transport of critically ill patients in need of extracorporeal life support. Crit Care Clin 1992;8:581–596.

Diekema DS. Unwinding the COBRA: new perspectives on EMTALA. Pediatr Emerg Care 1995;11:243–248.

Donnelly JA, Smith EA, Hope AT, Alexander RJT. An assessment of portable carbon dioxide monitors during interhospital transfer. Anaesthesia 1995;50:703–705.

Edge WE, Kanter RK, Weigle CGM, Walsh RF. Reduction of morbidity in interhospital transport by specialized pediatric staff. Crit Care Med 1994;22:1186–1191.

Fletcher MA, MacDonald. Atlas of Procedures in Neonatology. 2nd ed. Philadelphia: JB Lippincott, 1993.

Henning R, McNamara V. Difficulties encountered in transport of the critically ill child. Pediatr Emerg Care 1991;7:133–137.

Housel FB, Pearson D, Rhee KJ, Yomada J. Does the substitution of a resident for a flight nurse alter scene time? J Emerg Med 1995;13:151–153.

Hunt RC, Brown LH, Cabinum ES, et al. Is ambulance transport time with lights and siren faster than that without? Ann Emerg Med 1995;25:507–511.

Jaimovich DG, Vidyasagar D, eds. Handbook of Pediatric and Neonatal Transport Medicine. Philadelphia: Hanley and Belfus, 1996.

Kanter RK, Boeing NM, Hannan WP, Kanter DL. Excess morbidity associated with interhospital transport. Pediatrics 1992;90:893–898.

Kerr HD, Byrd JC, Stuart RW, Whitcomb J. Interhospital transfers, the COBRA Law (1985) and the use of a public assistance health plan. South Med J 1993;86:1210–1214.

Kronick JB, Frewen TC, Kissoon N, et al. Influence of referring physicians on interventions by a pediatric and neonatal critical care transport team. Pediatr Emerg Care 1996;12:73–77.

Lee A, Lum ME, Beehan SJ, Hillman KM. Interhospital transfers: decision-making in critical care areas. Crit Care Med 1996;24:618–622.

McCloskey KA, Hackel A, Notterman D, et al. Guidelines for Air and Ground Transport of Neonatal and Pediatric Patients. Task Force on Interhospital Transport, American Academy of Pediatrics, Washington, DC, 1993.

McCloskey KA, Orr RA, eds. Pediatric Transport Medicine. St Louis: Mosby, 1995.

Macnab AJ. Paediatric interfacility transport: organization and principles. Paedriatr Anaesth 1994;4:351–357.

McDonald TB, Berkowitz RA. Airway management and sedation for pediatric transport. Pediatr Clin North Am 1993;40:381–407.

Moront ML, Gotschall CS, Eichelberger MR. Helicopter transport of injured children: system effectiveness and triage criteria. J Pediatr Surg 1996;31:1183–1188.

Orr RA, Venkataraman ST, Cinoman MI, et al. Pretransport pediatric risk of mortality (PRISM) score underestimates the requirement for intensive care or major interventions during interhospital transport. Crit Care Med 1994;22:101–107.

Prasad NH, Brown LH, Ausband SC, et al. Prehospital blood pressures: inaccuracies caused by ambulance noise? Am J Emerg Med 1994;12:617–620.

Reinhart RO. Basic Flight Physiology. 2nd ed. New York: McGraw Hill, 1996.

Schneider C, Gomez M, Lee R. Evaluation of ground ambulance, rotor-wing, and fixed-wing aircraft services. Crit Care Clin 1992;8:533–564.

Seidel J. Preparing for pediatric emergencies. Pediatr Rev 1995;16: 466–472.

Spaite DW, Criss EA, Valenzuela TD. Air medical transport of trauma patients: air versus ground transport. Top Emerg Med 1994;16:73–80.

Strauss RH, Rooney B. Critical care pediatrician-led aeromedical transports: physician interventions and predictiveness of outcome. Pediatr Emerg Care 1993;9:270–274.

Strobos J. Tightening the screw: statutory and legal supervision of interhospital patient transfers. Ann Emerg Med 1991;20:302–310.

Szem JW, Hydo LJ, Fischer E, et al. High-risk intrahospital transport of critically ill patients: Safety and outcome of the necessary “road trip.” Crit Care Med 1995;23:1660–1666.

Thomas SH, Stone CK, Bryan-Berge D. The ability to perform closed chest compressions in helicopters. Am J Emerg Med 1994;12:296–298.

Tobias JD, Lynch A, Garrett J. Alterations of end-tidal carbon dioxide during the intrahospital transport of children. Pediatr Emerg Care 1996;12:249–251.

Weil TP. Health care reform and air medical transport services. J Emerg Med 1995;13:381–387.

Woodward GA. Transport case 1: a time to fly? A dilemma in pediatric transport. Pediatr Emerg Care 1996;12:122–125.

Woodward GA, Fein JA. Transport of Sedated Patients. In: Kraus B, Brustowicz R, eds. The Practice of Pediatric Sedation and Analgesia in the Outpatient Setting. Baltimore: Williams & Wilkins, 1999.

Woodward GA, King B. “Interfacility” transport from the home or office. Pediatr Emerg Care 1997;13:164–168.

Woodward GA, Levy RJ, Weinzimer SA. Diabetes and transport: a potentially bittersweet combination. Pediatr Emerg Care 1998;14:71–76.

Woodward GA, Moore KJ, Needle MN. Oncology and transport: beware the presentation and anticipate the clinical course. Pediatr Emerg Care 1996;12:454–459.

Woodward GA, Somogyvari Z. The Hungarian (Budapest) neonatal interfacility transport system: insight into program development and results. Pediatr Emerg Care 1997;13:290–293.

Woodward GA, Wernovsky G, Rhodes LA, et al. Sepsis, septic shock, acute abdomen? The ability of cardiac disease to mimic other illness. Pediatr Emerg Care 1996;12:317–324.

Yamamoto LG. Wireless teleradiology and fax using cellular phones and notebook PCs for instant access to consultants. Am J Emerg Med

1995;13:184–187.

Yamamoto LG, Inaba AS, DiMauro R. Personal computer teleradiology interhospital image transmission to facilitate tertiary pediatric telephone consultation and patient transfer: soft-tissue lateral neck and elbow radiographs. Pediatr Emerg Care 1994;10:237–277.

Youngberg BJ. Medical-legal considerations involved in the transport of critically ill patients. Crit Care Clin 1992;8:501–514.

CHAPTER 8

Acute Myocardial Infarction

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Although acute myocardial infarction (AMI) represents one of the most common diagnoses in adult emergency medicine, it is a rare event in the pediatric emergency department (ED). When it does occur, AMI is associated with a high morbidity and mortality if not recognized and treated. Five million patients with chest pain are seen in EDs in the United States each year. Of these patients, 1.5 million have an AMI, and approximately 450,000 die from the initial event. Almost half of these deaths occur in the prehospital setting. Early recognition and treatment are critical in lowering the morbidity and mortality associated with AMI. Pediatric EDs are not routinely set up to provide definitive treatment for patients with AMI. Therefore, pediatric emergency physicians must be prepared to recognize, acutely stabilize, and transfer the patient with an AMI. This chapter focuses on the recognition and initial stabilization of AMI in the pediatric ED.

HISTORICAL PERSPECTIVE

There have been two eras in the evolution of ED management of AMI: the prethrombolytic era (ending in the late 1970s) and the thrombolytic era (early 1980s to the present). In the prethrombolytic era, the mainstay of ED treatment of AMI was analgesics (morphine) and nitrates (nitroglycerin [NTG]). In the thrombolytic era, treatment focus shifted from antianginal treatment to clot lysis with an imperative to significantly reduce “door-to-drug” time.

The thrombolytic era has evolved in two phases: the intracoronary administration phase and the peripheral administration phase. In the early 1980s, multicenter clinical trials on the use of intracoronary thrombolytic agents demonstrated their ability to decrease morbidity and mortality associated with AMI by limiting infarct size through reperfusion of ischemic myocardium. During this period, patients with clinical criteria for AMI were taken to cardiac catheterization and given intracoronary streptokinase after demonstration of coronary occlusion by angiography. Once the benefits of thrombolytic therapy for AMI were clearly demonstrated, it was a relatively short time before large multicenter clinical trials showed that intravenous (IV) administration was as effective as intracoronary administration. In these studies, the patients who benefited most from thrombolytic therapy received treatment within 4 hours of onset of symptoms. Based on this data, a concerted effort was made to educate emergency physicians about the benefits of reducing door-to-needle time (time from presentation to the ED to the initiation of thrombolytic therapy). These studies lead to the current ED strategy for the management of AMI, which can be summarized as follows:

1. Early prehospital recognition of AMI (through sustained paramedic education and increasing availability of 12-lead electrocardiogram [ECG] in the prehospital setting)
2. National target to reduce door-to-needle time to 30 minutes or less
3. Peripheral thrombolytic administration in the ED
4. Enhanced cardiac specific serial markers for early detection of myocardial injury
5. ED 24-hour observation units and chest pain centers for management of patients with chest pain who are assessed to be at low risk for AMI (“soft rule-outs”)

PATHOPHYSIOLOGY

The pathophysiology of AMI evolves in three phases: progressive coronary artery narrowing from atherosclerotic plaque formation and deposition, acute reduction in coronary blood flow from platelet aggregation and subsequent thrombus

occlusion at the site of atherosclerotic narrowing, and myocardial injury.

The extent of damage to myocardial tissue from an acute reduction in coronary perfusion depends on multiple factors, including the extent of occlusion (partial or total), the location of the occlusion and the area of the myocardium supplied by the occluded vessel, the degree of existing collateral circulation around the occluded segment, and the myocardial oxygen demand at the time of occlusion (e.g., was the patient at rest or exercising at the time of occlusion).

CLINICAL MANIFESTATIONS

When the patient is suspected of having an AMI, a directed history and physical examination are essential in optimizing time to treatment and transfer. The key features of such an approach are discussed in the following sections and summarized in Table 8.1 and Table 8.2.

Presence of Pain Are you in pain right now?	
Time Frame When did the pain start?	
Activity Level What were you doing when the pain started? Did the pain occur at rest or with exertion?	
Pain Location Where is the pain?	
Pain Radiation Is the pain just in your chest or does it travel (to your shoulder, arm, neck, jaw, back)?	
Pain Characteristics What does the pain feel like (sharp, dull, heavy, crushing, squeezing, tight, crampy)?	
Associated Symptoms When the pain began, did you feel (sweaty, nauseous, short of breath, dizzy, weak)?	
Medical History Do you have any underlying medical problems? Have you ever had pain like this before? If yes, how many times and what was the frequency and pattern?	
Current Medications Do you take any medications regularly?	

Table 8.1. Acute Myocardial Infarction: Clinical History

Chest pain	Nausea/vomiting
Diaphoresis	Syncope or near-syncope
Pallor	Jaw pain or numbness
Anxiety	Neck pain
Confusion	Shoulder pain
Shortness of breath	Arm pain
Generalized weakness	Back pain

Table 8.2. Acute Myocardial Infarction: Initial Presentation

Presence of Acute Pain

Although acute chest pain is the most common presenting symptom in AMI, it may be atypical or absent. Classic chest pain associated with AMI is substernal or in the left side of the chest. Atypical chest pain may be in the right side of the chest, shoulder, upper arm, jaw, neck, or back. AMI without chest pain may occur in selected patient populations, including those who are elderly, diabetic, and hypertensive.

Time Frame

The time frame, from onset of pain to presentation in ED, in the patient with suspected AMI is a critical determinant of whether the patient is a thrombolytic candidate. A narrow window, of approximately 12 hours from onset of symptoms, exists during which patients have been shown to benefit from thrombolytic therapy. Furthermore, patients who receive thrombolysis within 4 hours of onset of symptoms have better outcomes in terms of myocardial salvage and reduced mortality than those patients who receive thrombolysis between 4 and 12 hours after onset of symptoms.

Activity Level

Pain may begin at rest or with exertion. Unlike angina pectoris, the exertional pain of AMI is usually not relieved by rest.

Location of the Pain

Substernal chest pain is the most common location of AMI pain. Atypical pain associated with AMI may occur only in the right side of the chest, one or both shoulders, one or both upper arms and/or elbows, the neck or jaw, the back (especially between the scapula), and even as a tight band around the upper abdomen. Periumbilical and lower abdominal pain are not usually associated with AMI.

Radiation of Pain

Radiation of the pain of AMI to the shoulder, neck, jaw, and back is common, and the patient should be specifically asked about pain radiating to any of these locations. Arm heaviness, weakness, or paresthesias are symptoms commonly associated with AMI.

Characteristics of the Pain

Numerous terms are used to describe and characterize the sensation of AMI pain (e.g., sharp, dull, heavy, crushing, squeezing, tight). It is important to find the appropriate metaphor or metaphors that fit with what the patient is experiencing. A patient may say that his or her chest or arm is sore, heavy, numb, or tight but deny having “pain.”

Associated Symptoms

Diaphoresis, nausea/vomiting, dizziness/light-headedness, dyspnea/shortness of breath, and fatigue/generalized weakness are all classically associated with AMI. Particularly worrisome symptoms include the following:

- Significant diaphoresis: This can range from the patient with cool and clammy skin to the patient literally drenched in sweat.
- Pallor: This is often described as an ashen appearance.
- Marked anxiety and restlessness: Some patients who are acutely infarcting may experience profound anxiety and even a premonition of doom.
- Dyspnea: This may herald the initial presentation AMI in the elderly as acute left ventricular failure. *Shortness of breath without chest pain may be the sole presentation of AMI in the elderly or diabetic patient.*
- Chest pain is preceded by syncope or near-syncope.
- Profound generalized weakness: Patients with AMI commonly report feeling as if all the energy had been suddenly drained from their body.
- Acute confusion or change in mental status: A spectrum from mild dizziness and light-headedness to confusion and disorientation is seen.
- Palpitations are present in the setting of chest pain.

Medical History

Establishing whether patients have had previous episodes of chest pain is useful in determining whether they are having a typical bout of angina (stable angina) or whether the pattern suggests a worsening from their baseline (unstable angina). Stable angina is characterized by a typical pattern of pain (e.g., one to two times per week with exertion, relieved by rest or a single dose of NTG). Unstable angina is defined as a worsening or escalating pattern from baseline (initially one to two times per week with exertion, relieved by rest or a single dose of NTG, now once a day with exertion relieved only by multiple doses of NTG or baseline chest pain with exertion now occurring at rest with increased frequency).

In addition, questions about smoking, family history of heart disease, other relevant medical history (Table 8.3), cholesterol level, diabetes, and hypertension, are helpful in assessing the patient's risk for coronary artery disease.

Congenital	Acquired
Antithrombin III deficiency	Patient's family history
Familial combined hyperlipidemia	Drug abuse (cocaine and amphet-amines)
Familial hypercholesterolemia	
Anomalies of the coronary arteries	Kawasaki disease
Homocystinuria	Transposition repair with coronary artery switch
Williams syndrome	

Table 8.3. Pediatric Populations at Risk for Acute Myocardial Infarction

Current Medications

Medications that may provide information about a patient's coronary risk include antihypertensive agents, cholesterol-lowering agents, and antianginal drugs (calcium channel blockers, nitrates, b-blockers).

Physical Examination

Because arrhythmias and heart failure are the most common complications of AMI, the physical examination should be focused on identifying the manifestations of these states (rhythm disturbances with or without hypotension, left ventricular failure with pulmonary edema or hypotension). Signs of hypoperfusion include diaphoresis, cool extremities, and confusion, and in the setting of persistent chest pain, these signs often herald the onset of cardiogenic shock. Rales, increased jugular venous distension (especially with right ventricular infarction and right-sided failure), hepatomegaly, and pitting ankle edema often indicate volume overload in the setting of singular or biventricular failure. An extra heart sound, either an S_4 (reflecting decreased left ventricular compliance) or an S_3 gallop (indicating heart failure), may be

heard.

In the uncomplicated AMI, tachycardia and elevated systolic blood pressure are usually present. A new murmur, particularly a new systolic murmur, is of special concern because it may signify acute mitral regurgitation and papillary muscle dysfunction or ventricular septal rupture.

The location of the ischemic myocardial segment often determines the presenting signs. Inferior infarction, affecting the right coronary and nodal arteries, may cause hypotension and bradycardia secondary to localized ischemia and enhanced parasympathetic discharge. Anterior infarction, affecting the left coronary artery and sympathetic system, may lead to the opposite signs (tachycardia and/or hypertension).

Vital Signs and ECG

The initial vital signs and ECG should be scrutinized carefully for rhythm disturbances (tachycardia and tachyarrhythmias, as well as bradycardia and bradyarrhythmias), conduction abnormalities (atrioventricular blocks and new bundle branch blocks), variations in blood pressure, changes in respiratory rate, and hypoxia.

Acute ischemic changes on the initial ECG are present in only 40 to 65% of patients with AMI. Therefore, repeat or serial ECGs should be considered in patients with symptoms suggestive of AMI with an initial normal ECG. The following information should be obtained from the ECG:

- *Rate*
  Sinus tachycardia without acute changes can indicate early ischemia before the presence of ST- or T-wave changes
- *Rhythm*
  Presence of arrhythmias
- *Intervals*
  Short P-R interval indicative of preexcitation syndromes
  Prolonged P-R interval indicative of atrioventricular block
  Widened QRS indicative of bundle branch block
- *Ischemic changes* (Table 8.4 and Fig. 8.1, Fig. 8.2, Fig. 8.3 and Fig. 8.4)
  Hyperacute T waves in the anterior and/or septal leads (early sign of anterior ischemia)
  Flipped or inverted T waves
  ST-segment elevation or depression
  Location of ischemic segment

Injury Location	ECG Leads	Coronary Artery	ECG
Anterior/ anteroseptal	V_1-V_4	LAD	Figs. 8.1, 8.2
Inferior	2, 3, and aVF	RCA	Fig. 8.3
Lateral	1, aVL, V_5, and V_6	LAD, circumflex artery	Fig. 8.4

LAD, left anterior descending; RCA, right coronary artery.

Table 8.4. Characteristics of Ischemia in AMI

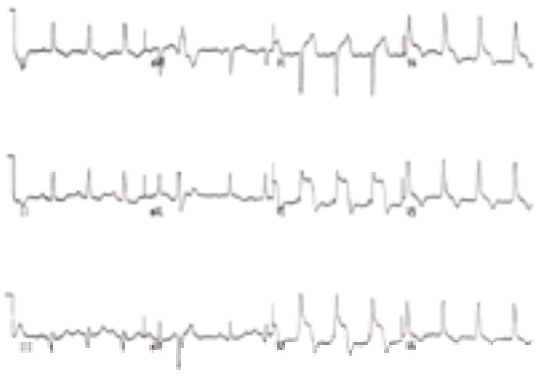

FIGURE 8.1. Anteroseptal MI with ST elevation in V_1–V_4 with reciprocal inverted T waves laterally in 1, aVL, and V_5–V_6. (Reproduced with permission from David F.M. Brown, MD.)

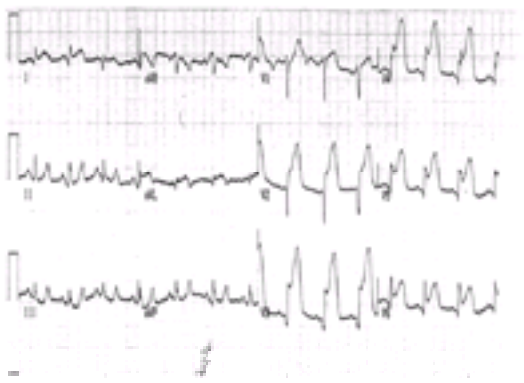

FIGURE 8.2. Anterior MI with ST elevation in V_1–V_6 and reciprocal inverted T waves laterally in 1 and aVL. (Reproduced with permission from David F.M. Brown, MD.)

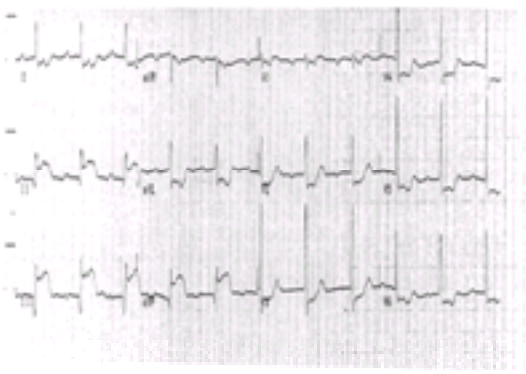

FIGURE 8.3. Inferior MI with ST elevation in 2, 3, and aVF, and reciprocal ST depression anterolaterally in 1, aVL, and V_1–V_6. (Reproduced with permission from David F.M. Brown, MD.)

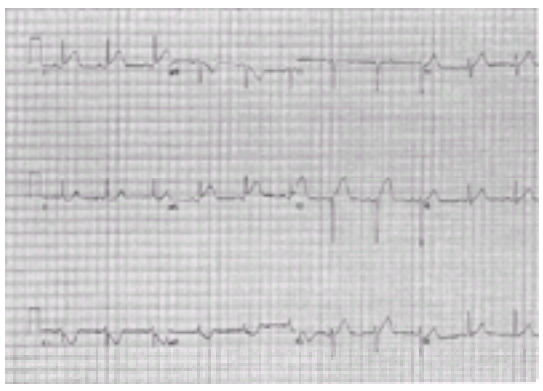

FIGURE 8.4. Lateral MI with ST elevation in 1 and aVL and reciprocal ST depression inferiorly in 3 and aVF. (Reproduced with permission from David F.M. Brown, MD.)

MANAGEMENT

The management of AMI in the pediatric ED consists of four sequential phases: early recognition of the clinical manifestations of AMI, acute stabilization, preparation for transfer, and transport.

Early Recognition

The strategy for managing patients with chest pain in the pediatric ED begins with the following steps: rapid recognition of the clinical manifestations of AMI, setting appropriate priorities, and mobilization of resources. The primary goal in managing AMI patients in the pediatric ED is acute stabilization followed by rapid transfer to the appropriate facility, whether intrahospital or interhospital, where definitive care can be provided. Management priorities should reflect this goal and time should not be wasted with unnecessary diagnostic tests or procedures that result in delay in transfer and delivery of definitive care. Resources (whether personnel, equipment, or information) from the treating facility, the receiving facility, and the prehospital system should be mobilized as soon as an AMI is recognized. Acute stabilization and treatment should be urgently initiated once the preliminary diagnosis of AMI has been made.

Acute Stabilization

The goals of acute stabilization are to decrease myocardial oxygen consumption, decrease preload, reduce afterload, and identify and treat complications.

During acute ischemia, oxygen supply to myocardial tissue is significantly compromised. Catecholamine activity increases secondary to pain and heightened anxiety. The catecholamine response further drives myocardial oxygen

consumption, which increases the rate of ischemia, creating a vicious circle. Decreasing myocardial oxygen consumption can reduce the extent of infarction. Initial treatment is therefore directed at relieving pain and anxiety and decreasing catecholamine response. Morphine sulfate (2 mg IV) and NTG (0.4 mg sublingual) are the first-line antianginal agents.

The main actions of NTG are venodilation (increased venous capacitance, leading to decreased venous return and a reduction in preload), coronary artery and collateral vessel dilation (with resultant increase in myocardial oxygen supply), and afterload reduction. A reflex tachycardia is commonly seen with NTG secondary to its potent vasodilatory properties. It is available in topical (ointment), oral, transmucosal (sublingual pills or spray), and IV formulations. In the ED treatment of AMI, the transmucosal and IV routes are the most useful. The transmucosal route provides rapid onset in 2 to 3 minutes. Expiration dates should always be checked because NTG has a relatively short shelf life. Patients usually experience a pounding sensation in the head or headache with the onset of NTG. If the patient reports no effects as described from NTG within 5 minutes, a different bottle of pills should be tried.

Topical NTG should never be used to treat acute chest pain. Its use should be restricted to maintenance therapy. Once the patient is pain free, 1 to 2 inches of NTG ointment can be applied for a sustained nitrate effect.

Morphine is an opioid that acts as a venous and arterial vasodilator, providing reduction in both preload and afterload as well as exerting a vagotonic effect resulting in a decrease in heart rate and myocardial oxygen consumption. Both NTG and morphine can cause significant vasodilation and hypotension, especially in patients with inferior wall or right ventricular infarctions, and must be used carefully. The simultaneous use of NTG and morphine should be avoided because severe hypotension may occur with a resultant increase in myocardial oxygen consumption and worsening ischemia.

Management of the uncomplicated AMI (Fig. 8.5) begins by placing the patient on a cardiac monitor, starting oxygen by nasal cannula at 2 to 4 L/minute, obtaining baseline vital signs including oxygen saturation, and securing peripheral IV access. A 12- to 15-lead ECG should then be obtained and a portable chest radiograph ordered (as long as the chest radiograph does not delay treatment). Treatment should then be initiated with chewable aspirin (160 mg) followed by NTG 0.4 mg (or $^{1}/_{150}$ grain) sublingual every 5 minutes times three or until symptoms subside. If the patient is symptom free after one to three doses of NTG, 1 to 2 inches of NTG ointment are applied and the ECG is repeated. This patient is now stabilized and ready for transfer.

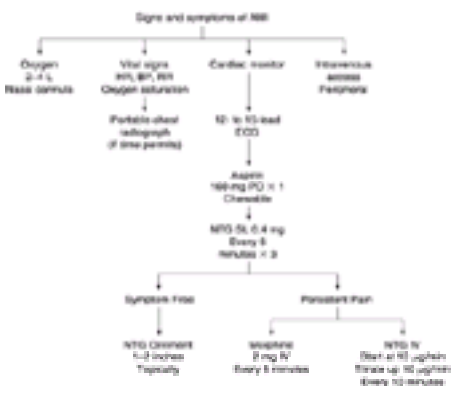

FIGURE 8.5. Treatment of uncomplicated AMI.

If the patient is not symptom free after the third NTG, two treatment options are available: morphine 2 mg IV every 5 minutes until pain subsides or NTG IV starting at 10 µg/minute and titrating upward in increments of 10 µg/minute every 10 minutes until symptoms subside. The patient whose pain or symptoms are not relieved with three sublingual NTG and who has been started on morphine or IV NTG is unstable but suitable for transfer as long as there are no active signs of heart failure or unstable cardiac rhythm or conduction disturbances (which if present, should be addressed before transfer).

Once the patient with an uncomplicated AMI is acutely stabilized, continued monitoring is essential to identify and rapidly treat emerging complications. Heart failure, cardiac arrhythmias, and conduction abnormalities can occur at any time during the course of an AMI. Patients with an uncomplicated initial course may go on to deteriorate into an unstable AMI. Figure 8.6, Figure 8.7, Figure 8.8, Figure 8.9 and Figure 8.10 illustrate the common complications of AMI and their treatment.

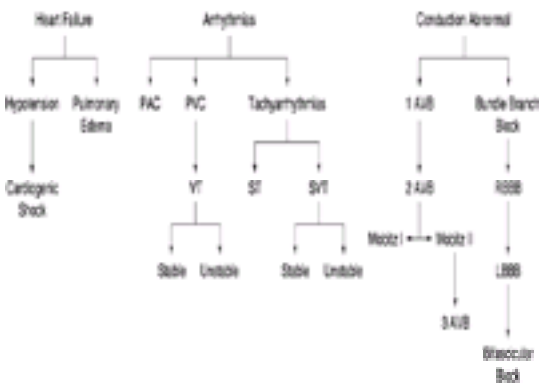

FIGURE 8.6. Complications of AMI. *1 AVB,* first-degree atrioventricular block; *2 AVB,* second-degree atrioventricular block; *3 AVB,* third-degree atrioventricular block; *PAC,* premature atrial contraction; *PVC,* premature ventricular

contraction; *ST,* sinus tachycardia; *SVT,* supraventricular tachycardia.

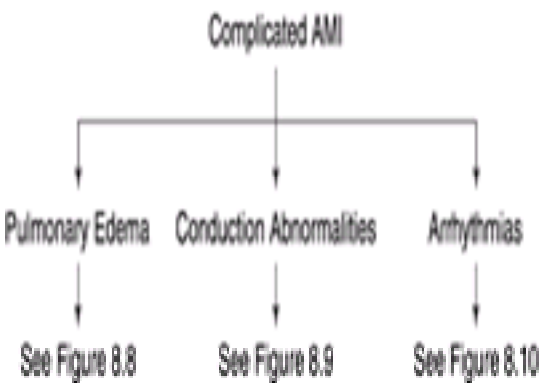

FIGURE 8.7. Treatment of complicated AMI.

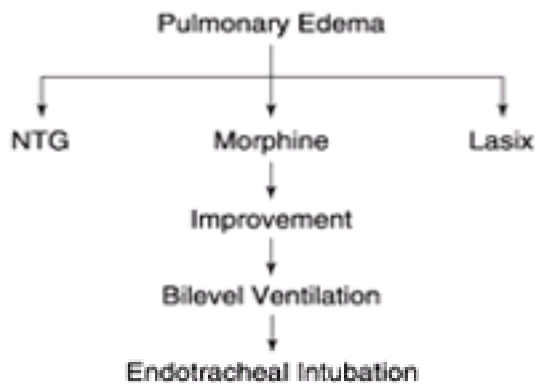

FIGURE 8.8. Pulmonary edema.

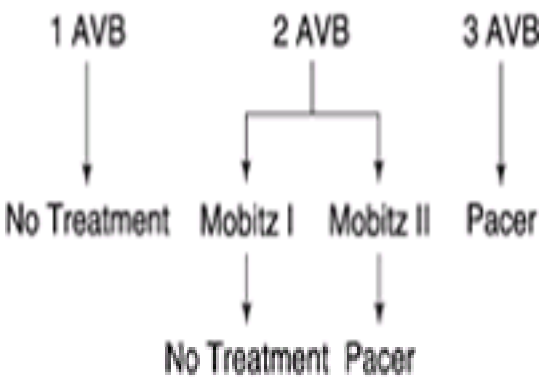

FIGURE 8.9. Conduction abnormalities. *1 AVB,* first-degree atrioventricular block; *2 AVB,* second-degree atrioventricular block; *3 AVB,* third-degree atrioventricular block.

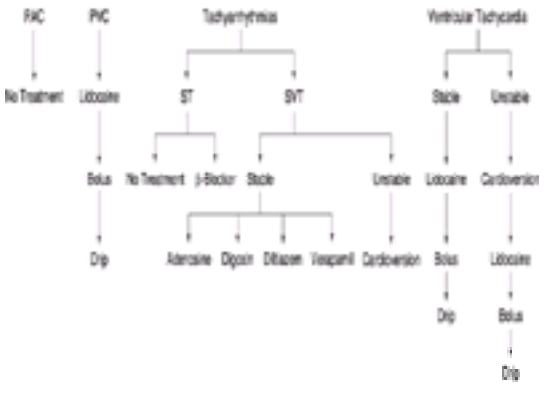

FIGURE 8.10. Arrhythmias. *PAC,* premature atrial contraction; *PVC,* premature ventricular contraction; *ST,* sinus tachycardia; *SVT,* supraventricular tachycardia.

A complicated AMI is defined, for our purposes, as AMI symptoms with heart failure and/or rhythm or conduction abnormalities. The initial management of these patients has a dual focus: global antianginal therapy and specific treatment of complications. The primary antianginal agents (NTG and morphine) can be used for global treatment of ischemia and for specific treatment of certain complications (e.g., pulmonary edema). In this case, the preload reduction

provided by NTG and/or morphine is useful to treat both ischemia and pulmonary edema.

Preparation for Transfer

While acute stabilization is under way, preparation for transfer should begin (Table 8.5). The nature and extent of the preparation depends on whether the transfer is interhospital (i.e., treating facility is not set up to provide definitive care) or intrahospital (intensive care unit, cardiac catheterization laboratory, angioplasty suite, operating room for bypass). If the transfer is interhospital, it is imperative that the treating physician alert the receiving facility that the patient is having an AMI so that the appropriate resources can be mobilized in advance of the patient's arrival. The patient's condition (stable or unstable) and whether he or she is a thrombolytic candidate (Table 8.6 and Table 8.7) must also be relayed to the receiving facility.

1. Begin arranging transfer as soon as AMI is recognized.
2. Determine mode of transport.
   - Intrahospital
   - Air
   - Ground
   - BLS with or without physician
   - ALS
3. Contact transport personnel.
4. Notify receiving facility

Table 8.5. Protocol for Transferring Patients with Acute Myocardial Infarction

1. Symptoms of AMI for at least 30 minutes and less than 12 hours
2. New ECG changes (any of the following):
   - ST elevations >1 mm in two of the anterior, inferior, or lateral leads
   - ST depressions in the anterior leads
   - New LBBB
3. No absolute contraindcations (see Table 8.7)

LBBB, left bundle branch block.

Table 8.6. Thrombolytic Therapy Candidates

Absolute	Relative
Active PUD	HTN (systolic >180 and/or diastolic >100)
Surgery or invasive procedure within 2 weeks	Brief CPR (<10 min)
Prolonged CPR (>10 min)	Chronic anticoagulation therapy
CVA within 1 year	Severe hepatic or renal disease
Suspected aortic dissection	
Gastrointestinal or genitourinary bleeding within 10 days	
Recent major trauma	

PUD, peptic ulcer disease; *CVA*, cerebrovascular accident; *CPR*, cardiopulmonary resuscitation; *BP*, blood pressure; *HTN*, hypertension.

Table 8.7. Contraindications to Thrombolytic Therapy

Transport

The treating physician in the pediatric ED must determine the appropriate mode of transport once the patient is acutely stabilized. The appropriate mode of transport will depend on the location and type of the treating facility (general or community ED, pediatric ED contiguous to adult ED, free-standing pediatric ED), the proximity of the treating facility to a facility with thrombolytic capability, and the length of transport. Transport options include ground versus air, advanced life support (ALS) transport with paramedics versus basic life support (BLS) transport with or without treating facility personnel accompanying the transport.

Suggested Readings

Amin M, Gabelman G, Karpel J, et al. Acute myocardial infarction and chest pain syndrome after cocaine use. Am J Cardiol 1990;66: 1434–1437.

Braunwald E. Acute myocardial infarction: the value of being prepared. N Engl J Med 1996;334:51–52.

Fibrinolytic Therapy Trials (FTT) Collaborative Group. Indications for thrombolytic therapy in suspected acute myocardial infarction: collaborative overview of early mortality and major morbidity results from all randomized trials of more than 1000 patients. Lancet 1994;343:311–322.

Gibler WB, Runyon JP, Levy RC, et al. A rapid diagnostic and treatment center for patients with chest pain in the emergency department. Ann Emerg Med 1995;25:1–8.

GISSI investigators. Effectiveness of intravenous thrombolytic treatment in acute myocardial infarction. Lancet 1986;8478:397–401.

Gusto Angiographic Investigators. The effects of tissue plasminogen activator, streptokinase, or both on coronary-artery patency, ventricular function, and survival after acute myocardial infarction. N Engl J Med 1993;329:1615–1622.

ISIS-2 (Second International Study of Infarct Survival) Collaborative Group. Randomized trial of intravenous streptokinase, oral aspirin, both, or neither among 17,187 cases of suspected acute myocardial infarction: ISIS-2. Lancet 1988;2:349–360.

Kato H, Ichinose E, Kawasaki T. Myocardial infarction in Kawasaki disease: clinical analyses in 195 cases. J Pediatr 1986;108:923–927.

Lee TH, Rouan GW, Weisberg MC, et al. Clinical characteristics and natural history of patients with acute myocardial infarction sent home from the emergency room. Am J Cardiol 1987;60:219–224.

Lee TH, Rouan GW, Weisberg MC, et al. Sensitivity of routine clinical criteria for diagnosing myocardial infarction within 24 hours of hospitalization. Ann Intern Med 1987;106:181–186.

McCarthy BD, Beshansky JR, D'Agostino RB, et al. Missed diagnoses of acute myocardial infarction in the emergency department: results from a multicenter study. Ann Emerg Med 1993;22:579–582.

National Heart Attack Alert Coordinating Committee. Emergency department: rapid identification and treatment of patients with acute myocardial infarction. Ann Emerg Med 1994;23:311–329.

Peters S, Vandenplas Y, Jochmans K, et al. Myocardial infarction in a neonate with hereditary antithrombin III deficiency. Acta Pediatr 1993;82:610–613.

Reeder GS, Gersh BJ. Modern management of acute myocardial infarction. Curr Probl Cardiol 1996;21:585–667.

Rouan GW, Lee TH, Cook EF, et al. Clinical characteristics and outcome of acute myocardial infarction in patients with initially normal or nonspecific electrocardiograms (a report from the multicenter chest pain study). Am J Cardiol 1989;64:1087–1092.

Ryan TJ, Anderson JL, Antman EM, et al. ACC/AHA Guidelines for the management of patients with acute myocardial infarction: executive summary. Circulation 1996;94:2341–2350.

Selig MB. Early management of acute MI: thrombolysis, angioplasty, and adjunctive therapies. Am J Emerg Med 1996;14:209–217.

Silber SH, Leo PJ, Katapadi M. Serial electrocardiograms for chest pain patients with initial nondiagnostic electrocardiograms: implications for thrombolytic therapy. Acad Emerg Med 1996;3:147–152.

Simoons ML, Boersma E, et al. Management of MI: the proper priorities. Eur Heart J 1997;18:896–899.

Sirois JG. Acute myocardial infarction. Emerg Med Clin North Am 1995;13:759–769.

Tatum JL, et al. Comprehensive strategy for the evaluation and triage of the chest pain patient. Ann Emerg Med 1997;29:116–125.

CHAPTER 9

Abdominal Distension

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For this discussion, *abdominal distension* is defined as an increase in the volume of the abdominal cavity. Apparent abdominal distension secondary to poor posture, the natural lordosis of childhood, abdominal wall weakness, obesity, and pulmonary hyperinflation is easily mistaken for true abdominal distension. Examination of the patient in both the supine and upright positions assists the clinician in recognizing these factors before diagnoses that truly increase the volume of the abdominal cavity are considered.

Abdominal distension is a nonspecific sign. That is, the causes of abdominal distension are numerous (Table 9.1). Even when the discussion is limited to emergent and urgent causes of abdominal distension, as is the case for this chapter, the list is long. When confronted with a patient with abdominal distension, one approach is to divide the causes into the following generic categories: distended bowel, free air, extraluminal fluid, massive hepatomegaly, massive splenomegaly, and other causes. This categorization is more easily described on paper than discerned at the bedside. A large cystic mass is easily mistaken for ascitic fluid. A Wilms' tumor may examine much like splenomegaly. Another difficulty in the clinical application of this categorization is that many pathologic processes that lead to abdominal distension do so through several of the previously mentioned categories. For example, kwashiorkor causes abdominal distension secondary to hepatosplenomegaly and ascites. For these reasons, the reader is urged to regard this initial categorization, when used at the bedside, as tentative, pending confirmation from plain radiograph, ultrasound, computed tomography (CT), or other imaging studies.

Table 9.1. Differential Diagnosis of Abdominal Distension

DIFFERENTIAL DIAGNOSIS

Bowel distension occurs secondary to mechanical or functional intestinal obstruction, aerophagia, malabsorption, or obstipation (Table 9.2). Mechanical obstruction most commonly occurs in infants secondary to congenital malformations (atresias, volvulus, incarcerated hernia) and intussusception and in all ages secondary to previous abdominal surgery with resulting adhesions (see Chapter 118). The lack of air in the rectum and sigmoid colon on a prone cross-table lateral of the abdomen supports the diagnosis of mechanical obstruction. Functional obstruction, or ileus, is suggested by tympanitic distension in the absence of bowel sounds. It may occur secondary to numerous causes. Some of the more important causes are described briefly (Table 9.3). Peritoneal irritation secondary to infection, pancreatic enzymes, bile, or blood is suggested by signs such as involuntary guarding and pain with movement. Fever without peritoneal signs suggests intestinal inflammation, systemic infection, or anticholinergic poisoning (see Chapter 84 and Chapter 88). Various poisonings (atroponics), toxins (botulism), and electrolyte disturbances (hypokalemia) may result in an ileus. These will most likely occur in the patient who has no abdominal findings other than tympanitic distension. In particular, the abdomen will be nontender. Finally, posttraumatic gastric distension is an extremely important entity that may result in significant respiratory embarrassment secondary to upward pressure on the diaphragm. It probably is secondary to a combination of aerophagia and ileus.

Aerophagia (crying, feeding)	Pneumonia/sepsis
Gastroenteritis	Peritonitis
Obstipation	Intra-abdominal bleeding
Pregnancy	Hemolytic disease
Traumatic ileus	Congestive heart failure
Intestinal obstruction (mechanical)	Hepatitis
Obstructive uropathy (infants)	

[a] Listed in approximate order of frequency

Table 9.2. Common Causes of Abdominal Distension[a]

Infectious	*Other*
Peritonitis	Intestinal obstruction (mechanical)
Sepsis/pneumonia	Electrolyte abnormality
Botulism	Renal failure
Pancreatitis	Poisoning
Congenital syphilis	Necrotizing enterocolitis
Hepatitis	Intestinal perforation
Tuberculosis	Shock
Congenital	Budd-Chiari syndrome
Tyrosinemia	Congestive heart failure
Galactosemia	Pericarditis
Hemolytic disease	Portal hypertension
Traumatic	Acquired immunodeficiency
Intra-abdominal bleeding	syndrome
Neoplastic	
Leukemia and other malignancies	

Table 9.3. Life-Threatening Causes of Abdominal Distension

Bulky, foul-smelling, or diarrheal stools suggest malabsorption secondary to many causes: formula enteropathies, bacterial overgrowth, parasites, cystic fibrosis, and celiac disease (see Chapter 84 and Chapter 93). Obstipation is a common cause of abdominal distension. The patient usually has a history of irregular stooling or chronic constipation. This may represent a severe functional disturbance or reflect many pathologic processes, including Hirschsprung's disease and other defects in bowel enervation and hypothyroidism.

Free peritoneal air results from intestinal perforation secondary to trauma or inflammation. It is demonstrated with an upright or cross-table lateral radiograph of the abdomen. The child will be toxic, and peritoneal signs will be present. An ileus generally contributes to the abdominal distension.

Extraluminal fluid accumulates in the abdominal cavity because of a decreased serum albumin, inflammation, bleeding, intraperitoneal chyle, and/or increased venous and lymphatic resistance through the portal and hepatic veins. A low serum albumin level is suggested by peripheral edema and pleural effusion (see Chapter 22). It occurs as a result of protein loss secondary to nephrotic syndrome or protein-losing enteropathy, or decreased protein synthesis like that which occurs in cirrhosis and malnutrition. Obstruction of blood flow through the liver is suggested by distended abdominal wall veins, a history of hemoptysis, and an enlarged spleen. The obstruction may occur at the prehepatic level (portal venous thrombosis), within the liver parenchyma (end-stage cirrhosis), at the hepatic veins (Budd-Chiari syndrome), or at the intrathoracic level (congestive heart failure [CHF], pericarditis). Obstruction at the portahepatis usually is idiopathic, although a history of umbilical venous catheterization or omphalitis in the newborn period should suggest this possibility. Obstruction at this level generally does not cause marked ascites. Although cirrhosis evolves gradually, its clinical presentation may be abrupt. It results from Wilson's disease, a $_1$-antitrypsin disease, biliary atresia, and other congenital problems, or occasionally, from chronic active hepatitis. Decreased clotting factors would be among the many laboratory findings of cirrhosis. Obstruction of flow at the hepatic veins or above occurs as a result of Budd-Chiari syndrome, CHF, or constrictive pericarditis (see Chapter 82). The liver is engorged, resulting in hepatomegaly and right upper quadrant tenderness in each of these entities.

A history of recent trauma and signs of shock point to intraperitoneal bleeding, usually secondary to a splenic or hepatic laceration. An ileus secondary to both peritoneal inflammation and shock likely contributes to the abdominal distension. Trauma in the recent past suggests chylous ascites. Finally, a diffusely tender abdomen suggests infectious peritonitis, pancreatitis, or bile peritonitis.

Extreme hepatomegaly that develops acutely occurs secondary to inflammation, engorgement, or trauma (see Chapter 93). There will be marked right upper quadrant tenderness and generally systemic toxicity. Causes include hepatitis, CHF, constrictive pericarditis, and congenital enzyme deficiencies. Other causes of extreme hepatomegaly include neoplastic disease, storage diseases, and congenital hemolytic anemias. Generally, hepatomegaly develops gradually in these conditions and is accompanied by many other signs of chronic illness. If neoplastic disease is suspected because of suspicious lymphadenopathy or other masses or if trauma is a consideration, an abdominal tomographic scan should be considered.

Extreme splenomegaly without marked hepatomegaly in the toxic-appearing child suggests intraparenchymal bleeding with an intact capsule, sickle cell sequestration crisis, or malaria (see Chapter 84 and Chapter 87). In the nontoxic child, portal hypertension, neoplastic disease, and chronic hemolysis should be suspected. Neoplastic disease often results in

a spleen with an irregular surface. Abdominal CT scan again is useful in identifying this possibility. Chronic hemolysis secondary to sickle cell disease, b-thalassemia, and hereditary spherocytosis may result in a very large spleen. In the case of hemoglobin SS disease, but not hemoglobin SC disease or sickle-thalassemia, splenic enlargement is followed by splenic atrophy beyond 5 years of age. A peripheral blood smear generally identifies this group of causes of massive splenomegaly (see Chapter 87).

Other causes of abdominal distension include cysts, tumors, uterine enlargement, obstructive uropathy, bowel duplication, and inflammation. Cystic lesions include ovarian cysts; mesenteric, omental, or peritoneal cysts; choledochal cysts; and polycystic kidneys. These conditions generally present with a subacute history and physical examination. The exception is torsion of the large ovarian cyst, which produces vomiting and marked abdominal pain. Abdominal ultrasound generally identifies intra-abdominal cysts readily. Of course, an abdominal CT scan also is diagnostic. Obstructive uropathy is probably the most common cause of abdominal distension in early infancy. Although often normal, an abnormal urinalysis or blood urea nitrogen (BUN):creatinine ratio would add support to the diagnosis of obstructive uropathy. Tumors such as Wilms' tumor, an ovarian tumor, and a teratoma generally can be palpated easily as firm, discrete abdominal masses by the time they are causing frank abdominal distension (see Chapter 100). Bowel duplication can be a subtle diagnosis until a complication such as mechanical bowel obstruction or hematochezia develops. A contrast CT scan of the abdomen, however, generally confirms this diagnosis once suspected. Regional enteritis with sufficient inflammatory mass to cause abdominal distension is preceded by a long history of obstructive and malabsorptive symptoms. Acute phase reactants such as the sedimentation rate are likely to be abnormal in regional enteritis. Finally, a midline pelvic mass should suggest pregnancy or hematocolpos.

EVALUATION AND DECISION

History

The history should attempt first to differentiate acute from chronic symptomatology, focusing on rate of progression, recent trauma, weight loss, or weight gain. Next, systemic signs such as fever, anorexia, edema, and lethargy further define the acuteness of the problem and, to some degree, narrow the diagnostic possibilities. One must always be on the alert, however, for an acute complication superimposed on a more subtle chronic condition. Next, symptoms relative to specific organs, including the gastrointestinal (GI), renal, cardiac, and gynecologic systems, should be pursued. These symptoms include vomiting (bilious or nonbilious), abdominal pain, stool history, shortness of breath, cough, hemoptysis, urine output (including strength of stream and any abnormality of urinary color or foamy urine), and menstrual history. Finally, a family history of anemia, early infant death among relatives or metabolic disease, a travel history, and a careful newborn history may be revealing.

Physical Examination

After ruling out life-threatening respiratory embarrassment and shock, the physical examination should focus on determining whether the generic cause of the abdominal distension is bowel (air or stool) (Fig. 9.1), free fluid (Fig. 9.2), massive hepatomegaly (Fig. 9.3), massive splenomegaly (Fig. 9.4), inspissated stool, or a discrete mass (Fig. 9.5). A tympanitic abdomen with intestinal outlines on the anterior abdominal wall suggests bowel distension as the cause of the abdominal enlargement. Tympani in a toxic child without intestinal outlines suggest free air. A fluid wave and dullness suggest ascites. Palpable loops of bowel or a palpable descending colon suggests stool. Massive hepatomegaly and splenomegaly generally are defined easily by palpation. The examiner must be cautious, however, because other masses may mimic hepatomegaly and, in particular, splenomegaly. Other key physical findings include signs of CHF, abdominal tenderness, edema, signs of trauma or easy bruising, lymphadenopathy, pallor, and jaundice. A rectal examination for a mass, tenderness, and the presence or absence of stool is also helpful. More specific findings may be pursued once an initial hypothesis is made based on the algorithms in this chapter.

FIGURE 9.1. Bowel distension.

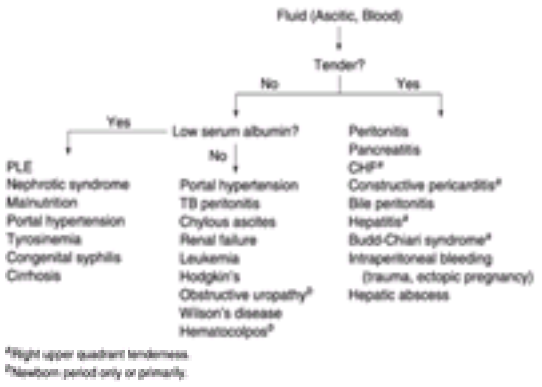

FIGURE 9.2. Fluid (ascitic, blood).

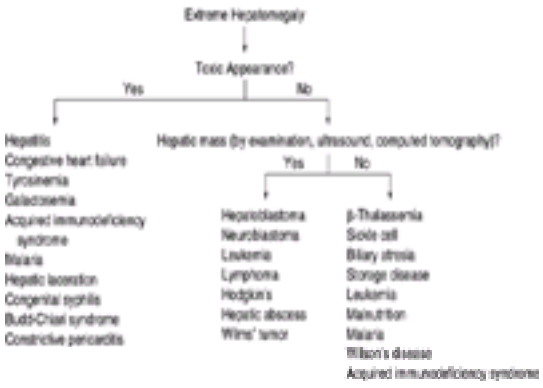

FIGURE 9.3. Extreme hepatomegaly.

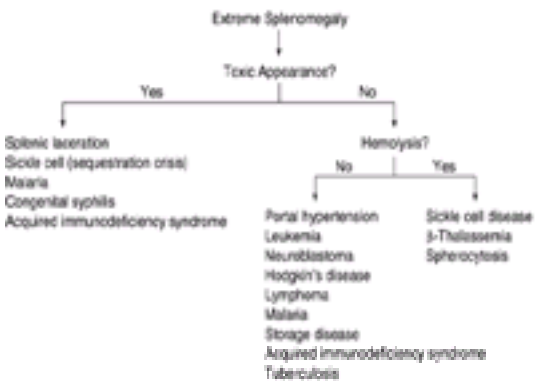

FIGURE 9.4. Extreme splenomegaly.

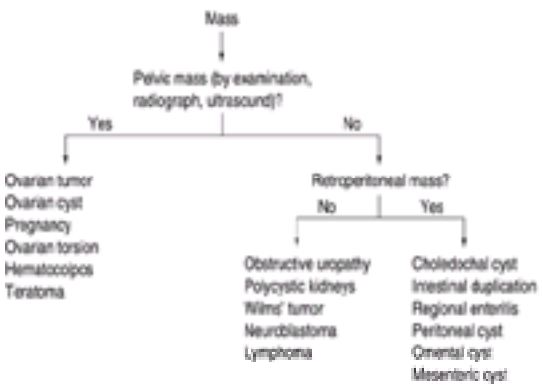

FIGURE 9.5. Mass.

Laboratory

The initial laboratory evaluation of abdominal distension is determined by the clinical findings and may include complete blood count with smear, erythrocyte sedimentation rate, reticulocyte count; liver function tests, including serum albumin and clotting studies; electrolytes with BUN, creatinine, and amylase; a urinalysis with reducing substances; and a chest radiograph and a two-view abdomen plain radiograph. If intestinal obstruction is suspected, one of the plain radiographs should be a prone cross-table lateral to determine the presence or absence of air in the rectum and sigmoid colon. It is preferable that this study be performed before a rectal examination is done.

Often, after the initial history, physical examination and laboratory evaluation imaging studies will be necessary. Ultrasound, if available, is an excellent first step because it does not involve radiation and often produces a definitive

answer. If intestinal obstruction is suspected, an upper GI series is the preferred study (or an air-contrast or barium enema if intussusception is suspected). An abdominal CT scan is the preferred study to evaluate abdominal distension thought to be secondary to trauma.

Management

Abdominal distension by itself may represent a medical emergency. First, this occurs when the distension is so severe that diaphragmatic excursion is compromised. For example, gastric and bowel distension secondary to aerophagia and ileus posttrauma may significantly impair a child's respiratory status. Massive ascites and free peritoneal air also may compromise respiration. Therefore, the first step in management is to assess and stabilize the child's respiratory status, including positive-pressure ventilation or emergent relief of distension or both. Passage of a nasogastric or orogastric tube may result in dramatic improvement in the child's respiratory status.

The second, far less common, situation in which abdominal distension may represent an emergent situation in itself is compression of the inferior vena cava (IVC), resulting in a compromised cardiovascular status. For example, occasionally, a child with severe obstipation may present with weak pulses and cool extremities. In this situation, a fluid bolus plus disimpaction will improve the patient's perfusion status rapidly. Managing the child in the lateral decubitus position may relieve pressure on the IVC. When the airway, breathing, and circulation have been stabilized, the diagnostic evaluation can proceed with laboratory and imagining studies as discussed previously.

Suggested Readings

Oski FA. Principles and Practice of Pediatrics. Philadelphia: JB Lippincott, 1990.

Tunnessen WW. Signs and Symptoms in Pediatrics. Philadelphia: JB Lippincott, 1988.

Walker WA, Durie PR. Pediatric Gastrointestinal Disease. Philadelphia, BC Decker, 1991.

Ziai M. Bedside Pediatrics. Boston: Little Brown, 1983.

CHAPTER 10

Apnea

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Apnea is the final manifestation of many pathophysiologic processes seen among patients of all ages, but neonates and infants can experience apneic episodes in response to a variety of physiologic and pathophysiologic processes not seen in later life. Differences in maturity of the central nervous system (CNS), respiratory reserve, and susceptibility to infectious agents are among the factors that interact to make the very young patient unique. The causes of apnea in older children are similar to those in adults although the susceptibility and reserve of the child, again, are different. In this chapter, the neonate and young infant are emphasized, but for completeness, the older child also is considered.

Apnea is defined as a respiratory pause of greater than 15 seconds or of any duration if there is associated pallor or cyanosis and/or bradycardia. Apnea must be distinguished from periodic breathing, which is a common respiratory pattern in young infants and is characterized by cycles of short respiratory pause followed by an increase in respiratory rate. Normal newborn infants display respiratory patterns that vary by sex and by conceptual age as well as by sleep state. Premature infants have more apneic episodes, defined for research purposes as respiratory pauses of greater than 2 seconds, than term infants. Normal-term infants experience significantly more episodes of nonperiodic apnea during active sleep than during quiet sleep, although respiratory failure occurs more often during quiet sleep. Severe apnea may be accompanied by change in color, muscle tone, or mental status, or by choking. Such an episode is described as an acute life-threatening event (ALTE).

PATHOPHYSIOLOGY

Respiration is controlled through respiratory centers in the pons and medulla. The output from these centers to the upper airway and bellows apparatus through the vagus, phrenic, and intercostal nerves is modulated by peripheral and cortical factors. The response of the neonate and young infant to these influences differs from that of the older child, thus accounting, in part, for the vulnerability of these small patients. Specifically, the adult response to hypoxemia is to increase respiratory rate in proportion to the decrease in oxygen partial pressure (P_{O_2}). This tachypnea is maintained for the duration of the hypoxic stimulus. In contrast, the neonate demonstrates a brief increase in respiratory rate followed by depression of respiratory drive and, often, apnea. During sleep, infants who are mildly hypoxic tend to breathe periodically or develop apneic spells. Furthermore, hypoxemia during sleep may not cause arousal. Hypoxemia also results in less of a response to arterial carbon dioxide tension ($PaCO_2$) and further depression of respiratory drive with worsening hypoxemia.

Feeding affects ventilation in young infants. Poor coordination of sucking and breathing can result in hypoxemia, apnea, and bradycardia. Babies also become more vulnerable during sleep, when oxygen tension falls. In fact, it has been shown that regurgitation while hypoxemic, either from feeding or during sleep, produces profound apnea and bradycardia as a result of an accentuated laryngochemoreflex. Finally, some infants who have experienced an ALTE have increased levels of b endorphin in their cerebrospinal fluid (CSF), suggesting a relationship between the ALTE and respiratory depression from endogenous opioids.

A number of exogenous factors, including toxins and metabolic derangements, express their influence on respiratory control by causing medullary depression. Clinical experience demonstrates that newborn and very young infants are particularly sensitive to these factors; for example, hypoglycemia can be manifested as apnea in young infants, and anemia often is related to apnea in premature babies. The young infant is susceptible to bellows failure on a purely mechanical basis. The infant's thoracic cage is extremely pliable, and the diaphragmatic muscles tire easily, resulting in greater vulnerability to respiratory failure as a result of respiratory distress.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of apnea is extensive (Table 10.1), and several categories are unique to newborns and young infants. For example, apnea may be the only clinical manifestation of seizure activity. This may be particularly difficult for emergency physicians to identify, however, because they did not witness the episode, and neurologic examination may be normal in the postictal period. Several infectious processes can cause apnea. Meningitis, for example, even in the absence of fever, must be included in the differential diagnosis. Respiratory syncytial virus is the predominant cause of bronchiolitis, which may cause apnea in children who were premature or have preexisting lung disease or congenital

heart disease. Infant botulism is a diagnosis that will be made, it is hoped, before apnea occurs and must be suspected on the basis of age, symptoms, and clinical findings. Recent data suggest that gastroesophageal reflux often occurs in infants with an ALTE despite the absence of a history of vomiting. Several systemic disease processes, including metabolic abnormalities that result in hypoglycemia, and sepsis, will become evident because the child develops apnea. The presence of congenital abnormalities always must be considered in newborns and in young infants. Finally, there have been well-substantiated reports of life-threatening child abuse as the etiology of ALTE.

	Neonate, Infant	Older Child
Central nervous system	Infection (meningitis, encephalitis)	Infection
	Seizure	Toxin
	Prematurity	Tumor
	Intraventricular hemorrhage	Seizure
	Increased intracranial pressure (ICP)	Increased ICP (trauma, hydrocephalus)
	Congenital anomaly (e.g., Arnold-Chiari)	Idiopathic hypoventilation ("Ondine's curse")
	Breath-holding spell	
Upper airway	Laryngospasm (e.g., gastroesophageal reflux)	Obstructive sleep apnea
	Infection (e.g., croup)	Infection (epiglottitis, croup)
	Congenital anomaly (e.g., Down syndrome)	Foreign body
Lower airway	Infection (pneumonia, bronchiolitis)	Infection
	Congenital anomaly	Asthma
Other	Infant botulism	Guillain-Barré syndrome
	Hypocalcemia, hypoglycemia	Spinal cord injury
	Anemia	Fat cheat
	Sepsis	Arrhythmias
	Arrhythmia	
	Sudden infant death syndrome (SIDS)	

Table 10.1. Differential Diagnosis of Apnea

Of great concern to both parents and physician is the risk of sudden infant death syndrome (SIDS) for an infant who has an unexplained ALTE. Although any of the diagnoses previously described can result in an ALTE, no cause is identified in about half of patients. No clear relationship exists between an “idiopathic” ALTE and SIDS; however, such a possibility is of grave concern to all parties. SIDS is implicated in approximately 1.5 to 2 deaths per 1000 live births in the United States. This rate fluctuates, based on epidemiologic variables such as socioeconomic status, ethnic origin, maternal drug addiction, discharge from a neonatal intensive care unit (ICU), season of the year (higher in winter), and vigor with which other diagnoses are pursued. The relationship between positioning during sleep and SIDS has been extensively studied. As a result of this research, the American Academy of Pediatrics, since 1992, has recommended that infants be placed supine or on the side during sleep. A coincident decline in the incidence of SIDS has been observed since that time. There have also been reports of ALTEs occurring while sitting in the upright position in a car seat. These factors must be taken into account in considering the differential diagnosis and subsequent management of an infant who has had a significant apneic episode.

EVALUATION AND DECISION

Initial Stabilization

The first priority of the emergency physician, after immediate resuscitation of the patient, is to identify a life-threatening condition—persistent or recurrent apnea (Fig. 10.1), hypoxia, septic shock, and hypoglycemia among others. In addition to assessment of the vital signs, including a rectal temperature and blood pressure (BP), the general appearance and mental status should be noted. Regardless of the cause, apnea is life threatening; therefore, even in the absence of abnormal physical findings, appropriate diagnostic studies should be performed to evaluate the child for several common etiologies (Table 10.2). The next phase of evaluation addresses two key questions: Is this episode of clinical significance? and What is the risk of recurrence? Factors to consider include signs of another acute illness, the age of the child, and other possible risk factors for clinically significant or recurrent apnea (Table 10.3).

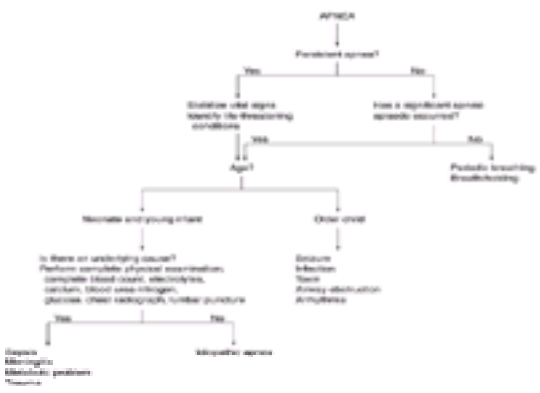

FIGURE 10.1. Approach to the diagnosis and management of apnea.

Pneumonia	Seizures
Sepsis/meningitis	Intracranial hypertension
Hypoglycemia	Shock

Table 10.2. Common Life-Threatening Conditions That Cause Apnea

History	Significant Apnea
Duration of event	Greater than 10 seconds
Was child asleep or awake?	Either, but apnea during sleep is more worrisome
Color change	Pallor or cyanosis
Associated movements, posture, or change in tone	Seizure activity Hypotonia "He looked dead"
Resuscitative efforts and response	Color change or hypotonia requiring cardiopulmonary resuscitation to improve
Interval since last feeding	If shortly after feeding, consider gastroesophageal reflux
Where event occured	Association with sleep, trauma

Table 10.3. Historical Features of Apnea

Has a Significant Apneic Episode Occurred?

The key to answering the questions previously outlined is invariably in the history (Table 10.3). Therefore, every effort should be made to obtain precise information from a firsthand observer. This may not be a simple task, considering the observer's recent stressful experience, but a clear initial history without the predictable influence of repeated questions is vital. The following details should be included: 1) where the event took place; 2) how long the event lasted; 3) whether the infant was awake or asleep; 4) whether there was an associated color change and, if so, to what colors and in what order; 5) description of associated movements, posture, or changes in tone; 6) what resuscitative efforts were made and the infant's response to them; and 7) when was the infant last fed? Attention to the response to these questions may provide the physician with a diagnosis. For example, if an 8-month-old infant was interrupted in a favorite activity, began to cry, turned red and blue, and finally had several seconds of tonic-clonic motor activity, the diagnosis of a breath-holding spell would be straightforward. In contrast, a story of 40 minutes of cyanosis and apnea in a now well-appearing child suggests that parts of the history are unreliable. Other recent events that should be documented are symptoms of other illnesses, including changes in behavior, activity, and appetite, as well as recent trauma and immunizations.

In many cases an absolute determination of significant apnea cannot be made in the emergency department (ED). Nevertheless, the description of the event may clearly suggest that significant apnea did occur, and hospitalization for further workup, as outlined next, is warranted. A typical case might be the previously well 2-month-old child who was noted by the parents to be apneic during a nap. The infant was described as limp and blue and “looked like he was dead.” There was no response to tactile or verbal stimulation for 5 to 10 seconds, but after 15 to 20 seconds of mouth-to-mouth breathing, the child coughed, gagged, and began to breathe. His color improved over the next 30 seconds, and the parents rushed him to the ED. Such a baby may look entirely normal on examination in the ED but be at grave risk for experiencing an ALTE.

The medical history also may provide important information regarding infants at risk for significant or recurrent apnea. The physician should ask specifically about previous similar episodes. Information about perinatal events, including gestational age (birth weight), labor and delivery, maternal health, and nursery course, is helpful. A family history with specific reference to seizures, infant deaths, and serious illnesses in young family members also should be included. Finally, information regarding poisons available in the household may be important in treating an older child.

Is There an Underlying Cause?

A careful physical examination identifies many treatable acute illnesses that can cause apnea; however, the likelihood of an underlying illness varies by age. One clue to serious systemic disease is fever or hypothermia. Tachypnea suggests either a respiratory or metabolic problem, and shock may be secondary to sepsis or hypovolemia from occult trauma. Evaluation of the nervous system should include notation of mental status, palpation of the fontanelles, and funduscopic examination. Dysmorphic features might suggest an underlying congenital abnormality; however, an entirely normal physical examination provides no reassurance that the described event was clinically insignificant and will not recur.

Laboratory evaluation should be guided by the history and physical examination. Tests to consider in the ED include a measurement of blood glucose and serum electrolytes. Any indication that the infant could have a serious infection should be pursued with cultures of blood and urine and by examination of CSF. Urine and blood for toxologic analysis

should be obtained on patients who may have been poisoned. In the child who has no pulmonary findings, an arterial blood gas serves primarily to indicate a persistent abnormality such as hypoxia or a metabolic derangement. For instance, in carbon monoxide poisoning, the Pa_{O_2} may be normal, but metabolic acidosis will be apparent. A significant apneic episode can occur, followed by recovery and a completely normal arterial blood gas determination. Therefore, the arterial or venous blood gas examination does not serve as a screening test for a serious event and should be obtained based on specific indications. Radiologic studies during the initial evaluation might include lateral neck, chest, abdomen, or skull films—again, as indicated by the history and physical examination.

The tasks of the emergency physician faced with a young patient who has had an apneic episode are to identify whether he or she should be hospitalized and to treat underlying conditions. If a careful history and physical examination suggest that a significant apneic episode has not occurred, the diagnosis of periodic breathing or breath-holding can be made, and the patient can be discharged after appropriate counseling of the parents and arrangements for follow-up. The evaluation of a young child with apnea, however, rarely will be so straightforward. If historical information indicates that significant apnea has occurred, the infant is at risk for a recurrence of this life-threatening event. An aggressive search for an underlying cause is necessary and often includes laboratory studies, lumbar puncture, chest radiograph, and electrocardiogram (ECG). Hospital admission should be arranged for observation and further diagnostic evaluation.

A significant apneic episode in the absence of systemic disease suggests a diagnosis of primary apnea. In a recent survey of pediatricians in varying types of practices, most reported that they would refer such children to a teaching hospital or to an established infant apnea study program for evaluation. There were considerable differences in opinion regarding the relationship between apnea and SIDS. This is not surprising considering that there is no single etiology for SIDS and little is known about how associated factors relate to the cause of death. This leaves the emergency physician in a quandary. There may not be an explanation for the event that satisfies the physician or the anxious parents. Thus, it is judicious to refer the family to an available specialist or center.

The standard evaluation that usually is pursued is designed to identify known causes of primary apnea. It generally includes in-hospital observation with monitoring as well as an evaluation of the CNS with an electroencephalogram, some type of sleep study, a chest radiograph, and an ECG. Respiratory function is evaluated with a pneumogram, and a barium swallow and esophageal pH study might identify gastroesophageal reflux. An ultrasound or computed tomogram (CT) of the head would be indicated if a central (CNS) cause for apnea is suspected. The decision to recommend home cardiorespiratory monitoring is beyond the scope of emergency practice because it necessitates a level of continuity of care and follow-up that cannot be provided by the emergency physician.

In many instances, a thorough history and careful physical examination with appropriate laboratory studies will suggest that a significant apneic event has not occurred and that there is no serious underlying illness. In this situation, the emergency physician should reassure and educate the family before discharging the patient. Good medical practice dictates that the parents also should be given specific instructions regarding indications for another ED visit and a follow-up visit to a primary care provider.

Suggested Readings

Arens R, Gozal D, Williams JC, et al. Recurrent apparent life-threatening events during infancy: a manifestation of inborn errors of metabolism. J Pediatr 1993;123:415–418.

Bass J, Mehta KA. Oxygen desaturation of selected term infants in car seats. Pediatrics 1995;96:288–290.

DiMario FJ. Breath-holding spells in childhood. Am J Dis Child 1992;146:125–131.

Hewertson J, Poets CF, Samuels MP, et al. Epileptic seizure-induced hypoxemia in infants with apparent life-threatening events. Pediatrics 1994;94:148–156.

Myer E, Morris D, Adams M, et al. Increased cerebrospinal fluid ß-endorphin immunoreactivity in infants with apnea and in siblings of victims of sudden infant death syndrome. J Pediatr 1987;111:660–666.

Rigatto H. Maturation of breathing. Clin in Perinat 1992;19:739–755.

See C, Newman L, Berezin S, et al. Gastroesophageal reflux-induced hypoxemia in infants with apparent life-threatening event(s). Am J Dis Child 1989;143:951–954.

Southall DP, Plunkett CB, Banks MW, et al. Covert video recordings of life-threatening child abuse: lessons for child protection. Pediatrics 1997;100:735–760.

Task Force on Infant Positioning. Positioning and sudden infant death syndrome (SIDS): update. Pediatrics 1996;98:1216–1218.

Wennergren G, Hertzberg T, Milerad M, et al. Hypoxia reinforces laryngeal reflex bradycardia in infants. Acta Paediatr Scand 1989;78:11–17.

CHAPTER 11

Ataxia

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Acute childhood ataxia is an uncommon presenting complaint in the emergency department. *Ataxia* is defined as a disturbance in coordination of movements and may be manifest as an unsteady gait. When it occurs, it is a distressing problem to both parent and clinician. It is important to establish the sign because true ataxia may be difficult to differentiate from clumsiness in toddlers. Parents are generally more sensitive to gait abnormalities in this age group. In older children, ataxia may be confused with weakness or vertigo. Life-threatening causes of pure ataxia are rare in children. After consideration of these, the problem may be approached in a cautious, step-wise fashion.

PATHOPHYSIOLOGY

The cerebellum coordinates complex activities such as walking, talking, and eye movements. Ataxia may be caused by a pathologic condition at either a focal or global level within the cerebellum or by disruptions in the afferent or efferent pathways. Anatomically, the cerebellum is located in the posterior cranial fossa, separated from the cerebrum by the tentorium. The ventral borders of the cerebellum form the roof of the fourth ventricle. Space-occupying lesions such as posterior fossa tumors and cerebellar hemorrhage may impede cerebrospinal fluid (CSF) flow, leading to hydrocephalus and increased intracranial pressure. Conversely, direct pressure on the cerebellar peduncles may cause ataxia to present.

The cerebellum links with other portions of the central nervous system (CNS) through the superior, middle, and inferior peduncles via the midbrain, pons, and medulla. Proprioceptive and sensory afferent impulses from muscles, joints, and tendons are carried via inferior peduncles to the cerebellar cortex. Labyrinthine afferent input is also conducted through the inferior peduncles. Connections from frontal motor cortex travel through the middle cerebellar peduncles. The superior peduncles carry efferent output to musculoskeletal tracts from the nuclei of the cerebellum.

The cerebellum is composed of two hemispheres. Because of the decussation patterns, a lesion that affects only one side of the cerebellum will result in movement abnormalities of the ipsilateral side, with distal movements more affected than proximal ones. Midline lesions lead to truncal ataxia, with swaying during standing, sitting, and walking, and/or with titubations of the head and neck. Finally, the intrinsic function of the cerebellum may be disrupted by autoimmune, metabolic, and toxic disorders.

DIFFERENTIAL DIAGNOSIS

Ataxia as a presenting sign invokes a broad differential diagnosis (Table 11.1). Distinguishing among acute, intermittent, and chronic progressive and chronic nonprogressive ataxia may be helpful, although some diagnoses have overlap in their time course at presentation.

Acute Ataxia	*Chronic, Progressive*
Acute cerebellar ataxia (postinfectious)	Hydrocephalus
Drug intoxication	Posterior fossa tumors
Labyrinthitis	Urea cycle defects
Vasculitis or Kawasaki disease	Multiple carboxylase deficiency
Meningitis	Vitamin E deficiency
Viral encephalitis	Abetalipoproteinemia
Intracranial hemorrhage	Refsum disease
Post-concussion syndrome	Hartnup disease
Benign paroxysmal vertigo	Familial periodic ataxia
Conversion reaction	Freidrich's ataxia
Intermittent	Ataxia telangiectasia
Migraine	Olivopontocerebellar atrophy
Epilepsy	*Chronic, Nonprogressive*
Transient ischemic attacks	Familial
Hartnup disease	Chiari I malformation
Wilson disease	Dandy-Walker malformation
Hereditary paroxysmal ataxia	Joubert syndrome
Maple syrup urine disease	Spastic cerebral palsy
Pyruvate decarboxylase deficiency	Cerebellar agenesis

Table 11.1. Differential Diagnosis

Fortunately, common causes of pure ataxia (Table 11.2) are not rapidly progressive. Acute cerebellar ataxia or postinfectious cerebellitis is truncal in nature and occurs 2 to 3 weeks after a viral illness (see Chapter 83). Children

ages 1 to 3 years are most commonly affected. Varicella is the classically identified culprit. This self-limiting illness is most severe at its onset, but complete recovery may not occur for several months. CSF may show mild lymphocytosis and increased protein. Imaging studies are normal. A small percentage of patients may show long-term sequelae such as learning disabilities or coordination problems.

Acute cerebellar ataxia
Drug ingestion
Guillain-Barré syndrome

Table 11.2. Common Causes of Acute Ataxia

Toxic ingestions of anticonvulsants, alcohol, and sedative-hypnotics generally cause ataxia and thus cause depressed mental status (see Chapter 88). However, for certain substances (phenytoin, carbamazepine, primidone), ataxia may be the most remarkable feature of intoxication.

When an ataxic patient presents with weakness and areflexia, Guillain-Barré syndrome may be present. If opthalmolplegia and areflexia are prominent, the Miller-Fisher variant can be suspected. Neuroimaging is normal, and the CSF may show a mild leukocytosis and elevated protein.

Ataxia may be an early prominent sign of posterior fossa tumors (especially medulloblastoma) and other conditions associated with increased intracranial pressure, including hydrocephalus and supratentorial tumors (see Chapter 100). Labyrinthitis and benign paroxysmal vertigo are rarely seen in young children but are occasionally encountered in adolescents. The sensation of loss of balance generally produces a classic wide-based gait. Conversion disorder should be suspected in a patient who walks with a narrow gait and has elaborate “near falls.”

Life-threatening causes of ataxia (Table 11.3) rarely present as ataxia alone. In a few cases, bacterial meningitis has been reported with ataxia as the first symptom. Viral cerebellitis may occur as a result of enteroviral disease. Neuroblastoma may present with titubations, myoclonic ataxia, and chaotic eye movements. The syndrome is immune-mediated. It should be suspected in patients with acute ataxia that waxes and wanes over several days. One should consider vertebrobasilar occlusion in a patient with neck trauma and ataxia, cerebellar hemorrhage with ataxia and headache, and vasculitis in a child with features of Kawasaki disease.

Meningitis	Neuroblastoma
Drug intoxication	CVA
Brain tumor	Intracranial hemorrhage

Table 11.3. Life-Threatening Causes of Ataxia

Migraine, seizure, transient ischemic attack, and metabolic disease are the most common causes for intermittent ataxia. Chronic progressive ataxias may have a basis in metabolic defects, some of which are treatable. When a progressive ataxia acutely worsens, this may signify severe hydrocephalus or hemorrhage into a posterior fossa tumor. A variety of familial, metabolic, and congenital causes exist for chronic nonprogressive ataxias.

EVALUATION AND DECISION

The approach to the problem should begin with a thorough history and physical examination. The duration and progression of the illness can be established and will help define the ataxia as acute, intermittent, or chronic. Chronic ataxia should be further divided into progressive or nonprogressive. Key historical points to cover include recent illnesses such as varicella or other viral diseases and access to medications or alcohol (Table 11.4). Family history may be helpful in recurrent or genetic causes.

Phenytoin	Dextromethorphan
Alcohol	Lead
Carbamazepine	5-Fluorouracil
Benzodiazepines	Ethylene glycol
Tricyclic antidepressants	Primidone
Antihistamines	Phenothiazines

Table 11.4. Drugs and Toxins That May Cause Ataxia

Physical examination should focus on signs of increased intracranial pressure (bulging fontanelle, papilledema, bradycardia, hypertension, abnormal respirations), meningeal irritation (nuchal rigidity, Kernig's or Brudzinski's sign), fever, rash, and evidence of middle ear disease. A detailed neurologic examination should document general level of consciousness, cranial nerve function, strength, tone, reflexes, sensation, and proprioception. Romberg's test will demonstrate a sensory deficit. Observation of the actual movements will help sharpen the diagnosis because particular syndromes have more truncal versus distal involvement, or unilateral versus bilateral involvement. Specific testing of cerebellar function is impossible in young children. However, a cooperative older child can be asked to perform a finger–nose–finger test, heel–shin test, and rapid alternating movements to further delineate neurologic dysfunction.

The decision to pursue specific laboratory testing is outlined in the algorithm shown in Figure 11.1. Patients with an acute presentation, focal neurologic deficits, recent head trauma, or signs of increased intracranial pressure warrant urgent evaluation via cranial computed tomography scan. Evidence of intracranial hemorrhage, hydrocephalus, or posterior fossa tumor provides an etiology for the ataxia. Neurosurgical involvement should be sought. If the imaging study is normal, the diagnosis may be postconcussion syndrome for patients with head trauma. Consultation with a neurologist may be indicated if physical examination findings other than ataxia persist.

FIGURE 11.1. The diagnostic approach to the child with ataxia.

If the patient appears “toxic” with fever or nuchal rigidity, an emergent imaging of the head is indicated because cerebellar tonsil herniation may cause neck stiffness. If imaging results are negative, a lumbar puncture can be performed safely. When bacterial meningitis is strongly suspected, appropriate antibiotics may be administered before the testing is done.

When other causes have been eliminated, it is prudent to suspect drug or alcohol ingestion (Table 11.4). With the exception of benzodiazepines and tricyclics, the routine toxicologic screen of urine will not detect these drugs. Thus, specific blood levels are indicated when intoxication is suspected.

Management of ataxia in children is directed at the underlying cause. Fortunately, the most common cause, acute cerebellar ataxia, is a self-limiting illness that resolves completely in most cases. During periods of significant ataxia, head protection may be warranted because of the risk of falling. Also, special caution with sedatives is necessary because their effect may be greatly heightened.

Suggested Readings

Connolly AM, Dodson WE, Prensky AL, et al. Course and outcome of acute cerebellar ataxia. Ann Neurol 1994;35:673–679.

Fenichel GM. Ataxia. In: Clinical Pediatric Neurology: A Signs and Symptoms Approach. 3rd ed. Philadelphia: WB Saunders, 1997:230–252.

Gieron-Korthals MA, Westbery KR, Emmanuel PJ. Acute childhood ataxia: 10-year experience. J Child Neurol 1994;9:381–384.

Schwartz JF. Ataxia in bacterial meningitis. Neurology 1972;22:1071–1074.

Stumpf DA. Acute Ataxia. Pediatr Rev 1987;8:303.

CHAPTER 12

Breast Lesion

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Complaints related to the breast usually involve pain, discharge, and discrete or diffuse enlargement. The evaluation of a breast lesion in a pediatric patient in the emergency setting is uncommon; however, pediatric emergency physicians must be able to distinguish problems that require immediate intervention from those that are most appropriately handled by referral to and close follow-up with either a general pediatrician or a subspecialist. Fortunately, most breast lesions that occur in children and adolescents are benign and self-limited. Many patients and their families, however, will benefit from the reassurance that neoplastic disease is extremely rare in any pediatric age group. This chapter covers the spectrum of disorders that pediatric emergency physicians are likely to encounter and focuses on an approach to the initial diagnosis and management of some of the more common causes.

DIFFERENTIAL DIAGNOSIS

Subsequent discussion of breast lesions in children is divided into the following categories: infections, benign cysts or masses, malignant masses, abnormal secretions, lesions associated with pregnancy and lactation, and miscellaneous causes, including both anatomic and physiologic entities (Table 12.1A and Table 12.1B). After completion of the history and physical examination, it is likely that the emergency physician will have a good idea into which category the lesion fits, and he or she can then decide how to proceed. With few exceptions, most lesions will be worked up as an outpatient procedure with referral to an appropriate subspecialist. The commonly encountered disorders (Table 12.2) are almost always benign, but consideration must be given to potentially life-threatening processes (Table 12.3).

I. Inflammatory Conditions
  A. Cellulitis
  B. Abscess
II. Noninflammatory Conditions
  A. Infancy
    1. Physiologic hypertrophy
    2. Tumor (rare)
  B. Childhood
    1. Premature thelarche
    2. Precocious puberty
    3. Prepubertal gynecomastia (male)
    4. Cancer (rare)
  C. Adolescence
    1. Male
      a. Postpubertal (physiologic) gynecomastia
      b. Exogenous hormonal stimulation
      c. Endocrinopathy
      d. Nipple cyst
      e. Cancer (rare)
    2. Female
      a. Isolated, benign cyst
      b. Fibroadenoma
      c. Fibrocystic disease
      d. Juvenile hypertrophy
      e. Hematoma/fat necrosis (posttraumatic)
      f. Papillomatosis
      g. Cystosarcoma phyllodes and other cancers (rare)

Table 12.1A. Breast Enlargement/Masses

Mastalgia

Galactorrhea

Miscellaneous

Table 12.1B. Other Complaints Related to the Breast

Newborn
Physiologic hypertrophy
Mastitis
Prepubertal Child
Premature thelarche (female)
Pubertal/Postpubertal Male
Pubertal gynecomastia
Pubertal/Postpubertal Female
Enlargement secondary to pregnancy
Cellulitis/abscess
Fibroadenoma
Fibrocystic disease
Benign, isolated cysts

Table 12.2. Common Breast Lesions

Newborn
Mastitis
Prepubertal Child
Breast enlargement with precocious puberty (secondary to hormonal secretion by a tumor)
Postpubertal Male
Breast enlargement with abnormal sexual development (secondary to hormonal secretion by a tumor)
Postpubertal Female
Neoplastic mass
Galactorrhea secondary to prolactin secreting tumor

Table 12.3. Life-Threatening Breast Lesions

Infections

Infection in the breast may take the form of a generalized cellulitis (mastitis) or an abscess. There is a bimodal age occurrence of breast infection with the first, less common peak seen in neonates and the second, in postpubertal females. Neonatal breast infection is most often seen in the first few weeks of life, when the breast bud is enlarged because of maternal estrogen stimulation. In some cases, excessive handling of the hypertrophied tissue by concerned parents or caretakers may lead to the introduction of bacteria. *Staphylococcus aureus* is the usual organism, but *Escherichia coli* has also been found. Neonatal mastitis generally requires a complete septic workup in the emergency department with initiation of broad-spectrum intravenous antibiotics for at least 48 hours because of the presence of fever in this age group. If an abscess develops, aspiration or, less commonly, incision and drainage is warranted, but care must be taken to avoid damaging the breast bud.

Mastitis in postpubertal females can be further classified as lactational and nonlactational. Lactational mastitis is discussed later. Nonlactational mastitis can develop in either the central or peripheral regions of the breast. It is often seen in women who are overweight, have large breasts, and possibly, practice poor hygiene. Peripheral mastitis can be associated with diabetes, rheumatoid arthritis, steroid treatment, granulomatous disease, and trauma. Other predisposing factors for mastitis include previous radiation therapy, sebaceous cysts, and hidradenitis suppurativa. Local signs and symptoms of infection in the breast include warmth, pain, tenderness, erythema, dimpling of the skin, and purulent nipple discharge. Fever may or may not be present. The organisms often isolated are *S. aureus*, enterococci, anaerobic streptococci, and bacteroides species.

Upon diagnosis of mastitis in the postpubertal female, the emergency physician should initiate oral antibiotics with an antistaphylococcal drug such as cephalexin or dicloxacillin. Patients should be instructed to keep the area as clean and dry as possible, to wear a clean cotton bra to avoid excessive sweating, and to avoid skin creams or talcum powders. Patients should have a follow-up appointment in 24 to 48 hours to ensure that the infection is clearing. Patients with systemic symptoms, those who appear toxic, or those who show no improvement should be admitted for intravenous antibiotics. If an abscess is suspected, it should be confirmed and treated with aspiration; incision and drainage are only occasionally necessary.

Benign Cysts and Masses

Enlargement of breast tissue may occur at any age beginning in the neonatal period. As previously discussed, the male and female neonatal breast bud is hypertrophied in the first few weeks of life because of maternal estrogen stimulation. No treatment is required because this condition abates over time; caretakers should avoid manual stimulation. In preschool-aged girls, there may be a temporary unilateral or bilateral enlargement of the breast bud. This is consistent with isolated premature thelarche as long as there are no other manifestations of developing secondary sexual characteristics. If premature thelarche is suspected, the girl can be referred for follow-up to her primary care physician;

the enlargement will most likely spontaneously resolve. If other secondary sexual characteristics are present (precocious puberty) or if this condition occurs in young boys (prepubertal gynecomastia), a specific cause should be aggressively pursued. The workup for this disorder generally includes a search for any adrenal, ovarian, or hypothalamic pathology, including tumors. Patients should be referred to an experienced pediatrician or endocrinologist if these conditions seem likely.

Fibroadenomas are the most common benign breast lesion in women less than 30 years of age. When present in adolescent girls, these lesions are sometimes called juvenile fibroadenomas. Fibroadenomas are usually discovered as solitary, mobile, and sometimes tender masses. Mammography is usually not indicated in adolescent girls unless there is a strong family history of breast malignancy or the contralateral breast needs to be evaluated. The treatment of choice for a fibroadenoma is excisional biopsy, so patients should be referred to a pediatric surgeon or breast surgeon. Reassurance may be given that the malignant potential of a fibroadenoma is very low. Hamartomas, benign breast tumors that can be mistaken for fibroadenomas, are also seen in pediatric patients and are also treated by excisional biopsy.

Fibrocystic disease is a benign, progressive process that is generally seen in women in their teens and twenties and may be encountered by emergency physicians. It usually presents as cyclically painful masses, sometimes bilateral and often most prominent in the upper outer quadrants of the breast. The masses often change in size or degree of nodularity during the course of the menstrual cycle, with the worst degree of pain occurring in the premenstrual phase. Nipple discharge may also be present and is nonbloody, green, or brown. Although not usually indicated, biopsy and histologic examination can provide a definitive diagnosis. These lesions are not considered precancerous. No specific treatment exists, but avoidance of caffeine and use of nonsteroidal anti-inflammatory drugs have been recommended for symptomatic relief.

Solitary or multiple breast cysts are also occasionally discovered on the breast examination of adolescent girls. These cysts are also benign lesions that can be confirmed easily by ultrasound. Referral of the patient to a primary care physician or surgeon should be done expediently so that aspiration can be performed with close follow-up.

Nipple masses represent another group of generally benign breast masses. Benign papillomatosis can be seen in prepubertal or pubertal boys and girls and often comes to attention because of bleeding. Occasionally, the lesion may obstruct the nipple and become more painful and possibly infected. In rare instances, a nipple mass can represent a carcinoma. Therefore, when detected, expedient referral to a breast surgeon or pediatric surgeon is indicated. Nipple masses may be observed for several weeks; if the mass or bleeding persist, they are usually excised.

Trauma to the breast can result in the formation of hematomas and fat necrosis, both of which can be palpated as firm, well-circumscribed breast masses. Initially, these lesions may be tender. If left untreated, they may develop into areas of scar tissue that are fixed to the skin. Breast trauma and fat necrosis are relatively common, but the differentiation from other more serious lesions may be difficult, requiring consultation with a surgeon in cases of uncertainty.

Malignant Masses

Cancers of the breast have been reported in young children; however, they are extremely rare. Often, these tumors are categorized as secretory carcinomas and behave more benignly than breast cancers in adults. Other histologic malignancies reported in children and adolescents are carcinomas, sarcomas, and the more common cystosarcoma phylloides, which can have both benign and malignant features. Characteristics that are suggestive of malignancy include a hard, nontender, solitary mass with ambiguous margins. There may be overlying skin changes (warmth, dimpling, or edema), bleeding from the nipple, and lymphadenopathy. The appropriate treatment is the same as that for a suspected benign mass—prompt referral to a pediatric surgeon or breast surgeon for definitive workup, which usually consists of excisional biopsy.

Abnormal Secretions (Nipple Discharge)

There are multiple causes for abnormal nipple secretions that can be divided according to whether they might require surgical treatment. Nonsurgical causes usually present as nonspontaneous discharges. The most common example is discharge fluid expressed during breast self-examination. The fluid may be milky, multicolored, and sticky. If infection is present, a purulent discharge may occur.

Galactorrhea is the most common spontaneous nipple discharge that usually occurs bilaterally. If it is not associated with pregnancy and lactation, increased prolactin secretion should be suspected. Both drugs and structural lesions of the hypothalamus and pituitary can cause high prolactin levels. Drugs that have been implicated include oral contraceptives, tricyclic antidepressants, phenothiazines, metoclopramide, reserpine, and a-methyldopa. Neonates may secrete a colostrum-like material that has been referred to as witch's milk. This discharge occurs temporarily, until maternal estrogen levels decline, and is not considered pathologic.

Other spontaneous nipple discharges have been described as multicolored, grossly bloody, serous, or clear and watery. Nonbloody discharges are usually not indicative of cancer. Mammary duct ectasia, traumatic nipple erosions (e.g., “jogger's nipple”), and eczema are some of the more common causes of nonbloody discharges. These disorders can be treated with nipple hygiene, warm compresses, and topical antibiotics if necessary. When nipple discharge is described as serosanguinous or frankly bloody or when it tests positive for occult blood, the association with cancer rises, particularly if a mass is palpable below the nipple. However, cancer is the cause of only 6% of bloody nipple discharges; more common causes include duct ectasia, papillomas, fibrocystic disease, and pregnancy. Any pediatric patient with spontaneous nipple discharge not explained by an obvious cause (e.g., jogger's nipples) should be referred to an appropriate specialist for close follow-up and further workup.

Lesions Associated with Pregnancy and Lactation

Significant changes occur in the female breast as a result of pregnancy, most prominently an increase in breast size and weight. Although pregnant patients may have any of the breast lesions seen in nonpregnant patients, they are prone to develop some unique conditions. The most common of these conditions occurs in lactating patients. Mastitis affects 2% of lactating women. If it occurs within several days of delivery, it is likely to be caused by *S. aureus*, which may be transmitted from infant to infant and then to the nursing mother through cracked skin in the nipple. Breast engorgement may exacerbate the symptoms. Therapy consists of warm compresses, continued breast-feeding, and an antistaphylococcal antibiotic. Mastitis that occurs 2 weeks or more after delivery is the result of either poor hygiene or inadequate emptying of the breast with subsequent milk stasis, engorgement, and colonization of bacteria within the milk. If an abscess develops, it usually requires both antibiotics and drainage, often by aspiration, for cure. Breast-feeding can usually proceed in the opposite breast, but not from the breast with an abscess because of the risk of the neonate acquiring infection. Pregnant patients may also have simple milk-filled cysts called galactoceles, which are often tender and located on the periphery of the breast. Ice packs, breast support, and aspiration may be needed to relieve the obstruction of the milk-filled ducts.

Nonlactating pregnant patients may have a bloody discharge from the nipple during the second or third trimester. This usually represents the benign condition of epithelial cell proliferation. If the discharge persists after delivery, a more thorough investigation should take place. Fibroadenomas often increase in size during pregnancy to the point of infarcting and causing significant pain. Excision is often advised for any solitary mass, and the patient should be expediently referred to a breast surgeon. The number of cases of breast malignancy diagnosed during pregnancy is small.

Miscellaneous Breast Lesions

Polymastia, Polythelia

Supernumerary breasts (polymastia) and supernumerary nipples (polythelia) are two congenital conditions that are unlikely to present as chief complaints in the emergency department but that may be discovered incidentally on examination. The incidence of polymastia is unknown, but it is more common in girls and results from failure of the embryonic mammary ridges to regress. Polymastia is present at birth, often resembling skin tags or nevi, and may not be noticed until the tissue is hormonally influenced during puberty, pregnancy, or lactation. Supernumerary breasts are most commonly found in the axillae but have been reported to occur in several locations. This ectopic tissue may become tender with menses and has been reported to develop the same range of pathology as normal breast tissue, necessitating excision under certain circumstances.

Polythelia is present in 0.6% of Caucasians and 1.5% of African-Americans. It is both sporadic and familial. Polythelia is most commonly found on the left, inferior to the normal nipple. In newborns, polythelia may appear as small, wrinkled lesions with or without pigmentation. The significance of polythelia is questionable. One series of patients with polythelia had a 23% incidence of associated unsuspected urologic anomalies. Other reported associations include pyloric stenosis, hypertension, congenital heart disease, and cardiac conduction defects. For this reason, patients with polythelia should be referred for at least a primary screening of underlying urologic disease. Otherwise, this disorder requires no treatment unless the diagnosis is uncertain (e.g., the lesion looks like a possible melanoma) or it is perceived as a cosmetic problem.

Juvenile Breast Hypertrophy

Juvenile breast hypertrophy is a rare disorder characterized by sudden, rapid, massive breast enlargement at a time of intense endocrine stimulation, usually around 8 to 16 years of age, just after menarche. It is thought to be caused by end-organ hypersensitivity to estrogen. The hypertrophy is usually bilateral and asymmetric and may progress at an alarming rate over 36 months. The differential diagnosis of this lesion includes cystosarcoma phylloides, juvenile fibroadenoma, and precocious puberty. Usually, no endocrine or neoplastic lesions are found. In some cases, the hypertrophy regresses in 1 to 3 years, but referral to a breast surgeon is always indicated because breast reduction or even total ablation may become necessary. This disorder is often associated with extreme emotional and psychosocial distress for patients and families.

Gynecomastia

Gynecomastia is a term commonly used to describe a broad spectrum of clinical breast lesions in boys, including excess breast tissue, breast enlargement, and a firm rubbery mass of tissue below the nipple that is discrete and nonadherent to the chest wall. Some texts assert that it is almost always unilateral; others assert that it is exclusively bilateral. Gynecomastia has been described as the male equivalent of fibrocystic changes in the female breast. Despite the confusion in the literature, histologically, there is a proliferation of dense periductal connective tissue with hyperplasia of ductal epithelial cells, which is the result of an increased effective estrogen:testosterone ratio in the serum or in the breast tissue. Causes of this relative hormone imbalance include physiologic changes (neonatal, puberty, aging); drug use; tumors of the testes, adrenal glands, and lungs; metabolic conditions (cirrhosis, hyperthyroidism, renal disease); and hypogonadism.

From a clinical perspective, gynecomastia occurs in about 50% of all boys between the ages of 11 to 18 years and lasts about 2 years. It can be associated with growth spurts and can also cause a significant degree of pain. More commonly, gynecomastia will come to a physician's attention because of the anxiety it causes in adolescent boys. If the patient with gynecomastia has normal-sized genitalia and none of the predisposing conditions listed earlier, reassurance is all that is needed. There is often particular concern about gynecomastia in obese boys because they may appear to have an overabundance of fatty tissue in the breast region. Surrounding fatty tissue may also give the illusion of small genitalia;

however, these patients have no higher incidence of gynecomastia than their nonobese counterparts. Gynecomastia is best dealt with by referral to the primary care physician for long-term follow-up.

Physiologic Mastalgia

During the first trimester of pregnancy, some teenage girls may complain of breast fullness. Occasional, nongravid patients may have breast pain for which no underlying cause is grossly apparent or suspected. These patients may have mastalgia that is likely related to the hormonal milieu of the breast throughout the menstrual cycle. Mastalgia is often described as a bilateral, poorly localized, heavy, dull, achy pain that radiates to the axillae. The pain is often worse with activity and relieved with the onset of menses. In general, there are no abnormal physical findings except tender, nodular breasts. The differential diagnosis includes costochondritis, Tietze's syndrome, cervical root syndromes, old breast trauma that has resulted in fat necrosis or hematoma, lung disease, and gallstones. Once these possibilities can be reasonably ruled out, mastalgia is the likely diagnosis. Most patients will improve with reassurance, analgesics such as nonsteroidal anti-inflammatory medications, warm compresses, and breast support. If the pain is refractory to these measures, other suggested therapies include caffeine avoidance, salt restriction, diuretics, and Danazol, a synthetic androgen, for severe, debilitating pain.

EVALUATION AND DECISION

History and Physical Examination

Initial evaluation of a breast lesion begins with a careful history and physical examination (Table 12.4). The two most common categories of breast lesions in children are infections and structural or mass lesions. When infection is suspected, especially in neonates or infants, particular attention should be given to the presence of systemic symptoms such as fever, chills, malaise, poor appetite, and lethargy. For the evaluation of mass lesions, it is imperative to obtain a detailed menstrual history and a chronology of the development of secondary sexual characteristics. Pregnant or lactating patients may also present to a pediatric emergency department. These patients should be asked about breast-feeding or breast-feeding attempts, as well as about general symptoms related to changes in the breast tissue. Medications may have an effect on the growth of certain breast lesions and may also affect hormonal pathways, leading to abnormal breast secretions. Therefore, a detailed drug history should be obtained. A few breast disorders may have a familial pattern, so a careful family history should also be taken.

History
Onset/duration of lesion
Presence/absence of pain
Presence/absence of nipple discharge
Change in lesion with menses
Complete menstrual and sexual development history including sexual activity and previous pregnancies
Family history of breast disease
Diet
Medications
Concomitant medical disorders
Systemic symptoms: fever, weight loss, sweating, headaches, visual changes
Physical Examination
Breasts: symmetry, skin appearance, temperature, areola, nipples, secretions, masses, chest wall, axillae
Lymph nodes
Hair distribution
Genitalia

Table 12.4. Important Historical and Physical Examination Components in the Evaluation of a Breast Lesion

A general physical examination should be performed on any pediatric patient who complains of a breast problem. Findings in other organ systems may be helpful in pinpointing the cause of the disorder. For example, premature secondary sexual characteristics, hirsutism, or abnormal skin coloring may indicate an endocrinopathy. A detailed evaluation of the breasts and adjacent structures is essential. The chest wall should be inspected for any gross deformities, asymmetry, or skin changes. The physician should have the patient lean forward with hands on hips and again observe for any asymmetry or skin retraction. With the patient supine with arms above the head, the physician should palpate each breast in a series of concentric circles radiating outward from the nipple, looking and feeling for nodules, cysts, masses, or inconsistencies in the breast tissue. Each areola should be gently compressed to assess for masses or nipple discharge. If present, the color, character, and odor of any discharge should be noted. The physician should feel for the presence of any masses or lymphadenopathy in both axillae.

Laboratory Testing

Very few laboratory tests will be helpful in the initial evaluation of pediatric patients with breast lesions. All postmenarchal girls should have a pregnancy test performed; breast tenderness and swelling may be some of the earliest signs of pregnancy. Chest radiographs are rarely helpful except in cases in which the examiner believes that signs and symptoms from the lungs or chest wall are referred to the breast. Mammography, cyst aspiration, and endocrinologic testing may be indicated for some breast lesions, although these generally take place outside the emergency setting.

Approach

The approach to the patient with complaints related to the breast depends first on whether the patient is prepubertal or pubertal/postpubertal. Among patients who are pubertal/postpubertal, the considerations vary greatly between boys and girls. Finally, unique considerations pertain to the pregnant or lactating girl, as discussed earlier.

Prepubertal Child (Fig. 12.1)

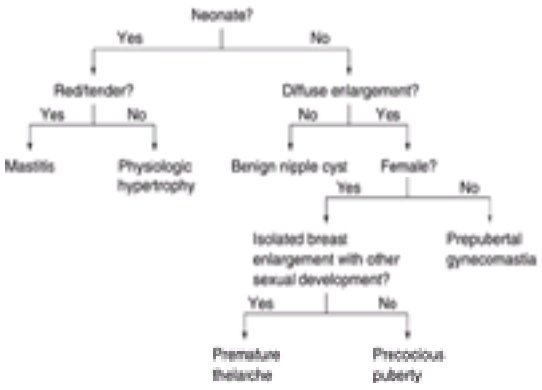

FIGURE 12.1. Approach to breast complaints in the prepubertal child.

Among prepubertal children, disorders of the breast occur commonly only in the immediate newborn period, usually because of physiologic hypertrophy. If erythema or tenderness is present, the physician should consider mastitis, a potentially serious bacterial infection. Among children, isolated lesions underneath the nipple are usually benign cysts. Diffuse enlargement calls for endocrinologic consultation.

Pubertal/Postpubertal Male (Fig. 12.2)

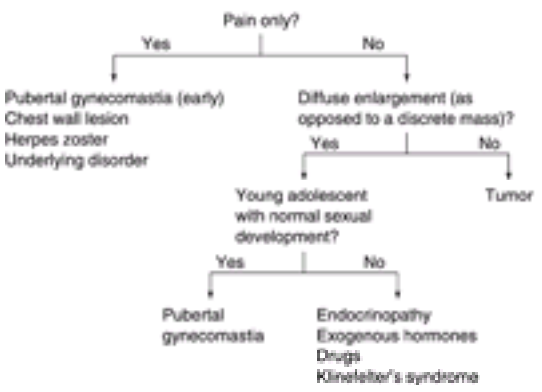

FIGURE 12.2. Approach to breast complaints in the pubertal/postpubertal boy.

The adolescent boy may complain of pain, yet have no clearly palpable breast enlargement. This sensation may be caused by minor chest trauma in a boy with early pubertal gynecomastia or may represent underlying chest pain (see Chapter 52). Most often, the complaint will be that there is bilateral (sometimes asymmetric) enlargement diffusely throughout the breast tissue, which represents (physiologic) pubertal gynecomastia in the boy who has normal sexual development. Either a unilateral, discrete mass or bilateral, diffuse enlargement with abnormal sexual development requires further evaluation.

Pubertal/Postpubertal Female (Fig. 12.3)

FIGURE 12.3. Approach to breast complaints in the pubertal/postpubertal girl.

The initial step in evaluating the adolescent girl is to obtain a pregnancy test, which when positive points specifically to a number of conditions that occur only in the gravid state (see earlier discussion). Both pregnant and nonpregnant girls may experience a myriad of disorders, related to the breast. The emergency physician's primary goal is to distinguish underlying disorders that are causing chest rather than breast pain (see Chapter 52) and to diagnose a few relatively

minor problems, including cellulitis, abscess, hematoma, and traumatic erosions. Other causes for breast enlargement, masses, and discharge require follow-up by an appropriate specialist.

Suggested Readings

Bradley AL, Sharp KW. Breast disease. Med Clin North Am 1995;79: 1443–1455.

Brawer-Ostrovsky Y, Kiderman A, Furst AL. Breast lumps in men: four case reports and literature review. J Fam Pract 1993;37:180–184.

Coleman WL. Recurrent chest pain in children. Pediatr Clin North Am 1984;31:1007–1026.

Dixon JM. Breast infection. Br Med J 1994;309:946–949.

Hansen N, Morrow M. Breast disease. Med Clin North Am 1998;82:203–222.

Klein TA. Office gynecology for the primary care physician, part II. Med Clin North Am 1996;80:321–336.

Kupfer D, Dingman D, Broadbent R. Juvenile breast hypertrophy: report of a familial pattern and review of the literature. Plast Reconstr Surg 1992;90:303–309.

Mansel RE. Breast pain. Br Med J 1994;309:866–868.

Scott EB. Fibrocystic breast disease. Am Fam Phys 1986;36:119–126.

Scott-Conner CE, Schorr SJ. The diagnosis and management of breast problems during pregnancy and lactation. Am J Surg 1995;170:401–405.

Velanovich V. Ectopic breast tissue, supernumerary breasts, and supernumerary nipples. South Med J 1995;88:903–906.

CHAPTER 13

Coma and Altered Level of Consciousness

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Consciousness refers to the state of being awake and aware of oneself and one's surroundings. It is a basic cerebral function that is not easily compromised; impairment of this faculty may therefore signal the presence of a life-threatening condition. An altered level of consciousness (ALOC) is not in itself a disease. It is a state caused by an underlying disease process, which must be addressed quickly to maximize a patient's chance of recovery. *Coma* refers to a state of complete unawareness and unresponsiveness (e.g., unconsciousness) from which a patient cannot be roused; this represents the most extreme form of ALOC. The term coma is often modified with descriptors such as light or deep. Lesser levels of impairment are described using other terms whose meaning may overlap. Lethargy refers to depressed consciousness resembling a deep sleep from which a patient can be aroused but into which he or she immediately returns. A patient is said to be stuporous or obtunded when he or she is not totally asleep but demonstrates greatly diminished responses to external stimuli. Because neurologic status may vary dramatically over time, it may be difficult to describe such symptoms using a single descriptor. Recording the comatose patient's specific response (body movement, type of vocalization) to a defined stimulus (e.g., a sternal rub) is usually preferable (Table 13.1).

Eye Opening	
Spontaneous	4
To speech	3
To pain	2
None	1
Best Motor Response	
Obeys verbal command	6
Localizes to painful stimulus	5
Flexion withdrawal	4
Flexion decorticate	3
Extension decerebrate	2
No response	1
Best Verbal Response[a]	
Oriented; converses	5
Disoriented; converses	4
Inappropriate words	3
Incomprehensible sounds	2
No response	1

[a]Preverbal children should receive full verbal score for crying with stimulation.

Table 13.1. Glasgow Coma Scale

PATHOPHYSIOLOGY

The state of wakefulness is mediated by neurons of the ascending reticular activating system (ARAS) located in the brainstem and pons. Neural pathways from these locations project throughout the cortex, which is responsible for awareness. If the function of these neurons is compromised or if both cerebral hemispheres are sufficiently affected by disease, an ALOC will result.

Proper function of the ARAS and cerebral hemispheres depends on many factors, including the presence of substrates needed for energy production, adequate blood flow to deliver these substrates, absence of abnormal serum concentrations of metabolic waste products or extraneous toxins, maintenance of body temperature within normal ranges, and the absence of abnormal neuronal excitation or irritation from seizure activity or central nervous system (CNS)

infection.

Disorders that produce coma by raising intracranial pressure (ICP) increase the volume of an existing intracerebral component such as brain, blood, or cerebrospinal fluid (CSF). Alternatively, a new component such as a tumor may be introduced. The brain can initially compensate for this altered volume relationship by regulating blood flow and CSF production. When the limits of these compensatory mechanisms are reached, ICP will rise abruptly, decreasing cerebral perfusion pressure (defined as mean arterial pressure minus ICP) and placing the patient at risk for herniation.

Herniation, the displacement of a part of the brain from its usual position into an unfamiliar intracranial compartment, can occur in several locations within the cranium, as shown in Figure 13.1. Central herniation results from an increase in volume and pressure in both cerebral hemispheres, compressing and displacing the midbrain and upper brainstem downward through the tentorium. Cingulate gyrus herniation occurs as a result of unilateral cerebral hemisphere volume increase when the gyrus is displaced laterally underneath the falx, crossing the midline of the cranium. This unilateral volume increase may instead cause uncal herniation because the lower midline portion of a cerebral hemisphere and adjoining hippocampal gyrus are directed downward through the tentorium. Foramen magnum (or tonsillar) herniation is a consequence of increased pressure in the posterior fossa, forcing the cerebellar tonsils through the foramen magnum at the base of the skull.

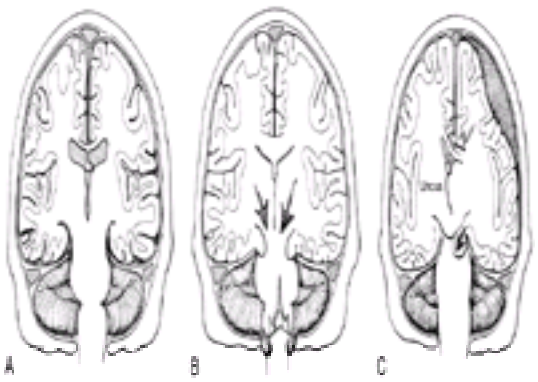

FIGURE 13.1. Intracranial contents. **A.** Normal relationships. **B.** Dark arrow, central herniation; light arrow, foramen magnum or tonsillar herniation. **C.** Dark arrow, uncal herniation; light arrow, cingulate gyrus herniation.

DIFFERENTIAL DIAGNOSIS

A differential diagnosis for children presenting in or near coma is shown in Table 13.2. Conditions arising from trauma or disease within the CNS are separated from those affecting the brain diffusely due to extracranial problems. The more commonly encountered causes of coma are listed in Table 13.3. These most likely causes of coma should be considered in every patient presenting in this condition. Life-threatening causes of ALOC are listed in Table 13.4 and must be considered in every patient. If present, these disorders require emergent treatment. More than one problem may be present simultaneously; for example, a drowning victim may incur a head injury when falling into a swimming pool, or a deeply postictal patient with known seizure disorder may have ingested a toxin.

Table 13.2. Etiology of Acute-Onset Coma/ALOC

Subdural hematoma	Posthypoxia
Epidural hematoma	Hypoglycemia
Cerebral edema	Toxic ingestions
Postictal state	Meningitis
Hypotension	

Table 13.3. Common Causes of Coma/ALOC

Epidural hematoma	Meningitis, encephalitis
Cerebral edema	Toxic ingestions
Brain neoplasms	Hypotension
Cerebral infarctions	Hypoxia
Cerebrospinal fluid shunt malfunction	Sepsis

Table 13.4. Life-Threatening Causes of Coma/ALOC

Primary Central Nervous System Disorders

Trauma

Coma-producing brain lesions that result from trauma include subdural and epidural hematomas, intraparenchymal and subarachnoid hemorrhage, penetrating injuries, cerebral contusion, diffuse cerebral edema, and concussion (see Chapter 105). Penetrating injuries are rare and of obvious origin. Most cases of pediatric head injury are blunt in nature, involving rapid deceleration against an automobile interior or the ground. Inflicted injury is also common in young children. The site of hemorrhage may be located opposite external signs of trauma due to the rebounding of the brain within the skull after impact (contrecoup injury). Patients may present in a comatose state or may be alert for variable periods after impact. All traumatic lesions of the brain may increase ICP, which is the chief cause of the resulting vomiting, lethargy, and/or coma seen in these patients. Increased ICP reduces cerebral perfusion pressure initially and may eventually result in herniation.

Epidural hematomas are caused by bleeding from cerebral arteries or veins; 85% are associated with an overlying skull fracture. Epidural hematomas may occur after relatively minor trauma; in one series, 24 of 53 children with epidural hematoma had fallen less than 5 feet. The classic location of the injury is the temporal lobe, due to tearing of the middle meningeal artery. Such arterial bleeding produces a faster onset of symptoms such as headache, vomiting, and decreased LOC than venous hemorrhage. Approximately 40% of these patients may appear neurologically normal on presentation, during the classically described “lucid interval.” On computed tomography (CT) scan, epidural hematomas usually appear sharply localized and are unilateral with a lenticular (lenslike) shape.

Subdural hematomas, produced by tearing of cortical bridging veins between the dura and arachnoid, can occur bilaterally and are 5 to 10 times more common than epidural bleeding. They may occur on a chronic basis in young, abused children and are associated with skull fractures in 30% of cases. Retinal hemorrhages may be found in 75% of patients with subdural hematomas. On neuroimaging, these lesions may appear classically crescent-shaped.

Diffuse cerebral edema is more common than focal lesions after brain trauma and is unfortunately less amenable to neurosurgical intervention. Characteristic CT findings of loss of gray–white interface may not be visible for 12 to 24 hours after the trauma was sustained. When radiographic abnormalities appear, they may be similar to those produced by hypoxia. *Concussion* is an inexact term for a transient alteration in normal neurologic function, often involving temporary loss of consciousness, after experiencing head trauma. A postconcussion syndrome may last for hours to days and is characterized by nausea, vomiting, dizziness, headache, and lethargy. Neuroimaging studies are normal, yet patients may be ill enough to require admission for observation and intravenous hydration.

Seizures

LOC is greatly diminished both during and after periods of seizure activity. Although generalized seizure activity is readily recognizable by the rhythmic motor activity accompanying an ALOC, partial or absence seizure activity may present more subtlely with staring, tremors, eye blinking, rhythmic nodding, or other inappropriate repetitive motor activity. Seizures of all types except petit mal may be followed by a postictal period, during which obtunded patients gradually regain responsiveness to and awareness of their surroundings.

The diagnostic approach toward a patient with ALOC from seizure activity varies based on whether seizures have occurred in the past and the progression of resolution of his or her neurologic abnormalities (see Chapter 70). Posttraumatic or new focal seizures are assumed to reflect an intracranial lesion until proven otherwise. Children taking anticonvulsants for known seizure disorders benefit from drug level measurement during an observation period. Subtherapeutic anticonvulsant levels result in convulsions with postictal ALOC, whereas supratherapeutic levels produce ALOC of a different appearance based on the medication involved. The presence of fever may indicate that a febrile seizure has occurred or that the patient has contracted a CNS infection such as meningitis or encephalitis (see Chapter 83 and Chapter 84). The new onset of afebrile generalized seizures requires a more elaborate evaluation, as detailed in Chapter 70.

Infection

Coma-inducing infections of the CNS may involve large areas of the brain and surrounding structures, as in meningitis or encephalitis, or they may be confined to a smaller region, as in the case of cerebral abscess or empyema (see Chapter 84). Bacterial meningitis remains the most common infection severe enough to produce profoundly diminished LOC; despite the overall decrease in total number of cases since the introduction of vaccines effective against *Haemophilus influenzae, Streptococcus pneumoniae* and *Neisseria meningitidis* are now the most common etiologic agents after the neonatal period. Meningitis may be caused by viral (enteroviruses, herpes), fungal (Candida, cryptococcus), mycobacterial (tuberculosis), and parasitic (cysticercosis) organisms as well. These nonbacterial infections usually have a slower onset of symptoms. The incidence of viral meningitis peaks in late summer, when enterovirus infections are most common.

Encephalitis, or inflammation of brain parenchyma, may involve the meninges as well (see Chapter 84). It occurs most commonly as a result of viral infection or immunologic mechanisms. Mumps and measles viruses were common etiologic agents before immunizations against these diseases, and they still occur in unimmunized individuals. Varicella encephalitis occurs 2 to 9 days after the onset of rash. The incidence of arthropodborne encephalitides varies by geographic location but usually peaks in late summer and early fall. The herpes simplex virus remains the most common devastating cause of encephalitis, causing death or permanent neurologic sequelae in more than 70% of patients. It affects the temporal lobes most severely, leading to seizures and parenchymal swelling, which can cause uncal herniation.

Focal CNS infections include brain abscesses, subdural empyemas, and epidural abscesses (see Chapter 84). Brain abscesses occur most often in patients with chronic sinusitis, chronic ear infection, dental infection, endocarditis, or congenital heart disease. One-fourth of cases of brain abscess occur in children less than 15 years of age, with a peak incidence between 4 and 7 years of age. Subdural empyema occurs secondary to chronic ear or sinus infection as well, but it is most commonly seen as a sequela of bacterial meningitis. Cranial epidural abscess is rare, but most cases occur from extension of sinusitis, otitis, orbital cellulitis, or calvarial osteomyelitis.

Neoplasms

Alterations in consciousness as a result of intracranial neoplasms (see Chapter 100) may be caused by seizure, hemorrhage, increases in ICP caused by interruption of CSF flow, or direct invasion of the ARAS by the malignancy (which is unlikely to cause coma of rapid onset). The location of the tumor determines additional symptoms: ataxia and vomiting for infratentorial lesions versus seizures, hemiparesis, and speech or intellectual difficulties resulting from supratentorial neoplasms. Acute hydrocephalus secondary to tumor growth most commonly presents with headache, lethargy, and vomiting.

Vascular

Coma of cerebrovascular origin is caused by interruption of cerebral blood flow (stroke) as a result of hemorrhage, thrombosis, or embolism (see Chapter 125). Hemorrhage is often nontraumatic, stemming from an abnormal vascular structure such as an arteriovenous malformation (AVM), aneurysm, or cavernous hemangioma. Rupture of an AVM is the most common cause of spontaneous intracranial bleeding among pediatric patients. The hemorrhage is arterial in origin and located within the parenchyma, but it can rupture into a ventricle or the subarachnoid space. Aneurysm rupture is less common and is unusual in that repetitive episodes of bleeding may occur (“sentinel bleeds”), with rising morbidity and mortality from each subsequent episode of bleeding. Subarachnoid blood may be present in either case, although more commonly with aneurysm rupture. Cavernous and venous hemangiomas are lower-flow lesions that produce a less acute onset of symptoms.

Stroke may also occur from thrombosis or embolism of a normal vessel. Cerebral infarction caused by occlusion of the anterior, middle, or posterior cerebral artery usually produces focal neurologic deficit, not coma. Acute occlusion of the carotid artery, however, may produce sufficient unilateral hemispheric swelling that herniation and coma may ensue; infarction may lead to hemorrhage as well. Central venous thrombosis is most commonly seen as a sequela of infections of the ear or sinus or hypercoagulable states.

Swelling or hemorrhage from infarcted brain can cause increased ICP, leading to interruption of blood flow to the ARAS and resultant coma. Less severe, often focal symptoms vary based on the size and location of brain denied adequate blood supply. Vascular accidents in the cerebellum present with combinations of ataxia, vertigo, nausea, occipital headache, and resistance to neck flexion. Coma is an unusual early sign of infarction of cerebral structures but becomes more common as lower anatomic centers are affected. Occlusion of the basilar artery may result in upper brainstem infarction, resulting in rapid onset of coma, as does hemorrhage or infarction of the pons.

Cerebrospinal Fluid Shunt Problems

Children with congenital or acquired hydrocephalus as a result of prematurity, neoplasm, or trauma depend on the continued function of a neurosurgical shunt to drain CSF and prevent rises in ICP (see Chapter 83). The most common shunt type is ventriculoperitoneal (VP), draining CSF from a lateral cerebral ventricle, up through a small hole in the skull, through a valve with an attached reservoir located beneath the scalp, and into the peritoneum via tubing placed under the skin of the neck, chest, and abdomen. CSF shunts may malfunction for many reasons, including tubing rupture, valve malfunction, tubing blockage, tubing disconnection, and shunt infection. The risk of failure is greatest during the first 6 months after shunt placement or revision.

Systemic Abnormalities

The second major category of disorders causing coma listed in Table 13.2 arise in organs other than the CNS and affect the brain diffusely. These abnormalities alter neuronal activity by a variety of means, including decreasing metabolic substrates required for normal function (e.g., hypoxia, hypotension, hypoglycemia, other electrolyte abnormalities), altering the rate of intracellular chemical reactions (e.g., hypothermia, hyperthermia), and introducing extraneous toxins into the CNS.

Hypoxia

Oxygen delivery to the brain may be adversely affected by disorders that compromise a patient's airway, breathing, or circulation. Neurons are the cells most sensitive to oxygen deprivation, and they will cease to function within seconds after being deprived of adequate levels of oxygen. Hypoxic coma may result from airway obstruction, pulmonary disease, severe acute anemia, severe methemoglobinemia, carbon monoxide poisoning, or asphyxia (e.g., drowning). Permanent CNS dysfunction results from total anoxia lasting more than 4 to 5 minutes at normal body temperatures; lesser degrees of hypoxia may be tolerated for longer periods. Submersion in cold water may cool the brain sufficiently to exert a neuroprotective effect. It is usually unclear in the emergency department how much permanent neurologic damage has taken place as a result of hypoxia. Hypercarbia may accompany hypoxia and be responsible for neurologic depression and coma as well.

Cardiovascular Abnormalities

Hypotension may be the product of numerous causes, including hemorrhage, dehydration, sepsis, arrhythmia, and intoxication, but the result is poor cerebral perfusion, which produces diminished mental status (see Chapter 3). Hypertensive encephalopathy is distinguished by headache, nausea, vomiting, visual disturbance, altered mental status, or coma in the presence of a blood pressure above the 95th percentile for age and sex (see Chapter 35). The acute onset of severe hypertension may reflect ongoing renal (e.g., unilateral renal artery stenosis, acute glomerulonephritis), endocrine (e.g., pheochromocytoma), or cardiac (e.g., aortic coarctation) pathology, or it may be the result of a toxic ingestion (e.g., cocaine). Cerebral hemorrhage may result. Hypertension accompanied by bradycardia may indicate increased ICP.

Disorders of Thermoregulation

Hypothermia or hyperthermia in the pediatric patient is usually caused by prolonged environmental exposure to temperature extremes such as those found in cold water or in a closed car in sunlight (see Chapter 89). The child made comatose as a result of abnormal core temperature will have multiple organ system abnormalities in addition to CNS dysfunction. Mental impairment is progressive as body temperature is lowered because each fall of 1°C produces a 6% decline in cerebral blood flow. At 29° to 31°C, confusion or delirium is present, as is muscular rigidity. Patients with core temperatures of 25° to 29°C are comatose with absent deep tendon reflexes and fixed, dilated pupils. CNS findings in hyperthermia include headache, vomiting, and obtundation, leading to coma and/or seizures, especially above 41°C.

Toxic Ingestions

Pediatric toxic ingestions are often not witnessed, may involve a large dose on a milligram per kilogram basis, are rarely intentionally inflicted, and are usually complicated by the young patient's inability to provide information regarding the quantity or identity of the substance ingested (see Chapter 88). Table 13.2 lists many drug classes that cause coma when an overdose is taken. Exogenous toxins may impair neuronal function directly or by causing hypoxia, acidosis, enzyme inhibition, hypoglycemia, or seizures.

Metabolic Alterations

Abnormal serum concentrations of any substrate or product involved in neuronal metabolism can produce obtundation, leading to coma. Hypoglycemia is the most common disorder in this category, especially in infants and young children, whose capacity for hepatic gluconeogenesis is limited. Disorders known to produce hypoglycemia include serious bacterial infections, sepsis, dehydration, and toxic ingestions (especially ethanol and oral hypoglycemics). Diabetes mellitus, especially of new onset, may present with profoundly depressed consciousness from the combination of hyperosmolarity, dehydration, hypotension, and lactic acidosis and ketoacidosis. Patients under treatment for diabetic ketoacidosis may also develop cerebrae edema.

Metabolic acidosis or alkalosis of sufficient degree produces altered mental status. Severe dehydration that leads to significant metabolic acidosis is the most common disorder of this type seen in children. Abnormal concentrations of any serum cation, including sodium, calcium, magnesium, and phosphorus, can produce altered mental status as well. The degree of resulting neurologic compromise will be affected by the duration of the problem and concurrent disorders. Severe dehydration alone, from any cause without significant electrolyte abnormalities, may produce profound lethargy in infants and children as well.

Other causes of metabolic coma in the pediatric age group include kidney or hepatic failure, both of which may result in progressive apathy, confusion, and lethargy. Urea cycle defects may present with coma and hyperammonemia in young infants. Acute toxic encephalopathy (Reye syndrome) is a rare but devastating illness caused by mitochondrial injury of unknown origin that affects all organs of the body, particularly the brain and liver (see Chapter 83). An epidemiologic association exists between the disorder and an antecedent viral illness (including varicella) from which a patient is recovering. Patients with Reye syndrome typically develop severe vomiting, followed by combative delirium that progresses to coma. Cerebral edema, increased ICP, and central herniation may occur with typically poor outcome.

Miscellaneous Conditions

Other causes of coma or depressed LOC in children are less easily categorized. Children with intussusception, the most common cause of bowel obstruction in childhood, may have significant apathy and lethargy in addition to vomiting, intermittent abdominal pain, and bloody stools. As a result, they are often treated for dehydration, sepsis, or meningitis before the appropriate diagnosis is discovered. The presence of “currant jelly” stools or a palpable abdominal mass suggests this condition.

Psychiatric disorders may produce a true stuporous state. More commonly, neurologically intact patients attempt to feign unresponsiveness for reasons known only to them, and they may be remarkably successful at remaining immobile despite painful stimuli. The nature of their “impairment” may be discovered by a detailed neurologic examination. Conscious patients will usually avoid hitting their face with a dropped arm, may resist eyelid opening, will raise their heart rate to auditory or painful stimuli, and will have intact deep tendon, oculovestibular, and oculocephalic reflexes.

A useful acronym incorporating the common causes of coma in children has been proposed by Schunk and is listed in Table 13.5; the acronym is based on the names of childhood immunizations: DPT (for *d*ehydration, *p*oisoning, *t*rauma), OPV (*o*ccult trauma, *p*ostictal or *p*ostanoxia, *v*entriculoperitoneal shunt problem), HIB (*h*ypoxia or *h*yperthermia, *i*ntussusception, *b*rain masses), and MMR (*m*eningitis or encephalitis, *m*etabolic, *R*eye syndrome, other *r*arities).

DPT
*D*ehydration
*P*oisoning
*T*rauma
OPV
*O*ccult trauma
*P*ostictal or *p*ostanoxia
*V*entriculoperitoneal shunt problem
HIB
*H*ypoxia or *h*yperthermia
*I*ntussusception
*B*rain masses
MMR
*M*eningitis or encephalitis
*M*etabolic
*R*eye syndrome, other *r*arities

Modified from Schunk JE. The pediatric patient with altered level of consciousness: remember your “immunizations.” J Emerg Nurs 1992;18:419–421.

Table 13.5. Mnemonic for Causes of Coma

EVALUATION AND DECISION

An approach for the evaluation of pediatric patients presenting with coma is summarized in Figure 13.2. All patients need rapid assessment of their airway, breathing, and circulation, followed by a focused history, physical examination, and consideration of laboratory and imaging studies. This approach is based on the selective use of the following critical clinical and laboratory findings: 1) vital signs; 2) a history of recent head trauma, seizure activity, or ingestion; 3) signs of increased ICP or focal neurologic abnormality; 4) fever; 5) laboratory results; 6) brain CT scan results; and 7) CSF analysis. The evaluation of the comatose patient should follow an orderly series of steps, addressing the more life-threatening problems of hypoxia, hypotension, and increased ICP before progressing to the investigation of less urgent disorders. If one or more of the former are present, immediate resuscitative efforts are begun.

FIGURE 13.2. Evaluation of the comatose child.

History and Physical Examination

Although open-ended questions have merit in medicine, goal-directed questioning pertaining to suspected diagnoses are required in cases of coma of unknown origin. Specific queries regarding current medications, medications available to ingest, seizures, fever, headache, irritability, vomiting, changes in gait, and behavioral abnormalities should be made. The most important historical finding in a comatose patient is a history of recent head trauma. If no history of head trauma is present, it should be considered as a cause of ALOC if a pediatric patient was unsupervised at any time within 24 hours of presentation, if all caregivers are not available, or if the veracity of caregivers is questionable.

A patient's vital signs will reveal the presence of fever, hypotension, or hypertension. The LOC of a neurologically impaired patient may initially be evaluated using a simple AVPU scale, representing four major levels of alertness: *a*lert, responsive to *v*erbal stimuli, responsive to *p*ainful stimuli, and *u*nresponsive. Elements of a more detailed neurologic

evaluation are discussed in the following section.

The patient should be carefully examined for physical findings consistent with head trauma, including retinal hemorrhage, hemotympanum, CSF otorrhea or rhinorrhea, postauricular hematoma (Battle's sign), palpable or visual damage to scalp or skull, and periorbital hematoma (“raccoon eyes”). Child abuse should be suspected if unexplained bruising is present or the stated mechanism of injury is disproportionate to the degree of physical damage present. Other significant physical findings include anisocoria, no pupil reactivity, papilledema, and nuchal rigidity. Purpuric or varicelliform rashes may signify the presence of systemic infections with CNS involvement. Incontinence of urine or stool may indicate that an unwitnessed seizure has occurred.

Neurologic Examination and Scoring

The neurologic examination of the comatose patient should include standard tests of eye opening, responsiveness to verbal and tactile stimuli, and deep tendon reflexes, as well as the more specialized examinations described below. Any focal (unilateral) abnormal finding is always significant because it may indicate a structural CNS lesion. Abnormal findings on neurologic examination reflect the underlying pathologic condition causing coma and may allow localization of a lesion within the brain.

Patients with ALOC benefit from quantification of their impairment using standard measurements. This allows evaluation of patients' changing neurologic status over time and the recording of this information in the medical record. The effect of medical interventions may then be more easily assessed. The use of accepted scoring systems also facilitates communication with consultants such as neurologists and neurosurgeons. In addition, many outcome measures of neurologically injured patients rely upon scales used to assess neurologic function. A widely used measurement of consciousness is the Glasgow Coma Scale (GCS) shown in Table 13.1. Patients are graded on three areas of neurologic function: eye opening, motor responses, and verbal responsiveness. A GCS score of 3 is the minimum score possible and represents complete unresponsiveness; a GCS score of 15 is assigned to fully alert patients.

Pupillary responses provide the most direct window to the brain of a comatose patient. A unilaterally enlarging pupil (greater than 5 mm) that becomes progressively less reactive to light indicates either progressive displacement of the midbrain or medial temporal lobe, or downward displacement of the upper brainstem. Bilateral enlarged and unreactive (“blown”) pupils indicate massive CNS dysfunction and are most commonly seen with posttraumatic increases in ICP. Conditions affecting the brain diffusely usually spare pupillary responses. Exceptions include opiate intoxication, which may cause pinpoint pupils whose constriction is so subtle it may be detected only with an ophthalmoscope. Intoxication with substances having anticholinergic effects, such as scopolamine, is accompanied by widely dilated pupils that may not react to light.

Other ocular signs noted in patients with depressed LOC are the roving side-to-side conjugate eye movements seen in lighter stages of metabolic coma. Persistent conjugate deviation of the eyes to one side may be caused by focal seizure activity, its resultant postictal state, or focal lesions within the brain. Ongoing seizure activity is usually apparent because of the jerking ocular movements present. Most structural brainstem lesions abolish conjugate eye movements, but it is rare for a metabolic disorder to do so. Oculocephalic reflexes (doll's eye movements) consist of conjugate turning of the eyes in the direction opposite brisk head rotation. They should not be checked in any patient who has suffered a traumatic injury because the cervical spine injury may be exacerbated. In the comatose patient, the presence of this reflex implies an intact brainstem and cranial nerves. Deepening ALOC may also be measured by the reduction and loss of spontaneous blinking, then loss of blinking caused by touching the eyelashes, and finally loss of blink with corneal touch. Both eyes should always be tested to detect asymmetry. Neurologically normal patients with 30 degrees of head elevation exhibit an oculovestibular (caloric) response to irrigation of each ear with 10 mL of ice water, consisting of slow conjugate deviation of the eyes toward the irrigated ear with fast beats of nystagmus away from that side. Comatose patients with intact brainstem lose the nystagmic component and have eyes that remain deviated toward the irrigated side for several minutes.

Limb movement and postural changes seen in comatose patients include the bilateral restless movements of the limbs of patients in light coma. Unilateral jerking muscular movements may indicate focal seizure activity or generalized convulsions in a patient with hemiparesis. Decerebrate rigidity refers to stiff extension of limbs with internal rotation of the arms and plantar flexion of the feet. It is not a posture that is held constantly; it usually occurs intermittently in patients with midbrain compression, cerebellar lesions, or metabolic disorders. Decorticate rigidity, when arms are held in flexion and adduction and legs are extended, indicates CNS dysfunction at a higher anatomic level, usually in cerebral white matter or internal capsule and thalamus. Signs of meningeal irritation include Kernig's sign, resistance to bent knee extension with the hip in 90 degrees flexion, and Brudzinski's sign, involuntary knee and hip flexion with passive neck flexion.

The abnormal breathing pattern most commonly seen in comatose patients is Cheyne-Stokes respirations, where intervals of waxing and waning hyperpnea alternate with short periods of apnea. Other abnormal breathing patterns that occur with brainstem lesions include central neurogenic hyperventilation, which can produce a respiratory alkalosis, and apneustic breathing, in which a 2- to 3-second pause occurs during each full inspiration.

Laboratory and Radiologic Studies

Laboratory tests commonly obtained on comatose patients include electrolytes, blood urea nitrogen, creatinine, glucose, blood gases, hemoglobin, hematocrit, and anticonvulsant levels. Toxicologic screening of both blood and urine should be obtained in patients with ALOC of unknown origin. If bedside glucose determination is available, it should be performed on every patient with nontraumatic ALOC. Comatose patients need intravenous access, and laboratory tests may often be obtained at the time of intravenous catheter placement. A noncontrast CT scan of the brain can reveal many of the lesions associated with coma, such as cerebral edema, hydrocephalus, malignancy, hematomas, and abscesses. Infarction and thromboses may require the additions of contrast or the use of magnetic resonance imaging (MRI)

scanning to be fully defined.

Vital Sign Abnormalities

Evaluation and treatment of airway, breathing, and circulatory compromise always take precedence over neurologic problems in the child with ALOC. Airway patency and respiratory effort are both compromised by decreased mental status and may result in hypoxia and/or hypercarbia. The former may be readily measured using pulse oximetry, although values will be inaccurate if a toxic hemoglobinopathy, such as methemoglobinemia or carboxyhemoglobinemia, is present. Hypoxia is usually evident by cyanosis of the lips and nailbeds and pulse oximetry values below 90% (see Chapter 16). Arterial blood gas analysis is useful in some cases to quantify respiratory status and to identify altered hemoglobin states. The treatment of hypoxia, regardless of the cause, always begins with supplemental oxygen administered via an appropriate route.

The numerical definition of hypotension varies with age, but pallor and evidence of poor peripheral perfusion, with capillary refill time greater than 4 seconds, is recognizable even before placement of a sphygmomanometer cuff. Immediate administration of intravenous crystalloid therapy starting with 20 mL/kg of normal saline or lactated Ringer's solution is indicated, followed by additional boluses and pressors if needed (see Chapter 3). Efforts should be made during IV placement to draw blood for laboratory tests. Of the empiric antidotal therapies often used in adults, only glucose 0.25 to 0.5 g/kg is routinely administered to children; an empiric trial of naloxone (1 to 2 mg) is often justified, whereas flumazenil and thiamine are given only when specific indications for their use exist (see Chapter 88).

Severe hypertension is less easily discerned on physical examination. If confirmed in more than one extremity, antihypertensives should be administered via the intravenous or sublingual route (see Chapter 35 and Chapter 86). Mental status should improve after blood pressure is lowered to high normal levels. Patients in hypertensive crises are at risk for hemorrhagic stroke and should be evaluated with a head CT scan if they are neurologically abnormal after blood pressure lowering. Note that hypertension in the comatose patient with increased ICP may represent a physiologic response to maintain cerebral perfusion pressure (by raising mean arterial pressure) and in this context should not be treated with antihypertensives.

Hypothermia and hyperthermia are readily recognized once a core (rectal) temperature less than 35°C or greater than 41°C are obtained. The mental status of these patients should begin to improve as body temperature approaches the normal range. A significant percentage of patients with abnormal core temperatures have drowned, fallen through ice, or were engaged in sporting activities in extreme environments. Head trauma, hypoxia, and/or cervical spine injury may be present in these patients.

History of Head Trauma

The patient with deeply depressed consciousness (GCS score less than 9) after head trauma is presumed to have increased ICP until proven otherwise. Rapid sequence intubation with 1 mg/kg of lidocaine added to standard paralytics and sedatives to blunt rises in ICP caused by laryngeal manipulation is indicated. Cervical spine injury should be assumed and cervical immobilization maintained at all times. An emergent noncontrast brain CT scan should be obtained and neurosurgery consulted.

History of Seizures

The patient with ALOC in the absence of trauma should be evaluated for recent seizure activity with current postictal state (see Chapter 70 and Chapter 83). A history of previous seizures, witnessed convulsive activity, and ALOC consistent with previous postictal periods are valuable clues to this etiology of coma. Ongoing seizure activity may be revealed by the presence of muscular twitching, nystagmus, or fluttering of the eyelids. Subtle forms of status epilepticus may require an electroencephalogram (EEG), to diagnose usually performed somewhat later in the evaluation. The mental status examination of the postictal patient should gradually improve over several hours. Although temporary focal neurologic deficits may follow seizures of any cause, they must be presumed to indicate the presence of focal CNS lesions until proven otherwise or resolved.

The evaluation of neurologically depressed patients with seizures varies based on the patient's history, type of seizure, and presence or absence of fever. Patients with a history of seizures should have serum anticonvulsant concentrations measured and be observed until they approach their neurologic baseline. Children who have had a simple febrile seizure (see Chapter 70) should return to their baseline state soon, usually within 1 hour. Those who remain lethargic or irritable past this point (especially after antipyretics have been administered) should be suspected of having meningitis and are candidates for lumbar puncture. Patients with new-onset generalized seizures who do not meet criteria for simple febrile seizures (see Chapter 28, Chapter 70, and Chapter 83) require more extensive evaluation, which may include measurements of electrolytes (especially sodium, glucose, and calcium), toxicologic screening, examination of CSF, and neurology consultation.

The new onset of focal seizures, with or without the presence of fever, should be evaluated with a head CT scan (using contrast when indicated) to determine the presence of a focal lesion such as a tumor, abscess, or hemorrhage. Only after the results of this study are known should a lumbar puncture be performed. If neuroimaging is unavailable and meningitis or encephalitis is a concern, empiric treatment for bacterial meningitis or herpetic encephalitis may be administered and lumbar puncture deferred (see Chapter 84).

History of Toxic Ingestions

If no history or physical examination findings suggestive of head trauma or seizures are present, a toxic ingestion should be considered, especially in toddlers and adolescents. The availability in the home of any substances capable of depressing CNS function should be thoroughly explored. In general, coma from toxic ingestions is of slower onset than

that from trauma and may be preceded by delirium or other abnormal behaviors.

Chapter 88 lists major toxidromes that result from ingestions that produce CNS depression. The pupils of a poisoned comatose patient are a particularly valuable source of information. Miosis occurs with ingestions of narcotics, clonidine, organophosphates, phencyclidine, phenothiazines, and occasionally, barbiturates and ethanol. Mydriasis is produced by ingestions of anticholinergic agents (e.g., atropine, antihistamines, and tricyclic antidepressants) and sympathomimetic compounds (e.g., amphetamines, caffeine, cocaine, LSD, and nicotine). Nystagmus may indicate the ingestion of barbiturates, ketamine, phencyclidine, or phenytoin. Pupillary responses are more likely to be preserved in toxic or metabolic comas. Systemic toxins do not cause unequal pupils; anisocoria should be pursued with neuroimaging.

A toxicologic screen of blood and urine should be considered in all children with coma of unknown origin. Specific assays for other chemicals may be ordered as suspected. A serum acetaminophen level should be ordered in all children with significant ingestions. Table 13.6 lists compounds capable of causing coma that are not typically detected by drug screening; the compounds are grouped by pupillary effects.

Miosis Present
Clonidine
Chloral hydrate
Organophosphates
Tetrahydrozoline
Bromide
Mydriasis Present
Carbon monoxide
Cyanide
Methemoglobinemia
LSD

Table 13.6. Poisons Undetected by Drug Screening That Cause Coma

The poisoned patient with depressed consciousness should be intubated with a cuffed endotracheal tube for airway protection before decontamination efforts are made. Ipecac-induced emesis is never indicated in the patient with depressed consciousness. Naloxone may be administered as empiric antidotal therapy for coma-producing toxic ingestions involving unknown medications. Flumazenil should not be given to these patients because seizures may result. Its use is limited to pure benzodiazepine overdoses in patients with no history of seizures.

Increased Intracranial Pressure or Focal Neurologic Defect

Nontraumatic causes of increased ICP or focal neurologic deficits include neoplasms, CSF shunt malfunction, and hemorrhage secondary to cerebrovascular disease (see Chapter 83). These patients may present with a history of headache, vomiting, confusion, lethargy, meningismus, focal neurologic dysfunction, seizure activity, or deep coma. Initial physical signs of increased ICP include a bulging fontanelle in infants and sluggishly reactive pupils. More severe and prolonged increases in ICP produce a unilaterally enlarged pupil, other cranial nerve palsies (III, IV, VI), papilledema, and Cushing's triad of hypertension, bradycardia, and periodic breathing. All may signal impending or progressive herniation. From the standpoint of the emergency physician, which type of herniation is present is unimportant; all are life-threatening, and the initial treatment is identical for all. Endotracheal intubation using rapid sequence induction (with cervical immobilization) is performed to minimize increases in ICP while gaining airway control. Evaluation should parallel that of the patient with a traumatic head injury, with the only change being the increased desirability of using intravenous contrast during CT scan. Urgent neurosurgical consultation is recommended for all patients in this category regardless of the focality of findings on CT scan. Comatose patients with a CSF shunt may need their shunt reservoir or ventricle tapped to decrease ICP.

Fever

Coma accompanied by fever indicates that CNS infection may be present (see Chapter 28 and Chapter 84). Resistance to neck flexion is the most important physical finding in meningitis, the most common infection of this type, although children less than 2 years of age may lack this finding. Historical data may also include a steadily increasing headache, irritability, vomiting, and worsening oral intake. Kernig's and Brudzinski's signs may be present. Other useful physical clues to CNS infection are the rashes that accompany meningococcemia, varicella, and Rocky Mountain spotted fever. The historical and physical findings in encephalitis are similar to those in meningitis; meningismus may be absent, however. Seizures are particularly common if herpes simplex is the causative agent.

A history of localized CNS dysfunction or seizures before the onset of febrile coma or the presence of concomitant focal neurologic signs may indicate the presence of a focal cerebral infection such as an abscess, granuloma, or subdural empyema. In addition, either diffuse or focal infections may present with signs of increased ICP secondary to abscess formation, cerebral edema, or blockage of CSF flow. If this is the case, a head CT scan should be obtained before lumbar puncture is performed. A contrast-enhanced study is desirable if concern about focal infection is present. The ill-appearing patient should receive antibiotics before neuroimaging is performed.

CSF analysis remains the key to establishing the diagnosis of CNS infection. Abnormalities of CSF white blood cell count (pleocytosis), glucose, and protein occur in roughly predicable patterns with bacterial or viral meningitis, and pathogens may be visible using Gram and other stains (see Chapter 84). CSF pleocytosis in encephalitis is variable and, if present, is usually mild (less than 500 cells/mm^3), with normal levels of glucose and protein being common. CSF in herpes simplex

encephalitis contains red blood cells in 50% of cases. Bloody or xanthochromic CSF under increased pressure in the absence of signs of infection indicates subarachnoid hemorrhage.

Metabolic Abnormalities

The presence of a metabolic disorder leading to coma is usually apparent once the results of routine laboratory tests are available. These values for glucose, sodium, potassium, bicarbonate, calcium, magnesium, and phosphorus make any deficiency or excess of these serum components readily apparent and treatable. Blood gas analysis for evaluation of acidosis or alkalosis from metabolic or respiratory causes may be indicated as well. Decreased LOC caused by diabetic ketoacidosis may initially worsen because of a paradoxical temporary decrease in CSF pH and/or cerebral edema complicating therapy.

Renal and hepatic function should be quantified with analysis of blood urea nitrogen, creatinine, and ammonia. Markedly elevated serum blood urea nitrogen and creatinine, oliguria, hypertension, anemia, acidosis, and hypocalcemia indicate the presence of uremic coma as a result of renal failure. Hyperammonemia with decreased mental status is most commonly caused by hepatic failure, acetaminophen ingestion with resultant hepatotoxicity, and Reye syndrome. The hyperammonemia of Reye syndrome is accompanied by a history of antecedent viral illness (possibly varicella), resolving within the past week, and likely treated with aspirin (see Chapter 83). Encephalopathy begins soon after unremitting vomiting; jaundice, scleral icterus, focal neurologic signs, and meningeal irritation are absent. Hyperammonemia without accompanying liver failure in the young infant may indicate the presence of a congenital urea cycle defect.

Coma of Unknown Origin

Patients with coma of unknown origin not falling into any of the diagnostic categories discussed previously usually benefit from a noncontrast brain CT scan, CSF analysis, and neurologic consultation, in that order. If meningeal irritation is present without fever or other signs of infection, a subarachnoid hemorrhage may be the cause. Common avoidable errors in the evaluation and management of children with coma are listed in Table 13.7. Patients presenting in a comatose state usually need admission for continuing treatment, observation, and specialized care, except when there is an easily recognized and reversible cause, such as hypoglycemia in a known diabetic.

Assuming no head trauma has taken place if no such history is given
Neglecting to secure the airway before imaging studies are performed
Hyperventilating intubated patients to a Pco_2 well below 35 mm Hg
Not sedating patients once they are paralyzed
Believing that a toxic ingestion has not occurred because the "tox screen" is negative

Table 13.7. Common Errors in the Evaluation and Management of Children with Coma

Suggested Readings

Adams RD, Victor M, Ropper AH. Coma and related disorders of consciousness. In: Adams RD, Victor M, Ropper AH, eds. Principles of Neurology. 6th ed. New York: McGraw-Hill, 1997:344–366.

Brust JCM. Coma. In: Rowland LP, ed. Merritt's Textbook of Neurology. Baltimore: Williams & Wilkins, 1995:19.

Ghajar J, Hariri RJ. Management of pediatric head injury. Pediatr Clin North Am 1992;39:1093–1125.

Schunk JE. The pediatric patient with altered level of consciousness: remember your “immunizations.” J Emerg Nurs 1992;18:419–421.

Schutzman SA, Barnes PD, Mantello M, Scott RM. Epidural hematomas in children. Ann Emerg Med 1993;22:535–541.

Shemie S, Jay V, Rutka J, Armstrong D. Acute obstructive hydrocephalus and sudden death in children. Ann Emerg Med 1997;29:524–528.

Wiley JF. Difficult diagnoses in toxicology. Pediatr Clin North Am 1991;38:725–737.

CHAPTER 14

Constipation

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Constipation is an important problem in the pediatric emergency department for many reasons. It is one of the most common pediatric complaints, accounting for 3% of primary care visits. There are many causes for constipation (Table 14.1), some rare and some very common (Table 14.2). Occasionally, the presentation of constipation is atypical, with chief complaints that superficially seem unrelated to the gastrointestinal tract (Table 14.3). Although relatively rare, some causes of constipation are potentially life-threatening and need to be recognized promptly by the emergency physician (Table 14.4). In addition, constipation may produce symptoms that mimic other serious illnesses such as appendicitis.

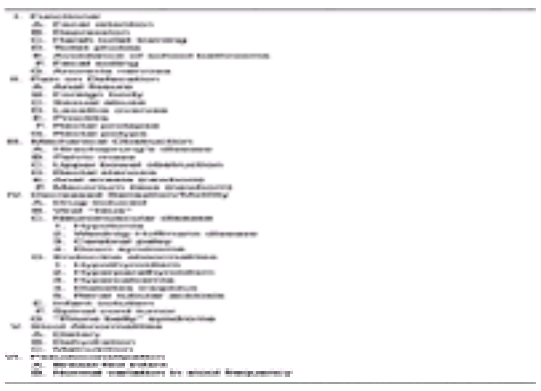

Table 14.1. Etiology of Constipation

Table 14.2. Common Causes of Constipation

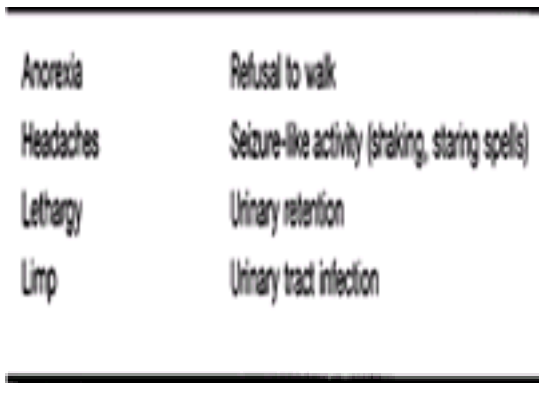

Table 14.3. Some Atypical Presentations of Constipation

Acute Constipation	Chronic Constipation
Mechanical obstruction	Hirschsprung's disease
Dehydration	
Infantile botulism	

Table 14.4. Life-Threatening Causes of Constipation

DEFINITION

Although constipation most commonly is defined as decreased stool frequency, there is not one simple definition. The stooling pattern of children changes based on age, diet, and other factors. Average stooling frequency in infants is approximately 4 stools per day during the first week of life, decreasing to 1.7 stools per day by 2 years of age, and approaching the adult frequency of 1.2 stools per day by 4 years of age. Nevertheless, normal infants can range from 7 stools per day to 1 stool per week. Older children can defecate every 2 to 3 days and be normal.

It is easier to define constipation as a problem with defecation. This may encompass infrequent stooling, passage of large and/or hard stools associated with pain, incomplete evacuation of rectal contents, involuntary soiling (called encopresis), or inability to pass stool at all.

PHYSIOLOGY

The passage of food from mouth to anus is a complex process. The intestine relies on input from intrinsic nerves, extrinsic nerves, and hormones to function properly. Normal defecation involves voluntary and involuntary components. Disruption of any of these can result in constipation.

The colon is specialized to transport fecal material and balance water and electrolytes contained in the feces. When all is functioning well, the fecal bolus arrives in the rectum formed but soft enough for easy passage through the anus.

Normal defecation requires the coordination of the autonomic and somatic nervous systems and normal anatomy of the anorectal region. The internal anal sphincter is smooth muscle, which is innervated by the autonomic nervous system. It is tonically contracted at baseline. It relaxes in response to the arrival of a fecal bolus in the rectum, allowing stool to descend to the portion of the anus innervated by somatic nerves. At this point, the external anal sphincter, striated muscle under voluntary control, tightens until the appropriate time for fecal passage. Before defecation, squatting straightens the angle between the rectum and the anal canal, allowing easier passage. Voluntary relaxation of the external anal sphincter allows passage of the feces, and increasing intra-abdominal pressure via Valsalva aids the process.

EVALUATION AND DECISION

The evaluation of the child presumed to have constipation should begin with a thorough history and physical examination, with special attention paid to changes in frequency and consistency of stool for the individual patient in question. A complaint of constipation is not sufficient. A decrease in stool frequency or the appearance of straining is often interpreted as constipation. The physician should be aware of the grunting baby syndrome in which an infant grunts, turns red, strains, and may cry while passing a soft stool. This is the result of poor coordination between Valsalva and relaxation of the voluntary sphincter muscles. Examination reveals the absence of palpable stool in the rectum or abdomen. Complaints of constipation not supported by history or physical examination are called pseudoconstipation (Fig. 14.1).

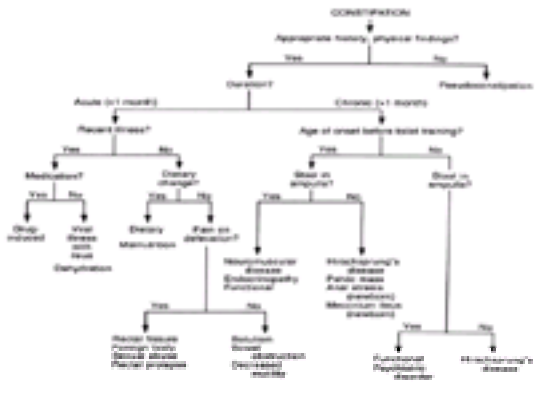

FIGURE 14.1. Approach to constipation.

Acute Constipation

Constipation is not a disease; it is a symptom of a problem. The patient's age and the duration of the constipation are important when determining the cause and significance of the problem. The infant less than 1 year of age with true constipation is particularly concerning.

Constipation of less than 1 month's duration is termed acute. In this case, history will identify many of the problems responsible. A recent viral illness is a common cause of short-lived constipation in the infant. Excessive water loss through vomiting, diarrhea, fever, and increased respiratory rate may cause hard stools. Adynamic ileus or decreased intake after gastroenteritis may cause slower transit time through the colon, and anal fissures after a bout of diarrhea may precipitate painful defecation, resulting in stool retention. The infant may be noted to assume a retentive posture consisting of extension of the body with contraction of the gluteal and anal muscles.

Excessive intake of cow's milk, inadequate fluid intake, and malnutrition should all be uncovered by a complete diet history. Recent courses of medication cannot be overlooked because many can cause constipation (Table 14.5). Ingestion of lead is also a potential and serious reason for constipation.

Aluminum	Iron
Amiodarone	Mesalamine
Amitriptyline	Omeprazole
Anticholinergic agents (benztropine, glycopyrrolate, promethazine)	Ondansetron
	Opioids
Antineoplastic agents (procarbazine, vincristine)	Phenobarbitol
	Phenothiazines and derivatives (prochlorperszine, promethazine, haloperidol)
Benzodiazepines	
β-Blockers	
Calcium salts	Phenytoin
Calcium-channel blockers	Ranitidine
Cholestyramine	Sucralfate
Diazoxide	Ursodiol

Table 14.5. Some Medications Associated with Constipation

Acute constipation can be caused by bowel obstruction, but it is normally a less prominent feature than other symptoms. Infantile botulism commonly presents with constipation, weak cry, poor feeding, and decreasing muscle tone.

Simple constipation in an infant should be treated initially with dietary changes (Table 14.6). Decreasing consumption of cow's milk and increasing fluid intake when appropriate may be enough to alleviate the symptoms. If not improved, constipation can be treated by supplementing the diet with barley malt (Maltsupex) or Karo syrup. Stool lubricants such as mineral oil can be administered orally when aspiration is not a concern. Good perianal care will eliminate stool retention from painful defecation. Follow-up is the most important aspect of treating simple constipation.

I. Dietary Change	IV. Lubricants
A. Increase fluids	A. Mineral oil (1-2 oz BID)
B. Increase carbohydrates (corn sugar syrup, Karo)	B. Kondremul
	C. Milkinol
C. Barley malt (Maltsupex)	V. Local Rectal Care
D. Bran and other fiber	A. Sitz baths
II. Bulk Stool Softeners	B. Vaseline ointment/A&D Ointment
A. Metamucil	
B. Hydrocil	VI. Other Important Treatments
C. Naturacil	A. Patient/parent education
III. Cleansing Enemas/Cathartics	B. Set bathroom time after meal
A. Hypertonic phosphate enema (Fleet) (1–3 only)	C. Proper size toilet with foot support
B. Polyethylene glycol—electrolyte solution (Golytely)	D. Vitamin supplementation
	E. Reward systems/positive reinforcement

Table 14.6. Treatment Steps for Functional Constipation

Acute constipation in the child older than 1 year of age occurs for many of the same reasons as in the infant. History may reveal recent viral illness or use of medication, as well as the presence of underlying illness, such as neuromuscular disease. Physical examination suffices to rule out anal malformations and other physical problems that could result in trouble defecating. Therapy for functional constipation should be the same as that for the infant, with dietary changes a mainstay; however, attention should also be paid to psychological factors such as recent stress that may be complicating the situation.

Chronic Constipation

Constipation of more than 1 month's duration in an infant is especially concerning. Spinal muscular atrophy, amyotonia, congenital absence of abdominal muscles, dystonic states, and spinal dysraphism, which cause problems with

defecation, can be readily diagnosed with history and physical examination.

Anorectal anomalies occur in approximately 1 in 2500 live births. Anal stenosis causes the passage of ribbonlike stools with intense effort. Diagnosis is made by anal examination, which demonstrates a tight, constricted canal. The condition is treated by repeated anal dilations, sometimes over several months. The anus can be covered by a flap of skin, leaving only a portion open for passage of stool. This “covered anus” may require anoplasty with dilation. Anterior displacement of the anus is thought to cause constipation by creating a pouch at the posterior portion of the distal rectum that catches the stool and allows only overflow to be expelled after great straining. The treatment may be medical or surgical.

Hirschsprung's disease, or congenital intestinal aganglionosis, is rare but must be considered in the constipated infant because it has the potential to cause life-threatening complications. The incidence is 1 in 5000 live births, with a male:female predominance of 4:1. As a result of failure of migration of ganglion cell precursors along the gastrointestinal tract, there is the absence of ganglion cells in the submucosal and myenteric plexuses of the affected segment. The absence of ganglion cells leaves the affected segment tonically contracted, blocking passage of stool. The segment proximal to the blockage dilates as the buildup of stool progresses. In most cases, the child never feels the urge to defecate because the blockage is proximal to the internal sphincter and anal canal.

In Hirschsprung's disease, abdominal examination often yields a suprapubic mass of stool that may extend throughout the abdomen. Rectal examination reveals a constricted anal canal with the absence of stool in the rectal vault, commonly followed by expulsion of stool when the finger is removed. The combination of palpable abdominal feces and an empty rectal vault is abnormal and must be further investigated.

Megacolon in Hirschsprung's disease can lead to enterocolitis characterized by abdominal distension; explosive stools, which are sometimes bloody; and fever progressing to sepsis and hypovolemic shock. Enterocolitis represents a major cause of mortality in this condition.

Of infants with Hirschsprung's disease, 80% are diagnosed within the first year of life. A history of late passage of meconium is often found (Table 14.7). However, if the involved segment is relatively short, the diagnosis may be delayed. If suspected, diagnosis is supported by unprepped barium enema, which typically demonstrates narrow bowel rapidly expanding to a dilated area. This transition zone represents the location where the aganglionic, tonically contracted bowel meets the dilated, innervated bowel. In disease where only a short segment of bowel is involved, barium enema may miss the transition zone and anal manometry aids in diagnosis. Confirmation is achieved by demonstration of aganglionosis on biopsy.

	Hirschsprung's	Functional
Onset in infancy	Common	Rare
Delayed passage of meconium	Common	Rare
Painful defecation	Rare	Common
Stool-withholding behavior	Rare	Common
Soiling	Rare	Common
Stool in rectal vault	Rare	Common
Failure to thrive	Common	Rare

Table 14.7. Findings in Hirschsprung's Disease and Functional Constipation

Hypothyroidism in the infant may present with constipation. Water-losing disorders such as diabetes insipidus and renal tubular acidosis may also contribute to this condition. Particularly among whites, cystic fibrosis merits consideration because it may lead to inspissated meconium or stool, presenting as constipation.

Chronic constipation in the older child is overwhelmingly likely to be functional constipation. Typically, a cycle of stool-withholding starts when the child disregards the signal to defecate and strikes a retentive posture—rising on the toes and stiffening the legs and buttocks. This maneuver forces the stool out of the anal canal and back into the rectum, which subjects the fecal bolus to further absorption of water. The longer the stool sits, the more likely defecation is to be painful and traumatic. This reinforces stool-withholding behavior, creating larger and harder stool in the rectum.

Over time in functional constipation, the rectum dilates and sensation diminishes. Eventually, the child loses the urge to defecate altogether. Watery stool from higher in the gastrointestinal tract can leak around the large fecal mass, causing involuntary soiling, or encopresis. This may be misconstrued as diarrhea or as regression in the toilet-trained child. Many parents consult a physician at this point. Other reasons parents seek medical attention for their children are abdominal pain, anorexia, vomiting, and irritability.

Peak times for constipation to develop are when routines change. Toilet training represents a major alteration in the toddler's routine. It also is a time when the child and caregiver battle for control. Another problematic time is after starting school, when a child may be uncomfortable using an unfamiliar bathroom or unable to adapt to a lack of privacy. Involvement with friends or games may distract a child from the signal to defecate. Painful defecation from streptococcal perianal disease or sexual abuse must be remembered as potential precipitants of stool-withholding.

A history supportive of functional constipation includes retentive posturing, infrequent passage of very large stools, and involuntary soiling during the peak ages. Physical examination normally reveals palpable stool in the abdomen. The back

should be inspected for skin changes over the sacral area that suggest spinal dysraphism. Normal deep tendon reflexes and strength in the lower extremities in conjunction with a normal anal-wink reflex virtually excludes neurologic impairment. The anus should be normal in placement and appearance. Rectal examination typically yields a dilated vault filled with stool. Abdominal flatplate can be helpful but is not necessary. Failure to thrive is not associated with functional constipation and, if present, should prompt further investigation.

The patient with functional constipation needs no further evaluation before induction of therapy. Treatment (Table 14.6) begins with evacuation of the stool remaining in the bowel. This is most readily accomplished with hypertonic phosphate (Fleet) enemas. A mineral oil enema administered the night before the first phosphate enema may soften existing stool, allowing less painful passage. The phosphate enemas should be given in doses of 3 mL/kg and administered as pairs 1 hour apart every 12 hours until clear. If there is no response after 2 days, more aggressive disimpaction under physician supervision is indicated. Phosphate enemas should be used with caution in patients with renal impairment. Tap water and soapsuds enemas should be avoided because of the possibility of water intoxication.

A lubricant such as mineral oil should be given orally in a dose adequate to overcome stool retention. Fat-soluble vitamins need to be supplemented during this phase of treatment. Reeducation of bowel habits is vital. Toilet training should be discontinued in the toddler. Regular toileting should be encouraged with positive reinforcement in the school-age child. Close follow-up is a mainstay of treatment. Successful therapy may take months to years to complete.

Although functional constipation encompasses most cases of chronic constipation in the child more than 1 year old, the less common causes must always be considered.

As in the infant, endocrine abnormalities can cause and present as constipation. Hypothyroidism is often associated with constipation, as well as with sluggishness, somnolence, hypothermia, weight gain, and peripheral edema. Increased serum calcium causes constipation through decreased peristalsis. Causes include hyperparathyroidism and hypervitaminosis D. Diabetes mellitus produces increased urinary water loss, possibly leading to harder stool.

Rarely, an abdominal or pelvic mass may present with chronic constipation. Careful abdominal examination will demonstrate the mass. Rectal masses may present similarly. Follow-up again is emphasized because a mass that does not resolve after clearance of impaction needs further evaluation. Hydrometrocolpos can present with constipation and urinary frequency; therefore, a genital examination is indicated in girls to document a perforated hymen. One must also remember that intrauterine pregnancy is a common cause of pelvic mass and constipation in adolescent girls.

Children with neuromuscular disorders often develop constipation. Myasthenia gravis, the muscular dystrophies, and other dystonic states can predispose children to constipation through a number of mechanisms. A detailed history and physical examination should recognize most neuromuscular problems, allowing symptomatic treatment to be provided.

Psychiatric problems must not be forgotten in the evaluation of constipation. Depression can be associated with constipation secondary to decreased intake, irregular diet, and decreased activity. Many psychotropic drugs can cause constipation. Anorexia nervosa may present with constipation because of decreased intake or metabolic abnormalities, and laxative abuse can cause paradoxical constipation.

Suggested Readings

Abi-Hanna A, Lake AM. Constipation and encopresis in childhood. Pediatr Rev 1998;19:123–131.

Fitzgerald JF. Constipation in children. Pediatr Rev 1987;8:10299–10302.

Lewis LG, Rudolph CD. Practical approach to defecation disorders in children. Pediatr Ann 1997;26:4260–4268.

Loening-Baucke V. Chronic constipation in children. Gastroenterology 1993;105:1557–1564.

Loening-Baucke V. Encopresis and soiling. Pediatr Gastroenterol 1996;43:1279–1297.

Scobie WG, Mackinlay GA. Anorectal myectomy in treatment of ultrashort segment Hirschsprung's disease. Arch Dis Child 1977;52:713–715.

Seth R, Heyman MB. Management of constipation and encopresis in infants and children. Pediatr Gastroenterol 1994;23:4621–4636.

Swenson O, Davidson FZ. Similarities of mechanical intestinal obstruction and aganglionic megacolon in the newborn infant. N Engl J Med 1960;262:64–67.

CHAPTER 15

Cough

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Cough is a common pediatric complaint with a variety of causes. Although cough is usually a self-limited symptom associated with upper respiratory illnesses, it occasionally indicates a more serious process. Under most circumstances, history and physical examination can accurately determine the cause.

PATHOPHYSIOLOGY

Cough is a reflex designed to clear the airway. Although a cough can be initiated voluntarily, it is usually elicited by stimulation of receptors located throughout the respiratory tract, from the pharynx to bronchioles. The receptors are triggered by inflammatory, chemical, mechanical, and thermal stimuli. Direct (central) stimulation of a cough center in the brain occurs more rarely. The reflex consists of a forced expiration and sudden opening of the glottis, which rapidly forces air through the airway to expel any mucus or foreign material.

DIFFERENTIAL DIAGNOSIS

The causes of cough differ in the type of stimulus and the site of involvement in the respiratory tract (Table 15.1). The common causes of cough are listed in Table 15.2. Potentially life-threatening causes are listed in Table 15.3.

Infection	*Neoplasm*
Upper respiratory infection	Pharyngeal or nasal polyp
Sinusitis	Hemangioma
Tonsillitis	Papilloma
Laryngitis	Lymphoma
Laryngotracheitis (croup)	Mediastinal tumors
Tracheitis/tracheobronchitis	*Congenital Anomalies*
Bronchiolitis	Cleft palate
Acute bronchitis	Laryngotracheomalacia
Pneumonia	Laryngeal or tracheal webs
Pleuritis	Tracheoesophageal fistula
Bronchiectasis/pulmonary abscess	Vascular ring
Inflammation/Allergy	Pulmonary sequestration
Allergic rhinitis	*Miscellaneous*
Laryngeal edema	Gastroesophageal reflux
Reactive airway disease	Congestive heart failure
Chronic bronchitis	Swallowing dysfunction
Cystic fibrosis	Granulomatous diseases
Mechanical or Chemical Irritation	Psychogenic cough
Foreign body aspiration	Foreign body in otic canal
Neck/chest trauma	
Chemical fumes	
Inhaled particulates	
Smoking	

Table 15.1. Causes of Cough in Children

Upper respiratory infection	Acute bronchitis
Sinusitis	Pneumonia
Laryngotracheitis	Allergic rhinitis
Bronchiolitis	Reactive airway disease

Table 15.2. Common Causes of Cough

Reactive airway disease	Laryngeal edema
Croup	Pertussis
Bronchiolitis	Toxic inhalation
Foreign body	Congestive heart failure
Pneumonia	

Table 15.3. Life-Threatening Causes of Cough

EVALUATION AND DECISION

The history and physical examination are the keys to establishing a diagnosis for cough. The first priority is to recognize and treat any life-threatening conditions: patients with significant respiratory distress should receive supplemental oxygen and rapid assessment of their airway and breathing (Fig. 15.1).

FIGURE 15.1. Approach to the child with cough.

History

Cough can occur as an acute or chronic symptom, depending on the underlying process. Most common and serious causes of cough have an acute onset (Fig. 15.1). Certain conditions, such as asthma, may present with an acute or a chronic history of cough.

The relationship of the cough to other factors is helpful. Cough in the neonate must raise the possibility of congenital anomalies, gastroesophageal reflux, congestive heart failure, and atypical pneumonia (e.g., *Chlamydia*). If the cough began with other upper respiratory tract symptoms or fever, an infectious cause is likely. A cough that started with a choking episode, especially in an older infant or toddler, suggests a foreign body aspiration. Cough associated with exercise or cold exposure, even in the absence of wheezing, may be a sign of reactive airway disease. A primarily nocturnal cough often stems from allergy, sinusitis, or reactive airway disease. Systemic complaints should also be considered in patients with a cough: headache, fever, facial tenderness or pressure (sinusitis), acute dyspnea (asthma, pneumonia, cardiac disease), chest pain (asthma, pleuritis, pneumonia), dysphagia (esophageal foreign body), dysphonia (laryngeal edema or tracheal mass), or weight loss (malignancy or tuberculosis).

The quality of the cough may also be helpful in localizing the process. A barking, seal-like cough with or without stridor supports the diagnosis of laryngotracheitis. A paroxysmal cough associated with an inspiratory “whoop,” cyanosis, or apnea is characteristic of pertussis. Tracheitis gives a deep “brassy” cough, whereas conditions accompanied by wheezing (asthma or bronchiolitis) typically produce a high-pitched “tight” (often termed bronchospastic) cough. Determining whether a cough is productive can be difficult in young children who often swallow, rather than expectorate, their sputum. However, many parents can convey whether the cough is “dry” or “wet.” Although a productive-sounding cough may be seen with uncomplicated upper respiratory infections (URIs), sinusitis and lower respiratory tract infections are commonly accompanied by a productive cough.

Typically, the onset of cough with rhinorrhea suggests a viral URI. However, if a child with an apparent URI becomes more ill or has persistent symptoms, secondary bacterial infections should be considered.

Physical Examination

Patients with a cough require evaluation of the entire respiratory system. Older patients may be able to initiate a typical cough for assessing the quality, and with younger children, gentle gagging with a tongue depressor can trigger a cough. Usually, the cause of the cough can be localized to the upper or lower respiratory tract by the physical examination. Rhinorrhea, congestion, swollen turbinates, sinus tenderness, and pharyngitis are all signs of upper respiratory tract involvement. Allergic features include boggy nasal mucosa, an allergic nasal crease, and allergic “shiners.” An otoscopic exam may reveal a small foreign body (e.g. hair) in the otic canal which may cause chronic cough. Laryngitis and/or

stridor generally imply inflammation or obstruction at the level of the trachea or larynx. Unequal breath sounds, wheezes, rhonchi, and rales are signs of lower respiratory tract disease. A careful cardiac evaluation should be performed, and any clubbing should be noted. Young infants may have respiratory distress with localized upper airway congestion, but older infants and children usually have lower respiratory tract disease if significantly distressed (except in the obvious case of stridor).

Ancillary Studies

In most circumstances of children with a cough, the history and physical examination should be sufficient to make a diagnosis. In patients with unexplained cough or significant or persistent pulmonary signs, a chest radiograph is warranted. In children with an uncomplicated exacerbation of their asthma, a radiograph is unnecessary. If a radiolucent foreign body is suspected, inspiratory and expiratory films or decubitus films should be obtained to detect air trapping (see Chapter 29). Other studies that could be useful in selected patients include sinus films, lateral neck radiographs, barium swallow, and computed tomography of the sinuses, neck, or chest.

In addition, laboratory tests may be necessary for specific diagnoses. Such tests include a complete blood count and differential, blood culture, tuberculin test, nasopharyngeal swab for rapid assays or culture (commonly for pertussis and respiratory syncytial virus), Wright stain of nasal secretions (eosinophils with allergic rhinitis, neutrophils with sinusitis), and sputum culture and Gram stain (neutrophils and Gram-positive diplococci with pneumococcal pneumonia). Pulmonary function testing can be useful to diagnose or follow obstructive airway disease. In cases of airway masses, airway anomalies, foreign bodies, or atypical pneumonias, bronchoscopy may be necessary.

Approach

The major considerations in evaluating a child with cough include the quality of the cough, associated choking or emesis, and the findings of lower respiratory tract signs or fever (Fig. 15.1). Obviously, any child with respiratory distress needs immediate attention to oxygenation and ventilation.

Most patients with cough of acute onset will have a simple upper respiratory tract infection, asthma, bronchiolitis, or pneumonia. A sudden onset with choking or gagging, especially in the preverbal child, is suspicious for a foreign body aspiration (see Chapter 29). A barky cough, with or without stridor, in a child 3 months to 3 years of age suggests laryngotracheitis. Paroxysms of coughing associated with perioral cyanosis, posttussive emesis, or apnea points to pertussis. Visualizing the posterior pharynx with a tongue blade will elicit an episode of coughing.

Physical examination should include inspection of the nares and oropharynx and auscultation of the chest. Wheezing indicates bronchiolitis, asthma, or, rarely, foreign body aspiration. Patients with asthma may complain only of cough and deny any wheezing. Careful auscultation during forced exhalation may detect wheezing or a prolonged expiratory phase. In an older child, significant lower airway obstruction can be measured with a handheld peak flow meter. Asymmetric, or focal, wheezing is seen with lower airway masses and foreign bodies. Rales, rhonchi, and decreased breath sounds are characteristic of lower respiratory tract infection.

The remaining patients with a cough of acute onset will have pneumonia or an upper respiratory tract infection such as viral nasopharyngitis, sinusitis, pharyngitis, or tracheitis. Although rales, decreased breath sounds, or focal wheezing are signs associated with pneumonia, a small proportion of patients with pneumonia may not have any findings by auscultation. Therefore, in cases of significant cough, especially in very young children and those with high fever or elevated white blood cell counts, a chest radiograph is useful to exclude the diagnosis of pneumonia.

Children with chronic cough are likely to have reactive airway disease, allergic rhinitis, or sinusitis. In young children with failure to thrive or recurrent pulmonary infections, cystic fibrosis (see Chapter 96) should be considered. Chronic cough with a history of recurrent pneumonias or chronic bronchitis can also be suggestive of immunodeficiency or anatomic lesions (see Chapter 119). Choking with feeding or emesis followed by cough or wheezing in young infants is typical of gastroesophageal reflux. Newborns who exhibit a cough deserve special consideration for airway anomalies, atypical pneumonias, and congestive heart failure (see Chapter 82). Persistent cough during the day that stops with distraction or sleep is supportive of a psychogenic cause.

TREATMENT

The primary goal should be to treat the underlying process rather than to try to suppress the cough. Patients with any distress need supplemental oxygen and immediate assessment of the airway and breathing. Wheezing from asthma or bronchiolitis is primarily treated with inhaled bronchodilators. In children with suspected reactive airway disease based on history alone, a trial of bronchodilator therapy is warranted. Follow-up with their primary care physician is crucial for establishing a treatment plan. Children with suspected foreign bodies or airway masses (intrinsic or extrinsic to the airway) need appropriate intervention for their removal. Croup treatment consists of mist therapy in mild cases, and racemic epinephrine, steroids, and oxygen for more severe episodes. Treatment of pneumonia depends on the age and suspected pathogen. Patients with pertussis require antibiotics for eradication of the organism, and young infants or any child with significant paroxysms need hospitalization.

Antitussive medications have limited value and should not be used routinely in young infants. It is better to give specific therapy (bronchodilators in asthma, antibiotics with sinusitis) and avoid suppressing a cough in conditions with increased sputum production (e.g., asthma, pneumonia). In older children with a nonproductive cough that interrupts sleep, antitussives can be prescribed. Using cool mist humidifiers and elevating the head during sleep can be beneficial for coughs associated with viral URIs.

Suggested Readings

Cairns-Bazarian AM, et al. Plastic bronchitis: an unusual cause of respiratory distress in children. Pediatr Emerg Care 1992;8(6):335–337 (Review).

Callahan CW. Etiology of chronic cough in a population of children referred to a pediatric pulmonologist. J Am Board Fam Pract 1996;9:324–374.

Cloud G, Cassell G, Tiller R, et al. Infant pneumonitis associated with cytomegalovirus, chlamydia, pneumocystis, and ureaplasma. Pediatrics 1987;79:76–83.

Cloutier MM, Loughlin GM. Chronic cough in children: a manifestation of airway hyperreactivity. Pediatrics 1981;67:6–12.

Cropp GJ. Special features of asthma in children. Chest 1985;87(1 Suppl):55S–62S (Review).

del Rosario JF, Orenstein SR. Evaluation and management of gastroesophageal reflux and pulmonary disease. Curr Opin Pediatr 1996;8(3):209–215 (Review).

Eigen H. The clinical evaluation of chronic cough. Pediatr Clin North Am 1982;29:67–71.

Evans ED, Kramer SS, Kravitz RM. Pediatric diseases of the lower airways. Semin Roentgenol 1998;33(2):136–150 (Review).

Fireman P. Diagnosis of sinusitis in children: emphasis on the history and physical examination. J Allergy Clin Immunol 1992;90(3 Pt 2):433–436 (Review).

Hancock BJ, Di Lorenzo M, Youssef S, Yazbeck S, Marcotte JE, Collin PP. Childhood primary pulmonary neoplasms. J Pediatr Surg 1993;28(9):1133–1136 (Review).

Mansell AL. Update: signs and symptoms of pulmonary disease in children. Clin Chest Med 1987;8(2):329–334 (Review).

Morgan WJ, Taussig LM. The child with persistent cough. Pediatr Rev 1987;8:249–253.

Nguyen KL, Corbett ML, Garcia DP, et al. Chronic sinusitis among pediatric patients with chronic respiratory complaints. J Allergy Clin Immunol 1993;92:824–830.

O'Connell EJ, Rojas AR, Sachs MI. Cough-type asthma: a review. Ann Allergy 1991;66(4):278–282 (Review).

Reisman J, Canny GJ, Levison H. The approach to chronic cough in childhood. Ann Allergy 1988;61:163–169.

Reisman JJ. Advances in respiratory therapeutics in children. Curr Opin Pediatr 1993;5(2):216–222 (Review).

Rencken I, Patton WL, Brasch RC. Airway obstruction in pediatric patients: from croup to BOOP. Radiol Clin North Am 1998;36(1):175–187 (Review).

Roback MG, Dreitlein DA. Chest radiograph in the evaluation of first time wheezing episodes: review of current clinical practice and efficacy. Pediatr Emerg Care 1998;14(3):181–184.

Wald ER, Pang D, Milmoe GJ, Schramm VL Jr. Sinusitis and its complications in the pediatric patient. Pediatr Clin North Am 1981;28:777–796.

CHAPTER 16

Cyanosis

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Cyanosis, a bluish-purple discoloration of the tissues, is a disturbing condition commonly confronted by the pediatric emergency physician. It is most easily appreciated in the lips, nailbeds, earlobes, mucous membranes, and locations where the skin is thin and may be enhanced or obscured by lighting conditions and skin pigmentation.

PATHOPHYSIOLOGY

Three factors that ultimately determine the occurrence of cyanosis are the total amount of hemoglobin (Hb) in the blood, the degree of Hb oxygen saturation or qualitative changes in the Hb, and the state of the circulation.

Oxygenated Hb is bright red, and deoxygenated Hb is purple. Cyanosis is evident when the reduced or deoxygenated Hb in the blood exceeds 5 g/100 mL or when oxygen saturation approaches 85%. When the total amount of Hb in the blood is increased, as in polycythemia, substantial contribution is evident in the overall appearance of the patient from the increased red blood cell mass, and the patient may appear ruddy. The relative increase in the amount of unsaturated Hb in the polycythemic patient will add a blue hue to the skin. Conversely, when the total amount of Hb is decreased, as in anemia, the patient appears pale, and even if Hb is desaturated, cyanosis may not appear.

The degree of Hb saturation is determined by several factors, including the partial pressure of oxygen (P_{O_2}) in the alveolus, the ability of oxygen (O_2) to diffuse across the alveolar epithelial cell wall into the capillary bed and subsequently into the red cell itself, and the Hb molecule. First, because the P_{O_2} in the alveolus is determined by a balance between the amount of O_2 added during alveolar ventilation and that removed by blood flow throughout the alveolar capillary bed, if the level of alveolar ventilation falls, so does the P_{O_2} of alveolar gas, causing a fall in arterial P_{O_2} and desaturation. Second, the ability of O_2 to diffuse across the alveolar wall into the red cell, or gas–blood barrier, is greatly affected by the circumstances of the barrier itself. According to *Fick's law,* the volume of gas per unit time moving across a tissue sheet is directly proportional to the area of the sheet and the difference in partial pressures between the two sides but inversely proportional to the thickness. Any condition that diminishes surface area or increases the thickness will decrease the amount of O_2 in the blood. Third, the Hb molecule itself has unique properties that affect the amount of oxygen it can carry. Although the complexities of Hb and its ability to carry O_2 are beyond the scope of this chapter, to understand cyanosis, it is critical to note that the color of whole blood is in part determined by the state of the Hb molecule. Oxygen binds reversibly to the iron molecule of the Hb subunit, changing its conformation, and oxygenated Hb is bright red. Consequently, factors that affect O_2 binding to Hb will affect the color of the blood. For example, carbon monoxide competitively binds to the ferrous portion of heme, but at an affinity 200 times more than that of oxygen. The change in conformation of Hb with carbon monoxide occupying all the iron in the heme tetramer gives carboxyhemoglobin a cherry red hue, despite the fact that little oxygen is bound to the Hb molecule. In addition, when heme iron is oxidized to the ferric state (it is normally in the ferrous state, even when bound to O_2), known as methemoglobin, it is also incapable of binding O_2. Therefore, Hb will remain deoxygenated, and methemoglobin itself is a brownish-purple color.

The state of the circulation plays an important role in the presence and degree of cyanosis. If a shunt is present, cyanosis can result. A *shunt* is defined as a mechanism by which blood that has not traveled through the ventilated alveolar capillary bed mixes with arterial blood. Deoxygenated blood mixing with oxygenated blood reduces the arterial P_{O_2}, and if the shunt is large, the reduction in P_{O_2} can be severe, leading to marked cyanosis. Another contribution from the state of the circulation on presence of cyanosis concerns blood as it travels through a capillary bed. Oxygen is unloaded to the tissues as blood travels through a capillary, with the relative concentration of unsaturated Hb increasing from one end of the capillary bed to the other. Factors that slow blood flow, such as poor perfusion states and cold temperature, favor the unloading of oxygen and thus increase the amount of unsaturated Hb in the tissue capillaries. A third contribution from the circulation concerns the ratio of blood flow to ventilation within the lung. Simply stated, in an upright lung, the apex is ventilated more than the base, and the base is perfused more than the apex. Because most of the blood flow in the lung then comes from the relatively less ventilated areas of the lung, depression of the blood P_{O_2} is inevitable. In normal healthy subjects, this depression is only a few millimeters of mercury; however, in patients with diseased lungs, the contribution of ventilation/perfusion inequality to lowering of blood P_{O_2} can be significant.

DIFFERENTIAL DIAGNOSIS

The most common causes of cyanosis are cardiac and respiratory diseases that lead to a decrease in the arterial P_{O_2}, but many other conditions can also cause a patient to appear blue (Table 16.1 and Table 16.2). Therefore, consideration of the pathophysiologic framework outlined previously allows an orderly approach to the differential diagnosis of cyanosis. Life-threatening causes of cyanosis are summarized in Table 16.3.

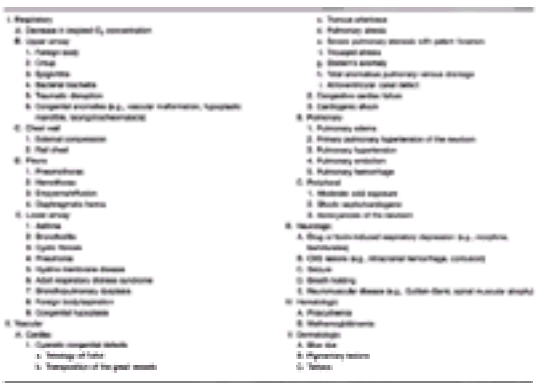

Table 16.1. Causes of Cyanosis

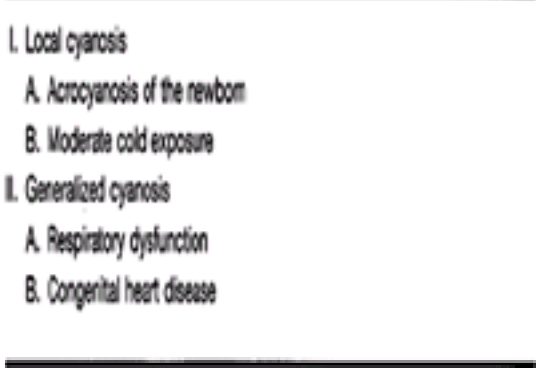

Table 16.2. Common Causes of Cyanosis

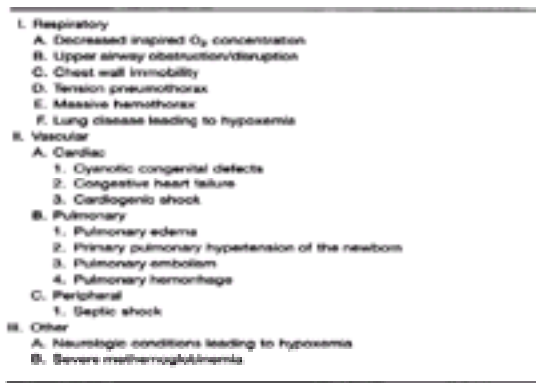

Table 16.3. Life-Threatening Causes of Cyanosis

With regard to the amount of Hb, polycythemia, as in newborns with twin–twin transfusion, infants of diabetic mothers, children with high erythropoietin states, or other conditions associated with increased red cell mass, may give the appearance of cyanosis because of the relative increase in the amount of unsaturated Hb.

The degree of Hb saturation is affected by many factors, which can be grouped conveniently by systems. First is the significant contribution from respiratory conditions. Any circumstance leading to a decrease in the concentration of inspired oxygen, such as a house fire where oxygen is consumed by combustion, confinement to a small unventilated space such as being locked inside a discarded refrigerator, or high altitude, can eventually lead to diminished P_{O_2} and cyanosis. Likewise, upper airway obstruction, as with a foreign body, croup, epiglottitis, bacterial tracheitis, tracheal/bronchial disruption, or congenital airway abnormalities, quickly leads to decreased alveolar ventilation and hypoxemia. Age, events leading to presentation, and examination features, such as barking cough, can help distinguish these. Cyanosis ensues rapidly when chest wall movement or lung inflation is impeded. This condition is often a result of trauma and includes external chest compression, flail chest, or hemothorax. Tension pneumothorax, whether traumatic or as a result of preexisting lung disease such as asthma or cystic fibrosis, is diagnosed by dyspnea, deviated trachea, and possibly distended neck veins with diminished breath sounds on the affected side. Empyema or pleural effusion caused by infection, malignancy, or large chylothorax may be associated with fever, respiratory distress, dullness to percussion, and an asymmetric examination on auscultation. Importantly, any lung dysfunction that directly affects pulmonary gas exchange can lead to cyanosis. The most common conditions in children are asthma, bronchiolitis, pneumonia, cystic fibrosis, pulmonary edema, and hyaline membrane disease. Other causes include bronchopulmonary dysplasia, foreign

body or substance aspiration, and congenital pulmonary lesions, to list a few.

Circulatory or vascular conditions leading to diminished arterial P_{O_2} are also associated with cyanosis. One of the most common causes of cyanosis in children is congenital heart disease. Although most newborns with cyanotic congenital heart disease are discovered while still in the newborn nursery, on occasion, such a newborn will initially present to the emergency department (ED) in the first few days or weeks of life with cyanosis. One condition particularly prone to such late presentation is tetralogy of Fallot, specifically in those infants with concomitant pulmonary atresia who have patent ductus arteriosis–dependent pulmonary blood flow. When the ductus closes, profound cyanosis ensues. Rarely, an infant with mild tetralogy of Fallot (or “pink tet”) may present with intermittent cyanosis during a “tet spell,” which is a 15- to 30-minute self-limited episode of cyanosis caused by increased right-to-left shunting and decrease in pulmonary blood flow. Diagnosis in the “pink tet” is facilitated by presence of a loud systolic murmur. The causes of cyanotic congenital heart disease are listed in Table 16.1, Section II, A. Although many mixing lesions are correctable, several congenital lesions remain with significant shunting of blood from right to left, and these cyanotic children will inevitably be seen in the ED over the course of their lives. Cyanosis may also be caused by pulmonary congestion from cardiac failure or left-to-right cardiac lesions leading to increased pulmonary blood flow and diminished diffusion of O_2 across the gas–blood barrier. (For a detailed discussion of cardiac disease, see Chapter 82.) Several pulmonary vascular abnormalities can lead to cyanosis as well. These include primary pulmonary hypertension of the newborn or pulmonary hypertension from other causes where, because of high pulmonary pressures, blood is shunted away from the lungs and the child becomes hypoxemic. Pulmonary embolism and pulmonary hemorrhage, although rare in children, also impair lung perfusion and must be considered.

Low perfusion states may lead to local cyanosis, particularly of the hands, feet, and lips. Moderate cold exposure slows transit time for red cells across capillary beds, leading to greater unloading of oxygen to the tissues and local blueness. Patients in septic or cardiogenic shock may have perfusion-related cyanosis with long capillary refill times as a result of vascular collapse of sepsis or pump failure. Acrocyanosis, or blueness of the hands and feet with preserved pinkness in the mucous membranes and elsewhere, is seen commonly in newborns and is related to variable perfusion in the extremities. It is seen in well-appearing babies and resolves within the first few days of life.

Neurologic conditions can lead to Hb desaturation and cyanosis as well. Patients who hypoventilate because of central nervous system (CNS) depression, whether from primary CNS lesions or drugs/toxins that depress the respiratory center, are often centrally cyanotic at presentation to the ED. Episodic blue spells in infants and young children who are otherwise well may be caused by breath holding, especially when associated with a sudden insult such as fright, pain, frustration, or anger. Vigorous crying is thought to cause cerebral ischemia via vasoconstriction from decreased P_{CO_2}, decreased cardiac output from Valsalva maneuver, and hypoxemia from apnea (see Chapter 131). Seizures are often associated with cyanosis from inadequate respiration during the convulsion. A variety of neuromuscular diseases that affect chest wall or diaphragmatic function may ultimately lead to hypoventilation.

With respect to the Hb molecule itself, methemoglobinemia is an unusual but not rare reason for admission to the pediatric ED. Methemoglobin can be either congenital or acquired. Congenital methemoglobinemia is caused by either Hb variants designated M hemoglobins or deficiency of NADH-dependent methemoglobin reductase. The more commonly acquired form occurs when red blood cells are exposed to oxidant chemicals and drugs. Young infants with gastroenteritis or oxidant toxin exposure are particularly susceptible to the development of methemoglobinemia as a result of immature enzyme systems required to reduce Hb. Symptoms, caused by decreased blood oxygen content and cellular hypoxia, include headache, dizziness, nausea, dyspnea, confusion, seizure, and coma. Even at low levels, skin discoloration is prominent, often with intense or “slate gray” cyanosis from the presence of methemoglobin as perceived through the skin. (For a more detailed discussion of methemoglobinemia, see Chapter 87.)

Other conditions leading to a blue appearance of the skin may be confused with cyanosis. A rare but perplexing presentation is that of the well-appearing child with unusually localized cyanosis, which after some head scratching, turns out to be related to blue dye of clothing. Certain pigmentary lesions such as mongolian spots can be confused with cyanosis, especially when uncharacteristically large or in unusual locations. Adolescents will occasionally “tattoo” areas of the body that may appear as local cyanosis.

EVALUATION AND DECISION

A careful yet rapid history and physical examination are critical to the approach to the cyanotic patient because timely correction may be lifesaving. Many historical features can help narrow the differential diagnosis and lead to prompt evaluation and treatment. The onset and pattern, location, quality, temporal nature, and presence of palliative or provocative features must be explored. Age of the patient with respect to onset of cyanosis, whether at birth, shortly after birth, or acquired later, is critical. In newborns, congenital cardiac and respiratory disease are the most common causes of cyanosis. Special attention must also be paid to known preexisting heart or lung disease that may predispose to the acute onset of cyanosis. History of exposure to environmental conditions or toxins, such as cold, trauma, clothing dye, smoke inhalation, confinement to an airtight space, drugs, or chemicals, is crucial. Known history or family history of M hemoglobin or deficiency of NADH-dependent methemoglobin reductase may lead directly to the cause of cyanosis. A history of sudden pain or fright with crying or seizure occurrence should be sought.

The physical examination must include a complete general examination, with special attention paid to the vital signs, oxygen saturation, and cardiovascular and pulmonary systems. An immediate and key physical examination feature is the presence or absence of respiratory distress. In general, children with respiratory distress are likely to have *respiratory* dysfunction, and careful examination of the airway, breathing, and circulation should be rapidly initiated. Presence of cough, “sniffing position,” stridor, retraction, or fever should be determined. Lung examination may reveal adventitious (e.g., wheezing or rales) or diminished breath sounds. Presence of a cardiac murmur suggests cardiac disease. Careful attention to the peripheral circulation, including pulses, capillary refill, and temperature, is helpful as well. A rapid neurologic examination should be done. Hypoventilation and subsequent hypoxemia can be the result of many conditions

affecting the CNS.

Location of cyanosis helps determine its cause. Central cyanosis is noted in the mucous membranes, tongue, trunk, and extremities. It is most often the result of decreased arterial P_{O_2} but can also result from severe methemoglobinemia or polycythemia. If the cyanosis is peripheral only (hands, feet, lips), moderate cold exposure, newborn acrocyanosis, shock states, or mild methemoglobinemia may be the cause. Local blue discoloration of a single extremity corresponds to compromise of distal circulation or autonomic tone as seen in traumatic vascular lesions or reflex sympathetic dystrophy. In addition, a local blue hue to the skin may also be a result of simple phenomena such as pigmentary lesions or blue clothing dye. If blue coloring appears on an alcohol swab wiped across the discolored area of skin, dye is responsible. Differential cyanosis of the lower body versus upper may indicate high pulmonary vascular resistance with right-to-left shunting via the ductus arteriosis Transposition of the great arteries with pulmonary-to-aortic shunt of oxygenated blood through the ductus arteriosis is represented in the rare instance that the upper body is blue and the lower body pink.

The path of the laboratory evaluation depends on the historical features and physical findings established on initial encounter (Fig. 16.1). All patients, except very well-appearing newborns and well-appearing cold-exposed patients with peripheral cyanosis only, require measurement of arterial P_{O_2}. (Oxygen saturation by pulse oximetry may be helpful in determining if hypoxemia is the cause of cyanosis, but it may also be misleading when forms of Hb are present other than oxyhemoglobin and deoxyhemoglobin.) If the P_{O_2} is normal, the laboratory evaluation is determined by the degree of ill appearance. Well-appearing oxygenated children with cyanosis usually have less urgent conditions, such as polycythemia, mild methemoglobinemia, cold exposure, newborn acrocyanosis, or dermatologic findings. In this case, laboratory evaluation might include a methemoglobin level and complete blood count or no further investigation may be warranted. Despite a normal P_{O_2}, an ill-appearing cyanotic patient may have a more emergent condition such as severe methemoglobinemia or septic or cardiogenic shock and may require aggressive laboratory investigation, including complete blood count, methemoglobin level, blood cultures, and blood chemistry. Blood with high methemoglobin content may appear very dark or “chocolate brown” and fails to turn red on exposure to air, such as in a drop on filter paper. Treatment is then directed at the underlying cause. Methemoglobinemia may improve with intravenous methylene blue. If the P_{O_2} is decreased, oxygen therapy should be instituted. In general, cyanosis caused by decreased alveolar ventilation or diffusional abnormalities often improves with delivery of 100% O_2. However, hypoxemia caused by decreased pulmonary perfusion responds little to oxygen therapy. Next, a chest radiograph should be obtained. Abnormalities of the lungs may confirm pulmonary disease as a major contributor to hypoxemia, and changes in the cardiac size or silhouette may suggest cardiac causes. If the chest radiograph is normal, other reasons for diminished arterial P_{O_2}, such as CNS or chest wall–related respiratory depression, upper airway obstruction, or pulmonary perfusion abnormalities, must be entertained. If a concomitant murmur or other concern for cardiac disease exists, an electrocardiogram (ECG) is essential. Abnormal ECGs suggest cardiac dysfunction, either congenital or acquired (see Table 16.1), and the addition of echocardiography will help establish the definitive diagnosis.

FIGURE 16.1. Laboratory evaluation of cyanosis.

Suggested Readings

Driscoll DJ. Evaluation of the cyanotic newborn. Pediatr Clin North Am 1990;37:123.

Grant JB, Saltzman AR. Respiratory functions of the lung. In: Baum GL, Wolinsky E, eds. Textbook of Pulmonary Diseases. 5th ed. Boston: Little, Brown, 1994.

Lundsgaard C, Van Slyke DD. Cyanosis. Medicine 1923;2:15.

Martin L, Khalil H. How much reduced hemoglobin is necessary to generate central cyanosis? Chest 1990;97:182.

Watcha M, Connor MT, Hing AV. Pulse oximetry in methemoglobinemia. Am J Dis Child 1989;143:845–847.

West JB. Gas transport to the periphery. In: West JB, ed. Physiological Basis of Medical Practice. 12th ed. Baltimore: Williams & Wilkins, 1990: 538–545.

West JB. Pulmonary gas exchange. In: West JB, ed. Physiological basis of medical practice. 12th ed. Baltimore: Williams & Wilkins, 1990:546–559.

CHAPTER 17

Crying and Colic in Early Infancy

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Crying is the means by which an infant may express discomfort, ranging from normal hunger and desire for company to severe, life-threatening illness. Many common minor irritations and illnesses are excluded by careful history and physical examination. Often, however, a normal, thriving baby will develop a chronic pattern of daily paroxysms of irritability and crying known as colic. The attacks usually have their onset in the second to third week of life and may last for several hours, more commonly in the late afternoon or evening. The typical episode is described as paroxysmal crying that develops into a piercing scream, as if the baby were in pain. The child may draw up the legs, the abdomen may appear distended, bowel sounds are increased, and flatus may be passed, leading parents to conclude that their child has abdominal distress. The emergency physician may be confronted with such a patient and the worried, occasionally hostile parents (usually no earlier than midnight). Colic cannot be cured in the emergency department (ED), and only when crying episodes are repeated and stereotypical and other causes of crying are excluded can the diagnosis of colic be considered. Establishing an orderly approach to the infant with unexplained crying is important to rule out the occasional physical illness and to provide preliminary guidance to the family.

PATHOPHYSIOLOGY

Any unpleasant sensation can cause an infant to cry. Pain or an altered threshold for discomfort (irritability) may be caused by diverse physical illnesses. Those most likely to present abruptly in a young infant are listed in Table 17.1. Numerous unproven theories abound about the etiology of colic. Cow's milk allergy, immaturity of the gastrointestinal tract or central nervous system, parental anxiety, poor feeding technique, and individual temperament characteristics all have been invoked. The search for a specific cause of colic continues. Recent publications suggest that a subgroup of infants who carry the diagnosis of colic may actually have “silent reflux” with resultant esophagitis causing irritability and crying paroxysms amenable to gastroesophageal reflux therapeutic modalities. Others have suggested that elevated serotonin levels found in colicky infants may contribute to the pathogenesis of colic. No single theory (or concomitant therapy) has gained uniform acceptance. Colic may be a syndrome that represents the manifestations of some or all of these factors in varying degrees in a normal population of babies whose tendency to cry varies along a normal distribution. Brazelton's original data on infant crying patterns has been supplemented by larger scale studies in Canada and England. All studies revealed crying levels to increase from birth to a peak of approximately 3 hours per day at 6 to 8 weeks, followed by a rapid decline.

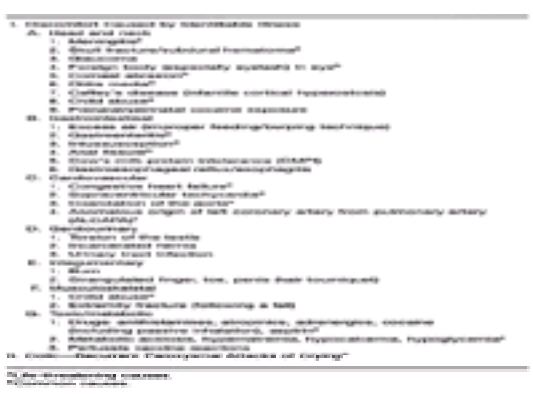

Table 17.1. Conditions Associated with Abrupt Onset of Inconsolable Crying in Young Infants

Early infant crying was also shown to cluster more commonly in afternoon and evening hours. These estimates of crying time over the first 12 weeks of life seem to reflect a certain degree of inconsolable crying behavior that normal infants are destined to exhibit in the first 3 months of life. An encounter with a health care provider is more likely if the infant is difficult to console or if the crying episode is thought to be associated with pain. The most persistent criers would be diagnosed by most pediatricians as having “colic.”

EVALUATION AND DECISION

A careful history and physical examination with emphasis on the head, eyes, ears, skin, abdomen, genitalia, and extremities, plus analysis and culture of a urine specimen, will usually enable the physician to diagnose identifiable illnesses or injuries causing severe paroxysms of crying (Table 17.1). Initially, this clinical evaluation must focus on those conditions that are potentially life-threatening: meningitis, child abuse, intussusception, incarcerated hernia, severe intoxication, and metabolic disturbance. Other less critical but more common conditions should be sought next; corneal abrasion or foreign body, otitis media, aerophagia, teething, gastroenteritis, and anal fissure are most commonly seen. As noted in Table 17.1, other diagnoses are encountered occasionally.

The history should include special attention to the onset of crying and any associated events—particularly recent immunization (“screaming spells” lasting up to 24 hours have been described after pertussis vaccine), trauma, fever, or use of medications. Physical examination must be thorough, with the baby completely undressed. Vital signs may reveal fever, suggesting infection (although not always present in young infants with serious infections), or hyperpnea, suggesting metabolic acidosis.

The head should be explored for evidence of trauma, and the fontanelle should be palpated. Eyes must be examined with fluorescein to look for corneal abrasion, even in infants with no symptoms referable to the eyes. In addition, eversion of the upper eyelids can exclude a foreign body. Funduscopy should be attempted (retinal hemorrhages are common signs of abuse, especially in shaken baby syndrome). Careful otoscopy is required to visualize the tympanic membranes. The heart should be evaluated for signs of congestive failure, arrhythmia, or rare ischemia-producing lesions (Table 17.1, Section I, C.). Abdominal and rectal examinations must be done to look for signs of anal fissure or intussusception. The diaper must be removed, and a careful search should be made for incarcerated hernia, testicular torsion, or strangulation of the penis or clitoris by an encircling hair. Crying may be the primary symptom of an occult urinary infection, so a suitable specimen of urine should be obtained for urinalysis and culture. Careful palpation of all long bones despite the absence of obvious signs of trauma can detect fracture sites that might otherwise have been overlooked. Each finger and toe should be inspected closely to rule out strangulation by hair or thread. Further consideration of laboratory evaluation is made in light of the clinical findings. A low threshold for urine toxicology screening is warranted in the persistently irritable baby, given the escalation in illicit drug use.

Many infants will have a completely negative examination, and the history (or subsequent follow-up) will be suggestive of colic. Over the time in which the crying attacks recur, the infant must demonstrate adequate weight gain (average 5 to 7 ounces/week in the first months of life) and absence of physical disorders on several examinations before underlying illnesses can be excluded and colic can be diagnosed confidently (Fig. 17.1). When it becomes clear that a given infant is experiencing colic, the practitioner faces a vexing problem. No dramatic cure is currently available, but the symptoms almost invariably resolve within 3 months of onset. Dicyclomine hydrochloride, an anticholinergic drug, is the only drug that has been shown to be effective in the treatment of colic; however, this drug has been associated with apnea, and its use is discouraged. Simethicone, although often prescribed, has been shown to be no more effective than placebos in most studies. In general, no safe and effective medical treatment for colic is available. Elimination of cow's milk protein through formula changes is only useful in the small subset of cases (4%) with cow's milk protein intolerance (CMPI). A study by Taubman found that parental counseling (to be more responsive to infant crying with immediate efforts at consoling) was far more effective than dietary manipulation. The safest and most effective course of treatment at this time seems to be counseling and empathy. The physician can reassure the parents that their baby is thriving and will outgrow the colic and develop normally.

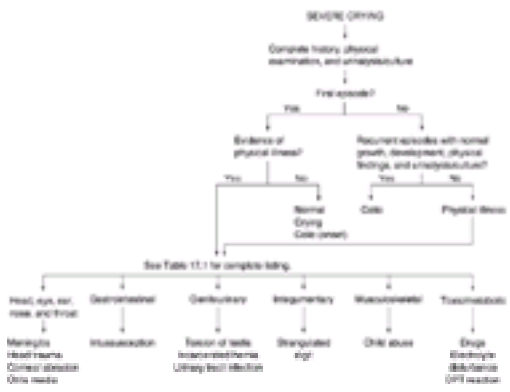

FIGURE 17.1. Approach to abrupt onset of severe crying in infancy. *DPT,* diphtheria-pertussis-tetanus (vaccine).

The emergency physician must be aware of colic as an entity to initiate the evaluation already described, to rule out acute treatable illness, and to refer the family to a pediatrician for follow-up. Colic is not serious and does not last forever, but it probably will be a nuisance for several weeks to come. The physician should stress to the family members their responsibility in the care of the baby so that the mother can get some periods of relief and rest from the child's care. The emergency physician is responsible for investigating the vulnerability of families and children who present with excessive crying. Assessment of the parents' emotional state and the status of available support systems is mandatory. Exhaustion of the parents may be dangerous for the infant, both psychologically and physically.

For immediate amelioration of crying at the time of the ED visit, no drug therapy or feeding change is recommended. Rather, most colicky babies derive some temporary relief from rhythmic motion, such as rocking, being carried, or riding in a car, and from continual monotonous sounds like those from a washing machine or electric fan. A purposefully chosen

circuitous route for the car ride home (one that combines motion and sound) should suffice as therapy for the first visit.

Suggested Readings

Berezin S, Glassman M, Bostwick H, et al. Esophagitis as a cause of infant colic. Clin Pediatr 1995;34(3):158–159.

Berkowitz D, Naveh Y, Berant M. “Infantile colic” as the sole manifestation of gastroesophageal reflux. J Pediatr Gastroenterol Nutr 1997;24(2): 231–233.

Brazelton TB. Crying in infancy. Pediatrics 1962;29:579–588.

Du JNH. Colic as the sole symptom of urinary tract infection in infants. Can Med Assoc J 1976;115:334–339.

Farron L, Farron D. The screaming baby blues. Parents Magazine 1981;July:56–60 (an excellent reference for parents).

Hardoin RA, Henslee JA, Christenson CP, et al. Colic medication and apparent life-threatening events. Clin Pediatr 1991;30:281–285.

Kurtoglu S, Üzüm K, Hallac IK, et al. 5-Hydroxy-3-indole acetic acid levels in infantile colic: is serotoninergic tonus responsible for this problem? Acta Paediatr 1997;85(4):764–765.

Lehtonen LA, Rautava PT. Infantile colic: natural history and treatment. Curr Probl Pediatr 1996;26:79–85.

Mahle WT. A dangerous case of colic: anomalous left coronary artery presenting with paroxysms of irritability. Pediatr Emerg Care 1998; 14(1):247.

Miller AR, Barr RG. Infantile colic: is it a gut issue? Pediatr Clin North Am 1991;38(6):1407–1425.

Mullen N. The problem patient—an irritable infant in respiratory distress. Hosp Prac 1984;January:209–213.

O'Donovan JC, Bradstock AS. The failure of conventional drug therapy in the management of infantile colic. Am J Dis Child 1979;133:999–1001.

Parkin PC, Schwartz CJ, Manuel BA. Randomized controlled trial of three interventions in the management of persistent crying of infancy. Pediatrics 1993;92(2):197–201.

Poole SR. The infant with acute, unexplained, excessive crying. Pediatrics 1991;88(3):450–455.

Singer JI. A fatal case of colic. Pediatr Emerg Care 1992;8(3):171–172.

St. James Roberts. Persistent infant crying. Arch Dis Child 1991;31(3): 653–655.

Taubman B. Parental counseling compared with elimination of cow's milk or soy milk protein for the treatment of infant colic syndrome: a randomized trial. Pediatrics 1988;81:756–761.

Wessel MA. Paroxysmal fussing in infancy, sometimes called “colic.” Pediatrics 1975;14:421–435.

CHAPTER 18

Dehydration

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Dehydration is a physiologic disturbance caused by the reduction or translocation of body fluids and is a type of hypovolemic shock. Infants have higher morbidity and mortality and are more susceptible to dehydration because of their larger water content, higher metabolic turnover rate of water, renal immaturity, and inability to meet their own needs independently. Because dehydration is not a disease itself, children with various illnesses and circumstances will present to the emergency department (ED) with signs of dehydration (Table 18.1).

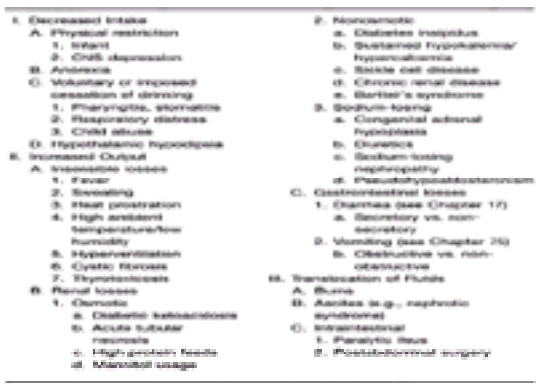

Table 18.1. Causes of Dehydration

Dehydration often is categorized by the patient's osmolarity (the disturbance of distribution of water among body spaces) and severity (degree of fluid deficit), which is helpful in determining fluid therapy. Based on the initial serum sodium, most children have isotonic dehydration (130 to 150 mEq/L), whereas others have hypertonic dehydration (greater than 150 mEq/L) or hypotonic dehydration (less than 130 mEq/L). The terms *isonatremia, hypernatremia,* and *hyponatremia* would be more appropriate. Severity is judged by the amount of body fluid lost or the percentage of weight loss, and the three categories include mild (less than 50 mL/kg, or less than 5%), moderate (50 to 100 mL/kg, or 5 to 10%), and severe (greater than 100 mL/kg, or greater than 10%).

DIFFERENTIAL DIAGNOSIS

Fluid losses in dehydration result from 1) decreased intake; 2) increased output secondary to insensible, renal, or gastrointestinal (GI) losses; or 3) translocation of fluid such as occurs with major burns or ascites (Table 18.1). Diarrhea (see Chapter 19) is the most common cause of dehydration in infants and children and is the leading cause of death worldwide in children less than 4 years of age. In the United States, an average of 300 children under 5 years of age die each year, and an additional 200,000 are hospitalized secondary to diarrheal illnesses with dehydration. Other common causes of dehydration in children include vomiting, stomatitis or pharyngitis with poor intake secondary to pain, febrile illnesses with increased insensible losses and decreased intake, and diabetic ketoacidosis (Table 18.2). More severe or life-threatening causes are listed in Table 18.3.

Table 18.2. Common Diagnoses

Gastroenteritis (especially infants)	Heat prostration
Diabetic ketoacidosis	Gastrointestinal obstruction
Burns over 25% of body surface area	Cystic fibrosis
Thyrotoxicosis	Diabetes insipidus
Congenital adrenal hyperplasia	Child abuse

Table 18.3. Life-Threatening Diagnoses

EVALUATION AND DECISION

The first step in evaluating a child with dehydration is to assess the severity or degree of dehydration, regardless of the cause (Table 18.4). Most children with clinically significant dehydration will have two of the following four clinical findings: 1) capillary refill greater than 2 seconds; 2) dry mucous membranes; 3) no tears; and 4) ill appearance. Dehydration is a type of hypovolemic shock. Mild, moderate, and severe dehydration correspond to impending, compensated, and uncompensated states of shock, respectively (see Chapter 3). If there is severe dehydration or uncompensated shock, the child must be treated immediately with isotonic fluids to restore intravascular volume, as detailed later in this chapter.

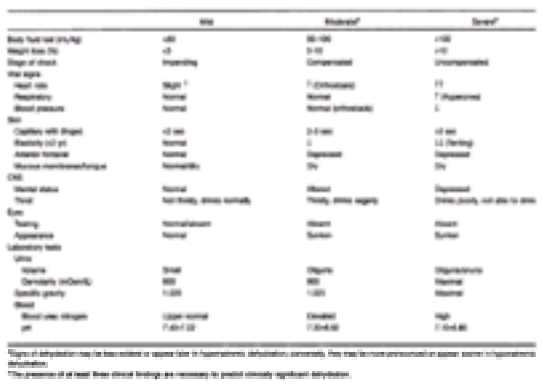

Table 18.4. Clinical Estimation of Degree of Dehydration[a]

History

A thorough history is needed to assess the child with dehydration to determine the cause and degree of dehydration (Fig. 18.1). Particular attention should be paid to the child's output and intake of fluids and minerals. Because overt GI losses from diarrhea and vomiting are the most common causes of dehydration in children, information about the amount and character of these losses is critical in determining a cause (see Chapter 19 and Chapter 78). The child may not be drinking because of physical restriction (e.g., dependence on a caregiver, pain, altered consciousness, anorexia). Fever, high ambient temperatures or bundling a baby, sweating, and hyperventilation may cause increased insensible losses. It is important to note whether there is any underlying disease that would contribute to dehydration (e.g., cystic fibrosis, diabetes, hyperthyroidism, renal disease).

FIGURE 18.1. Suspected dehydration.

Asking the parents about documented weight loss, amount of urine output, and the presence or absence of tears is helpful in determining the severity of the dehydration. All ingested fluids should be noted because diluted juices or water can be associated with hyponatremic dehydration, whereas excess salt intake or low liquid intake may indicate hypernatremic dehydration.

Physical Examination

Vital signs are important and objective parts of the evaluation of the child with dehydration (Table 18.4). The first sign of mild dehydration is tachycardia, whereas hypotension is a late sign of severe dehydration. In mild to moderate dehydration, the respiratory rate usually is normal. As a child becomes more acidotic and fluid depleted, the respiratory rate increases and the breathing pattern becomes hyperpneic. Unfortunately, vital signs alone are not always reliable. Tachycardia also may be caused by fever, agitation, or pain; respiratory illness affects respiratory rates; and orthostatic signs are difficult to obtain in babies and young children.

Age of the child, nutritional status, and type of dehydration also may affect clinical assessment, which is critical to effective management of the acutely dehydrated child. In general, older children show signs of dehydration sooner than babies do because of their lower levels of extracellular water. Fat babies may look less dehydrated than they really are, whereas severely malnourished babies may appear to be more dehydrated secondary to wasted supporting tissues. Signs of dehydration may be less evident or appear later in hypernatremic dehydration. Excessive irritability with increased muscle tone and doughy or smooth and velvety skin often are noted with this type of dehydration. Conversely, signs of dehydration may be more pronounced or appear sooner in hyponatremic dehydration. Keeping these caveats in mind, particular attention should be paid to the overall appearance, mental status, eyes, and skin on physical examination. The mildly dehydrated child usually appears well and may have decreased tearing and a slightly dry mouth. Dry mucous membranes are an early sign of dehydration, but this condition is affected by rapid breathing and ingestion of fluids. Conversely, the severely dehydrated baby classically appears quite ill with lethargy or irritability, a dry mouth, sunken fontanelle, and absent tears. More moderate states of dehydration, however, require more careful evaluation.

The skin is a reliable organ to assess for signs of peripheral perfusion because it is an indicator of the child's systemic vascular resistance and degree of shunting that is occurring to maintain blood pressure. Peripheral and central pulses and skin temperature should be compared. Cool peripheral extremities are an early sign of poor perfusion, whereas weak distal pulses are a very late sign. One of the more objective measures of dehydration is assessment of skin perfusion by measuring capillary refill time. Although the child's body temperature does not affect capillary refill time, it may be falsely prolonged when measured on the foot or in a cool room. Thus, the test should be performed on the fingertip or nailbed in a warm room. Light pressure is applied to blanch the fingernail bed, and the time is measured until color returns (Fig. 18.2). Delays of only 2 to 3 seconds indicate moderate dehydration, and a measurement of more than 3 seconds occurs with severe fluid losses. Skin elasticity can be assessed by determining whether there is a delay in return of skin to its original state after it is pinched into folds (tenting). This is a less reliable finding in older children and malnourished babies who have less subcutaneous tissue.

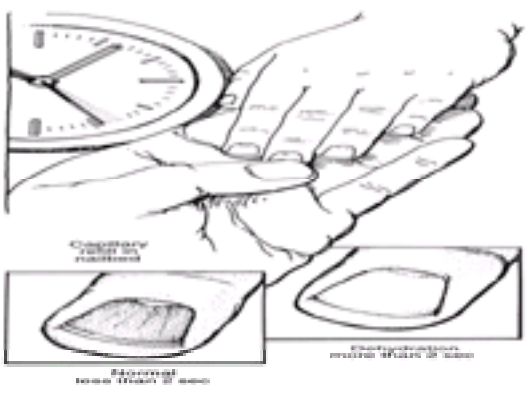

FIGURE 18.2. Assessing dehydration by capillary refill.

Laboratory

The quantity and quality of urine produced also are important indicators of the cause or degree of dehydration. Progressive decrease in urine output and increase in specific gravity and osmolarity are expected with increasing severity of dehydration. If the physical examination indicates significant dehydration and there is dilute or copious urine, a renal or adrenal origin is most likely. The presence of glucose or ketones may indicate diabetic ketoacidosis, and a history of disorders of the central nervous system (CNS) suggests diabetes insipidus.

In children who are judged to have moderate to severe dehydration that requires intravenous (IV) rehydration, laboratory tests of electrolytes, glucose, blood urea nitrogen, and creatinine usually are obtained to determine osmolarity and renal function. The acid-base status may be assessed further with an arterial or venous blood gas.

Diagnostic Approach

In approaching the patient with presumed dehydration, the initial assessment serves to determine whether shock is present. If the child appears to be in shock, resuscitation is called for immediately and a number of life-threatening disorders need to be considered, as listed in Table 18.3 and discussed in Chapter 3. Patients with obvious burns or

diseases that disrupt the integument in the same way (e.g., scalded skin syndrome) are presumed to have become dehydrated through transudation of fluids through the skin.

If the patient does not have an obvious cutaneous source for dehydration, GI losses provide the most likely explanation. A history of vomiting (see Chapter 78) or diarrhea (see Chapter 19) should be sought. Most children with vomiting or diarrhea have viral gastroenteritis, but many diseases (see Table 19.1 and Table 78.1) produce these symptoms. Additional history serves to establish the adequacy of oral intake. Several common minor infections, such as pharyngitis and stomatitis, as well as more serious disorders of the CNS, cause dehydration as a result of voluntary or involuntary limitation of fluids taken orally.

Next, the history should address the nature and quantity of the urine output. With dehydration, one expects to find oliguria or anuria, as seen with many renal diseases (see Chapter 86). The unexpected discovery of polyuria points to diabetes mellitus or insipidus.

By this point, the physician will have established a diagnosis in most patients. In hot weather or when there is prolonged fever, skin losses must be considered. Patients with cystic fibrosis (see Chapter 96) in particular are prone to dehydration under these conditions because of a high concentration of sodium in the sweat (the finding of hyponatremic dehydration seemingly unexplained by the estimated fluid loss should suggest this diagnosis). Additional considerations are listed in Table 18.1.

Initial Management

The dehydrated child must be examined immediately for the degree of dehydration or state of hypovolemic shock. If there is severe dehydration or uncompensated shock, the patient is treated acutely with isotonic fluids to restore intravascular volume regardless of serum osmolarity or cause of the dehydration. Normal saline or Ringer's lactate is given in 20-mL/kg aliquots over approximately 15 to 30 minutes or as quickly as possible if there is uncompensated shock. Reassessment is paramount after each fluid bolus. When blood pressure is restored, heart rate returns to normal, distal pulses strengthen, and skin perfusion improves, isotonic fluids may be safely discontinued. Urine output is the most important indicator of restored intravascular volume and should be a minimum of 1 mL/kg per hour. If dextrose is needed initially for low serum glucose, 0.5 g/kg is given in a single bolus of 10% or 25% dextrose and the serum level is rechecked.

Once the initial resuscitation phase is completed, an IV stock is determined (see Chapter 86). The initial stock often is D5¼NS or D5½NS with 20 mEq/L of potassium chloride. Notable exceptions include major burn patients who continue to require isotonic fluids (see Chapter 114), children with diabetic ketoacidosis who do not require dextrose initially (see Chapter 97), and children with severe electrolyte disturbances such as may occur with pyloric stenosis or severe hypernatremic dehydration (see Chapter 86).

The rate of the chosen stock is determined by the estimated fluid losses. If there are two documented recent weights on the same scale, each kilogram of acute weight lost is equivalent to 1 L of body fluid loss. If previous weights are unavailable, the current weight is assumed to be a certain percentage of the child's ideal or rehydrated weight. For example, if a 9-kg child is assessed to have moderate dehydration with an estimated 10% weight loss, the ideal or rehydrated weight would be 10 kg (9 kg is 90% of 10 kg). Therefore, the child has a 1-L or 1000-mL (100 mL/kg) deficit. Usually, 50% of the child's fluid deficit is given over the first 8 hours in addition to one-third of the maintenance fluid requirements. In the example given, the child would be given 500 mL of fluid replacement plus 333 mL of maintenance fluids over the first 8 hours. In hypertonic states, after initial stabilization with isotonic fluids, the replacement solution is given more slowly to allow equilibration across the blood–brain barrier (see Chapter 86 and Chapter 97).

Initially, oral rehydration should be considered strongly in children with mild or moderate dehydration who do not have uncompensated shock, severe vomiting, high stool output of more than 20 mL/kg per hour, or poor compliance. Oral rehydration also may be instituted after the initial resuscitation phase if the child is feeling well enough to take oral fluids. Initial rehydration solutions that contain 75 to 90 mEq/L of sodium and replacement or maintenance solutions that contain 40 to 60 mEq/L of sodium are available. Both solutions have approximately 20 mEq/L of potassium and a low glucose concentration of 2 to 2.5% to use the sodium–glucose cotransport system in the small intestine. The child is given a volume of rehydration solution equal to the estimated total fluid deficit to drink over approximately 4 to 6 hours. Then, the maintenance solution is begun or the initial rehydrating solution is alternated on a 1:1 basis with a no-sodium or low-sodium fluid such as water, low-carbohydrate juices, or breast milk. Disadvantages to oral rehydration therapy, including the need for prolonged, close observation and frequent feedings, often thwart this effective and safe approach when attempted in the busy EDs of developed countries.

In all types of dehydration and their methods of treatment, the patient must be reassessed continually, urine output monitored closely, ongoing losses quantified and replaced, and therapy individualized. Because dehydration has many causes, the origin of the problem needs to be determined and addressed.

Suggested Readings

American Academy of Pediatrics, Provisional Committee on Quality Improvement, Subcommittee on Acute Gastroenteritis. Practice Parameter. The management of acute gastroenteritis in young children. Pediatrics 1996;97:424.

Duggan C, Santosham M, Glass R. The management of acute diarrhea in children: oral rehydration, maintenance and nutritional therapy. MMWR 1992;41(RR16):120.

Finberg L, Kravath RE, Fleischman AR, eds. Water and Electrolytes in Pediatrics. Philadelphia: WB Saunders, 1982.

Glass RL, Lew JF, Gangarosa RE, et al. Estimates of morbidity and mortality rates for diarrheal diseases in American children. J Pediatr 1991;118(suppl):S27.

Gorelick MH, Shaw KN, Baker MD. Effect of temperature on capillary refill in normal children. Pediatrics 1993;92:699.

Gorelick MH, Shaw KN, Murphy KO. Validity and reliability of clinical signs in the diagnosis of dehydration in children. Pediatrics 1997;99:1. URL: http://www.pediatrics.org/cgi/content/full/99/5/e6 .

Gorelick MH, Shaw KN, Murphy KO, Baker MD. Effect of fever on capillary refill time. Pediatr Emerg Care 1997;13:305.

Hochman HI, Grodin MA, Corne RK. Dehydration, diabetic ketoacidosis, and shock in the pediatric patient. Pediatr Clin North Am 1979; 26:803.

Mackenzie A, Barnes G, Shann F. Clinical signs of dehydration in children. Lancet 1989;2:605.

The Treatment of Diarrhoea. A manual for physicians and other senior health workers. Division of Diarrhoeal and Acute Respiratory Disease Control. World Health Organization. WHO/CDD/SER/80.2, 3rd ed.

CHAPTER 19

Diarrhea

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Diarrhea refers to a softening in the consistency of the stool with or without an increase in the number of stools. Because of the variability in the frequency and type of stools among children, absolute limits of normalcy are difficult to define. Rather, any deviation from the child's usual pattern should arouse concern, regardless of the actual number of stools or their water content. Some infants, particularly those who are breast-fed, often have five or six loose stools daily as their normal routine; other healthy infants may produce only one formed stool every other day.

DIFFERENTIAL DIAGNOSIS

Diarrhea, with or without vomiting, often prompts a visit to the emergency department (ED). An estimated 15 to 20 million children less than 5 years of age have between 20 and 40 million episodes of diarrhea annually in the United States. Approximately 12% of all hospitalizations of children 1 month through 4 years of age include diarrhea among one of the top three positions on the list of discharge diagnoses. Although most bouts of diarrhea seen in the ED in developed countries result from self-limiting infections, diarrhea may be the initial manifestation of a wide spectrum of disorders, as outlined in Table 19.1.

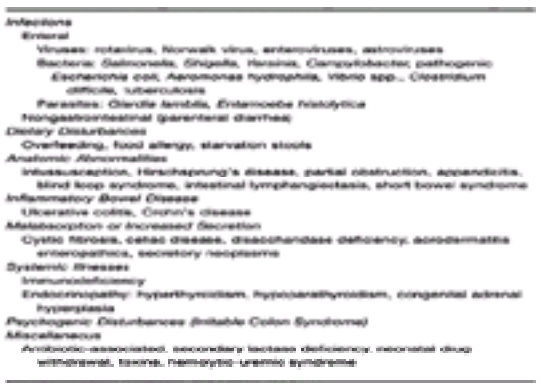

Table 19.1. Causes of Diarrhea

Of the many causes of diarrhea, a few are particularly common: infections with viruses and bacteria, parenteral diarrhea, and diarrhea associated with antibiotic administration (Table 19.2). The single most common disorder seen in the ED is viral gastroenteritis.

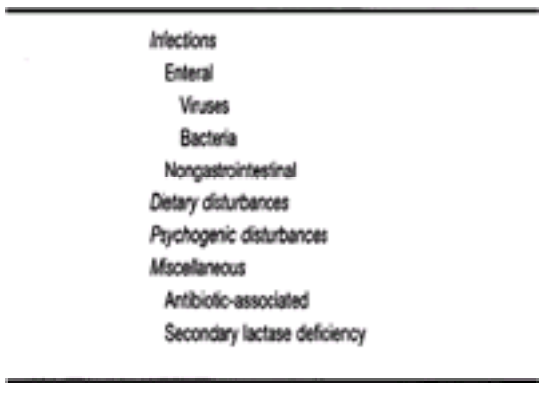

Table 19.2. Common Causes of Diarrhea

Any cause of diarrhea may produce a fatality secondary to dehydration. Approximately 400 young children die annually in the United States from gastroenteritis; however, most of the disorders, particularly viral gastroenteritis, are mild. The emergency physician must be vigilant in recognizing the few children who have diseases that are likely to be life-threatening from among the majority of children who have self-limiting infections. Particularly urgent are intussusception, pseudomembranous colitis, hemolytic uremic syndrome (HUS), and appendicitis (Table 19.3).

Intussusception
Hemolytic uremic syndrome
Pseudomembranous colitis
Appendicitis
Salmonella gastroenteritis (with bacteremia in the neonate or compromised host)
Hirschsprung's disease (with toxic megacolon)
Inflammatory bowel disease (with toxic megacolon)

Table 19.3. Life-Threatening Causes of Diarrhea

Intussusception is the most common of these serious disorders to have diarrhea as a predominant symptom. Although many children with intussusception primarily have rectal bleeding or severe abdominal pain, a significant number are brought to the hospital with the complaint of bloody diarrhea. Intussusception peaks in frequency between 5 and 10 months of age and tapers off rapidly after 2 years of age, unless there is a predisposing pathologic condition. Although the occasional child may be febrile, the temperature usually is within the normal range. Classically, intussusception causes colicky abdominal pain, vomiting, and an abdominal mass, in addition to a “currant jelly” stool. The finding of a mass in association with colicky abdominal pain, vomiting, and a currant jelly stool is pathognomonic for intussusception, but a mass actually is palpable in fewer than 50% of cases. Thus, the physician should consider this diagnosis even in the absence of a mass when a child in the first year or two of life has the combination of bloody diarrhea and severe, colicky abdominal pain. In addition, the child who is flaccid or lethargic out of proportion to the degree of dehydration should arouse the examiner's suspicions; intussusception can evoke “neurologic” signs. Plain films of the abdomen may be diagnostic (intussusception seen on the basis of air contrast), suggestive (mechanical obstruction), or nonspecific (normal or ileus); thus, a high index of suspicion mandates a contrast enema with air or barium.

HUS, although uncommon, merits consideration in the child with bloody diarrhea because it is a potentially fatal illness. Children are affected most often in the first 3 years of life. Over the course of several days, an initially mild gastroenteritis becomes complicated first by hematochezia and then by pallor (anemia), purpura (thrombocytopenia), hematuria (nephritis), and finally renal failure. When HUS is suspected, a complete blood count (CBC), urinalysis, and coagulation studies should be performed. The peripheral blood smear, in addition to reduced numbers of platelets, shows evidence of intravascular hemolysis, including helmet cells and red blood cell fragments. The urine tests positive for blood and may contain casts on microscopic examination.

Another serious disorder that may cause bloody diarrhea is pseudomembranous colitis. This disease results from an overgrowth of toxin-producing clostridial organisms in the bowel and must be considered after a course of antibiotic therapy, which can decimate the normal flora of the gut. It may occur at any age but is uncommon in early childhood. Although the incidence of pseudomembranous colitis is highest after treatment with clindamycin, an infrequently prescribed antibiotic, any of the antibacterial drugs may be the culprit. In fact, because of its frequent use, amoxicillin is responsible for most cases of pseudomembranous colitis in childhood, even though the incidence after therapy with this agent is low. Clinically, the patient with pseudomembranous colitis usually appears ill with prostration, abdominal distension, and significant amounts of blood in the stool. Microscopic examination of the feces, sigmoidoscopy, and stool toxin analysis are all useful tests.

Appendicitis manifests primarily with abdominal pain, followed by vomiting, often in association with constipation. Less commonly, appendicitis may cause diarrhea. The presumed mechanism for the diarrhea is irritation of the colon by the inflamed appendix. In most cases, careful questioning about the nature of the diarrhea will reveal a description of frequent, very low volume stools, with mucus. Particularly in very young children or among patients of any age who have a perforated appendix and a long duration of illness, the diagnosis of appendicitis as the cause of diarrhea may be delayed because the classic constellation of findings is often absent. However, the examiner will usually be able to elicit abdominal tenderness greater than that which would be expected with gastroenteritis.

EVALUATION AND DECISION

The initial evaluation of the child with diarrhea should serve the dual purpose of exploring the possible causes and assessing the degree of illness. Preexisting conditions in the child may account for the diarrhea or predispose him or her to unusual causes; in particular, the emergency physician should search for a history of gastrointestinal surgery or chronic illnesses, such as ulcerative colitis or regional enteritis. Immunodeficiency syndromes, neoplasms, and immunosuppressive therapy all lead to an increased susceptibility to infection. Institutionalized children and those recently returning from underdeveloped countries are more likely to harbor bacterial or parasitic pathogens.

A history of abdominal pain, particularly if severe, raises the index of suspicion for intussusception and appendicitis. Bloody diarrhea points particularly to bacterial enteritis but occasionally occurs with viral infections and may also herald

the onset of HUS or pseudomembranous colitis. The combination of episodic abdominal pain and blood in the stool characterizes intussusception. Vomiting in association with diarrhea is very suggestive of viral gastroenteritis, whereas vomiting in isolation (see Chapter 78) is more concerning.

With the initial interview, the physician should attempt to reconstruct historically the child's intake and output during the course of the illness. Detailed questions about the number and size of stools, the frequency of emesis, and the amount of liquid taken orally allow for an estimate of fluid balance. Decreases in the frequency or volume of urination (or the number of diaper changes in the infant) suggest an inadequate output, reflecting the development of dehydration.

The general physical examination can provide clues to an underlying illness in the child who appears malnourished or small for his or her age. The weight of the child always should be measured carefully and compared with weights previously recorded in the chart. If fever is present, infectious causes are most likely. The pulse and blood pressure, together with the turgor of the skin and mucous membranes, are useful in assessing the degree of dehydration (Table 19.4), except in the child who has hypernatremia. On abdominal examination, the finding of a mass (regional enteritis, intussusception) or evidence of obstruction is important. A rectal examination should be performed in the child who has chronic diarrhea. With overflow stools secondary to prolonged constipation, the rectal ampulla contains a large amount of hard stool, but it often is empty in the patient with Hirschsprung's disease. For selected children, laboratory measurements may assist in the evaluation of dehydration, but they often fall in the normal range despite marked loss of fluids.

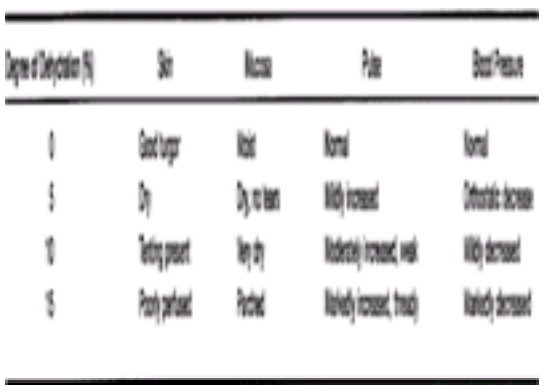

Table 19.4. Clinical Findings in Dehydration

A diagnostic approach to the pediatric patient with diarrhea is outlined in Figure 19.1. The physician should first determine whether the child appears seriously ill or has signs of a surgical abdominal process. Once it has been determined that immediate signs of a life-threatening process are absent, more than any other feature, the duration of diarrhea dictates the initial diagnostic considerations. Children with chronic diarrhea (more than 5 days) are likely to have irritable bowel syndrome, infections, inflammatory bowel disease, or various malabsorptive disorders. Such conditions, if uncomplicated, do not require a definitive diagnosis emergently but rather an extensive evaluation over time.

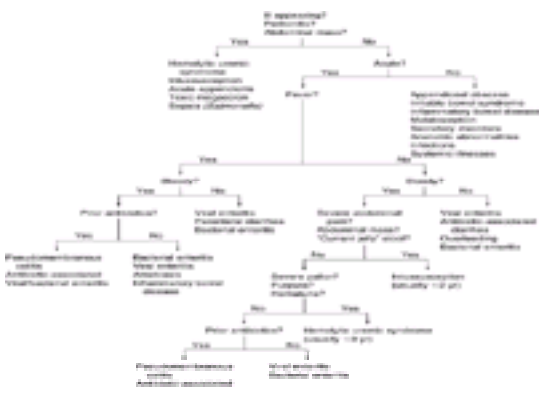

FIGURE 19.1. Diagnostic approach to the immunocompetent child with diarrhea.

Acute Diarrhea

With the acute onset of diarrhea, most children have an infectious cause for their disorder and will require evaluation in the ED. Fever, the hallmark of infection, serves as the first branch point in the approach to such patients. Although not all children with infectious enteritis have fevers, the finding of an elevated temperature points strongly in this direction. At the same time, the absence of fever, particularly in the presence of bloody stools, should alert the physician to the possibility of one of several serious noninfectious diseases, particularly intussusception and HUS.

The next question is whether hematochezia (bloody stool) is present. Blood is seen in the stool of approximately 10% of children with diarrhea. In most cases, the blood appears in small quantities as drops on the surface of the stool and should not be construed as ominous. A small percentage of children with diarrhea, however, have more profuse rectal bleeding. In these patients, one must exclude life-threatening disorders such as intussusception, HUS, and pseudomembranous colitis.

Febrile children with bloody diarrhea (Fig. 19.1) almost invariably have an infectious enteritis. Pseudomembranous colitis should be considered in patients who have received antibiotic therapy, but this diagnosis usually can be discarded on clinical grounds in the absence of systemic toxicity, abdominal distension, and gross blood in the stools. If pseudomembranous colitis is strongly suspected, admission to the hospital and a full diagnostic evaluation should be considered. Bacterial diarrhea should be sought by culture in febrile children with frankly bloody diarrhea but will be found only in 15 to 20% of cases; viral enteritis is much more common. In the first few months of life, in the infant for whom *Salmonella* gastroenteritis represents a more serious illness, a stool smear for polymorphonuclear leukocytes is useful because the finding of sheets of inflammatory cells strongly points to a bacterial origin. Amebiasis merits consideration only in endemic areas and among travelers. Finally, an occasional child with inflammatory bowel disease may present with an initial episode of acute, bloody diarrhea. With most of these children, the physician can elicit a preceding history of weight loss or recurrent abdominal pain; in the remainder, the diagnosis emerges when bloody diarrhea persists in the face of negative cultures.

Most febrile children with nonbloody diarrhea (Fig. 19.1) have viral enteritis. The physician must perform a thorough examination because nonenteric infections, particularly otitis media, may cause “parenteral” diarrhea. For similar reasons, a urine culture is indicated if any historical factors point to an infection of the urinary tract. Although a small percentage of these patients have a bacterial enteritis, routine cultures of stool are not recommended for nonbloody diarrhea of brief duration in otherwise healthy children. Immunocompromised patients, such as those with acquired immunodeficiency syndrome (AIDS) (see Chapter 85), require a more thorough evaluation, including bacterial cultures and examination for ova and parasites.

Afebrile children with bloody diarrhea (Fig. 19.1) represent the most worrisome category because most patients with intussusception, HUS, and pseudomembranous colitis have this symptom constellation. In particular, intussusception should be considered carefully in any child less than 1 year of age with bloody diarrhea that does not appear to have an infectious cause. Although the finding of a mass or a currant jelly stool is pathognomonic, a history of severe, colicky abdominal pain in a lethargic child warrants a contrast enema. Obvious pallor, purpura, and hematuria point to HUS, an unusual but potentially life-threatening disease. Once again, prior antibiotic therapy raises the possibility of pseudomembranous colitis. The most common diagnosis, infectious enteritis, should be assigned only after exclusion of the more serious disorders by history, physical examination, and occasionally, laboratory or radiographic studies.

Afebrile children with nonbloody diarrhea (Fig. 19.1) usually are judged to have viral enteritis. Those who receive antibiotic agents, such as amoxicillin, may be suffering from a drug-related gastrointestinal disturbance but not usually from pseudomembranous colitis. During the first 6 to 12 months of life, overfeeding may manifest as diarrhea. The tip-off to this diagnosis is the history of excessive intake in the overweight child. Bacterial enteritis, although a possibility, does not merit a stool culture in the usual clinical circumstances.

Chronic Diarrhea

Chronic diarrhea precipitates a child's visit to an ED less often than acute gastroenteritis does. An apparent worsening of a long-standing disease may be a final frustration on the part of the parents, however, particularly on a weekend when the family's usual physician may be unavailable. The evaluation of chronic diarrhea usually requires a period of observation and laboratory evaluation beyond the scope of the ED. In the management of these children, the role of the emergency physician is to select those few children who have urgent conditions and refer the remainder to their regular source of care. Particularly in the infant, consideration must be given to Hirschsprung's disease and to cystic fibrosis. A history of delayed passage of meconium, constipation since birth, and abdominal distension are compatible with Hirschsprung's disease. Malabsorptive stools and respiratory infections suggest cystic fibrosis. Failure to thrive, thrush, and pneumonia occur in association with human immunodeficiency virus (HIV) infection. A stool culture and examination for parasites serve to diagnose the serious infections of the gastrointestinal tract and provide a head start on the evaluation to the physician who subsequently sees the child.

The child who returns to the ED with the persistence of an acute diarrheal illness, presumed to be viral in origin and with no evidence of malnutrition or dehydration, often may be managed without an extensive evaluation. Three causes are common: 1) bacterial infections; 2) secondary lactase deficiency from mucosal sloughing; and 3) starvation stools in the child who inadvertently has been continued on a clear liquid diet for several days. A stool culture should be obtained, and testing for clostridial toxin is indicated in the presence of ongoing antibiotic therapy. If the child has remained on a clear liquid diet, gradual refeeding is recommended. Milk and all milk products should be proscribed temporarily when secondary lactase deficiency is suspected.

TREATMENT

The treatments for the myriad causes of diarrhea are covered in the medical and surgical sections of this book; however, the therapy for viral gastroenteritis or parenteral diarrhea merits a brief summary. Although all children with circulatory compromise and many children with moderate to severe dehydration need intravenous fluids, many patients with gastroenteritis can be managed with oral solutions. Most children, even those with vomiting, will tolerate frequent, small feedings, but occasionally delivery of fluids via a nasogastric tube may be helpful.

Optimal oral therapy emphasizes the use of appropriate glucose and electrolyte solutions as well as the early reintroduction of feeding. The ideal solutions, based on formulas carefully tested by the World Health Organization, have a carbohydrate:sodium ratio that approaches 1:1. Generally, initial rehydration is accomplished with a solution that contains 75 to 90 mEq/L of sodium (i.e., Rehydralyte®) and subsequent maintenance with a more hypotonic formulation (i.e., Pedialyte®). Older children with mild gastroenteritis tolerate juices and other household products, even though the carbohydrate:sodium ratio deviates from the standard. Recent studies have suggested that the use of glucose polymers (i.e., Ricelyte®) leads to a decrease in the amount and duration of diarrhea, compared with glucose alone; presumably,

these newer solutions will find more widespread use, particularly for patients with voluminous or prolonged diarrhea.

In general, antidiarrheal agents are ineffective and have no role in the treatment of infectious gastroenteritis during childhood; agents that decrease intestinal mobility (i.e., Lomotil ®) carry an additional risk of toxicity.

Preliminary studies that point to a decrease in stool output with bismuth subsalicylate (Pepto-Bismol) require confirmation of efficacy and safety before recommendations for use in children can be made. When selected pathogens are isolated or strongly suspected, antimicrobial agents should be prescribed.

Suggested Readings

American Academy of Pediatrics, Committee on Nutrition. Use of oral fluid therapy and posttreatment feeding following enteritis in children in a developed country. Pediatrics 1985;75:358.

Bonadio WA, Hennes HH, Machi J. Efficacy of measuring BUN in assessing children with dehydration due to gastroenteritis. Ann Emerg Med 1989; 18:755.

Brown KH. Dietary management of acute childhood diarrhea: optimal timing of feeding and appropriate use of milk and mixed diets. Pediatrics 1991;118:S92.

Cohen MB. Etiology and mechanisms of acute infectious diarrhea in infants in the United States. Pediatrics 1991;118:S34.

Dewitt TG, Humphrey F, McCarthy P. Clinical predictors of acute bacterial disease in young children. Pediatrics 1985;76:551.

Finkelstein JA, Schwartz JS, Torrey S, Fleisher GR. Common clinical features as predictors of bacterial diarrhea in infants. Am J Emerg Med 1989;7:469.

Glass RI, Lew JF, Gangarosa RE, et al. Estimates of morbidity and mortality rates for diarrheal diseases in American children. Pediatrics 1991;118:S27.

Gorelick MH, Shaw KN, Murphy KO. Validity and reliability of clinical signs in the diagnosis of dehydration in children. Pediatrics 1997;99:E6.

Gremse DA. Effectiveness of nasogastric rehydration in hospitalized children with acute diarrhea. J Pediatr Gastroenterol Nutr 1995;21: 145–148.

Ho MS, Glass RI, Pinsky RF, et al. Diarrheal deaths in American children: are they preventable? JAMA 1988;260:3281.

Horwitz JR, Gursoy M, Jaksic T, Lally KP. Importance of diarrhea as a presenting symptom of appendicitis in very young children. Am J Surg 1997;173:80–82.

Huicho L, Sanchez D, Contreras M, et al. Occult blood and fecal leukocytes as screening tests in childhood infectious diarrhea: an old problem revisited. Pediatr Infect Dis J 1993;12:474–477.

Lebenthal E, Lu RB. Glucose polymers as an alternative to glucose in oral rehydration solutions. Pediatrics 1991;118:S62.

Picus D, Shackelford GD. Perforated appendix presenting with severe diarrhea. Radiology 1983;149:141–143.

Reid SR, Bonadio WA. Outpatient rapid intravenous rehydration to correct dehydration and resolve vomiting in children with acute gastroenteritis. Ann Emerg Med 1996;28:318–323.

Rowe PC, Orrbine E, Lior H, et al. Risk of hemolytic uremic syndrome after sporadic *Escherichia coli* O157:H7 infection; results of a Canadian collaborative study. J Pediatr 1998;132:777–782.

Snyder JD. Use and misuse of oral therapy for diarrhea: comparison of U.S. practices with American Academy of Pediatrics recommendations. Pediatrics 1991;87:28.

Sorrano-Brucker H, Avendano P, O'Ryan M, et al. Bismuth subsalicylate in the treatment of acute diarrhea in children: a clinical study. Pediatrics 1991;87:18.

Torrey S, Fleisher G, Jaffe D. Incidence of Salmonella bacteremia in infants with Salmonella gastroenteritis. J Pediatr 1986;108:715.

CHAPTER 20

Disturbed Child

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This chapter presents an approach for the diagnosis of the acutely disturbed child who manifests agitation and aggression or withdrawal. Additional details of management of the conditions discussed here are found in Chapter 129.

Human beings generally respond to stress or personal threats by developing a fight-or-flight response. They become aggressive in an attempt to confront the threat or they withdraw to maintain safety. Throughout their development, regardless of age or developmental stage, children respond to threats through fight (agitation and aggression) or flight (withdrawal). Although these behaviors differ in their manifestations at different ages, the underlying responses are similar throughout childhood.

At times, the child's ways of responding to external events and changes in the environment are inadequate. In other situations, a previously supportive environment no longer provides security and protection. As a result, the child may no longer be in control of his or her social and emotional responses. It is usually at this point of crisis that the emergency physician meets the child and his or her caretakers.

Although agitation and withdrawal are distinctly different behaviors, it is important to recognize that a child in crisis can fluctuate easily from one to the other. For example, a sullen, withdrawn, and uncooperative adolescent may become agitated, angry, and disruptive in the face of additional stress. Also, both agitation and withdrawal can result from the same underlying physical and psychologic causes. Thus, both agitation and withdrawal as presenting symptoms in children in crisis are signs of significant emotional stress that the emergency physician should recognize and be prepared to treat.

The agitated child is typically anxious, upset, and unresponsive to attempts at support. The child may pace back and forth and may threaten staff or family. Speech usually is loud and may be abusive. Some children also may be disoriented and out of contact with reality. When agitated, younger children may be out of control, running about the examining room and having severe temper tantrums. They may cry or strike out at the parent or physician. Older children or adolescents may be distraught, sullen, and angry as they meet the examiner. It is important for the emergency physician to be aware of his or her own responses to agitated children; feelings of anger and frustration in the physician often reflect the amount of stress that the child is experiencing. Some children or adolescents, however, may appear to be calm and under control when seen in the emergency department (ED). Information from the parents may reveal significant destructiveness at home before coming to the hospital. In many cases, the improvement in the child's behavior in the ED is a response to the structure provided and the sense that help will be forthcoming.

The withdrawn child primarily demonstrates significant unresponsiveness to the demands of the situation. He or she is inappropriately quiet and does not attempt to develop any relationship with the emergency physician. In younger children, this withdrawal may be demonstrated by clinging behavior, whining, and crying. The child may be unresponsive to parents as well as to the physician, instead responding to internal stimuli and demonstrating inappropriate affect. Older children may be sullen, unresponsive, or apathetic when asked about the precipitant of the current ED visit. It is important to distinguish emotional withdrawal from shyness, which is a temperamental quality within the range of normal behavior.

DIFFERENTIAL DIAGNOSIS

A wide variety of medical and psychiatric conditions can lead to a child's development of significant agitation and withdrawal. These disorders are listed in Table 20.1 and include severe psychiatric disturbances, life-threatening medical conditions, and minor aberrations in the child's ability to respond to stressful events. Regardless of the underlying cause, an emotional state develops that disrupts the child's daily routine and the family's usual ways of coping with stress. Resumption of normal growth and development for the child requires that the underlying cause of the presenting behavior

be appreciated and addressed.

I. Psychosis, caused by:
  A. Medical illness
  B. Ingestion of toxic substance
  C. Pervasive developmental disorder (i.e., autism)
  D. Adult type of schizophrenia
  E. Manic depressive illness
II. Depression
III. Conduct Disorder
IV. Adjustment Reaction of Childhood or Adolescence
V. Attention Deficit Disorder
VI. Medical Illness in the Absence of Psychosis (i.e., thyrotoxicosis, temporal lobe epilepsy)
VII. Sensory Deficit: blindness, deafness
VIII. Severe Communication Disorder (e.g., childhood aphasia)

Table 20.1. Differential Diagnosis of Agitation and Withdrawal in Childhood

Psychosis

An acutely psychotic child may present to the ED as anxious and agitated or as preoccupied and withdrawn. *Psychosis* refers to a mental state in which major disturbances in thinking, relating, and reality testing occur. Psychotic patients do not express themselves clearly and have difficulty answering direct questions. They also may be extremely suspicious and hostile. Psychosis may be the result of a psychiatric disorder or a physical cause. Psychiatric causes of psychosis include early childhood autism, childhood schizophrenia, adolescent-onset schizophrenia, and manic depressive disorder. Children who have been previously well adjusted or may have had mild to moderate emotional problems rarely develop an acute psychotic reaction after a severe, overwhelming trauma, such as severe abuse, or threatened or witnessed violence. Physical or organic causes of psychosis include an ongoing medical illness or an acute intoxication with an exogenous substance. The emergency physician must be able to determine the cause of psychosis in a child because treatment presupposes proper diagnosis. Certain aspects of the child's mental status help distinguish between psychiatric and organic psychosis. These differentiating features are outlined in Table 20.2.

Evaluation Feature	Organic Psychosis	Psychiatric Psychosis
Onset	Acute	Gradual
Pathologic autonomic signs[b]	May be present	Absent
Vital signs	May be abnormal	Normal
Orientation	Impaired	Intact
Recent memory	Impaired	Intact
Intellectual ability	May be impaired	Intact
Hallucinations	Visual	Auditory

[a]Children with both functional and organic psychoses will have impaired reality testing, inappropriate affect, thought disorder, poor behavior control, and disturbed relating ability.
[b]Increase or decrease in heart rate, respiratory rate, blood pressure and temperature, miosis or mydriasis, skin color changes.

Table 20.2. Differentiating Features of Organic and Psychiatric Psychosis[a]

The child or adolescent who has developed an organic psychosis is likely to be disoriented, particularly with regard to time and place. The child's recent memory also is typically impaired in organic psychoses, and the child may be unable to describe the onset of his or her problems coherently. In addition, hallucinations, when present, usually are visual or tactile rather than auditory (although the latter also may be present). In contrast, the child with a psychiatric psychosis is likely to be oriented to person, place, and time and should be able to report recent events accurately. Other intellectual and cognitive functions usually remain intact. Hallucinations, when present, tend to be auditory in nature, and a greater sense of suspiciousness is common. The rate of onset of psychosis is also revealing. Organic psychosis is more likely to be acute in onset or be the result of acute deterioration in an ongoing chronic condition. A psychiatric psychosis is more likely to be gradual in onset, following a prolonged period of progressive social and emotional withdrawal. Table 20.3 lists medical conditions in childhood that may induce, at some point in their course, acute psychosis with agitation or withdrawal. As can be noted from Table 20.3, many of these illnesses are chronic conditions that may have been present for some time and may have been already under treatment. Therefore, with all psychotic children, it is extremely important to obtain an accurate history of current and previous medical problems. Other medical conditions that may cause psychosis, such as head injury and Reye syndrome, can develop acutely and may progress rapidly to unconsciousness and death unless identified and treated.

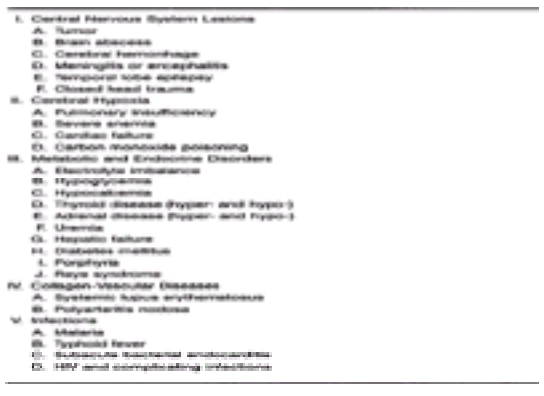

Table 20.3. Medical Conditions That May Lead to Psychosis

Table 20.4 lists drugs that may lead to psychosis characterized by agitation or withdrawal. Some of these drugs are used illicitly, and others are prescription drugs ingested either accidentally or on purpose. In obtaining a history, a frequent important clue in drug intoxication is the acute onset of disordered thinking in the presence of visual hallucinations. A history of drug abuse and the availability of toxic substances are other important historical clues to the diagnosis of acute drug intoxication. Intoxications with alcohol, sedatives, antidepressants, anticholinergic agents, and heavy metals all can be life-threatening if enough of the agent has been ingested and absorbed. Appropriate medical therapy must be instituted as rapidly as possible.

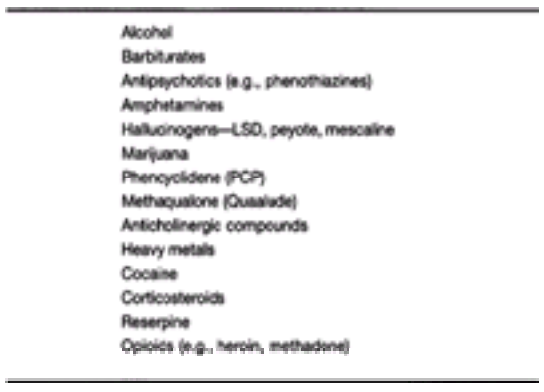

Table 20.4. Exogenous Substances Causing Psychosis after Ingestion of Significant Quantity

Psychiatric conditions that lead to psychosis in children and adolescents often are distressing and may also be life-threatening. This threat to life can occur when the child's hallucinations are so disturbing that suicide is sought for relief. Furthermore, as a part of psychosis, a child may develop a delusional system that leads him or her to attack another person who is seen as threatening. The psychotic child or adolescent may neglect his or her physical well-being through sleep deprivation and malnutrition. These deprivations themselves may intensify the child's vulnerability and exacerbate the psychosis. When dealing with the psychotic child or adolescent, it is imperative that the emergency physician evaluate the suicidal and homicidal potential of the patient as well as the overall physical condition and ability to maintain self-care.

Depression

Other children who may be brought to the ED for agitated or withdrawn behavior are those who are severely depressed. These children are more likely to be oriented, coherent in their thinking, and able to discern reality from fantasy. However, there may be a history of deterioration in social and intellectual performance over the preceding months and evidence of withdrawal from important activities and relationships. The child may be described by his or her parents as being apathetic and may be extremely sad. The child may present as withdrawn and hopeless and may make it difficult for the emergency physician to engage him or her in conversation. Other depressed children may present primarily as anxious and angry, with minor concerns that cause preoccupation and outbursts. Precipitants of depression and a sense of hopelessness may include parental divorce or separation, loss of a parent through death, a recent devaluation of personal abilities through poor academic performance, peer rejection, or the onset of significant physical illness. Once depression is identified as the cause of the child's agitation or withdrawal, it is extremely important for the emergency physician to inquire about the presence and nature of suicidal ideation. The physician also should ask about disturbances in appetite and sleep.

Other Conditions

Other psychiatric conditions—conduct disorders with severe behavioral disruption, adjustment reactions with interruption of normal coping mechanisms, and attention deficit disorder—also may lead a child to present in the ED with significant agitation or withdrawal. With each of these problems, the emergency physician's first task is to screen for suicidal or homicidal ideation and intent. Then, after evaluating the problem, the physician can consider and pursue alternatives for further treatment and referral. The child or adolescent who has a conduct disorder often will have a history of significant behavioral disruption, such as aggressiveness, stealing, fire-setting, truancy, and other forms of delinquency. The child presents as oriented and responsive but also may be angry and resentful about being brought to the ED. The child's

behavior may improve in the ED when it is made clear that misbehavior or temper tantrums will not be tolerated.

Children and adolescents who have been victims of past or ongoing physical or sexual abuse or other severe trauma may develop acute agitation or withdrawal brought on by posttraumatic stress disorder. The symptoms of this disorder include fluctuating behavior with episodes of excitement, fearfulness, or irritability; recurrent nightmares or flashbacks; and lack of involvement in usual friendships or activities. Children who experience posttraumatic reactions often avoid or refuse to talk about the trauma, and thus, parents may be confused about the reasons for the child's disturbed and disturbing behavior. If parents are aware of the traumatic event, they may be upset or feel guilty about its occurrence. In this situation, the parents' reaction may make it difficult for them to support and comfort the child and the emergency room physician may be faced with the need to calm the parents to evaluate the situation further.

A child who has an emotional disturbance from an adjustment reaction generally becomes calm as he or she recognizes the physician's concern and desire to understand the situation. An adjustment reaction is characterized by a deterioration of functioning from a previously higher level in the presence of some precipitating event or situation. The child with an adjustment reaction is oriented and usually can explain his or her problems well. At times, the precipitant may be a developmental event, such as enrollment in a new school, increased peer pressure, or the emergence of secondary sexual characteristics during puberty. The precipitant also may be an acute event such as the loss of a parent through death or divorce.

A child with an attention deficit disorder is likely to have a history of impulsivity with associated distractibility, as well as a history of learning difficulties at school. Acute agitation or withdrawal that requires an ED visit is likely to result from some consequence of the child's difficulties at school or at home.

Certain medical conditions, such as temporal lobe epilepsy and thyrotoxicosis, also can cause agitation and withdrawal without psychosis. The child with thyrotoxicosis will have tachycardia, appetite and sleep disturbances, weight loss, and possibly, exophthalmos in association with the disordered behavior. The child with temporal lobe epilepsy may be identified through the history of seizures and auras and the possible presence of abnormal neurologic findings on physical examination. Differentiation between temporal lobe epilepsy and psychiatric behavioral disturbances may, at times, be difficult and may require neurologic psychiatric consultation. Children with sensory deficits, such as blindness, deafness, or severe communication difficulties, including developmental aphasia, occasionally may present in the ED with concomitant agitation or withdrawal, and it is important for the physician to bear these possibilities in mind.

EVALUATION AND DECISION

The emergency assessment of the agitated or withdrawn child or adolescent involves three complementary areas. In actual practice, the physician gathers data from all three areas at the same time; however, for conceptual clarity, they are discussed separately here. The first area involves determination of whether the problematic behavior is caused by some medical condition or organic state and, if so, what diagnosis to make. To achieve this, the physician gathers information about the acute events that led to the ED visit, any possible drug ingestions or drug use, and the child's medical history. This is followed later by a complete physical examination. Potential life-threatening effects of the medical condition must be recognized and treated. Second, the psychiatric manifestations of the presenting condition, whether organic or psychiatric, are assessed. To do this, the physician performs a thorough mental status examination. Of particular concern is the determination of whether the patient is suicidal or homicidal. To complete the psychiatric evaluation of the child, the previous adjustment and the psychiatric history of the child are determined. These data should facilitate the diagnosis of psychiatric presenting problems. Third, the family system and social support for the child are assessed. This is done by observing the family's responses to the child during the ED visit and by assessing previous family functioning. Once these three areas have been evaluated, the physician can make an appropriate decision regarding disposition and further treatment.

Medical Conditions

First, to determine whether the child's agitation or withdrawal is organically based, the physician should bear in mind the differential diagnosis of these behaviors, which include psychiatric as well as organic origins (Table 20.1). A complete history of the acute events that led up to the ED visit, including any changes in behavior or functioning of the child, should be obtained. This information will help the psychiatric as well as medical assessment. The possibility of drug use or drug ingestions should be explored with the parents as well as with the child. The child's medical history should be documented carefully, and any previous episodes of the current behavior should be reviewed. In general, organically based problems are acute in onset and result from an ingestion, an injury, or the worsening of a medical condition. At times, the child's age may be helpful in suggesting certain diagnoses. For example, with children 12 years of age and older, drug abuse is common, whereas with younger children, acute ingestion of medications or other toxic substances is more common.

The differentiating features of organic psychoses and psychiatric psychoses already have been discussed and are listed in Table 20.2. Important information will come from the history (acuteness of onset of psychosis, presence of drug ingestion), the mental status examination (orientation, memory, intellectual function, nature of hallucinations), and the physical examination (presence of pathologic autonomic signs, abnormal vital signs, physical findings on medical and neurologic examinations). With an organic psychosis, the physician must consider the possible medical causes of psychosis (Table 20.3) as well as those that result from ingestion (Table 20.4).

The medical evaluation of agitation and withdrawal requires that every child who presents to the ED with these behaviors receive a complete physical examination, including full neurologic evaluation. This makes it possible to detect most significant ongoing organic illnesses and neurologic disease of traumatic, infectious, or structural origin. Pupillary abnormalities, when found in conjunction with abnormalities of pulse, temperature, and blood pressure, suggest an acute intoxication. Abnormal reflexes suggest central nervous system disease and may warrant further diagnostic study. Mild incoordination, abnormalities of rapid alternating movements, and impaired tandem gait may be present in children with

an attention deficit disorder. In situations in which an acute intoxication is being considered, blood and urine should be obtained and sent for specific drug determination or toxic screening, as appropriate. Additional laboratory studies should be pursued in accordance with the findings of the physical examination and may include a complete blood count, sedimentation rate, urinalysis, electrolytes, blood glucose, calcium, blood urea nitrogen, ammonia, and liver function tests. Thyroid studies are indicated when ongoing thyroid disease is suspected. With this information in hand, medical conditions, when present, can be diagnosed and appropriate treatment can be initiated. A computed tomographic (CT) scan or magnetic resonance imaging (MRI) examination may be helpful when trauma or a mass lesion is being considered.

Psychiatric Evaluation

The second major area in the ED approach to an agitated or withdrawn child involves assessment of psychiatric manifestations of the presenting condition. This is achieved through the mental status examination, in conjunction with an evaluation of the child's previous level of adjustment and the past psychiatric history. The ED mental status examination is the most important mechanism for determining the behavioral and emotional condition of the child, including the possible presence of suicidal or homicidal intent. The child's mental status can be determined through direct interaction with the child in the presence of the parents. The physician should approach the mental status examination systematically. With the various categories in mind, the physician can obtain much of the data during the history and physical examination. Other areas will require direct questioning of the child by the physician.

The categories of the mental status examination, as described in Chapter 129, also are summarized here. The child's appearance will have already been noted by the physician. Orientation to person, place, time, and situation should be determined. Short- and long-term memory should be tested, as should cognitive functions, which include intelligence, fund of knowledge, and the ability to reason and think (much of this information can be determined from the flow of the interview). The child's behavior should be assessed for activity level and age appropriateness. The child's capacity for relating to the physician can be determined by the physician's noting not only the child's behavior with him or her but also his or her own internal responses to the child's behavior. Particularly important in the emergency assessment of the child are affect and thinking. Affect refers to the predominant feelings displayed by the child. The examiner should observe the nature of the affect (e.g., happy, sad, angry, flat), its degree of appropriateness to the situation, and how it changes as various subjects are discussed. Thinking includes thought processes and thought content. The coherence and goal-directedness of verbal communication are assessed, and loose associations and speech that lacks internal consistency are noted.

Evaluation of thought content involves identifying the child's major themes and concerns. Preoccupations, such as hallucinations, delusions, and ideas of reference (present in psychosis), or sadness, hopelessness, and feelings of depression (present in depression), should also be sought. The child's strengths can be assessed from spontaneous statements as well as from forthrightness in answering specific questions. The child's insight into the current problem should be noted, and his or her capacity to suggest a plan to deal with the present crisis should be evaluated.

Determining the presence or absence of suicidal or homicidal ideation and intent is an essential part of the mental status examination and provides an opportunity to ask about past attempts. The circumstances and intent of any previous suicidal or homicidal attempts should be explored thoroughly. The most effective way of determining such intent is by asking the child directly. Such an approach opens the subject in a way that often is reassuring, thereby enabling the discussion to proceed.

As the mental status examination is carried out, the physician develops a picture of the child that leads to certain diagnostic possibilities. For example, the psychotic child will have bizarre or inappropriate affect, speech that is not goal-directed, and possibly, hallucinations or delusions. Such a child typically relates poorly to the physician, avoiding eye contact, failing to respond to the physician's attempts to empathize, and perhaps, engendering in the physician feelings of confusion and uneasiness. If the psychotic child is oriented with intact memory and cognitive functions, it is likely that the psychosis is psychiatric in origin. If the child's orientation, memory, and cognitive functions are significantly impaired, it is probable that the psychosis is organic in nature. When the child's thinking is coherent and the affect is not bizarre or inappropriate, it is likely that psychosis is not present. With depressed children, themes of sadness occur, and the physician may find himself or herself attracted to the child and feeling sorry for him or her. This same reaction by the physician also may occur with children who present with an adjustment reaction. Children who are upset about a previous trauma may be particularly difficult to evaluate. They will appear frightened, may be erratic in their behavior, and may be uncomfortable with relating and discussing previous traumatic events. These children may be helped to explain their thoughts and fears in a quiet environment, with gentle support from the physician. Younger children who are frightened and who do not easily separate from their parents should raise suspicion about past or current trauma. With a conduct disorder, the history usually is informative, and the child's manner of relating may be distant or manipulative. The physician may feel angry at such children. At times, the child with an attention deficit disorder may show distractibility and impulsivity in the ED, but often, these behaviors do not occur in one-to-one settings and will be revealed only by the history. The child who has some insight into his or her problem is more likely to have depression or an adjustment reaction than psychosis or a conduct disorder.

The child's previous level of adjustment offers important information for a complete profile. The physician can ask about the child's family relations, peer relations, and school performance before the onset of the crisis. The child's major areas of interest and special competences should be appreciated. A child in an age appropriate grade in a regular classroom who has had satisfactory involvement with friends and social activities has a good prognosis in general. The child from a chaotic family who is a loner and who attends school sporadically has a poorer prognosis. In all instances, however, it is important that the physician uncover some areas of strength in the child and family because this will serve as the basis for resolving the current crisis and for pursuing further treatment.

Previous psychiatric hospitalization or outpatient treatment should be determined, as well as any past incidents of suicidal or homicidal behavior. Knowledge of past and current medication also is crucial. Family history of psychiatric

disorder can be obtained at the same time that the child's history is taken.

Evaluation of Support Systems

Finally, emergency evaluation includes assessment of the family and social support system. To master a crisis, a child needs consistency and support within his or her environment. Information about who lives at home with the child, the nature of their relationships with each other, and any recent changes in family composition or in the child's living situation help in understanding the current problem and in determining treatment.

The physician can gain information about the family through observation and direct questioning. In so doing, information about family relationships, including the parents' level of concern and their ability to appreciate the child's current situation, is obtained. Family structure often is revealed by noting which family members accompany the child to the ED. Typically, the family members with the child are more involved. The parents' description of the child during the history taking offers insight into how the child is perceived in the family. The extent to which the parents try to engage a withdrawn child or to calm and set limits with an agitated child should be noted, as well as the child's response to these efforts.

As the child is asked a question by the physician, parents' responses are also informative. Do the parents answer for the child and interrupt when he or she tries to speak, or do they give their child an opportunity to form a relationship with the physician? The parental response suggests the degree to which the child's independent thinking and behavior are encouraged. If the child is not cooperative during the psychiatric or physical examinations, how effective are the parents in telling the child that he or she must cooperate? The parents' success in gaining the child's cooperation during the ED visit may offer a valuable clue about their ability to manage their child effectively at home.

The physician can assess the degree of coping by the family in part by the way in which the family members describe problems. Responses that suggest that the parents are overwhelmed and disorganized should lead the physician to consider psychiatric consultation and possible hospitalization. The openness of the family in discussing recent difficulties also is important. Some families are extremely guarded and deny problems, despite the presence of a major crisis that they are unable to manage. Other families offer a more balanced view of family functioning, instilling greater confidence in the physician. If the child's parents are divorced, assessing the relationship between the parents is important. Arguments and disagreements as well as the possibility of violence lead to a lack of safety and security for the child in crisis. The degree of support that a single parent receives from extended family and neighbors is also an important factor in evaluating family support and capacity.

The family should be asked about the adjustment of the child in the past and how earlier family difficulties were handled. Family history of emotional difficulties also should be obtained. The physician should be clear about who the major caretakers of the child have been through his or her life. In obtaining the family's perception of the crisis, it is best to begin by posing an open-ended question to each family member (e.g., “What do you think is going on with your child and in your family?”). After the family has responded, the physician can ask specific questions about recent significant events or changes that could be influencing the current situation. If the physician has any suspicion that previous or ongoing trauma or abuse underlies the child's acute difficulties, he or she should ask parents and child alike. (For further discussion of the management of child abuse, see Chapter 128). Before discharge, the physician should be confident that the child is safe; otherwise, social work or psychiatric consultation should be obtained. In all situations of posttraumatic stress reactions, outpatient psychiatric treatment generally is necessary.

In determining the disposition of a child with a psychiatric problem in the ED, the physician should be guided by the severity of the problem and by the ability of the family to manage the child on an outpatient basis. The physician should inquire about what social supports are available to the parents. If extended family or close friends are available and the parents believe that their participation would be helpful, the physician should encourage the parents to enlist such help. If other agencies are working with the family, their efforts should be coordinated with those of the hospital or mental health facility at which the child receives treatment. Many families express a clear preference about whether their child should be hospitalized. The physician should keep this preference in mind but should make the decision based on the data about the child's physical and emotional well-being and the assessment of the family support system.

Disposition

As depicted in Figure 20.1, agitation and withdrawal can occur in the presence or absence of psychosis. When psychosis is present, the first task is to establish whether an organic cause is present. Organic psychosis always requires a full medical evaluation, observation, and treatment of the underlying condition, all of which are best accomplished through medical hospitalization. Psychiatric consultation is indicated in all cases of psychiatric psychosis. Psychotic patients who are not suicidal or homicidal may be referred for ongoing outpatient treatment after a positive response to antipsychotic medication (see Chapter 129).

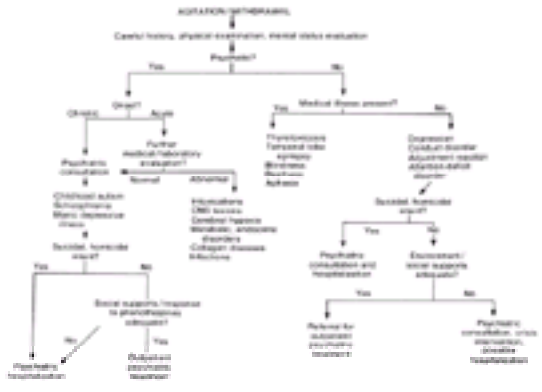

FIGURE 20.1. Approach to the diagnosis and initial disposition of the acutely disturbed child.

Psychotic patients who have suicidal or homicidal intent usually are hospitalized. Psychiatric consultation also is indicated in the presence of active suicidal or homicidal ideation in the absence of psychosis. Persistent suicidal or homicidal intent usually is an indication for psychiatric hospitalization. When suicidal or homicidal thoughts are absent, the ability of the family and social support system to control the child's behavior and prevent further emotional and physical harm should be assessed. If the support system is adequate, referral to outpatient psychiatric treatment may be appropriate. The physician who makes a referral for outpatient psychiatric treatment should help the family develop short-term measures to manage the child and relieve his or her distress until outpatient psychiatric treatment begins. When the support system is not adequate, psychiatric hospitalization may be necessary, especially with such behaviors as fire-setting, repeated running away, and persistent aggressiveness.

Suggested Readings

Benoit MB. Emergency assessment. In: Noshpitz JD, ed. Handbook of Child and Adolescent Psychiatry. Vol 7. Advances and New Directions. Adams PA, Bleiberg E, eds. New York: John Wiley & Sons, 1998:349–356.

Benoit MB. Suicide assessment in child and adolescent psychiatry, pp 369–379. In: Noshpitz JD, ed. Handbook of Child and Adolescent Psychiatry. Vol 7. Advances and New Directions. Adams PA, Bleiberg E, eds. New York: John Wiley & Sons, 1998:349–356.

Kahn AU. Psychiatric Emergencies in Pediatrics. Chicago: Year Book Medical Publishers, 1979.

Sadka S. Psychiatric emergencies in children and adolescents. New Dir Ment Health Serv 1995;67:65.

Sargent J, Silver M, Hodas G, Liebman R. Crisis intervention in children and adolescents. Clin Emerg Med 1984;4:245–262.

Terr LC. Childhood traumas: an outline and overview. Am J Psychiatry 1991;148:10.

Tomb DA. Child psychiatric emergencies. In: Lewis M, ed. Child and Adolescent Psychiatry: A Comprehensive Textbook. 2nd ed. Baltimore: Williams & Wilkins, 1996:929–934.

CHAPTER 21

Dizziness

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“Dizziness” is a common complaint in the pediatric emergency department. True vertigo, the perception that the environment is rotating relative to the patient or that the patient is rotating relative to the environment, arises in the peripheral or central vestibular system. It can be immensely disturbing, even frightening, to patients and their families. Preverbal children, unable to articulate the sensation, may merely be irritable, vomit, and prefer to lie still.

Unfortunately, most patients who use the term *dizziness* are in fact describing one of numerous nonvertiginous disturbances (pseudovertigo), which may be difficult for the practitioner to distinguish from true vertigo. Light-headedness, presyncope, intoxication, ataxia, visual disturbances, unsteadiness, stress, anxiety, and fear from any of numerous causes can initially be described as dizziness.

Therefore, when evaluating a child complaining of dizziness, the practitioner must listen carefully to the details of the history that will allow him or her to distinguish true vertigo from pseudovertigo. The key element in the history that distinguishes true vertigo is the subjective sense of rotation. Often, the best response to a chief complaint of being dizzy is to say, “Tell me what you mean by ‘dizzy.’” Initially vague complaints often become increasingly concrete, and the underlying diagnosis becomes increasingly clear.

PATHOPHYSIOLOGY

True vertigo arises from a disturbance in either the peripheral or central components of the vestibular system. The two peripheral sensory organs of the system (together known as the labyrinth) are the semicircular canals (stimulated by rotary motion of the head) and the vestibule (stimulated by gravity). Both organs lie near the cochlea within the petrous portion of the temporal bone. The proximity of the vestibular and cochlear apparatus explain the frequent association of vertigo with hearing impairment.

Afferent impulses from these organs travel via the vestibular portion of the eighth cranial nerve to the vestibular nuclei in the brainstem and to the cerebellum. Cortical projections terminate in the superior temporal gyrus and the frontal lobe. Efferents from the cerebellum and vestibulospinal tract to the peripheral muscles complete the circuit by which the vestibular system helps maintain balance and position sense. Additional impulses from the vestibular nuclei ascend within the medial longitudinal fasciculus to cranial nerves III, IV, and VI, accounting for the oculovestibular reflexes. Almost all patients complaining of true vertigo should have nystagmus, at least when the vertiginous symptoms are peaking. If not, then a vestibular defect is much less likely.

DIFFERENTIAL DIAGNOSIS

As discussed earlier, dizziness is best divided into vertiginous conditions (true vertigo) and nonvertiginous conditions (pseudovertigo). Table 21.1 lists the differential diagnosis of true vertigo and highlights the life-threatening causes. Table 21.2 lists the most common causes of vertigo. Table 21.3 lists numerous nonvertiginous conditions that may initially be described as dizziness. Because the spectrum of nonvertiginous conditions is so broad, the following discussion will concentrate on true vertigo.

Peripheral Causes	Central Causes
Suppurative or serous labyrinthitis	Tumor*
External ear impaction	Meningitis*
Ramsay Hunt syndrome	Encephalitis*
Cholesteatoma	Increased intracranial pressure*
Perilymphatic fistula	Multiple sclerosis
Vestibular neuronitis	Trauma*
Benign paroxysmal vertigo	Seizure (usually complex partial)
Ingestions*	Migraine
Temporal bone fracture*	Stroke*
Posttraumatic vestibular concussion	Motion sickness
Ménière's disease	Paroxysmal torticollis of infancy

*Life-threatening causes of vertigo.

Table 21.1. Causes of Vertigo in Children

Suppurative or serous labyrinthitis	Ingestions
Benign paroxysmal vertigo	Seizure
Migraine	Motion sickness
Vestibular neuronitis	

Table 21.2. Common Causes of Vertigo

Depression	Cardiac disease
Anxiety	Anemia
Hyperventilation	Hypoglycemia
Orthostatic hypotension	Pregnancy
Hypertension	Ataxia
Heat stroke	Visual disturbances
Arrhythmia	

Table 21.3. Common Causes of Pseudovertigo

Vertigo follows a dysfunction of the vestibular system within the semicircular canals, vestibule, or vestibular nerve (peripheral vertigo), or within the brainstem, cerebellum, or cortex (central vertigo). It can also be divided into conditions in which hearing is impaired (usually peripheral causes) and conditions in which hearing is spared (usually central causes). Finally, vertigo can be divided into acute (usually infectious, postinfectious, traumatic, or toxic) and chronic-recurrent groups (usually caused by seizures, migraine, or benign paroxysmal vertigo of childhood).

Infections

Both acute and chronic bacterial and viral infections of the middle ear with or without associated mastoiditis may cause both vestibular and auditory impairment (see Chapter 32). Severe, untreated, acute suppurative otitis media with effusion may extend directly into the labyrinth. Even without direct invasion of the pathogens, inflammatory toxins can cause a serous labyrinthitis.

Chronic and recurrent otitis media can produce a cholesteatoma of the tympanic membrane, an abnormal collection of keratin caused by repeated cycles of perforation and healing. Cholesteatomas can erode the temporal bone and the labyrinth, producing a draining fistula from the labyrinth that presents as vertigo, nausea, and hearing impairment. Computed axial tomography (CAT) scans show destruction of the temporal bone.

Viral infections can directly affect the labyrinth or the vestibular nerve; together these conditions are known as vestibular neuronitis. Known pathogens include mumps, measles, and the Epstein-Barr virus. More commonly, a nonspecific upper respiratory tract infection may precede the illness. Onset is usually acute and can be severe. Nystagmus is usually present. Patients prefer to lie motionless with their eyes closed. Recovery is from 1 to 3 weeks. Early use of prednisone may shorten the course.

Migraine

Vertigo may be a prominent feature of classic migraine or migraine equivalent, in which there is no associated headache

(see Chapter 83). Up to 19% of children with migraine may have vertiginous symptoms during their aura. Basilar migraine presents as a throbbing occipital headache following signs and symptoms of brainstem dysfunction (including vertigo, ataxia, tinnitus, and dysarthria). Vertigo from migraine equivalent (without pain) is typically seen in patients with a family history of migraine headache and is associated with other transient neurologic complaints (e.g., weakness, dysarthria). Symptoms may suggest temporal lobe epilepsy. The latter is distinguished by altered consciousness.

The differential diagnosis of headache and vertigo includes a brainstem or cerebellar mass, hemorrhage, and infarction. These uncommon disorders are best assessed by magnetic resonance imaging (MRI).

Benign Paroxysmal Vertigo

Considered by many to be a form of migraine, benign paroxysmal vertigo is most common in children between the ages of 1 and 5 years. Patients have recurrent attacks, usually one to four per month, and occasionally in clusters. Onset is sudden—the child often cries out at the start of each episode—and is associated with emesis, pallor, sweating, and nystagmus. Episodes are brief, lasting up to a few minutes, and may be mistaken for seizures. Consciousness and hearing are preserved, and the neurologic examination is otherwise normal. The disorder spontaneously remits after 2 to 3 years.

Ototoxic Drugs

Most agents that disturb vestibular function will also disturb auditory function. Specific agents include aminoglycoside antibiotics, furosemide, ethacrynic acid, streptomycin, minocycline, salicylates, and ethanol. Toxic doses of certain anticonvulsants and neuroleptics can produce measurable disturbances of vestibular function, although associated complaints of vertigo are rare.

Posttraumatic Vertigo

Several mechanisms account for posttraumatic vertigo. The most obvious is fracture through the temporal bone with damage to the labyrinth. Presentation includes vertigo, hearing loss, and hemotympanum. Computed tomography (CT) scanning of the temporal bone should be obtained when there is hemotympanum or posttraumatic evidence of vestibular dysfunction.

More subtle causes of posttraumatic vertigo include trauma-induced seizures, migraine, or a postconcussion syndrome. The latter disorder (vestibular concussion) typically follows blows to parieto-occipital or temporoparietal regions and presents with headache, nausea, vertigo, and nystagmus. Hyperextension and flexion (“whiplash”) injuries can be associated with vestibular dysfunction, probably caused by basilar artery spasm with subsequent impairment of their labyrinth and cochlear connections. Symptoms may mimic basilar artery migraine.

Seizures

Two types of seizures are associated with vertigo: vestibular seizures (seizures causing vertigo) and vestibulogenic seizures (“reflex” seizures brought on by stimulating the semicircular canals or vestibules by sudden rotation or caloric testing). Vestibular seizures, the more common type, consist of sudden onset of vertigo with or without nausea, emesis, and headache and are invariably followed by loss or alteration of consciousness. The electroencephalogram (EEG) is abnormal. Anticonvulsants may be of benefit.

Motion Sickness

Motion sickness is precipitated by a mismatch in information provided to the brain by the visual and vestibular systems during unfamiliar rotations and accelerations. The most common situation occurs when a child travels in a car or airplane and is deprived of visual stimulus that confirms movement. Symptoms include vertigo, nausea, and nystagmus. Attacks can be prevented by allowing patients to watch the environment move in a direction opposite to the direction of body movement. In car travel, encouraging children to “look out the window” is helpful.

Ménière's Disease

Uncommon in children younger than 10 years, Ménière's disease is characterized by episodic attacks of vertigo, hearing loss, tinnitus, nystagmus, and autonomic symptoms of pallor, nausea, and emesis. Between episodes, patients may complain of impaired balance. The underlying cause is believed to be an overaccumulation of endolymph within the labyrinth, which causes a rupture (endolymphatic hydrops). Typical attacks last from 1 to 3 hours and usually begin with tinnitus, a sense of fullness within the ear, and increasing hearing impairment. Attacks are intermittent and unpredictable, often lasting for years and, at times, evolving to permanent hearing loss.

Miscellaneous Causes

Vertigo may occur at any point in the clinical course of multiple sclerosis when the central demyelination interferes with the vestibular nuclei in the brainstem or its efferents or afferents. Diagnosis is confirmed by MRI and lumbar puncture. Paroxysmal torticollis of infancy consists of spells of head tilt associated with nausea, emesis, pallor, agitation, and ataxia. Episodes are brief and self-limited and may recur for months or years. The cause is unclear, although some authors see it as a prelude to later benign paroxysmal vertigo (considered by some to be a migraine variant). Perilymphatic fistula is an abnormal communication between the labyrinth and the middle ear, with leakage of perilymphatic fluid through the defect. It may be congenital or acquired by trauma, infection, or surgery. Diagnosis is by middle ear exploration. Finally, vertigo may be associated with diabetes mellitus and chronic renal failure.

EVALUATION AND DECISION

Differentiation of True Vertigo and Pseudovertigo

Evaluation of children complaining of dizziness begins by separating those with true vertigo from those with pseudovertigo (Table 21.1 and Table 21.3). True vertigo is always associated with a subjective sense of rotation of the environment relative to the patient or of the patient relative to the environment. Acute attacks are usually accompanied by nystagmus. Pseudovertigo is suggested by complaints of light-headedness, flushing, weakness, ataxia, unsteadiness, weakness, fatigue, pallor, anxiety, stress, and fear.

True Vertigo (Fig. 21.1)

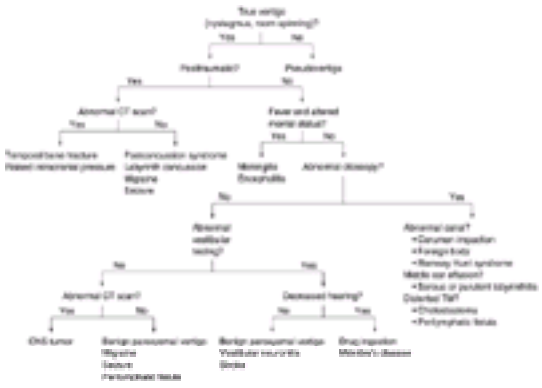

FIGURE 21.1. Approach to the child with true vertigo.

History and Physical Examination

Once true vertigo is identified, its severity, time course, and pattern must be established. In general, the most severe attacks of vertigo are due to peripheral causes, whereas central causes tend to be more recurrent, chronic, and progressive. Sudden onset of sustained vertigo suggests central or peripheral trauma, infection, central stroke, or ingestion. Chronic recurrent episodes suggest seizures, migraine, or benign paroxysmal vertigo. More persistent episodes suggest brainstem or cerebellar mass lesions. Recurrent, transient, altered mental status suggests seizure or basilar migraine. Episodes of prior head injury suggest concussion syndromes. Recent upper respiratory tract infections may suggest vestibular neuronitis. History of ototoxic drugs or intoxicants is important, as is a family history of migraine. Age of the patient is especially helpful—benign paroxysmal vertigo is unusual after age 5 years, whereas Ménière's disease is unusual before age 10 years.

The physical examination focuses on the middle ear and on neurologic and vestibular testing. Perforation or distortion of the tympanic membrane should be noted. A pneumatic bulb will enable the examiner to see whether abrupt changes in the middle ear pressure trigger an episode of vertigo, a suggestion that a perilymphatic fistula may be present (Hennebert's sign).

The neurologic examination must be complete, focusing closely on the auditory, vestibular, and cerebellar systems. Both vestibular and cerebellar disorders will present with an unsteady gait. In both situations, when there is a unilateral lesion, the child will fall toward the side of the lesion. The two can be distinguished by the presence or absence of nystagmus and, in the verbal child, by the presence or absence of a sense of rotation. In addition, if cerebellar dysfunction is present, there will be a breakdown of rapid alternating movements and finger-to-nose movements.

Nystagmus is a highly specific sign for both central and peripheral vertiginous disorders. A patient complaining of dizziness from vertigo may not have nystagmus at the time that he or she is examined. Tests to elicit positional vertigo and nystagmus can therefore be helpful in identifying and even distinguishing central and peripheral vestibular dysfunction, particularly if the tests elicit or increase the patient's complaint.

Initially, nystagmus should be sought in all positions of gaze and with changes in head position. Peripheral vestibular disorders are characterized by a “jerk” nystagmus with the slow component toward the affected side. Central lesions are characterized by nystagmus with the fast component toward the affected side and reversal of the fast component when changing from right to left lateral gaze. The Nylen-Barany test is performed by moving a child rapidly from a sitting to a supine position with the head 45 degrees below the edge of the examining table and turned 45 degrees to one side. Nystagmus and a vertiginous sensation may result as the vestibular system is stressed. Certain features of the nystagmus elicited may be helpful in distinguishing central from peripheral vestibular dysfunction. In central dysfunction, for example, onset of nystagmus is immediate; in peripheral vestibular disorders, it is delayed.

The cold caloric response tests for integrity of the peripheral vestibular system. Slow and careful irrigation of either 100 mL of tap water 7°C below body temperature or 10 mL of ice water into the external ear canal through a soft plastic tube, with the child lying about 60 degrees recumbent, should induce a slow movement of the eyes toward the stimulus and a fast movement away. Vestibular damage will suppress the response on the affected side. The test is contraindicated if the tympanic membrane is perforated.

Laboratory Data

Laboratory investigations have a limited role in the evaluation of vertigo. Useful initial tests include a complete blood count, a serum glucose, and an electrocardiogram. Together, these may help identify patients with pseudovertiginous conditions caused by anemia, hypoglycemia, and rhythm abnormalities. Further laboratory testing may reveal diabetes or renal failure, both of which have been associated with vertigo. Toxicologic testing including specific anticonvulsant levels and an ethanol level, if indicated, may be helpful. A lumbar puncture is indicated in cases of suspected meningitis or encephalitis.

Radiologic imaging of the central nervous system, preferably by MRI for adequate visualization of the posterior fossa and brainstem, is indicated in cases of chronic and recurrent vertigo to exclude mass lesions. Children with vertigo and an underlying bleeding diathesis or a predisposition toward ischemic stroke (i.e., sickle cell disease) may also need an emergent cranial CT or MRI. Posttraumatic vertigo, especially when accompanied by hearing loss or facial nerve paralysis, is best assessed by computed axial tomography including adequate images of the temporal bone.

Some children with true vertigo will require referral for more extensive testing. An EEG is indicated when vertigo accompanies loss of consciousness or other manifestations of a seizure. Audiometry is indicated when vertigo accompanies otalgia, hearing loss, or tinnitus. Specialized testing for nystagmus, including electronystagmography (ENG), which measures eye movements at rest and at extremes of gaze, can separate central from peripheral vestibular disorders.

Management

Specific disorders causing vertigo are treated directly. Suppurative or serous labyrinthitis, for example, is treated with antibiotics. An erosive cholesteatoma may require surgical removal. Anticonvulsants may diminish vestibular and vestibulogenic seizures. Motion sickness may respond to simple behavioral changes (e.g., encouraging children to look out the window). Other causes of vertigo spontaneously remit without therapy and merit only close monitoring. Vestibular neuronitis and benign paroxysmal vertigo are examples.

Subspecialist consultation is indicated in certain situations. Neurosurgical evaluation after trauma may be indicated in cases of suspected basilar skull fracture. Suspected perilymphatic fistula, cholesteatoma, or complicated otitis media may merit otorhinolaryngologic evaluation. Neurologists may be helpful in cases of suspected seizure or migraine.

Children with severe or recurrent attacks of vertigo may require treatment with specific antivertiginous medications. The antihistamines dimenhydrinate (5 mg/kg per day orally divided every 6 hours) and meclizine (25 mg orally every 12 hours in children over 12 years of age) may be helpful. Concomitant use of a benzodiazepine such as diazepam (0.1 to 0.3 mg/kg per day orally divided every 6 to 8 hours) as a sedative may be necessary in severe cases.

Pseudovertigo (Fig. 21.2)

FIGURE 21.2. Approach to the child with pseudovertigo.

Pseudovertigo refers to a broad array of diagnoses that present with symptoms such as light-headedness, presyncope, intoxication, ataxia, visual disturbances, unsteadiness, stress, anxiety, and fear. Uniformly absent are a sense of rotation and ocular nystagmus. Underlying causes are numerous; several of the most common causes are listed in Table 21.3. Careful consideration of the patient's age, sex, detailed history, and physical examination, together with a limited number of ancillary tests, may help establish the specific diagnosis.

Suggested Readings

Bower CM, Cotton RT. The spectrum of vertigo in children. Arch Otolaryngol Head Neck Surg 1995;121:911–915.

Busis SN. Dizziness in children. Pediatr Ann 1988;17:648–655.

Busis SN. Vertigo. In: Bluestone CD, ed. Pediatric Otorhinolaryngology. 3rd ed. Philadelphia: WB Saunders, 1996:285–301.

Casselbrant ML, Riera-March A, Furman JM. Vestibular evaluation in children. In: Bluestone CD, ed. Pediatric Otorhinolaryngology. 3rd ed. Philadelphia: WB Saunders, 1996:227–234.

Dunn DW. Dizziness: when is it vertigo? Contemp Pediatr 1987;4: 67–88.

Eviatar L. Vertigo. In: Swaiman KF, ed. Pediatric Neurology: Principles and Practice. 2nd ed. St. Louis: Mosby-Year Book, 1994: 297–307.

Feinchel GM. Clinical Pediatric Neurology: A Signs and Symptoms Approach. 3rd ed. Philadelphia: WB Saunders, 1997:359–364.

Tusa RJ, Saada AA, Niparko JK. Dizziness in childhood. J Child Neurol 1994;9:261–274.

CHAPTER 22

Edema

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Edema, the abnormal swelling of tissues from accumulation of fluid in the extravascular space, is a common emergency problem. This fluid characteristically appears either in the dependent portions of the extremities or lower back; in distensible tissues such as the eyelids, scrotum, or labia; or in organs or extremities at the site of tissue damage.

PATHOPHYSIOLOGY

The major mechanisms that lead to the formation of edema are decreased intravascular oncotic pressure (clinically indicated by a decreased serum albumin), increased venous or lymphatic pressure, and vasculitis from an allergic or hypersensitivity reaction. Hypoalbuminemia arises from decreased production of proteins caused by hepatic disease or increased renal or gastrointestinal (GI) losses. When the albumin level is less than 2.5 g/dL, the oncotic activity in the vascular space is reduced enough for fluid to move into the soft tissues and, eventually, the dependent extremities.

In the face of normal vascular permeability, edema formation without hypoalbuminemia requires an increased hydrostatic pressure that overcomes the oncotic pressure of intravascular protein and sodium, forcing fluid out of the vascular space. Hypervolemia from cardiac failure, salt retention, or estrogen–progesterone excess is the general mechanism responsible for the increased hydrostatic pressure that leads to edema. Renal tubular insensitivity to the natriuretic and diuretic actions of atrial natriuretic peptide is also a cofactor in the pathogenesis of edema.

In the hypersensitive or allergic state, the formation of edema may be rapid and localized. This may be life-threatening if the edema is formed near the airway or may be merely uncomfortable if restricted to the eyelids or ankles, as seen with insect bites. If the basis of edema is decreased oncotic pressure or elevated hydrostatic pressure, the onset of symptoms is sometimes gradual and may exist for weeks or months before edema is appreciated by the patient or physician. Usually, a 10 to 15% weight gain occurs before the patient comes to medical attention.

DIFFERENTIAL DIAGNOSIS

Numerous diseases may cause localized or generalized edema on the basis of the three major pathophysiologic mechanisms already discussed (Table 22.1). The most common origin for localized edema in children is an allergic reaction. Idiopathic nephrosis, although unusual (occurring in just 2 of every 100,000 children annually), is the most likely source for generalized edema (Table 22.2). Although most children who develop edema will have self-limiting disorders, potentially life-threatening conditions (Table 22.3) may be seen, including allergic reactions that involve the upper airway, bacteremic cases of cellulitis (see Chapter 84), thrombophlebitis, and dysfunction of the kidney, liver, or heart.

Table 22.1. Causes of Edema

Table 22.2. Common Causes of Edema

Table 22.3. Life-Threatening Causes of Edema

EVALUATION AND DECISION

When evaluating the child with edema, initially the physician must determine whether the patient has a localized or generalized process. Even if the complaint appears to be confined to one anatomic area, a thorough examination is necessary, checking in particular for edema around the eyes, the scrotum, and the feet. If there is any suspicion of generalized swelling, a urinalysis is indicated to rule out proteinuria (renal disease often leads to the formation of edema without other clinical findings). Acquired hypoproteinemia from hepatic or intestinal disease is unusual, and congestive heart failure (CHF) produces myriad findings.

LOCALIZED EDEMA

Children seek care in the emergency department (ED) more often with localized than generalized edema (Fig. 22.1). Rarely, an infant will have unexplained, localized swelling of an extremity since birth. In this situation, congenital lymphedema (Milroy's disease) should be considered; Turner's syndrome may be associated with bilateral leg edema and Noonan's syndrome with pedal edema. More commonly, the swelling arises over a few days. Most of these lesions result from minor trauma, infection of the subcutaneous tissues, or allergic (urticarial) reactions. Rapid onset of painful swelling typically follows pit viper envenomation (see Chapter 91). Tenderness to palpation points to trauma or infection, and fever with warmth over the lesion often occurs with the latter (Table 22.4). On the eyelids and over the ankles, insect bites are likely to produce a noticeable swelling, which can be difficult to distinguish from cellulitis. A therapeutic response to an antihistamine (diphenhydramine) or to a subcutaneous dose of epinephrine can aid in the diagnosis of an allergic reaction. When local edema occurs on the face, the physician should evaluate the child carefully for concurrent laryngeal involvement. When facial edema is severe or recurrent or when there is a family history of a similar problem, hereditary angioedema (see Chapter 92) is a consideration. Other causes of facial edema include acute sinusitis, dental abscess, orbital or buccal cellulitis, and cavernous sinus thrombosis. A history of environmental exposure can be helpful in the diagnosis of sunburn, frostbite, and plant-induced dermatitis. Sickle cell anemia may cause painful limb edema, as in the case of toddlers with dactylitis. Thrombophlebitis rarely occurs in the prepubertal child but may affect the adolescent; weightlifting and the use of oral contraceptive pills predispose teenagers to this condition. Finally, pregnant young women may develop edema in the lower extremities.

FIGURE 22.1. Edema in children.

Table 22.4. Differentiation among the Common Causes of Localized Edema

GENERALIZED EDEMA

Generalized edema, usually an indication of significant underlying disease, occasionally occurs with less serious conditions. Certain drugs (oral contraceptive pills, lithium, nonsteroidal anti-inflammatory agents) may cause some people to become edematous; cessation of the drug produces a resolution of the swelling. Just before menstruation, young women may complain of “bloating.” The cyclical nature of this problem usually provides a clue to the diagnosis.

The evaluation of the edematous child requires careful attention to the cardiovascular examination. The clinical manifestations of CHF or pericarditis are rarely subtle when these conditions are hemodynamically severe enough to produce edema. Tachycardia, tachypnea, adventitial pulmonary sounds, and hepatomegaly suggest CHF (see Chapter 82). In the child with a pericardial effusion, a pulsus paradoxus greater than 10 to 20 mm Hg, muffled heart tones, and jugular venous distension may occur. A chest radiograph often shows an enlarged cardiac silhouette with both conditions. An electrocardiogram with ST-segment elevation and generalized T-wave inversion on the tracing may suggest pericarditis with a pericardial effusion. Echocardiography is diagnostic.

Generalized edema that is not cardiovascular in origin most often arises secondary to diseases of the kidneys, particularly idiopathic nephrotic syndrome (see Chapter 86). Occasionally, other forms of the nephrotic syndrome, glomerulonephritis, hemolytic uremic syndrome, or Henoch-Schönlein purpura (HSP) are responsible. The detection of significant proteinuria (3+ or 4+) confirms the diagnosis of nephrotic syndrome or nephritis, and affected children require admission to the hospital for evaluation and treatment. In the child with HSP, the edema usually affects the lower extremities predominantly and often is accompanied by a purpuric eruption (despite normal platelet count and

coagulation studies).

In the absence of proteinuria or signs of CHF, occult diseases of the GI tract or liver remain considerations. An initial laboratory evaluation, including liver function tests, electrolyte levels, and measurement of total protein and albumin, should be performed. Abnormalities in these studies often suggest involvement of a specific organ system or severe vasculitis with protein leak.

By process of exclusion, the child with normal screening laboratory tests and no abnormal physical findings except generalized edema is likely to have some type of vasculitis. The finding of a normal albumin does not rule out vascular disease. In teenage girls, otherwise unexplained edema may occur in the premenstrual period and carries a benign prognosis.

Suggested Readings

Aebi C, Ahmed A, Ramilo O. Bacterial complications of primary varicella in children. Clin Infect Dis 1996;23:695–705.

Baliga R, Lewy JE. Pathogenesis and treatment of edema. Pediatr Clin North Am 1987;34:639–648, 1987.

Charlesworth EN. Urticaria and angioedema: a clinical spectrum. Ann Allergy Asthma Immunol 1996;76:484–495.

Fisher DA. Obscure and unusual edema. Pediatrics 1966;37:506.

Green ML. Edema. In: Pediatric Diagnosis: Interpretation of Symptoms and Signs in Infants, Children, and Adolescents. 5th ed. Philadelphia: WB Saunders, 1992:399–403.

Holliday MA, Barratt TM, Avner ED. Edema. In: Pediatric Nephrology. 3rd ed. Baltimore: Williams & Wilkins, 1994:200–204.

Salcedo JR, Thabet MA, Latta K, et al. Nephrosis in childhood. Nephron 1995;71:373–385.

Schrier RW. Pathogenesis of sodium and water retention in high-output and low-output cardiac failure, nephrotic syndrome, cirrhosis, and pregnancy. Parts I and II. N Engl J Med 1988;319:1065–1071, 1127–1133.

CHAPTER 23

Epistaxis

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Epistaxis (nosebleeding) is a common symptom in young children and may be alarming to parents who often overestimate the amount of blood loss. It usually is noted first at about age 3 years and increases in frequency with age, until peaking before or in adolescence. An orderly approach to the history and physical examination is necessary to identify the small minority of patients who require emergent hemorrhage control, laboratory investigation, or referral to an otorhinolaryngologist (ORL) for special treatment.

PATHOPHYSIOLOGY

Minor trauma, nasal inflammation, desiccation, and congestion, as well as the rich vascular supply of the nose, contribute to the frequency of nosebleeds in otherwise normal children. The nose is a favored site for recurrent minor trauma, especially habitual, often absentminded, picking. The small vessels that supply the nasal mucous membrane have little structural support because the mucosa is closely applied to the perichondrium and periosteum of the nasal septum and lateral nasal walls. Furthermore, the nasal mucosa is richly supplied with vessels that form plexiform networks. One such anastomosis of common etiologic significance is Kiesselbach's plexus in Little's area of the anterior nasal septum, about 0.5 cm from the tip of the nose (see Fig. 121.4). Any factors that tend to cause congestion of the nasal vessels or drying of the mucosa will enhance the likelihood of epistaxis, resulting from a given degree of trauma.

DIFFERENTIAL DIAGNOSIS

Many types of local and systemic disorders may cause epistaxis (Table 23.1). Local factors predominate in etiologic importance (Table 23.2). In addition to minor accidental trauma and habitual picking, any cause of acute inflammation will predispose the nose to bleeding. Acute upper respiratory infections, whether localized as in colds or secondary to more generalized infections such as measles, infectious mononucleosis, and influenzal illnesses, contribute to the onset of epistaxis. Allergic rhinitis may also be a factor. Staphylococcal furuncles, foreign bodies, telangiectasias (Osler-Weber-Rendu disease), hemangiomas, or evidence of other uncommon tumors may be found on inspection. Juvenile nasopharyngeal angiofibroma usually is seen in adolescent boys with nasal obstruction, mucopurulent discharge, and severe epistaxis. These tumors may bulge into the nasal cavity but often require examination of the nasopharynx to be identified. Although benign, they can cause severe problems through local invasion of adjacent structures. A rare childhood malignant tumor, nasopharyngeal lymphoepithelioma, may cause a syndrome of epistaxis, torticollis, trismus, and unilateral cervical lymphadenopathy. *Rhinitis sicca* refers to a condition that is common in northern latitudes during the winter, in which low ambient humidity, exacerbated by dry hot-air heating systems, leads to desiccation of the nasal mucosa with concurrent tendency to frequent bleeding. Other rare local causes of epistaxis include nasal diphtheria and Wegener's granulomatosis.

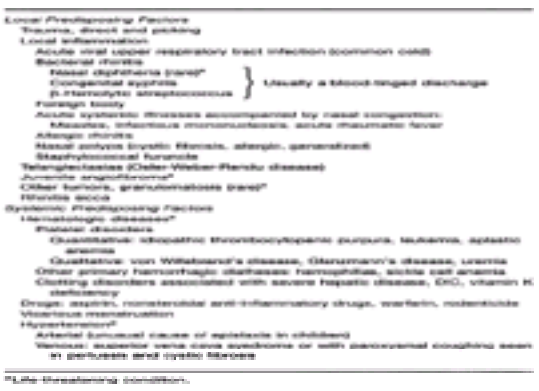

Table 23.1. Differential Diagnosis of Epistaxis

Trauma	Rhinitis sicca
Foreign body	Viral rhinitis
Allergic rhinitis	

Table 23.2. Common Causes of Epistaxis

Children rarely present with a nosebleed as their only manifestation of a more systemic disease. In children with severe or recurrent nosebleeds, a positive family history, or constitutional signs and symptoms, the physician should consider a systemic process. Von Willebrand's disease and platelet dysfunction are two of the more common systemic diseases that cause recurrent or severe nosebleeds. Other less common systemic factors include hematologic diseases such as sickle cell anemia, leukemia, hemophilia, and clotting disorders associated with severe hepatic dysfunction or uremia. Arterial hypertension rarely is a cause of epistaxis in children. Increased nasal venous pressure secondary to paroxysmal coughing, such as that which occurs in pertussis or cystic fibrosis, occasionally may cause nosebleeds. *Vicarious menstruation* refers to a condition occasionally found in adolescent girls in whom monthly epistaxis related to vascular congestion of the nasal mucosa occurs concordant with menses and is presumably related to cyclic changes in hormone levels.

EVALUATION AND DECISION

Rarely are nosebleeds in children life-threatening or do they require more than simple measures to gain control of hemorrhage. However, one's evaluation should begin with hemorrhage control and identification of children who are unstable by noting alterations in the patient's general appearance, vital signs, airway, color, and mental status.

Most childhood nosebleeds are anterior in origin. However, because posterior bleeds may require more extensive therapy, it is important to identify the site of bleeding. In general, posterior sites bleed more profusely, although parents may underestimate the volume because much of the blood is often swallowed. Blood seen in the oropharynx, blood in both nares, difficulty controlling bleeding despite adequate anterior pressure, and a normal anterior exam are more characteristic of a posterior nasal bleed but can be found with an anterior causative site.

After treating any emergent problems, the evaluation of the child with epistaxis begins with a thorough history. Specific features to be sought include frequency of occurrence, difficulty in control (and adequacy of simple at-home first aid), history of trauma, nose picking, frequent upper respiratory infection, allergic and chronic discharge, and obstructive symptoms. Often, asking children which finger they pick their noses with will elicit a more honest answer. Often, parents will note hematemesis or melena, prompting them to seek urgent medical attention. Specific questions regarding evidence for any systemic hemorrhagic disorder or family history of bleeding are asked. In adolescent girls, relation to menses is noted.

Physical examination must include a complete general examination with special attention paid to vital signs, including blood pressure, evidence of hematologic disease (enlarged nodes, organomegaly, petechiae, or pallor), and of course, inspection of the nasal cavity after reasonable efforts to stop the bleeding. When examining a child with a nosebleed, one will need a good light source, suction, and adequate body fluid precautions. Nasal inspection begins with clearing the passages by having the child blow his or her nose or by using gentle suction. On examination, one is looking for the site of bleeding, mucosal color, excoriations, discharge, a foreign body or other mass, and septal hematomas. Using one's thumb, the tip of the nose is pushed upward to allow examination of the vestibule, the anterior portion of the septum, and anterior portion of the inferior turbinate. If the mucosa is too boggy for adequate visualization, a topical vasoconstrictor or decongestant may be beneficial. A more thorough examination requires the use of a nasal speculum. Using one's nondominant hand, the speculum is passed vertically into the nares and opened, allowing examination of the septum, turbinates, and middle meatus. A topical anesthetic and restraints may be necessary for such an examination in young children.

Because most cases of bleeding in children are from the anterior nasal septum, the simplest way to stop the hemorrhage is to apply direct pressure on the bleeding site for 5 to 10 minutes by external compression of the nares between two fingers. In addition, a cotton (dental) roll may be placed under the upper lip to compress the labial artery. Occasionally, the addition of cotton pledgets moistened with a few drops of epinephrine (1:1000) or application of topical thrombin will help achieve hemostasis. The child should be sitting up, with his or her head tilted slightly forward during these procedures. If an anterior site of bleeding is identified and there is no evidence of a hemorrhagic diathesis—and particularly if bleeding has been recurrent—cautery with a silver nitrate stick may be warranted (see Section VII). For management of severe epistaxis not responsive to such measures, nasal packing or surgical ligation of vessels may be necessary (see Procedures 7.2). The recent advent of expandable nasal tampons has simplified the procedure of anterior nasal packing for the emergency physician, especially in patients who relapse after a successful cautery. However, nasal tampons pose the risk of toxic shock syndrome, necessitating careful patient instructions and follow-up. For children with nosebleeds from a hemorrhagic diathesis, one must also correct the underlying disorder. No laboratory workup is indicated in children without clinical evidence of severe blood loss in whom systemic factors are not suspected, and for whom an anterior site of bleeding is identified and stopped readily with local pressure. Reassurance and

education about appropriate at-home management needs to be provided.

Occasionally, recurrent epistaxis during an acute upper respiratory infection or flare-up of allergic rhinitis may be lessened with use of an antihistamine–decongestant preparation, although care must be taken not to dry the nose excessively, which can cause epistaxis related to dry mucosa. During the winter, especially in the context of forced hot-air heating systems, a cool mist vaporizer may lessen crusting and drying of nasal mucosa with its subsequent predisposition to recurrent bleeding. Petroleum jelly, placed in the nostrils twice daily, and saline nasal spray also are useful for maintaining normal moistness of the nasal mucosa. Keeping fingernails short may also be helpful.

All patients discharged from the emergency department (ED) after evaluation for significant epistaxis should be given specific instructions on nares compression and indications for repeat evaluation. For patients with specific local abnormalities, such as tumors, polyps, or telangiectasias, referral to an ORL is necessary. Such referral might also be considered, even with questionable findings on the ED nasal examination, if bleeding was severe, recurrent, or suspected to be posterior in origin.

Finally, evaluation for hemorrhagic diathesis should be done in any child with pertinent positive findings on history, family history, or physical examination. This usually would include prothrombin time, partial thromboplastin time, complete blood count, and bleeding time. Although the yield would be low in the absence of corroborative clinical features, some children with isolated epistaxis that seems particularly severe or frequently recurrent might also deserve such screening. Katsanis et al. found that 8 of 36 such children with isolated recurrent epistaxis had mild bleeding abnormalities; however, the diagnosis of these patients may be difficult, requiring more sophisticated laboratory evaluation (e.g., postaspirin bleeding time, factor VIII–related antigen, ristocetin aggregation study). These considerations are outlined in the epistaxis algorithm (Fig. 23.1).

FIGURE 23.1. Approach to diagnosis of epistaxis. *ORL,* otorhinolaryngologist; *CBC,* complete blood count; *PT,* prothrombin time; *PTT,* partial thromboplastin time.

Suggested Readings

Alvi A, Joyner-Triplett N. Acute epistaxis: how to spot the source and stop the flow. Postgrad Med 1996;99:83–96.

Katsanis E, Luke KH, Hsu E, et al. Prevalence and significance of mild bleeding disorders in children with recurrent epistaxis. J Pediatr 1998;113:73–76.

Mulbury PE. Recurrent epistaxis. Pediatr Rev 1991;12:213–216.

Pimpinella PJ. The nasopharyngeal angiofibroma in the adolescent male. J Pediatr 1964;64:260.

Potsic WP, Handler SD, Wetmore RF, Pasquariello PS. Nose and paranasal sinuses. In: Primary Care Pediatric Otolaryngology. 2nd ed. Andover, NJ: J. Michael Ryan, 1995:64–70.

Ritter FN. Vicarious menstruation. In: Strome M, ed. Differential Diagnosis in Pediatric Otolaryngology. Boston: Little, Brown, 1975:216.

Schulman I. Clinical conference: the significance of epistaxis in childhood. Pediatrics 1959;24:489.

CHAPTER 24

Eye—Red

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When approaching the problem of “red eye,” it is important to determine which tissues are involved. This chapter is confined to disorders in which the conjunctiva, episclera, or sclera are inflamed, causing the “white of the eye” to appear red or pink. Red eyes may be caused by local factors, intraocular disease, or systemic problems. Table 24.1, Table 24.2, and Table 24.3 list common and life-threatening causes of red eye. Discussion of chemical conjunctivitis or irritation caused by agents such as smoke or trauma is limited because the history is almost always known in these situations, making the diagnosis clear. The management of these disorders is discussed in Chapter 111 and Chapter 120.

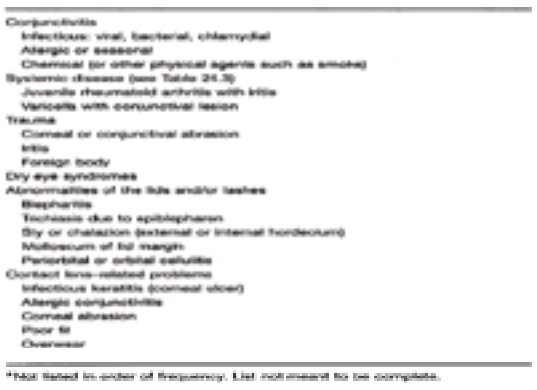
Conjunctivitis
Infectious: viral, bacterial, chlamydial
Allergic or seasonal
Chemical (or other physical agents such as smoke)
Systemic disease (see Table 24.3)
Juvenile rheumatoid arthritis with iritis
Varicella with conjunctival lesion
Trauma
Corneal or conjunctival abrasion
Iritis
Foreign body
Dry eye syndromes
Abnormalities of the lids and/or lashes
Blepharitis
Trichiasis due to epiblepharon
Stv or chalazion (external or internal hordeolum)
Molluscum of lid margin
Periorbital or orbital cellulitis
Contact lens–related problems
Infectious keratitis (corneal ulcer)
Allergic conjunctivitis
Corneal abrasion
Poor fit
Overwear
[illegible] listed in order of frequency. List not meant to be complete.

Table 24.1. Common Causes of Red Eye[a]

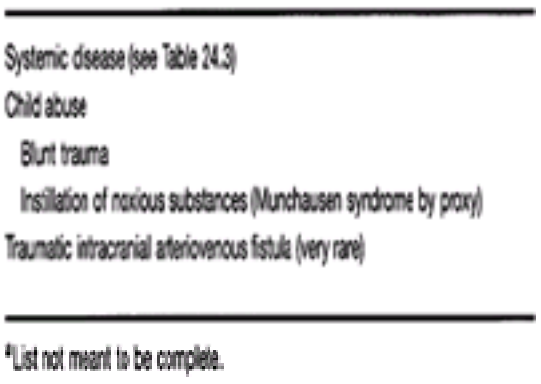
Systemic disease (see Table 24.3)
Child abuse
Blunt trauma
Instillation of noxious substances (Munchausen syndrome by proxy)
Traumatic intracranial arteriovenous fistula (very rare)
[a]List not meant to be complete.

Table 24.2. Life-Threatening Causes of Red Eye[a]

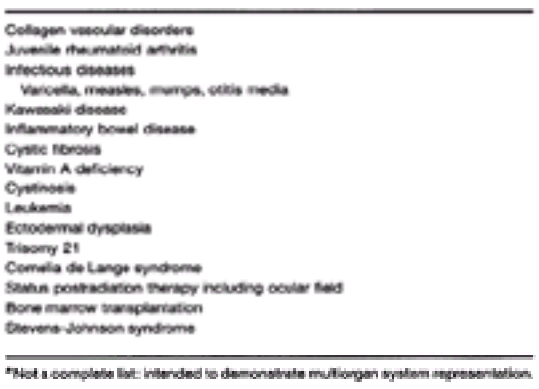
Collagen vascular disorders
Juvenile rheumatoid arthritis
Infectious diseases
Varicella, measles, mumps, otitis media
Kawasaki disease
Inflammatory bowel disease
Cystic fibrosis
Vitamin A deficiency
Cystinosis
Leukemia
Ectodermal dysplasia
Trisomy 21
Cornelia de Lange syndrome
Status postradiation therapy including ocular field
Bone marrow transplantation
Stevens-Johnson syndrome
[a]Not a complete list; intended to demonstrate multiorgan system representation.

Table 24.3. Systemic Conditions That May Be Associated with Red Eye[a]

Often, the cause of a red eye can be identified based on the history alone. Attention must be paid to documenting whether the inflammation is unilateral or bilateral, diffuse or sectorial, and acute or chronic. When bilateral, it is helpful to know whether both eyes were involved simultaneously or sequentially.

PATHOPHYSIOLOGY

With the exception of the cornea, the eye is covered by conjunctiva, a modified mucous membrane covered by nonkeratinized stratified squamous epithelium with goblet cells. The conjunctiva is contiguous with the corneal epithelium. The conjunctiva extends from the surface of the eyeball above and below onto the inner surface of the upper and lower eyelids, creating an upper and lower fornix as it reflects off the eyeball. These fornices may become repositories for foreign material or exudate. The conjunctiva overlying the eyeball (bulbar conjunctiva) may become inflamed without involvement of the conjunctiva lining the inner aspect of the eyelid (palpebral conjunctiva). The palpebral conjunctiva contains lymphoid follicular tissues that may become particularly prominent during ages when benign lymphoid hypertrophy (e.g., tonsils, adenopathy) is common. Benign lymphoid hyperplasia would appear as small bumps on the palpebral conjunctiva. A follicular reaction also may occur in some forms of red eye—in particular, viral conjunctivitis. The conjunctiva also covers the caruncle, a small lump of tissue containing glands located in the medial corner of the eye at the junction of the upper and lower eyelids.

The sclera (“the white of the eye”) is a largely avascular dense collagenous tissue that provides a tough fibrous outer wall for the eyeball. Four of the six extraocular muscles insert into the anterior sclera, 4 to 8 mm away from the cornea: superior, medial, inferior, and lateral recti (see Chapter 25). Although these insertions are not easily visible on the normal eye, inflammation of the muscles makes them apparent. Knowledge of this anatomy may be helpful in the diagnosis of myositis.

The sclera may become inflamed (scleritis). An intermediate layer, the episclera, lies beneath the conjunctiva, where it is firmly attached to the sclera. The episclera does not extend onto the eyelids. It is more vascularized than the sclera and may become inflamed either in a diffuse or localized fashion (diffuse, sectorial, or nodular episcleritis).

The term *conjunctivitis* should be reserved for disorders in which the conjunctiva is inflamed. Inflammation may be caused by direct irritation, infection, inflammation of underlying or contiguous structures, immune phenomena, or processes secondary to abnormalities of the lid and lashes. Disorders of the cornea may result in conjunctival inflammation. Inflammation within the anterior chamber between the cornea and iris (iritis) also may result in inflammation of the conjunctiva.

A tear film, which prevents desiccation, is constantly present over the surface of the eye. The tear film is made up of three components: an inner mucinous layer secreted by the goblet cells of the conjunctiva; a middle aqueous layer secreted by the lacrimal glands within the eyelid, orbit, and superior conjunctival fornix; and an outer layer secreted by glands in the body of the eyelids which empty at the eyelid margins. Each eyelid contains 20 to 30 glands. A disruption in the function of any three of these anatomic structures may result in an abnormal tear film with secondary desiccation of the ocular surface, resulting in irritation and inflammation (dry eye syndrome).

Innervation of the conjunctiva and cornea comes from the first division of the trigeminal nerve (V1). Abnormalities on the ocular surface may give rise to pain or a foreign body sensation. The efferent arm of a reflex arc that involves the trigeminal nerve (afferent limb) and the facial nerve results in a rapid blink, with contraction of the orbicularis oculi muscle, to protect the surface of the eye in response to noxious stimuli.

Two other reactions to noxious stimuli may occur: tearing and discharge. Tearing (epiphora) may accompany virtually any conjunctival inflammation or irritation. Tearing may even be a part of the dry eye syndrome, as the lacrimal gland attempts to compensate for ocular surface dessication due to disruption of the other layers of the tear film. Discharge results either from conjunctival exudation or precipitation of mucous out of the tear film. The latter occurs when the tear film is not flowing smoothly (e.g., nasolacrimal duct obstruction), causing misinterpretation as infection when the problem is actually mechanical. Although discharge may be a nonspecific finding, the nature of the discharge may be helpful in determining the cause of an infection. The presence of membranes or pseudomembranes (Fig. 24.1) on the palpebral conjunctiva is also helpful in establishing a cause. For example, these membranes are more common with adenovirus infection or Stevens-Johnson syndrome. These white or white-yellow plaques are caused by loosely or firmly adherent collections of inflammatory cells, cellular debris, and exudate.

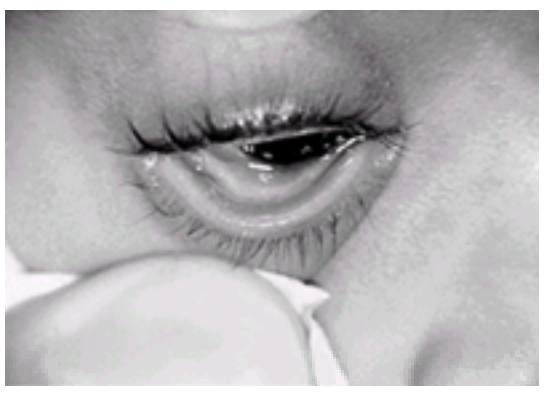

FIGURE 24.1. Pseudomembrane on lower lid palpebral conjunctiva and extending into inferior fornix in patient with epidemic keratoconjunctivitis. *Please see the color-tip insert (Color Plate 24.1).*

EVALUATION AND DECISION

The approach to the child who presents in the emergency department with a red eye is outlined in the flow chart shown in Figure 24.2. Any child who wears contact lens regularly, even if the lens is not in the eye at the time of the examination, should be referred to an ophthalmologist within 12 hours if he or she has red eye. Red, and often painful, eyes of a person who wears contact lens may represent potentially blinding corneal infection or the breakdown of the corneal epithelium, which subsequently would predispose the person to corneal infection. Other than removing the contact lens when possible (topical anesthesia may be helpful), further diagnostic or therapeutic interventions by the pediatric emergency physician are not indicated in these patients. It is recommended that empiric antibiotics not be started because the ophthalmologist may wish to culture the cornea.

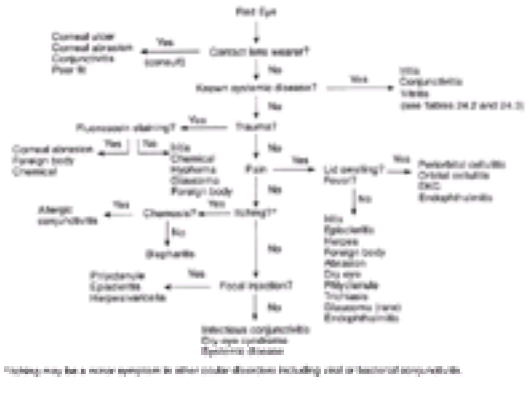

FIGURE 24.2. Approach to the child with red eye. *EKC,* epidemic keratoconjunctivitis (adenovirus).

Numerous systemic diseases may be associated with ocular inflammation. A representative sample can be found in Table 24.3. In some systemic diseases, the associated ocular abnormality involves intraocular inflammation (iritis, vitritis), which might cause secondary conjunctival infection. Patients with these diseases also may have coincidental ocular inflammation unrelated to their underlying conditions. Ophthalmologic consultation may be helpful in making this distinction.

Traumatic injury may result in a red eye because of corneal or conjunctival abrasion, hyphema, iritis, or rarely, traumatic glaucoma. (The diagnosis and treatment of these disorders are summarized in Chapter 111.) Very rarely, head injury may cause the development of an intracranial arteriovenous fistula that may present with proptosis, chemosis, red eye, corkscrew conjunctival blood vessels, and decreased vision. If there is no fluorescein staining of the conjunctiva or cornea and there is no obvious evidence of severe intraocular injury (e.g., hyphema, ruptured globe), the examiner may need to consider the possibility of noxious material coming in contact with the eyeball at the time of trauma. Both acidic and alkaline substances may cause a red eye. (The treatment of these disorders is summarized in Chapter 120.) Likewise, a foreign body may cause ocular pain and inflammation. Foreign bodies often can be difficult to see on brief, superficial examination. All the recesses and redundant folds of the conjunctiva must be inspected. The upper eyelid should be everted (see Chapter 111) and the lower eyelid should be pulled down from the globe as the patient looks upward so that the inferior fornix can be inspected. The patient should be asked to adduct the affected eye when the lateral canthus (junction of the upper and lower eyelid laterally) is stretched laterally to allow inspection of the lateral fornix. There is no analogous medial fornix.

It often is wise to inspect the position of the eyelashes before performing these manipulations. Eyelashes that turn against the ocular surface (trichiasis) may cause a red eye that is accompanied by pain or foreign body sensation. Although corneal fluorescein staining may reverse the irritation, the condition may be so mild that slitlamp biomicroscopy would be required despite significant symptoms. It is particularly common in patients who have had prior injury or surgery to the eyelid and in patients of Asian background. In the latter case, a prominent fold of skin (epiblepharon) may be found medially just below the eyelid margin, causing the eyelashes to rotate toward the eyeball (Fig. 24.3).

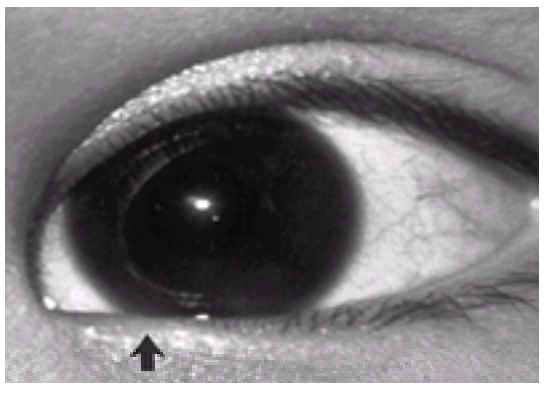

FIGURE 24.3. Epiblepharon. Extra skin fold medially on lower lid *(arrow)* rotates eyelashes back toward eyeball surface.

In the absence of cornea/conjunctiva abrasion, foreign body, and trichiasis, the painful red eye caused by trauma may have iritis. This may not present for up to 72 hours after the trauma was sustained. Photophobia and visual blurring also

may occur. The pupil may be smaller. Occasionally, one will see a cloudy inferior cornea caused by the deposition of inflammatory cells and debris on the inner surface. Iritis also may occur in association with systemic disease or as an isolated idiopathic ocular finding. Traumatic iritis and nontraumatic iritis often are indistinguishable except by history. All causes of iritis, regardless of the etiology, require ophthalmologic consultation and follow-up. The diagnosis of iritis requires slitlamp examination by a skilled observer. Topical steroids should not be prescribed by the primary care physician.

Episcleritis and scleritis also may cause a painful red eye. Although episcleritis usually is an isolated ocular abnormality, scleritis often is associated with an underlying systemic disease, particularly the collagen vascular disorders. Both entities may present with focal or diffuse inflammation. A nodular elevation may be seen involving the sclera. The eye may be tender, and the inflamed area may have a bluish hue. There also may be pain on attempted movement of the eye. Diagnosis and treatment require slitlamp examination and ophthalmologic consultation.

Another cause of painful ocular inflammation is herpetic corneal infection. This may be caused by the simplex or varicella-zoster viruses. Usually, there is no concomitant dermatologic manifestation except in association with chickenpox, when a unilateral or bilateral lesion may be seen on the conjunctiva (usually near but not on the cornea) with focal injection (Fig. 24.4) Usually, no treatment is required. However, herpetic corneal ulcers require urgent treatment to prevent corneal scarring and vision loss. Patients with herpetic corneal ulcers may have a history of prior recurrent painful red eye, although herpes occasionally can be painless because of induced corneal hypoesthesia. Herpes simplex is virtually always unilateral. Fluorescein staining of the cornea may reveal a linear branching pattern (see Fig. 120.10). If the infected area is located eccentrically on the corneal surface, the injection may be localized to the quadrant of conjunctiva adjacent to the lesion.

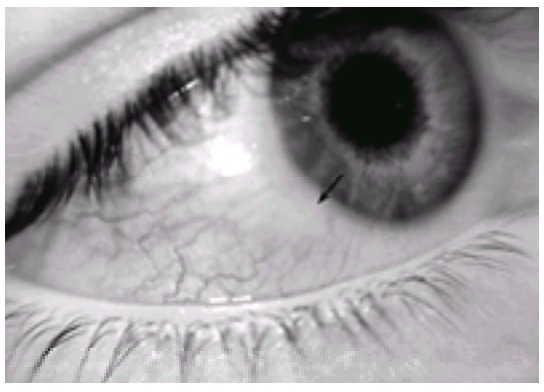

FIGURE 24.4. Red eye caused by chickenpox (varicella) involvement of conjunctiva. Note sectorial injection of conjunctiva. White area *(arrow)* at junction of conjunctiva and cornea is the pox lesion. *Please see the color-tip insert (Color Plate 24.4).*

If eye pain is relieved by a drop of topical anesthetic (see Chapter 111), the patient must have a surface problem (e.g., foreign body, abrasion). If the pain is not relieved and periorbital swelling and fever are present, the red eye may be caused by periorbital or orbital cellulitis. These are emergent conditions, the treatment and diagnosis of which are reviewed in Chapter 120. Eye pain and marked lid swelling also may be associated with epidemic keratoconjunctivitis (EKC) secondary to adenovirus (see Fig. 120.8). When questioned further, patients may reveal that they actually have a sandy foreign body sensation rather than true ocular pain. Pseudomembranes are a fairly diagnostic sign when present (Fig. 24.1). Low-grade fever and tender preauricular adenopathy also may occur, making it difficult to distinguish EKC from periorbital cellulitis. However, EKC usually affects the eyes consecutively and bilaterally. There also may be associated prominent photophobia and tearing, which is not usually seen in cellulitis.

Itching is another important diagnostic symptom. When it is associated with swelling of the conjunctiva, giving it the appearance of a blisterlike elevation (see Fig. 120.9), one should suspect allergic conjunctivitis. Often, there is no known causative agent, and there may be associated periocular swelling. The condition may be unilateral or bilateral and usually has an acute or hyperacute onset. Photophobia, tearing, and lid swelling also may occur. The emergency physician can prescribe topical antihistamines and/or vasoconstrictors as well as a cool compresses to relieve the symptoms.

Itching also may accompany blepharitis, an idiopathic disorder in which there is suboptimal flow of secretions from the glands normally present in the eyelids. Because these glands participate in the formation of the lubricating tear film that normally covers the eye, the deficiency of flow may result in an abnormal tear film and rapid corneal desiccation. Symptoms are aggravated by activities associated with prolonged staring and a decreased blink rate (reading, television viewing, and video games) or going outside on windy days. Patients may have photophobia and a sandy foreign body sensation. To compensate for the tear film deficiency, reflex excess tearing may occur from the lacrimal gland. The most characteristic sign is erythema of all four eyelid margins and flaking and crusting at the base of the eyelashes (Fig. 24.5). Slitlamp examination is helpful in making this diagnosis and is necessary to rule out the presence of corneal involvement.

FIGURE 24.5. Blepharitis. Note crusts and flakes at base of eyelashes.

Although itching and pain may be minor symptoms associated with several types of conjunctivitis, it is usually the absence of these symptoms that should lead one to suspect an infectious cause. Conjunctivitis usually causes diffuse inflammation of the conjunctiva, either unilaterally or bilaterally. The differentiation of bacterial, viral, chlamydial, and other types of conjunctivitis sometimes is difficult (see Chapter 120). Purulent discharge is particularly characteristic of bacterial infection. Diffuse injection may also be a sign of a previously unrecognized underlying systemic disease.

If the injection is localized, the examiner should consider a specific list of diagnostic possibilities. A phlyctenule appears as a small nodule on the conjunctiva at or near the corneal–conjunctival junction, usually with concomitant blepharitis. It usually represents an immune response to bacteria located on the surface of the eye or eyelid. Episcleritis and scleritis also may present with focal involvement, as previously discussed. Localized injection of the conjunctiva may be an indicator of an imbedded foreign body, herpes, chickenpox, or other focal processes that require the attention of an ophthalmologic consultant.

Although much feared, glaucoma in children is very rare. Congenital glaucoma usually is not associated with a red eye or pain. Small children who have glaucoma often have enlarged eyes (buphthalmos) with tearing, photophobia, and sometimes, heterochromia. Acute acquired glaucoma causes a painful red eye, perhaps associated with corneal clouding and decreased visual acuity. Acquired glaucoma, however, usually is associated with trauma, other anatomic abnormalities, or iritis that would be apparent on examination. Because it is difficult to determine intraocular pressure in children, ophthalmologic consultation is required in all cases.

Suggested Readings

Fisher MC. Conjunctivitis in children. Pediatr Clin North Am 1987;34: 1447–1456.

Levin AV. Eye emergencies: acute management in the pediatric ambulatory setting. Pediatr Emerg Care 1991;7:367–377.

Matoba A. Ocular viral infections. Pediatr Infect Dis 1984;3:358–368.

CHAPTER 25

Eye—Strabismus

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Strabismus refers to any ocular misalignment. *Esotropia* refers to eyes that are turned in (crossed eyes). *Exotropia* refers to eyes that are turned out (wall eyed). The terms *hypertropia* and *hypotropia* refer to a higher or lower eye, respectively. By convention, vertical misalignment of the eyes always is categorized by the higher eye (e.g., right hypertropia), unless it is known that a specific abnormal process is causing one eye to be held in a lower position (e.g., left hypotropia). Many children with strabismus require a formal evaluation by an ophthalmologist for definitive diagnosis and management, but the emergency physician should attempt to answer two questions: 1) Is the strabismus an emergency? and, if so, 2) What is the most likely cause?

PATHOPHYSIOLOGY

Six muscles surround each eyeball (Fig. 25.1). Although several of these muscles may individually move the eye in more than one direction, knowledge of the primary action of these muscles allows for the definition of diagnostic positions of gaze (Table 25.1). This can be helpful in pinpointing specific muscle dysfunction. For example, if a muscle that primarily governs abduction (lateral rectus) is impaired, the eye is unable to abduct and may lie in a position of adduction (esotropia). Likewise, if a muscle that is involved with downgaze (superior oblique muscle) is impaired, the eye will have a tendency to remain in relative upgaze (ipsilateral hypertropia).

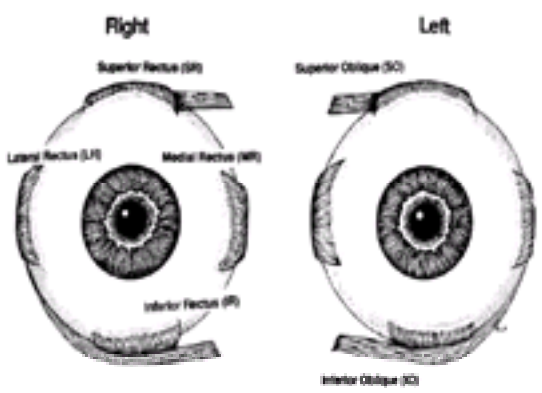

FIGURE 25.1. Normal extraocular muscle anatomy.

Muscle[a]	Cranial Nerve	Action[b]	Eye Position in Palsy
Medial rectus	III (inferior division)	Adduction	Exotropia
Inferior rectus	III (inferior division)	Downgaze	Hypertropia
Lateral rectus	VI	Abduction	Esotropia
Superior rectus	III (superior division)	Upgaze	Hypotropia
Superior oblique	IV	Downgaze	Hypertropia
Levator palpebrae[c]	III (superior division)	Eyelid	Ptosis (lid)

[a]Inferior oblique not included for simplicity. Isolated palsy of the inferior oblique is extremely rare.
[b]Action in the horizontal or vertical field only.
[c]By definition, not truly an extraocular muscle.

Table 25.1. Extraocular Muscles

Although the interactions of these muscles are complex, the eyeballs should always move symmetrically, both quantitatively and qualitatively, into each direction of gaze. On lateral gaze, the abducting and adducting eye should

move far enough that no sclera is visible laterally or medially, respectively (Fig. 25.2). On upgaze, the eyelids also should move up involuntarily. Likewise, on downgaze, the eyelids should move down symmetrically. In the primary position (straight ahead), no sclera should be visible superiorly. The upper eyelid margins should just cross over the iris without crossing over a significant portion of pupil. The lower eyelid margin usually crosses within 1 mm above or below the 6 o'clock position of the inferior iris.

FIGURE 25.2. Patient's head is being rotated passively to patient's left as he looks straight ahead. This causes displacement of eyes into right gaze. Left eye adducts fully, showing no visible sclera medially. Subtle right sixth nerve palsy demonstrated by failure of right eye to abduct fully: sclera is still visible laterally on right eye.

In general, there are two acute reasons why the function of a particular muscle might be impaired: neurogenic palsy or muscle restriction. Three cranial nerves are responsible for the innervation of the six extraocular muscles (Table 25.1). The sixth cranial nerve innervates the ipsilateral lateral rectus muscle. This nerve exits the ventral pons and then travels on the wall of the middle cranial fossa (clivus), reaching the sphenoid ridge, along which it travels until entering the cavernous sinus. The course of this nerve allows it to be injured by vascular or neoplastic changes in the midbrain, increased intracranial pressure (ICP), anterior midline craniofacial tumors (e.g., nasopharyngeal carcinoma), otitis media (OM), and any abnormality that involves the cavernous sinus.

The fourth cranial nerve innervates the superior oblique muscle. It is the only ocular cranial nerve that completely decussates and has a dorsal projection over the midbrain. This position renders the fourth cranial nerve particularly vulnerable to blunt head trauma, one of the most common causes of fourth nerve palsy. The fourth cranial nerve also has the longest intracranial course, which makes it particularly susceptible to increased ICP and parenchymal shifts caused by cerebral edema. It also runs through the cavernous sinus. Fourth cranial nerve palsy may be congenital, but asymptomatic for several years during childhood until the brain is no longer able to compensate. The eyes become misaligned vertically (ipsilateral hypertropia) in the same fashion that might result from a traumatic fourth nerve palsy. Ophthalmic consultation may allow differentiation of congenital and acquired palsy.

The third cranial nerve supplies the remaining four extraocular muscles. It is involved with downgaze, upgaze, and adduction. Parasympathetic innervation to the pupil (see Chapter 26) and innervation to the eyelid muscle (levator palpebrae) also are carried in the third cranial nerve. A complete third cranial nerve palsy results in an eye that is positioned down (from the remaining action of the unaffected superior oblique muscle) and out (from the remaining action of the unaffected lateral rectus muscle) with ipsilateral ptosis and ipsilateral pupillary dilation (Fig. 25.3). Because the third cranial nerve divides into a superior and an inferior division just as it enters the orbit from the cavernous sinus and because the fibers to individual muscles are segregated within the nerve throughout its course, partial third cranial nerve palsies may occur with or without ptosis and/or pupillary dilation. This may leave the patient with complex strabismus, which is best left to the ophthalmologic consultant. (The differential diagnosis of third cranial nerve palsies is summarized in Chapter 26.)

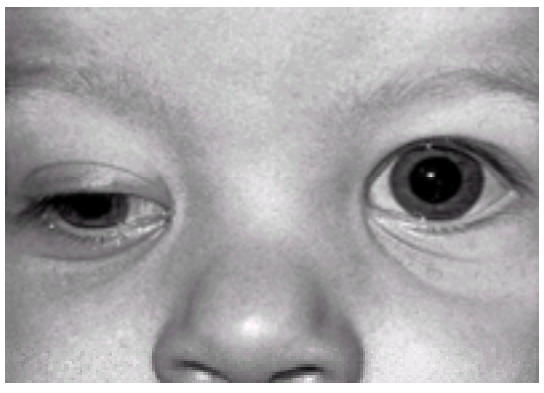

FIGURE 25.3. Right third cranial nerve palsy. When looking straight ahead with left eye, right eye rests in a hypotropic and exotropic position. Note right ptosis. Right pupil is involved (mydriatic), but both pupils were dilated pharmacologically by examiner just before this photograph was taken.

The action of a muscle also may be impaired by restriction. The muscle can become infiltrated with substances that might restrict its action or cause fibrosis. Children with hyperthyroid eye disease have large, tight eye muscles. An eyeball also may be restricted in its movements by tumors or infection in and around the globe. Orbital tumors, cellulitis, or abscesses

that cause restriction may be associated with proptosis or a displacement of the entire eyeball, either vertically or horizontally. Although it is beyond the scope of this chapter to discuss all the causes of restrictive strabismus, it is important to focus on the pathophysiology of strabismus that occurs after a blowout fracture. After blunt trauma to the eyeball, the globe may be translocated posteriorly, causing an increased intraorbital pressure that may result in fracture of the bony orbital wall. Limitations of movement caused by blowout fractures may not be noticeable until the eye attempts to move. In other words, the eyes may be parallel in the straight ahead position but may be misaligned when they attempt to look in a direction opposite the fracture site. When an orbital wall fracture occurs, the muscle that runs along that wall may become entrapped within that fracture, tethering the eyeball so that it cannot look in the direction opposite the fracture. For example, fractures of the orbital floor may entrap the inferior rectus muscle, tethering the eye downward so that upgaze is restricted (Fig. 25.4). Sometimes, the eye also may have a limitation of movement in the direction of the fracture.

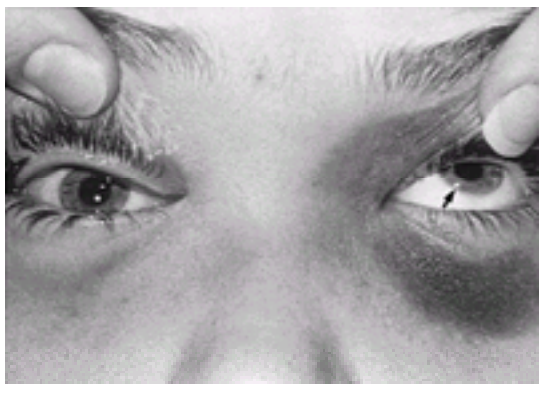

FIGURE 25.4. Patient is looking upward. Right inferior orbital wall blowout fracture causes restriction of upgaze in right eye. Note light reflexes (Hirschberg test). Left reflex *(arrow)* is lower in reference to pupil than right reflex, indicating the presence of a right hypotropia.

Orbital wall fractures may be associated with enophthalmos, in which the eye appears to be sunken in the orbit, or proptosis caused by orbital hemorrhage. There usually is a history of trauma and, perhaps, other evidence of ocular injury. All patients with orbital fractures must receive a complete ophthalmic examination to rule out accompanying ocular injury. The most common fracture involves the inferior and/or medial walls of the orbit. The lateral wall is rarely fractured. Fracture of the superior wall (orbital roof) is particularly worrisome because it may allow communication between the intracranial space and orbit.

EVALUATION AND DECISION

The Hirschberg light reflex test can be helpful in determining whether strabismus is present. The physician should shine a penlight or direct ophthalmoscope light at the patient's eyes from 2 to 3 feet away while the patient is told to look at the other end of the room. In younger children, the patient may choose to look at the light itself. The examiner should observe the white dot light reflex that appears to be located on the cornea, overlying the iris or pupil of each eye. This reflex should be located in a nearly symmetric position in each eye (Fig. 25.5). In the normal state, the light reflex actually falls slightly off center in the nasal direction in both eyes (Fig. 25.5). If the eye were to be misaligned, this symmetry would not be preserved (Fig. 25.4 and Fig. 25.6).

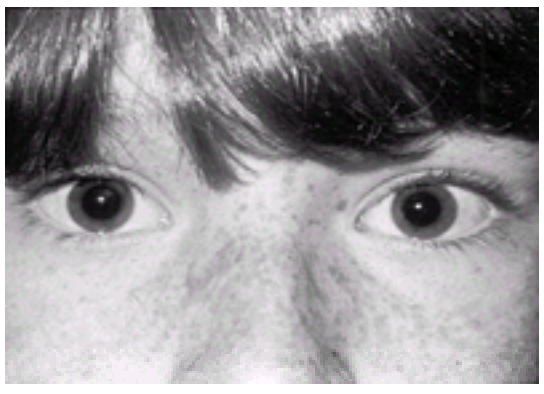

FIGURE 25.5. Normal Hirschberg light reflex test. Light reflexes fall symmetrically in each eye. The reflex in the patient's left eye is a bit nasal to the center.

FIGURE 25.6. Left esotropia. Note lateral displacement of Hirschberg light reflex in the left eye. Photograph demonstrates right ptosis and orange-red reflex in the left eye with black reflex in the right eye. Pupils are pharmacologically dilated. Asymmetry of red reflex is caused by misalignment of the eyes. *Please see the color-tip insert. (Color Plate 25.6)*

Two findings are helpful in assessing whether strabismus is emergent: 1) the presence or absence of double vision and 2) the status of the eye movements. Although young children may not complain of diplopia, this symptom often indicates an acute or subacute onset of ocular misalignment. Nonemergent childhood strabismus usually is not associated with double vision because the brain becomes adept at suppressing the misaligned nonfixing eye. If a child complains of diplopia, ophthalmologic consultation is appropriate, even if no strabismus appears on examination by the pediatric emergency physician.

If the eye movements are completely full and symmetric, one can be virtually certain that the strabismus is not emergent. Problems that cause emergent strabismus do so by impairing the action of one or more muscles. A neurogenic palsy or restrictive phenomenon cannot be present if the eye movements are full. If there are any questions about subtle reductions in extraocular movement or if there is prominent nystagmus elicited in one particular field of gaze (more than the few beats of normal end point nystagmus), ophthalmologic consultation is most likely appropriate.

Because of fear and noncompliance, some children will not follow the examiner's target that is presented to assess eye movement. If they will not follow the target at all but look at the examiner only, the examiner should ask the parent to gently move the patient's head to each side and then up and down. The examiner also can do this by putting one hand on the child's head, although this may serve only to heighten the child's anxiety. As the patient continues to look straight ahead when the head is being turned, the eyes are moving passively in reference to the head and orbit. When the head is turned to the left, the eyes move into right gaze to maintain fixation straight ahead (Fig. 25.2). If the head is tilted up, the eyes are moved into relative downgaze. Essentially, this is the “doll's eye” maneuver used in the assessment of comatose patients. If the eyes move symmetrically and fully on passive movement of the head, this rules out the presence of a neurogenic or restrictive problem with the same accuracy as if the patient had voluntarily followed a target. Although the neuro-ophthalmologic contributions to the assessment of the comatose patient are beyond the scope of this chapter, the presence of a cranial nerve palsy can be ruled out even in the patient who is experiencing an altered mental status related to central nervous system disease if the eyes move fully on the doll's eye maneuver as the head is rotated by the examiner.

When an orbital blowout fracture is suspected, confirmation may be obtained by a computed tomography (CT) scan of the orbit. Some controversy exists about the need to perform this test emergently. Some ophthalmologists prefer to image only if the strabismus and diplopia do not spontaneously resolve over 1 to 2 weeks. To evaluate the extraocular muscles, it is essential that coronal views be obtained. Contrast enhancement is unnecessary. Plain skull radiographs play virtually no role in the diagnosis and management of orbital fractures. Should there be a concern about intracranial injury or orbital roof fracture as the cause of strabismus, a CT scan of the head also is important.

The causes of pediatric strabismus are summarized in Table 25.2, Table 25.3 and Table 25.4. If the physician has identified an impairment of the eye to look in a given direction, an emergent condition may be present. The first considerations (Fig. 25.7 and Fig. 25.8) are restrictive strabismus (e.g., trauma) and neurogenic palsies. Myasthenia gravis and thyroid ophthalmopathy (hyperthyroidism), however, can mimic virtually any strabismus with deficiency of extraocular movement and must always be considered in the differential diagnosis in any pattern of ocular misalignment. Myasthenia may cause ptosis, whereas thyroid disease causes retraction of the upper lid. The pupils are not involved in either condition.

Neurogenic Palsies
III cranial nerve palsy (partial or complete)
IV cranial nerve palsy
VI cranial nerve palsy
Traumatic extraocular muscle palsy
Myasthenia gravis
Internuclear ophthalmoplegia
Skew deviation
Restrictive Strabismus
Orbital wall fracture
Orbital hemorrhage, tumor, infection, or abscess
Thyroid eye disease
Nonthyroid extraocular muscle infiltration (e.g., metastasis)
Orbital cellulitis
Nonneurogenic Nonrestrictive Strabismus
Idiopathic childhood strabismus
Strabismus caused by refractive errors (e.g., accommodative esotropia)
Sensory strabismus (unilateral visual loss)

[a] Not listed in order of frequency.

Table 25.2. Differential Diagnosis of Strabismus [a]

Exotropia
Congenital or acquired (with or without farsightedness), nonparalytic, nonrestrictive
Medial orbital wall fracture
VI cranial nerve palsy
Orbital mass, hemorrhage, or infection
Long-standing unilateral visual loss
Esotropia
Nonparalytic nonrestrictive idiopathic childhood exotropia
III cranial nerve palsy
Orbital mass, hemorrhage, or infection
Long-standing unilateral visual loss
Hypertropia
Dissociated vertical deviation (a nonparalytic nonrestrictive childhood deviation)
Inferior or superior orbital wall fracture
IV cranial nerve palsy: congenital or acquired
Orbital mass, hemorrhage, or infection
Hypotropia
Inferior or superior orbital wall fracture
Orbital mass, hemorrhage, or infection

[a]Not listed in order of frequency.

Table 25.3. Common Causes of Strabismus[a]

Intracranial mass	Head trauma
Elevated intracranial pressure	Meningitis
Myasthenia gravis	Neoplastic infiltration of extraocular muscles
Orbital tumor	Superior orbital wall fracture
Orbital cellulitis	Retinoblastoma

[a]Not listed in order of frequency.

Table 25.4. Life-Threatening Causes of Strabismus[a]

FIGURE 25.7. Evaluation of horizontal strabismus. *EOM,* extraocular muscle movements; *MR,* medial rectus; *LR,* lateral rectus; *LW,* lateral orbital wall; *MW,* medial orbital wall; *fx,* fracture; *ICP,* increased intracranial pressure; *INO,* internuclear ophthalmoplegia; *boldface type,* most likely fracture.
[a] With head held in straight ahead position.

FIGURE 25.8. Vertical strabismus. *EOM,* extraocular muscle movements; *IR,* inferior rectus; *SR,* superior rectus; *SW,* superior orbital wall; *IW,* inferior orbital wall; *boldface type,* most likely fracture.
[a]With head held in straight ahead position.

Esotropia Emergencies

An eye that rests in the crossed position is esotropic. The Hirschberg light reflex will fall laterally in reference to the

central pupil, compared with the unaffected eye (Fig. 25.6). Figure 25.7 summarizes the approach to a patient with esotropia. Patients with a restrictive or neurogenic esotropia (deficiency of abduction) may adopt an abnormal head position to place the eyes in the position of best alignment to avoid double vision. By turning the face in the direction of the deficiency (e.g., right face turn for right sixth nerve palsy) when looking straight ahead, the eyes actually may be straight. The patient's head must be held in the straight ahead position to notice that the affected eye actually is crossed.

In the presence of proptosis or a history of eye trauma, one must be concerned that an orbital process is causing the esotropia. Fracture of the medial orbital wall may cause entrapment and restriction of the medial rectus. Fracture of the lateral wall—usually part of a tripod fracture that involves the zygoma and inferior lateral wall—may cause orbital hemorrhage that would displace the eye medially. Likewise, an orbital tumor or abscess can push the eye toward the nose or restrict abduction. Any infiltrative process that involves the eye muscles may also cause esotropia through restriction. Orbital cellulitis can cause any type of misalignment, including esotropia, with or without abscess formation. A CT scan of the orbit with coronal and axial views is the diagnostic procedure of choice in these situations.

Lateral rectus palsy (sixth cranial nerve palsy) occurs most commonly secondary to head trauma (see Chapter 105) or increased ICP. Other central nervous system signs, such as papilledema, may be present. Magnetic resonance imaging (MRI) of the brain is the procedure of choice. Sixth cranial nerve palsy also can occur rather precipitously after the placement of ventricular shunts designed to relieve increased ICP. Sixth cranial nerve palsy may be bilateral, in which case both eyes will be in the crossed position. OM also may be associated with sixth cranial nerve palsy (Gradenigo syndrome).

Exotropia Emergencies

Orbital cellulitis, thyroid eye disease, and orbital tumors also may cause exotropia. Trauma very rarely results in exotropia because lateral wall fractures rarely cause entrapment. Orbital hemorrhage (with or without medial wall fracture) can have a mass effect, causing the eye to be turned out.

Isolated paresis of the medial rectus muscle, resulting in a deficiency of adduction and a turned out eye, is quite unusual because other muscles also are innervated by the third cranial nerve. One should look for accompanying ptosis, pupillary dilation, or deficiencies of upgaze or downgaze to confirm third cranial nerve involvement, even if these findings are subtle.

Unilateral isolated deficiency of adduction may be the result of an intranuclear ophthalmoplegia secondary to a brainstem injury that involves the interconnecting pathways between the third and sixth cranial nerves. Bilateral isolated deficiency of adduction is virtually diagnostic of this condition. MRI of the brainstem should be ordered emergently. The observer also should look for prominent nystagmus on attempted abduction of the contralateral eye.

Hypertropia/Hypotropia Emergencies

Any vertical eye muscle imbalance must be referred to an ophthalmologist. Figure 25.8 summarizes the approach to the patient with vertical ocular misalignment. To determine whether it is the higher or lower eye that is abnormal, the examiner must have the patient look upward then downward. If one eye is unable to look downward fully, the patient has a hypertropia of that eye. If one eye is unable to look upward fully, that eye is hypotropic (Fig. 25.4). Patients may adopt abnormal head positions to compensate for this misalignment. By lifting the chin to look straight ahead, the eyes are placed in relative downgaze, thus indicating that the strabismus is worse when the patient looks up. Likewise, the patient may adopt a chin down position to look straight ahead, indicating that the strabismus is worse in downgaze.

An eye may become hypertropic for several reasons. Any mass underneath the eyeball—for example, an orbital tumor or a mucocele extending upward from the maxillary sinus—may push the eye upward. A tightened superior rectus is unusual but may be seen in thyroid eye disease or after trauma. An inferior orbital wall fracture could injure (weaken) the inferior rectus or cause hemorrhage that would push the eye up into a hypertropic position. A CT scan of the orbit would be the proper diagnostic modality.

Perhaps the most important cause of ipsilateral hypertropia is a lesion that involves the fourth cranial nerve. Although the eye may be able to look straight down fully, there may be a restriction of gaze in the down-and-in position relative to the other eye (which then would be looking down and out). Because of the torsional forces of the superior oblique muscle on the eyeball, the patient may adopt a head position with a face turn and a head tilt away from the affected eye. One must always consider the possibility that a new fourth nerve palsy actually represents a decompensated congenital abnormality. Old pictures that show a previous head tilt can be helpful. The head tilt is one of the mechanisms used by the patient unconsciously to help compensate for the muscle imbalance.

Although rare, neurogenic palsies of the inferior oblique or inferior rectus with resultant vertical misalignment may occur. These have been reported after viral illnesses, including varicella. As with exotropia of neurogenic origin, however, it would be more likely to have other branches of the third cranial nerve involved with other findings. Another type of vertical eye muscle imbalance, skew deviation, can be the presenting sign of a midbrain lesion. Ophthalmologic consultation can be helpful in deciding whether MRI is appropriate.

Hypotropia can be caused by an orbital roof fracture with superior hematoma that pushes the eye down. Alternatively, hypertropia from a deficiency of downgaze due to tethering of the superior rectus muscle also can occur. Orbital roof fracture is an emergent condition. Neuroradiologic evaluation must be obtained to rule out communication between the orbit and the intracranial cavity. Pulsating proptosis is a particularly ominous sign.

Traumatic hypotropia most commonly results from inferior wall blowout fractures (Fig. 25.8). The eye often is enophthalmic and there may be associated numbness in the distribution of the infraorbital nerve as it innervates the ipsilateral infraorbital and malar region. Orbital lesions, including those that may have extended from the intracranial

cavity, also may push the eyeball downward and prevent it from looking upward. Thyroid eye disease also can cause hypotropia due to tightening of the inferior rectus.

Suggested Readings

Catalano JD. Strabismus. Pediatr Ann 1983;12:550–555.

Cibis GW, Waeltermann JM. Rapid strabismus screening for the pediatrician. Clin Pediatr 1986;25:304–307.

Dutton JJ, Mason PN, Iliff N, Putterman AM. Management of blow-out fractures of the orbital floor. Surv Ophthalmol 1991;35:279–298.

Nelson LB. Pediatric Ophthalmology. Major Problems in Clinical Pediatrics. Vol. 25. Philadelphia: WB Saunders, 1984:94–106.

CHAPTER 26

Eye—Unequal Pupils

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Abnormalities of the pupils can be helpful diagnostically when assessing central nervous system (CNS), autonomic nervous system (ANS), orbital, and ocular problems. The pupillary dilator muscle receives sympathetic innervation. The pupillary sphincter receives parasympathetic innervation that also supplies the ciliary muscle of the eyeball that governs focusing (accommodation) of the lens. Pupillary disorders can be divided into two categories: disorders in which the size of one or both pupils is abnormal and disorders in which the shape of one or both pupils are abnormal. When the size of both pupils is affected, the pupils usually are symmetrically abnormal. When the pupils are different in size, the term applied is *anisocoria.* An abnormally dilated pupil is called *mydriasis. Miosis* refers to an abnormally constricted pupil. Figure 26.1 represents a flowchart approach to anisocoria.

FIGURE 26.1. Unequal pupils. *CNS,* central nervous system; *DTR,* deep tendon reflex; *EOM,* extraocular muscle movement; *SLE,* slitlamp examination; *III,* third cranial nerve.

PATHOPHYSIOLOGY

The first-order sympathetic neurons extend from the hypothalamus through the midbrain, pons, and medulla into the spinal cord. There, they synapse with the second-order neurons at the ciliospinal center of Budge-Waller, just before exiting the cord at roots C8–T2. The thoracic sympathetic trunk then travels over the apex of the lungs to the superior cervical ganglion, where synapses are made with the third-order neurons. Sympathetic innervation to the face departs from the superior cervical ganglion or at the bifurcation of the common carotid artery. Therefore, complete unilateral anhidrosis in association with unilateral miosis suggests damage to the second-order neurons or superior cervical ganglion. The third-order neurons travel with the internal carotid artery into the cranial vault, where the fibers gain access to the orbit via the nasociliary branch of the first division of the trigeminal nerve. They then travel through the ciliary ganglion in the orbit without synapse. Fibers extend to the iris dilator via the ciliary nerves. Disruption of sympathetic innervation anywhere along the course results in ipsilateral miosis (Horner syndrome) and often is accompanied by ptosis, enophthalmos, and ipsilateral anhidrosis.

Parasympathetic neurons originate in the Edinger-Westphal nuclei, located on the dorsal aspect of the third cranial nerve nucleus in the anterior dorsal mesencephalon at the level of the superior colliculus, ventral to the sylvian aqueduct. These neurons travel with the third cranial nerve, exiting the midbrain on its ventral aspect and passing between the posterior cerebral artery and the superior cerebellar arteries as they arise from the posterior communicating artery in the circle of Willis. The nerve then runs anteriorly and enters the cavernous sinus superiorly and laterally. Just before entering the posterior orbit through the superior orbital fissure, the third cranial nerve splits into a superior and an inferior division. The latter contains the parasympathetic fibers that then pass into the ciliary ganglion, where they synapse. Short ciliary nerves then carry the postsynaptic fibers to the pupillary dilator muscle and the ciliary muscle (behind the iris). The

ciliary muscle governs accommodation of the lens. Unilateral mydriasis can be caused by damage to the parasympathetic fibers anywhere along their course. With the exceptions noted next, however, it is distinctly unusual for the parasympathetic fibers to be damaged without other evidence of third cranial nerve palsy (a deficit in the ability of the eye to adduct, look upward, or look downward).

Local factors also can cause physical changes in the iris or in the surrounding structures. This may result in unilateral miosis or mydriasis.

EVALUATION AND DECISION

When a child with unequal pupils arrives in the emergency department (ED), the initial concern is cerebral herniation leading to compression and stretching of the third cranial nerve. A rapid neurologic assessment usually is sufficient to diagnose herniation because most patients will have a decreased level of consciousness, focal findings in addition to a dilated pupil, and abnormal vital signs. Once the physician is certain that increased intracranial pressure (ICP) is not present, a more careful evaluation is appropriate.

When testing pupillary size, it is essential that the patient be instructed to look at a distant target that does not involve reading letters or numbers. This prevents the eyes from needing to accommodate. Because the innervation for accommodation is the same as that for the pupillary sphincter, the accommodating patient also has reflex contraction of the pupils. Likewise, focusing on the examiner's face, or on any other near object that would stimulate convergence of the eyes toward each other, also stimulates pupillary constriction. This method of examination is not pertinent for assessing the pupils of a comatose patient. Crying or forced eyelid closure also may induce miosis.

ANISOCORIA

When evaluating the patient with anisocoria, the examiner must answer two critical questions: 1) Which pupil is abnormal, the smaller or the larger? and 2) Is this abnormality acute or chronic?

To establish which pupil is abnormal, the relative difference in pupillary size should be noted under conditions of bright illumination and dim illumination. With the largest diameter circle used on the direct ophthalmoscope or a bright penlight, both pupils should be illuminated simultaneously in a room with the lights on. The room lights should then be turned off and the handheld light source held tangentially from below or from above so that the eyes are illuminated only enough that the examiner can note the pupillary size.

Normally, the pupils constrict in response to bright illumination and dilate in dim illumination. If the relative difference in pupillary size increases under bright illumination, the larger pupil is the abnormal pupil: the larger pupil is not constricting normally (Fig. 26.2). If the relative difference in pupillary size increases under dim illumination, the smaller pupil is the abnormal pupil: the smaller pupil is not dilating normally. If the relative difference in pupillary size is the same in both dim and bright illumination, the patient does not have an abnormal pupil (Fig. 26.3). Rather, the patient has physiologic anisocoria. Approximately 20% of people with normal pupils have a difference in the size of their pupils in excess of 0.4 mm.

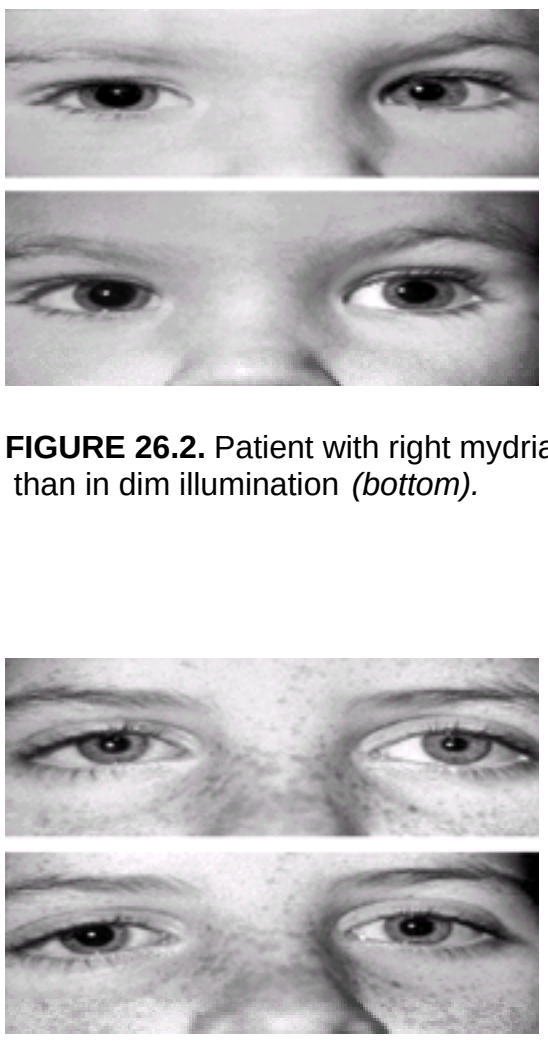

FIGURE 26.2. Patient with right mydriasis. Relative difference between the pupil size is greater in bright illumination *(top)* than in dim illumination *(bottom).*

FIGURE 26.3. Physiologic anisocoria. Relative difference in pupil size is the same in bright illumination *(top)* and dim illumination *(bottom).*

When trying to establish whether the anisocoria is of relatively recent or acute onset, as opposed to long-standing anisocoria, it is helpful to view old photographs. Sometimes, chronic physiologic anisocoria had not been noticed previously. The direct ophthalmoscope can be used to provide magnification of the photograph, as well as illumination. The focusing dial should be set on the highest black (or green) number and then the direct ophthalmoscope looked through at a distance that allows the photograph to be in focus. It also is important to note any other symptoms that accompanied the onset of anisocoria (headaches, eyeball pain, double vision, or blurred vision). The causes of anisocoria are summarized in Table 26.1, Table 26.2 and Table 26.3.

Physiologic anisocoria
Pharmacologic (miotics or mydriatics)
Local factors
Miosis: iritis, surgical trauma
Mydriasis: trauma
Abnormal pupil shape from scar formation following prior iritis or trauma
Neurologic causes
Miosis: Horner syndrome
Mydriasis: third cranial nerve palsy, Adie's pupil
Congenital abnormalities
Iris coloboma
Anterior chamber dysgenesis syndromes

[a]Not listed in order of frequency

Table 26.1. Differential Diagnosis of Unequal Pupils [a]

Physiologic anisocoria
Miosis
Iritis secondary to trauma, juvenile rheumatoid arthritis, or idiopathic
Abnormal pupil shape from scar formation following prior iritis or trauma
Horner syndrome (see Table 26.5)
Mydriasis
Trauma
Third cranial nerve palsy
Adie's pupil
Congenital abnormalities
Iris coloboma

[a]Not listed in order of frequency.

Table 26.2. Common Causes of Unequal Pupils [a]

Miosis
Intracranial mass lesion or vascular insult
Spinal cord tumor or compression
Intrathoracic tumor
Aneurysm
Cavernous sinus inflammation, thrombosis, or tumor
Mydriasis
Increased intracranial pressure
Intracranial mass lesion
Aneurysm
Cavernous sinus inflammation, thrombosis, or tumor
Orbital tumor

[a]Not listed in order of frequency.

Table 26.3. Life-Threatening Causes of Unequal Pupils [a]

MIOSIS

Local Factors

An irritated or inflamed iris sphincter muscle will result in miosis. Iritis, secondary to trauma or other factors, is a common cause. The eye usually is injected, and there are symptoms of eye pain, photophobia, tearing, and possibly, decreased vision. Injection may surround the cornea for 360 degrees, creating a ring of erythema (“ciliary blush”). More diffuse injection also may occur. Children with juvenile rheumatoid arthritis may not have these classic symptoms associated with their iritis; in fact, they may have no symptoms at all. Traumatic iritis is often not apparent for 12 to 72 hours after eye trauma. The diagnosis of iritis is confirmed by slitlamp biomicroscopy. This technique is described in Chapter 111. Ophthalmologic consultation is important for subsequent evaluation and treatment.

Other local factors include surgical irritation of the iris and pharmacologically induced unilateral miosis. Mechanical

contact with the iris during any intraocular surgical procedure may result in transient postoperative unilateral miosis.

Parasympathomimetic or sympatholytic drops also can result in transient miosis. A list of commonly used topical miotics is found in Table 26.4. These drops rarely are used in children, with the exception of their occasional bilateral use to treat crossed eyes or unilateral or bilateral use for glaucoma. Systemic drugs from the same categories may result in bilateral miosis. It is helpful to remember that most topical ophthalmic miotics are supplied in bottles that have green caps.

Generic Name	Trade Names
Cholinergics	
Pilocarpine	Adsorbocarpine, Akarpine, Almocarpine, Isopto Carpine, Miocarpine, Pilagan, Pilocar, Pilocel, Pilogel, Pilominin, Piloptic, Ocusert Pilo
Carbachol	Carbacel, Isopto Carbachol
Anticholinesterases	
Physostigmine	Eserine Sulfate, Isopto Eserine
Demecarium	Humorsol
Echothiopate iodide	Echodide, Phospholine Iodide
Isoflurophate (DFP)	Floropryl

Table 26.4. Topical Ophthalmic Miotics (Drops and Ointments)

Neurologic Factors

Miosis caused by damage to the sympathetic pathways (Horner syndrome) often is associated with ipsilateral mild ptosis, an upward displacement of the lower lid (“upside-down ptosis”), and the appearance of mild enophthalmos, with or without ipsilateral facial anhidrosis.

Congenital Horner syndrome may result from brachial plexus injury and often is associated with ipsilateral iris hypopigmentation. This sign is not helpful in the first few months of life.

More than 50% of children with congenital Horner syndrome have a history of difficult extraction at delivery. Congenital varicella infection may also be the cause. Knowledge of the sympathetic anatomy previously discussed can be exploited through the use of topically applied diagnostic agents to localize the site of the lesion. This testing is best performed by ophthalmologic or neurologic consultants. If the presence of Horner syndrome is questioned, one drop of topical 4% cocaine can be instilled into both eyes. Because cocaine prevents reuptake of norepinephrine at the terminal myoneural junction of the sphincter muscle, pupillary dilation will occur normally. Failure of the miotic pupil to dilate is diagnostic of Horner syndrome. Table 26.5 summarizes the causes of acquired Horner syndrome in children. All children who have Horner syndrome should receive a complete evaluation unless congenital Horner syndrome is present, based on history, old photographs, and examination.

First-order neuron
Brainstem glioma or other tumor
Brainstem vascular insult (aneurysm, infarct)
Spinal cord tumor
Syringomyelia
Poliomyelitis
Head or spinal trauma
Postsurgical
Second-order neuron
Intrathoracic tumor (neuroblastoma, ganglioneuroma, metastatic)
Intrathoracic aneurysm
Cervical tumor or adenitis
Trauma (especially brachial plexus trauma)
Postsurgical
Third-order neuron
Internal carotid thrombosis or aneurysm
Internal carotid or head trauma
Otitis media
Nasopharyngeal malignancy
Cavernous sinus thrombosis, tumor, or inflammation
Postsurgical

[a]Not listed in order of frequency

Table 26.5. Causes of Acquired Horner Syndrome in Children[a]

MYDRIASIS

Local Factors

Both trauma and topical agents can cause unilateral mydriasis. Blunt trauma (and, less commonly, intraocular surgery) can result in a fixed dilated pupil. Traumatic mydriasis usually occurs in a setting in which a clear history of trauma and other intraocular injuries, such as hyphema, is noted. The pupil may be somewhat irregular in shape if the sphincter is not damaged uniformly. Sometimes, pigment deposition can be seen on the anterior surface of the lens.

Topical parasympatholytics and sympathomimetics also can cause mydriasis. Systemic medications from the same classes can cause bilateral pupillary dilation. A list of topical agents is found in Table 26.6. Most topical mydriatics are supplied in bottles that have red caps. Pharmacologic mydriasis can be diagnosed by the instillation of pilocarpine 1% into both eyes. The pharmacologically dilated pupil will not constrict or will constrict only minimally.

Generic Name	Trade Names
Sympathomimetics	
Phenylephrine	Ak-Dilate, Ethicel, Mydfrin, Neo-Synephrine, Phenoptic
Cocaine (see text)[b]	
Parasympatholytics	
Atropine	Atropisol, Isopto Atropine, Ocu-Tropine
Cyclopentolate	Ak-Pentolate, Cyclogyl, Pentolair
Homatropine	Homatrocel, Isopto Homatropine
Scopolamine	Isopto Hyoscine, Mydramide
Tropicamide	Mydriacyl, Mydriafair, Tropicacyl

[a]Combination products may also be available.
[b]Diagnostic and anesthetic use only—not prescribed for outpatient use.

Table 26.6. Topical Ophthalmic Mydriatics[a] (Drops and Ointments)

Neurologic Factors

Unilateral mydriasis caused by disruption of parasympathetic innervation is often accompanied by other signs of third cranial nerve palsy: ptosis and/or abnormal eye muscle movements (see Chapter 25). Examination by an ophthalmologist often is indicated to help define patterns of extraocular muscle deficit and strabismus, as well as to assess for the possible presence of papilledema. Meningitis and increased ICP have been associated with mydriasis from third cranial nerve involvement without abnormalities of the extraocular muscles. In children, head trauma is the most common cause of acquired third cranial nerve palsy. Other causes are listed in Table 26.7. Neuroradiologic investigation is almost always indicated.

Head trauma
Congenital (isolated or with other cranial nerves involved)
Brain/meningeal tumor
Meningitis/encephalitis
Postviral syndromes
Hydrocephalus
Migraine
Cavernous sinus thrombosis
Aneurysm
Benign idiopathic ("cryptogenic")

[a]Not listed in order of frequency.

Table 26.7. Causes of Third Cranial Nerve Palsy in Children[a]

Other problems may mimic the eye muscle imbalance of third nerve palsy. For example, an inferior orbital wall blowout fracture (see Chapter 25) may cause a deficiency in the eye's ability to look up. Trauma also may result in unilateral mydriasis. Together, these findings may mimic a third cranial nerve palsy. This scenario underscores the need for ophthalmologic consultation when pathologic unilateral mydriasis and/or eye muscle deficits are present. Diagnostic clues to the presence of a long-standing third cranial nerve palsy include phenomena associated with aberrant regeneration of the oculomotor nerve. Examples include eyelid elevation when the patient looks down and pupillary constriction when the patient looks upward, downward, or into adduction.

Adie's pupil is most often unilateral. It is caused by parasympathetic denervation at the myoneural junction of the pupillary dilator muscle. It may be associated with deep tendon hyporeflexia. Also known as tonic pupil, an Adie's pupil constricts slowly to near convergence and then redilates deliberately. Slitlamp examination may reveal serpentine microundulations or asymmetries on constriction to light. Adie's pupil usually has an acute or subacute onset. It has been reported after trauma and viral illnesses, including varicella. The denervation is best demonstrated by instillation of weak pilocarpine (0.125 or 0.1%) into both eyes. This concentration is too weak to cause constriction of the normal pupil; however, the denervated Adie's pupil will become miotic. Acutely, this test may be falsely negative. Perhaps it is best that this test be left to the ophthalmologist so that complete ophthalmic examination can be conducted before pharmacologic alterations in pupil size are made.

A common misconception is that poorly seeing eyes have large pupils. Even an eye with very poor vision (light perception) or total blindness may have normal pupil size.

CORECTOPIA AND IRREGULAR PUPILS

Pupils can be unequal by virtue of irregularities in pupillary shape or position. Pupils that are located eccentrically (corectopia) rather than centrally usually are bilateral and represent congenital anomalies. When a unilateral pupil is located in a different position than its counterpart, however, this may indicate a progressive change in iris anatomy. Such changes may be associated with formation of other holes in the iris, abnormal iris strands that may become adherent to the cornea, or changes in iris color. These abnormalities also may be associated with glaucoma.

The direct ophthalmoscope can be helpful in identifying these rare anomalies. The focusing dial should be turned so that

the iris is in focus (less than 6 inches away from the patient). The dial will be turning in the direction of increasingly higher black (or green) numbers to provide increasing magnification with a shorter focal distance.

Pupils also may be irregular in shape despite having a central location. This often is caused by trauma or prior inflammation. After trauma, the presence of corectopia, or a teardrop-shaped pupil, is particularly ominous because this may indicate an underlying associated rupture of the eyeball (see Chapter 111). A history of iritis or undiagnosed recurrent ocular inflammation and pain may be associated with scar formation (posterior synechiae) between the edge of the pupil and the lens of the eye, which sits immediately behind the pupil. This can cause abnormal pupillary shapes.

Perhaps the most familiar disorder of pupillary shape and/or location is the congenital iris coloboma. This “keyhole” pupil (Fig. 26.4) represents a failure of proper embryologic development of the iris tissue. By itself, iris coloboma usually is asymptomatic and not associated with a functional deficit. However, associated colobomatous defects of the retina or optic nerve may exist, and these can result in serious visual compromise.

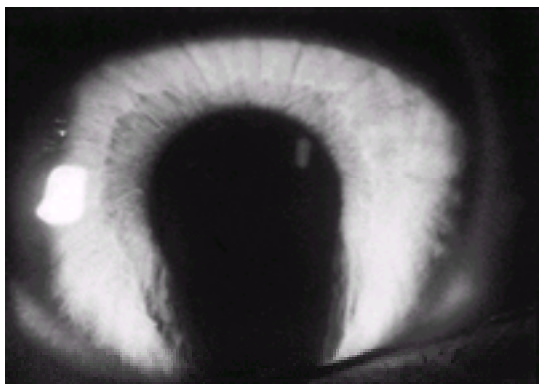

FIGURE 26.4. Iris coloboma creating a “keyhole” pupil.

It is wise to seek ophthalmologic consultation in all situations of unilateral corectopia or irregular pupillary shape. Occasionally, when dilating drops are instilled initially, the pupil may begin to dilate irregularly and asymmetrically. This is of no concern provided that the ultimate shape of the dilated pupil is round.

UNEQUAL PUPILLARY REACTIVITY

Both pupils should be equally brisk in their constricting reaction to a penlight (or direct ophthalmoscope light). When asymmetry in pupillary reactivity is found, it always is the more sluggish pupil that is abnormal. Often, the more sluggish pupil will be a unilaterally dilated pupil (for which the previous discussion applies). If both pupils are symmetric in their baseline positions, an abnormally sluggish pupil may indicate the presence of a serious retinal or optic nerve problem that is impairing the ability of the affected eye to perceive the light source equally. Testing visual acuity is essential under these circumstances.

The pupil should not be pharmacologically manipulated in the ED. Rather, direct referral to an ophthalmologist is appropriate so that the pupils may be observed unaltered.

Suggested Readings

Brazis PW. Localization of lesions of the oculomotor nerve: recent concepts. Mayo Clin Proc 1991;66:1029–1035.

Burde RM, Savino PJ, Troobe JD. Clinical Decision in Neuro-Ophthalmology. Princeton: CV Mosby, 1985;221–245.

Hamed LM. Associated neurologic and ophthalmologic findings in congenital oculomotor nerve palsy. Ophthalmology 1991;98:708–714.

Jeffery AR, Ellis FJ, Repka MX, Buncic JR. Pediatric Horner syndrome. JAAPOS 1998;2:159–167.

Miller NR. Solitary oculomotor nerve palsy in childhood. Am J Ophthalmol 1977;83:106–111.

Sauer C, Levinsohn MW. Horner's syndrome in childhood. Neurology 1976;26:216–220.

Thompson HS, Pilley SF. Unequal pupils: a flow chart for sorting out the anisocorias. Surv Ophthalmol 1976;21:45–48.

Young TA, Levin AV. The afferent pupillary defect. Pediatr Emerg Care 1997;13:61–65.

CHAPTER 27

Eye—Visual Disturbances

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Sudden loss or deterioration of vision (or diplopia) can be caused by numerous diseases and injuries (Table 27.1, Table 27.2 and Table 27.3). Although most cases are rare in the pediatric patient, a systematic approach is necessary to reach a correct diagnosis and to minimize permanent visual impairment. The patient's age, underlying disease conditions, visual history, and history of possible injury must be determined. It is important to remember that up to 5% of abused children present with ocular injuries and 40% of abused children will have ocular findings. The recent increase in survival of extremely low–birth weight infants has led to many children with varying degrees of visual disturbances that may not be readily noticed. The extent of the visual impairment and the rapidity of its onset also are vital pieces of information. A careful eye examination, including gross and ophthalmoscopic examination, determination of extraocular movement, and visual acuity, together with the history, leads to correct diagnosis and management of the patient (see Chapter 120).

	Traumatic	Nontraumatic
Periorbital	Eyelid hematoma, edema from trauma	Orbital or periorbital cellulitis, tumor, allergic edema
Cornea and conjunctiva	Chemical burns, thermal burns, ultraviolet or infrared burns, laceration of cornea	Conjunctivitis (bacterial, viral, fungal)
Anterior chamber	Traumatic iritis, hyphema, posttraumatic cataract, dislocation of lens, glaucoma	Acute iritis, glaucoma, uveitis
Posterior chamber	Vitreous hemorrhage	Endophthalmitis
Retina	Severed retinal artery, retinal tears or detachment, commotio retinae	Retinal vein or artery obstruction, spontaneous
Cortex	Head trauma	Optic neuritis, toxins, hysteria, hypoglycemia, melanoma, cerebrovascular accidents, migraine
Other	Carotid artery trauma	Poisoning Shunt malfunction Vitamin A deficiencies Measles

Table 27.1. Causes of Acute Visual Disturbances

Trauma	Rupture of globes
Chemical burns	Periorbital infection
Hyphema	Conjunctivitis

Table 27.2. Common Conditions That Cause Acute Visual Disturbances

Blowout fractures	Exercise
Poisoning	Arnold-Chiari malformation
Central nervous system pathology (tumor, bleed)	Myasthenia gravis
Shunt malfunction	Head trauma

Table 27.3. Causes of Acute Diplopla

Few ocular conditions in the pediatric population are truly emergent (Table 27.4), but many are urgent; most can be treated by the emergency physician or can be referred for appropriate follow-up with an ophthalmologist. Many conditions that a pediatric ophthalmologist sees are not discussed here because they rarely are seen in the emergency department (ED). Such conditions include congenital eye disorders and amblyopia. Sudden onset of diplopia may be secondary to many of the conditions listed, either as a direct effect of trauma, infection, central nervous system (CNS) pathology (tumor or shunt malfunction), or hysterical reaction. Likewise, head tilt may represent a visual disturbance and requires a complete ophthalmologic evaluation, although the condition also may be caused by musculoskeletal problems in the neck. Conditions that are more likely to be seen in the ED are emphasized in this chapter.

Alkali or acid burns
Central retinal artery occlusion

Table 27.4. Emergent Conditions That Cause Visual Disturbances

PATHOPHYSIOLOGY

Vision may be impaired through interference at any point in the visual pathway. Light must reach the eye, pass through the cornea and the anterior chamber, be focused by the lens, pass through the posterior chamber, and reach the retina. The retina must react to the visual stimuli, generate impulses, and pass these impulses along the optic nerve and eventually to the visual cortex for interpretation. In addition, for binocular vision, the movement of both eyes must be coordinated and smooth. If any step in this pathway is interrupted or damaged, visual impairment will occur. Loss of clarity of the visual media or damage to the conductive tissues leads to decreased vision.

DIFFERENTIAL DIAGNOSIS

Trauma and infections are the two most common causes of acute visual impairment in otherwise healthy children seen in the ED, and these two processes can interfere with any part or all of the visual pathway (Table 27.1 and Table 27.2). Children with shunts for hydrocephalus may have a variety of visual disturbances—from complete blindness to transient diplopia. Any child with an acute visual disturbance and a shunt must have the functioning of the shunt evaluated. The total spectrum of diseases that cause visual impairment can be understood best if the visual pathway is divided into its parts, and each part is considered sequentially (Table 27.1).

Vision may be limited by periorbital diseases such as orbital cellulitis, tumor, or infection or swelling of the eyelids. The history and physical examination usually make these diagnoses obvious.

Blunt trauma to the eye may cause a blowout fracture of the orbit. The weakest portion of the orbit, the floor, most commonly breaks, and this may entrap the extraocular muscles. Visual impairment may be limited to double vision when looking in a certain direction, particularly upward. Testing the extraocular movements reveals the limitation. Careful inspection of the globe also is necessary.

Diseases of the cornea that cause visual impairment are predominantly infectious or traumatic. Infections of the cornea and conjunctiva can be caused by bacteria, viruses, and fungi (see Chapter 24 and Chapter 120). All of these diseases may present as a unilateral or bilateral process, usually affecting only the conjunctiva and cornea. Onset is variable but usually occurs over 1 or 2 days, and vision is not greatly impaired. In the newborn period, gonococcal and chlamydial infections must be considered. With a recent eye injury or foreign body intrusion, fungal infections are possible. In the United States, the most common corneal infection that causes permanent visual impairment is herpes simplex, whereas trachoma is the most common cause worldwide. A careful ophthalmoscopic or slitlamp examination will reveal the

characteristic dendritic ulcers of herpes simplex infection after the eye has been stained with fluorescein. Unless this disease is excluded, steroid-containing medications should not be used. Herpes simplex infection may be recurrent, so if a child with a history of previous herpes simplex keratitis complains of a red eye on the previously infected side, recurrent herpes infection must be considered.

Traumatic injuries to the cornea include one of the true ophthalmologic emergencies: alkali burns. Alkali burns in general carry a worse prognosis than acid burns. Rapid treatment of this condition is imperative to prevent permanent visual impairment. The cause of the chemical injury usually is obvious from the history. Self-inflicted thermal injuries of the cornea from curling irons are increasing and should be sought in a child who has facial burns. Both ultraviolet and infrared light can cause damage to the cornea, resulting in severe pain and photophobia within 24 hours of exposure. Lacerations with perforation of the cornea usually affect other parts of the eye as well and can lead to significant visual impairment. Careful inspection of the globe with associated lid trauma is mandatory.

The anterior chamber of the eye consists of the aqueous humor, the iris, and the lens. Acute iritis is rare in children, and the cause often is uncertain. There is a sudden onset of pain, redness, and photophobia that usually affects one eye only. The degree of visual impairment varies with the severity of the inflammation. Certain diseases have associated iritis, such as juvenile rheumatoid arthritis, but these seldom are seen in the ED. Blunt trauma also can cause iritis, but vision is only slightly impaired unless other structures are involved. Trauma also can cause a hyphema or hemorrhage into the anterior chamber. This can result in little to severe visual impairment in the affected eye, depending on the extent of bleeding and associated trauma. Traumatic injuries can lead to cataract formation, usually within a few days of injury, but onset may be delayed for years. Dislocation of the lens after trauma causes significant visual impairment but can be recognized easily with a careful examination. Glaucoma and a retinal detachment may be late complications of blunt trauma. Congenital glaucoma is a major preventable cause of blindness in children; most cases manifest within the first 6 months of life and occasionally present to the ED. Corneal clouding, buphthalmos, or asymmetry in eye size may be the chief complaint. If any one of these is noted as a primary complaint or an incidental finding, immediate referral is required.

Injury or infection that involves the anterior chamber may lead to increased intraocular pressure. Glaucoma may become evident days to years after the initial trauma or infection. Pain around the eye, blurred vision, and occasionally, nausea and vomiting in a patient with glaucoma or with a recent eye injury may represent an acute attack of glaucoma. The diagnosis is made easily with tonometry.

The uvea consists of the iris, ciliary body, and choroid. One or all portions of the uvea may become inflamed, causing uveitis. Iritis and iridocyclitis may be called anterior uveitis, whereas inflammation of the choroid is often called posterior uveitis. The etiologies may be divided into infectious and noninfectious. Infectious uveitis may be caused by viruses, bacteria, fungi, or helminths. The most common cause of posterior uveitis in children is toxoplasmosis with *Toxocara canis* second. Noninfectious causes include sarcoidosis and sympathetic ophthalmia. Measles, mumps, and pertussis may be associated with uveitis that is not the result of invasion by the agent causing the infection. Vogt-Koyanagi-Harada syndrome is a panuveitis with meningeal and cutaneous findings. Prompt treatment of this syndrome is necessary for optimal visual outcome.

In addition to blurred vision in one or both eyes, anterior uveitis also is associated with pain in the affected eye, headache, photophobia, and conjunctival injection. Anterior uveitis may be confused with conjunctivitis or an acute attack of glaucoma. In posterior uveitis, the pain and photophobia may be less pronounced, but there may be a more pronounced visual impairment.

The posterior chamber is composed of the vitreous humor. The vitreous gel usually is clear, and any diseases that affect the clarity will impair vision. Certain chronic conditions such as uveitis can cause deposits in the vitreous humor, but the visual impairment is very gradual. Infections inside the eye (endophthalmitis) usually result from a penetrating injury, surgery, or an extension of a more superficial infection. Bacterial infections develop more rapidly than do fungal infections. The child will have severe pain in or around the eye and, with bacterial infections especially, may have fever and leukocytosis. The process usually is unilateral, and vision is severely compromised. Purulent exudate is formed in the vitreous humor, and ophthalmoscopic examination may reveal a greenish color with the details of the retina lost. A hypopyon—accumulation of pus in the anterior chamber—usually is present.

Either penetrating or blunt trauma (see Chapter 111) to the eye can lead to vitreous hemorrhage, but this is uncommon in children. Diabetes mellitus, hypertension, sickle cell disease, and leukemia may cause vitreous hemorrhage as well as retinal tears, central retinal vein occlusion, and tumor. There is a sudden loss or deterioration of vision in the affected eye. Findings on examination depend on the degree of hemorrhage. Blood clots may be visible with the ophthalmoscope, or the fundus reflex may be black, obscuring the retina in more severe cases.

Retinal vein and artery obstruction also are uncommon in pediatric patients. With central retinal artery occlusion, there is a sudden, painless, total loss of vision in one eye. If only a branch is occluded, a field loss will result. Ophthalmoscopic examination reveals the cherry-red spot of the fovea, the optic nerve appears pale white, and the arteries are narrowed significantly. A Marcus-Gunn pupil (relative afferent defect) may be present and may be diagnosed by shining a light in one eye, then in the other. When the light is shone in the normal eye, both pupils will constrict. When light is shone in the damaged eye, the pupil will dilate.

The retinal artery may be severed by trauma or obstructed by emboli, as in a patient with endocardial thrombi or arterial obstructions in systemic lupus erythematosus (SLE) and in diseases with hypercoagulability, such as sickle cell disease. The arterial spasm associated with migraine may lead to retinal artery obstruction.

As with retinal artery occlusion, retinal vein occlusion causes a painless loss of vision. Visual loss may be severe, with total occlusion of the central retinal vein, or less pronounced, with branch obstruction. Examination of the retina reveals multiple hemorrhages with a blurred, reddened optic disc. The arteries are narrowed, the veins engorged, and patchy

white exudates may be evident. These findings will be limited to one area in branch occlusion. Retinal vein obstruction, although rare, may occur with trauma or diseases such as leukemia, cystic fibrosis, or retinal phlebitis.

As mentioned, a tear in the retina may lead to vitreous hemorrhage, causing decreased vision in the affected eye. If the tear is in the macula, the visual loss will be severe. A tear in the retina may not cause immediate visual impairment. Retinal detachment from a retinal tear may be delayed for years. The visual impairment may go unnoticed if the detachment is peripheral. As the detachment progresses or when it involves more central areas, the patient will complain of cloudy vision with lightning flashes (photopsia). This may be followed by a shadow or curtain in the visual field. Visual acuity may remain normal if the macula is not involved. Examination of the eye will reveal a lighter appearing retina in the area of detachment, and it may have folds. Flashing lights or visual field defects, after trauma, should raise the suspicion of retinal detachment. Retinoschisis, splitting of the layers of the retina, may be seen in the shaken baby syndrome.

Commotio retinae, or Berlin's edema—that is, edema of the retina—may follow blunt ocular trauma by 24 hours. The visual loss is variable, and the retina will appear pale gray because of the edema, but the macula usually is spared.

The optic nerve transmits visual signals to the cortex. Optic neuritis is involvement of the optic nerve by inflammation or demyelinization. The process usually is acute and may be unilateral or bilateral. Loss of vision may take from hours to days, and visual impairment ranges from mild loss to complete blindness. Patients often complain of disturbance of color vision. Pain may be absent or present on movement of the eye or palpation of the globe. It rarely is an isolated event in children. Causes include meningitis, viral infections, immunizations, encephalomyelitis, Lyme disease, and demyelinating diseases. Exogenous toxins and drugs (e.g., lead poisoning, long-term chloramphenicol treatment) also may cause optic neuritis.

Various toxins are capable of causing impaired vision. The loss may be gradual or sudden, depending on the particular toxin. Toxins usually act on ganglion cells of the retina or on optic nerve fibers, causing contraction of the peripheral field, central visual defect, or a combination. Methyl alcohol, when ingested, may cause bilateral sudden blindness, which may be complete and permanent or may have a more gradual onset. With methyl alcohol ingestion, associated symptoms include nausea, vomiting, abdominal pain, headache, dizziness, delirium, and convulsions. Other toxins include halogenated hydrocarbons, sulfanilamide, quinine, and quinidine. Large doses of salicylates may cause amblyopia. Digitalis may cause transient amblyopia, visual blurring, or the perception of yellow halos around light (xanthopsia).

Visual impairment also may result from interference with the visual cortex of the brain. Cortical blindness has many causes (Table 27.5). Head trauma (see Chapter 38 and Chapter 105) may cause total loss of vision soon after the event. This has been called “footballer's migraine” because of its association with head trauma in soccer. Even trivial head trauma has been known to cause blindness. The apparent hysterical reaction that follows head trauma, especially in young children, may represent complete blindness in a child unable to express the problem or who is too frightened by the experience. The physical examination may be completely normal. There may be a delay of onset, but the entire course is usually brief, lasting minutes to hours. This form of blindness often is confused with hysterical blindness, the latter being a diagnosis of exclusion. Monocular blindness may be caused by trauma to the carotid artery on the affected side.

Cardiac arrest	Hydrocephalus
Status epilepticus	Shunt malfunction
Hypoxia	Head trauma
Perinatal asphyxia	Cardiac surgery
Cerebral infarction	Cerebral or vertebral angiography
Meningitis	Drugs (steroids)
Encephalitis	Carbon monoxide poisoning
Subacute sclerosing leukoencephalitis	Occipital epilepsy
Hypoglycemia	Postictal states
Uremia	

Table 27.5. Causes of Cortical Blindness

Migraine headaches are a common cause of bilateral field loss (hemianopsia) in adolescents. This field loss usually lasts less than 1 hour. It almost always is misidentified by the patient as loss of vision in only one eye. Headache and nausea after such an episode are the rule but occasionally may be absent.

EVALUATION AND DECISION

The absolute ophthalmologic emergencies are alkali burns, a ruptured globe, and retinal artery occlusion. The diagnosis of the first is by history, and therapy must be initiated promptly to minimize the damage to the eye. If there is any doubt about the actual substance to which the eyes have been exposed, treatment for an alkali burn is always prudent. A ruptured globe must be suspected with any possible penetrating injury to the eye. The injury may be subtle, and the vision may be normal. If a possibility of a ruptured globe exists, the eye should be protected and the patient should have immediate evaluation by an ophthalmologist. Retinal artery thrombosis is rare in children, but it should be suspected when there is sudden, unilateral painless loss of vision and a predisposing condition. Predisposing conditions include those associated with emboli, such as endocardial thrombi or amniotic fluid; conditions with arteritis leading to obstruction, as in SLE; disease states associated with hypercoagulability, such as sickling hemoglobinopathies; and conditions with arterial spasm, such as severe hypertension.

If alkali burns, a ruptured globe, and retinal artery occlusion can be excluded, the patient may be evaluated more carefully before instituting therapy. When visual acuity is being determined, the history may be obtained. Significant historical information includes episodes of recent trauma, unilateral or bilateral nature of the loss, and association of pain in or around the eye (Fig. 27.1). Child abuse may present with any of the variety of traumatic injuries. Retinal hemorrhages in a child are almost always caused by intentional trauma. Most children seen in the ED will have a traumatic or infectious process.

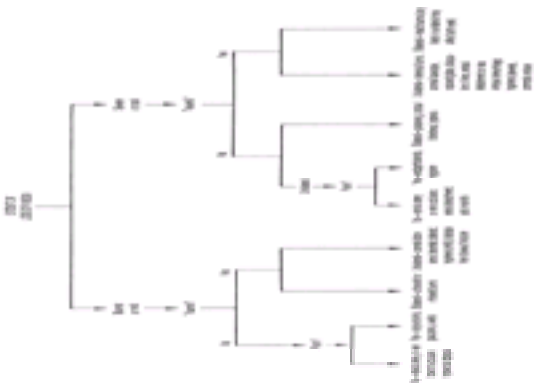

FIGURE 27.1. Diagnostic approach to visual disturbances.

Severe Visual Loss Associated with Trauma

Severe bilateral visual loss associated with trauma is the result of head trauma, causing cortical blindness. This condition usually is totally reversible in less than a few hours.

Any of the traumatic injuries that cause severe unilateral loss of vision may cause bilateral loss if both eyes are involved. The mechanism of injury should be elicited. If there is any possibility of a penetrating injury or rupture of the globe, the involved eye should be protected from further damage by shielding until careful examination can be performed by a skilled physician. If the globe is intact and no penetration by a foreign body occurred, an ophthalmoscopic or slitlamp examination usually leads to the correct diagnosis. These conditions include chemical burns of the cornea, hyphema, dislocation of the lens, vitreous hemorrhage, detachment or tear of the retina, and commotio retinae.

Severe Visual Loss Not Associated with Trauma

With severe bilateral visual loss not associated with trauma, the possibility of toxins must be explored. Also, cortical blindness may cause a similar picture, but this is rare and generally associated with another problem, such as hypoglycemia, leukemia, and cerebrovascular or anesthetic accidents. If severe visual loss is unilateral and painful, endophthalmitis must be suspected, but once again, such loss usually is the result of a previous penetrating injury or an extension of a local infectious process. If a headache is associated with the visual loss, migraine may be implicated. If the severe loss is unilateral and painless, retinal artery or vein occlusion or retinal detachment may be diagnosed by ophthalmoscopic examination. Optic neuritis will also present this way.

Mild Visual Loss with Trauma

If the visual loss is unilateral, not severe, and if trauma recently occurred, corneal abrasions, traumatic cataracts, and small hyphemas should be sought. A blowout fracture may cause diplopia, but if each eye is examined individually, the visual acuity should be normal. If the process is bilateral, exposure to ultraviolet or infrared light should be considered.

Mild Visual Loss without Trauma

When the visual loss is mild and nontraumatic, and if the process is unilateral and painful, conjunctivitis, uveitis, and acute attacks of glaucoma are possible. If the process is painless, retinal vein or artery branch occlusion may be suspected. Any of these processes may be bilateral as well.

Suggested Readings

Andrews AP, Quillen. Ocular manifestations of child abuse. Pennsylvania Medicine, Supplement, March 1996.

Beauchamp GR. Cause of visual impairment in children. Pediatr Ann 1980;9:414–418.

Bradley WG, Whitty CWM. Acute optic neuritis: its clinical features and their relation to prognosis for recovery of vision. J Neurol Neurosurg Psychiatry 1967;30:531–538.

Burgess P, Johnson A. Ocular defects in infants of extremely low birth weight and low gestational age. Br J Ophthalmol 1991;75:84–87.

Cedzich C, Schramm J, Wenzel D. Reversible visual loss after shunt malfunction. Acta Neurochir (Wien) 1990;105:121–123.

Chew E, Morin JD. Glaucoma in children. Pediatr Clin North Am 1983;30:1043–1059.

Datner EM, Jolly BT. Pediatric ophthalmology. Emerg Clin North Am 1995;13:669–679.

Foster A. Childhood blindness. Eye 1988;2:27–36.

Franks W, Taylor D. Congenital glaucoma—a preventable cause of blindness. Arch Dis Child 1989;64:649–650.

Frey T. External diseases of the eye. Pediatr Ann 1977;1:23–40.

Gayle MO, Kissoon N, Hered RW, Harwood-Nuss A. Retinal hemorrhage in the young child: a review of etiology, predisposed conditions, and clinical implications. J Emerg Med 1995;13:233–239.

Gilbert JA, Pollack ES, Pollack CV. Vogt-Koyanagi-Harada syndrome: case report and review. J Emerg Med 1994;12:615–619.

Gombos GM. Handbook of Ophthalmologic Emergencies. Flushing, NY: Medical Examination Publishing, 1973.

Harley RD. Pediatric Ophthalmology. 2nd ed. Philadelphia: WB Saunders, 1983.

Harrison DW, Walls RM. Blindness following minor head trauma in children: a report of two cases with a review of the literature. J Emerg Med 1990;6:21–24.

Holmes GL. Prolonged cortical blindness after closed head trauma. South Med J 1978;71:612–613.

Huber M, Poon C, Buchanan N. Acute glomerulonephritis presenting with acute blindness. Med J Aust 1981;5:595.

Kattan JC, Dagi TF. Compensatory head tilt in upbeating nystagmus. J Clin Neuro-ophthalmol 1990;10:27–31.

King MA, Barkovich AJ, Halbach VV, et al. Traumatic monocular blindness and associated carotid injuries. Pediatrics 1989;84:128–132.

Kompf D, Neundorfer B, Ehret W, Wallescl CW. Transitory impairment of vision after light head trauma in childhood. Neuropediatrics 1977;8: 354–359.

Kwartz B, Leatherbarrow B, Davis H. Diplopia following head injury. Injury 1990;21:351–352.

Mukamel M, Weitz R, Nissenkorn I, Yassor I, Varsano I. Acute cortical blindness associated with hypoglycemia. J Pediatr 1981;98:583–584.

Oesterle CS. Exercise induced esotropia. J Pediatr Ophthalmol Strabismus 1989;26:150–151.

Oglesby R. Eye trauma in children. Pediatr Ann 1977;1:5–22.

Weaver FA, Wagner WN, Yellin AE, Cohen JL. Monocular blindness after penetrating trauma to the carotid artery. J Vasc Surg 1989;10:89–92.

Williams JR. Optic neuritis in a child. Pediatr Emerg Care 1996;12(3): 210–212.

Wong VC. Cortical blindness in children: a study of etiology and prognosis. Pediatr Neurol 1990;7:179–185.

Woodward G. Posttraumatic cortical blindness: are we missing the diagnosis in children? Pediatr Emerg Care 1990;6:289–292.

CHAPTER 28

Fever

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Fever, the abnormal elevation of body temperature, has been recognized for centuries by physicians as a sign of disease. Furthermore, the problem of the febrile child is one of the most commonly encountered in clinical pediatrics, accounting for as many as 20% of pediatric emergency department (ED) visits. Despite such considerable clinical experience, only recently has much progress been made in our understanding of the pathogenesis of fever. The problem of appropriate clinical and laboratory evaluation of febrile children, however, remains a major challenge to the pediatrician and emergency physician. The approach outlined in this chapter helps the physician treat a febrile child in the ED and proceed systematically with the appropriate diagnostic steps and the institution of therapy. The principal causes of fever in children are listed in Table 28.1.

Table 28.1. Principal Conditions in Children Associated with Fever

PATHOPHYSIOLOGY

Fever is a complex process, involving the highly coordinated interplay of autonomic, neuroendocrine, and behavioral responses to a variety of infectious and noninfectious inflammatory challenges. It is believed to represent an adaptive response to such threats and is manifested by nearly all vertebrate species. The febrile reaction is relatively stereotyped and independent of precise causation. Exogenous pyrogens (e.g., toxins, infectious agents, antigen–antibody complexes) from many sources produce fever in humans by inducing the production of proteins, collectively termed endogenous pyrogens, by phagocytic leukocytes. These are now identified as large proteins, with the average size being 15,000 to 30,000 daltons, and include interleukin-1, tumor necrosis factor-a, and several interferons. These proteins enter the circulation after their synthesis and interact with specialized receptor neurons in the organum vasculosum of the anterior hypothalamus, one of the “circumventricular organs” of the brain, which are now understood to have no blood–brain barrier and to function as neurohumoral receptors for blood borne hormones. Signaling at this site leads to the production of prostaglandins, particularly PGE_2, monoamines, and probably cyclic adenosine monophosphate. These mediators in turn initiate signals that reset the thermostatic set point in the hypothalamus and result in several responses. The principal effect is on the vasomotor center and results in peripheral vasoconstriction of cutaneous beds with redirection of blood flow to deeper tissues, thus minimizing skin heat loss. In addition, sweating is decreased; vasopressin secretion falls, which results in lowered extracellular fluid volume that requires heating; and behavioral adjustments such as shivering and seeking a warmer environment are stimulated. These effects all combine to elevate body temperature. There is some evidence that increased body temperature impairs replication of many microbes and may aid phagocytic bactericidal activity. The febrile response includes further adaptive neuroendocrine effects. Glucose metabolism is curtailed in favor of that based on lipolysis and proteolysis, thus depriving bacteria of their preferred substrate. Fever-induced anorexia further diminishes glucose availability to microbes. Hepatic production of acute-phase reactant proteins may result in binding of divalent cations, which are also growth factors for microorganisms. All of these effects combine to further enhance the host's response to microbial invasion. Very rarely, central nervous system (CNS) dysfunction (e.g., hypothalamic tumor, infarction) alters the thermostatic set point directly.

It is difficult to pinpoint the lowest temperature elevation considered to be definitely abnormal for all children under all circumstances. Some children normally have rectal temperatures as low as 36.2°C (97°F) or as high as 38°C (100.4°F).

Children, like adults, also have diurnal variations in temperature, with the peak usually occurring between 5 PM and 7 PM. This variation is less pronounced in infants. In the 2- to 6-year age range, the temperature may vary by 0.9°C (33.6°F), and in children over 6 years of age, diurnal variation may span 1.1°C (34°F). Factors such as excessive clothing, physical activity, hot weather, digestion of food, and ovulation can raise temperature in the absence of disease. For the appropriately dressed child who has been at rest 30 minutes, a rectal temperature of 38°C (100.4°F) is defined as fever for this discussion. Using the proper technique to record rectal temperature is important for optimal accuracy. Proper technique includes proper positioning and restraint in infants (prone, supine, or on the side with hips slightly flexed), depth of insertion (about 2 to 3 cm), and time for equilibration (2 to 3 minutes with glass thermometers, several seconds with electronic digital probes). The thermometer should not be placed directly into a fecal mass because the temperature may not have equilibrated with rapid fluctuations in core temperature and thus may be falsely low as temperature rises rapidly. Oral and axillary temperatures usually are about 0.6°C (33.1°F) and 1.1°C (34°F) lower than rectal temperatures, respectively. Recent attempts to measure temperature with a less invasive technique include temperature-sensitive pacifiers and forehead strips, both of which have been found to be unreliable in young children. However, one new technique that has been found to be acceptable to parents and reliable in most settings is that of infrared tympanic membrane thermometry. The tympanic membrane shares vascular supply with the hypothalamus, and it has been shown in adult intensive care settings that there is excellent correlation between core temperature (e.g., pulmonary artery catheter) and tympanic membrane readings. Several studies in children have tended to confirm the reliability of this technique compared with rectal temperature, although others have questioned the reliability of tympanic measurements in young infants, especially those less than 3 months of age. The presence of otitis media (OM) or cerumen does not seem to affect reliability adversely. Because even low-grade fever may be clinically significant in young infants and there is at least some doubt about the reliability of tympanic measurements in this age group, it would seem prudent to rely on rectal temperatures in this population.

EVALUATION AND DECISION

The importance of fever lies in its role as a sign of disease. The physicians caring for a febrile child should concentrate on discovering the cause of the fever and treating the underlying illness. Any fever may signify serious infection; however, severe hyperpyrexia, defined as a temperature of 41.1°C (106°F) or higher, is more often associated with diagnoses of pneumonia, bacteremia, or meningitis. The magnitude of reduction of a fever in response to antipyretics does not distinguish children with serious bacterial illnesses from those with viral diseases. If no specific treatment for the determined diagnosis is necessary, the physician's goal is then to provide appropriate supportive care and follow-up. Because many parents have “fever phobia,” instructions that explain the importance of fever as an indicator of disease, not as an inherently harmful entity, should be given.

A complete history and physical examination will provide most important clues in determining the diagnosis of children with febrile illnesses. The general impression obtained in the first few moments of an evaluation is extremely important in the recognition of potentially life-threatening causes of fever (Table 28.2). A great deal of information can be attained by visual assessment of the child while in the arms or lap of his or her parent. The severity of the illness may become apparent if the child is agitated or uninterested in the surroundings while in this comfortable, safe position. If the child appears nontoxic, observation of the child while the history of the present illness is being discussed with the parent may provide further insight into the diagnosis. Fever has different management implications for distinct subsets of children. Therefore, a clear understanding of the degree, mode of measurement, and duration of fever is especially important in the initial evaluation. The physician should ask questions concerning associated signs and symptoms, medications being given (including antipyretics and antibiotics), presence of ill contacts, travel history, and pet or insect exposures. The medical history should focus on recurrent febrile illnesses and the presence of any diseases or drug regimens that would compromise normal host defenses, such as sickle cell anemia, asplenia (functional, congenital, or surgical), malignancy (noting particularly chemotherapeutic or radiation treatments), human immunodeficiency virus (HIV), renal disease, prolonged steroid use, or indwelling catheters or ventriculoperitoneal shunts. Immunization status should also be determined. An understanding of prior evaluation and treatments during this illness may be helpful.

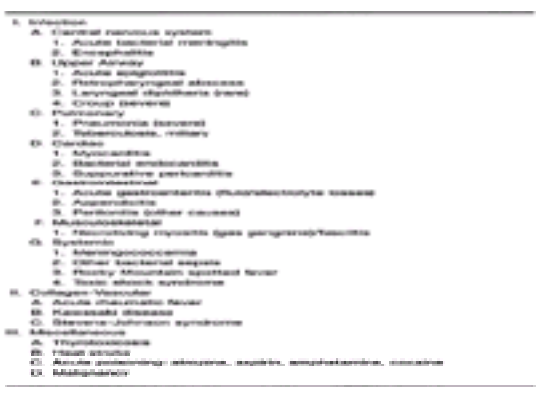

Table 28.2. Life-Threatening Acute Febrile Illnesses

As stated previously, the physical examination of the young febrile patient begins during the historical interview with the caregiver. The physician should note the child's alertness, responsiveness to persons and objects, work of breathing, color, feeding activity, and age-related appropriateness of social interaction and gross motor functions. If a child is noted to be playing with toys or smiling at his or her parent, the febrile illness is most likely not immediately life-threatening. However, the febrile infant who appears irritable and/or lethargic while being held by a parent before the examination has high probability of having a serious infection such as meningitis or sepsis. The complaint or observation that a child's crying increases with parental attempts to comfort is critical because “paradoxical irritability” is an important sign of

meningitis in infancy.

Other signs of severe or life-threatening infections heralded by fever should be sought early in the examination. CNS infections may be marked by fever with altered sensorium, convulsion, meningismus, or focal neurologic deficits. However, infants younger than 2 years of age with meningitis often do not have meningismus, but they may instead have irritability, anorexia, lethargy, vomiting, or bulging fontanelle. Severe upper airway infections may present with stridor, excessive drooling, and tripod positioning. A child with pneumonia, pericarditis, endocarditis, or sepsis syndrome may display dyspnea or tachypnea, cyanosis or pallor, tachycardia, and hypotension, as well as altered mental status. Hemorrhagic rashes may signal bacterial or rickettsial infections such as meningococcemia or Rocky Mountain spotted fever.

Although the index of suspicion for serious febrile illness must be high throughout the evaluation of each child, most childhood illnesses with fever are minor and self-limiting. Once the physician has ascertained that the child is not in immediate danger, the examination should focus on sites of common pediatric infections, including the ears, nose, and throat; cervical lymph nodes; respiratory, gastrointestinal, and genitourinary tracts; and skin, joints, and skeletal system (Table 28.3). Evaluation of each child is developed upon an understanding of the common infectious entities that affect that child's age group and the presenting signs and symptoms or lack thereof in each infectious entity (see Chapter 84 for a full discussion of infectious diseases).

Table 28.3. Common Causes of Fever

Many febrile exanthems are characteristic enough to be diagnostic (see Chapter 62, Chapter 63, Chapter 64, Chapter 65, Chapter 67, and Chapter 84). Varicella, rubeola, scarlet fever, and Coxsackie virus can all be identified by their pathognomonic rashes. However, if a child with chickenpox presents several days into the illness with a new fever, the possibility of group A b-hemolytic streptococcal coinfection needs to be fully evaluated. Children with fever and petechiae may have invasive meningococcal disease, disseminated streptococcal infection, or Rocky Mountain spotted fever. However, they may simply have a less serious viral infection or streptococcal pharyngitis. Differentiation of these entities is crucial and is based on clinical appearance of the patient and laboratory evaluation. A child with petechiae only above the nipple line, normal white blood cell (WBC) count, and well appearance is less likely to have invasive disease. However, any child who appears ill, has a laboratory abnormality, or has progressive petechial rash needs a full evaluation, including lumbar puncture and antibiotic administration.

On physical examination, acute OM is identified by changes in the tympanic membranes, such as redness, bulging, decreased mobility, loss of landmarks and light reflex, and purulent drainage from a perforation. Careful examination of the head and neck may reveal rhinorrhea and signs of inflammation, indicating a viral upper respiratory infection (URI). The oropharynx may reveal findings suggestive of acute pharyngitis or stomatitis (see Chapter 49 and Chapter 71). Children with a history of a recent respiratory infection may have tender swollen cervical lymph nodes characteristic of a subsequent adenitis. Croup is readily identified by a barky cough in young children, whereas a distinctive “hot potato voice” with unilateral tonsillar swelling in adolescents indicates a peritonsillar abscess. Wheezing, tachypnea, and fever in infants mark bronchiolitis. Pneumonia often presents with tachypnea, fever, and nasal flaring or retractions. Mild abdominal pain, tenderness, vomiting, and/or diarrhea may suggest viral gastroenteritis or early hepatitis or pancreatitis. More severe findings, particularly the occurrence of peritoneal signs, may indicate intra-abdominal abscess, peritonitis, or appendicitis (see Chapter 50 and Chapter 118). However, in children, fever with abdominal pain may also represent lower lobe pneumonia, streptococcal pharyngitis, or urinary tract infection (UTI). Additional findings in UTI may include suprapubic or costovertebral angle tenderness. Adolescent girls with pelvic or abdominal pain and fever should be evaluated for pyelonephritis and/or pelvic inflammatory disease (see Chapter 94). Differentiation of these diverse diagnoses depends on a thorough history, physical examination, and at times, well-directed laboratory evaluation (see Chapter 84).

Recent public health measures have changed the frequency and risk of certain febrile illness in children. In a recently published report from the Centers for Disease Control and Prevention, Schuchat et al. have revealed that the *Haemophilus influenzae* type B vaccine has drastically changed the risk and causative agents for meningitis in children. There has been a 94% reduction in the incidence of *H. influenzae* meningitis and a shift in the median age of those affected from 15 months to 25 years of age. The current rarity of epiglottitis in children is also due to this decline in *H. influenzae* infections. Recognition of this epidemiologic change is crucial in evaluating and treating the febrile child. Children between 2 and 18 years of age who have bacterial meningitis will most likely be infected with *Neisseria meningitidis*. Seventy percent of invasive *Streptococcus pneumoniae* infections in children less than 5 years of age present as bacteremia without a focus of infection. In this recent study and others, the risk of meningitis in the setting of invasive *S. pneumoniae* infections has been found to be between 2 and 6%. These findings obviously influence the evaluation and treatment of febrile children with signs of meningitis, as well as those young children without an identified

source of infection after thorough historical and physical examination.

Occult bacteremia is the presence of pathogenic bacteria in the blood of a well-appearing febrile child in the absence of an identifiable focus of infection (see also Chapter 84). Children most commonly suspected to be at risk for occult bacteremia are those between the ages of 3 and 36 months with a fever of 39°C (102°F) or higher. Infants less than 3 months of age are at increased risk for invasive disease and therefore necessitate different evaluation and treatment. The reported incidence of occult bacteremia among children 3 to 36 months of age with a temperature higher than 39°C before the initiation of the *H. influenzae* type B vaccine program was between 3 and 10%. However, Lee and Harper have recently shown that the prevalence of occult bacteremia in children 3 to 36 months of age is currently 1.6%, with *S. pneumoniae* accounting for 92% of infections. Preliminary work in children 3 to 24 months of age by Alpern et al. correlates with this study, showing a prevalence of occult bacteremia of 1.8%. Both studies failed to identify *H. influenzae* bacteremia in any of the children considered to be at risk for occult bacteremia.

Given these general considerations, an algorithmic approach to the child with an acute (less than 5 days) febrile illness can be formulated, using the following key features: overall degree of toxicity and presence of signs or symptoms of life-threatening disease, immunocompromised host status, patient's age, degree of fever, and presence of localizing features on history and physical examination (Fig. 28.1). Laboratory studies are indicated only for selected situations as defined by these clinical features. Most older febrile children seen in the ED need no laboratory testing.

FIGURE 28.1. Approach to the evaluation of the febrile child.

Infants less than 2 months of age are at increased risk of serious bacterial infections and bacteremia and are more difficult to assess clinically than older children. Thus, all children with fevers of 38°C (100.4°F) or higher who are less than 2 months of age should receive full laboratory investigation for serious infection (“sepsis workup”), including complete blood count (CBC), blood culture, urine analysis, urine culture, and lumbar puncture with cerebral spinal fluid (CSF) for cell count, glucose, protein, and culture. These infants less than 1 month old are usually admitted to the hospital for observation with presumptive antibiotic therapy. Herpes simplex virus polymerase chain reaction (PCR) or culture with presumptive antiviral treatment should be considered in neonates with historical concerns or physical findings of skin, eye, or mouth lesions; respiratory distress; seizures; or signs of sepsis. Stool for leukocytes and culture should be obtained if diarrhea is present. In a recent meta-analysis, Bramson et al. have shown that respiratory findings are good predictors of clinically significant positive chest radiographs in children less than 3 months old. Therefore, chest radiographs may be obtained only when there are clinically evident respiratory symptoms. Baker et al. showed that in children 1 to 2 months of age, a standardized observation scale (Yale Observation Scale) is not sensitive enough on its own to identify serious illness. Many recent studies (Baker et al., Baskin et al., Dagan et al., and Jaskiewicz et al.) have found that children between 1 and 2 months of age, not pretreated with any antibiotics, and who have a pristine physical examination and completely benign laboratory evaluation may be safely discharged home with careful observation and close follow-up. For such a disposition, parents should be able to watch the infant closely for changes in symptoms, should have ready access to health care, and should be willing to return for evaluation. These studies have also found that both empiric intramuscular ceftriaxone (e.g., Baskin et al.) and close observation without antibiotics (e.g., Baker et al.) are safe and effective management strategies in this age group.

An additional dilemma involves the young baby who presents to the ED with a description of either tactile fever alone or fever confirmed by rectal temperature at home but who is afebrile on arrival. This situation was studied by Bonadio et al., who found that the history of tactile fever in such infants did not correlate with subsequent fever, whereas an elevated rectal temperature at home correlated with subsequent fever in 20% of such patients. However, all infants who were found to have serious bacterial infections (including five who were afebrile on presentation) were observed to have had an abnormal initial clinical profile and/or laboratory workup. Although there is no consensus on the approach to this situation, it seems prudent to consider a careful clinical evaluation in all young infants with a history of fever, including one or more repeat temperatures over 1 to 2 hours in the ED after the baby is unbundled. If there is a reliable history of elevated rectal temperature, a sepsis workup should be considered seriously along with a subsequent disposition based on the evolution of temperature pattern, clinical findings, and laboratory results. The infant with only a history of tactile fever whose repeated temperatures are normal and who has an entirely normal clinical evaluation may be assessed as not requiring laboratory studies. All such infants discharged home warrant close follow-up and appropriate short-term monitoring of rectal temperature.

Children between 2 and 3 months of age are evaluated clinically for degree of fever and degree of irritability. Occasionally, an infant at this age may truly “look great,” despite significant fever, and be judged as requiring symptomatic treatment only without aggressive laboratory investigation or hospitalization. However, a sepsis workup should be done for infants with high fever and mild to moderate irritability and/or a full fontanelle. They should be hospitalized if signs of a serious bacterial infection (e.g., cellulitis, pneumonia, septic arthritis, osteomyelitis, UTI) are

present. If all laboratory results are normal, such a child may be discharged on symptomatic treatment and/or antibiotics, if a minor infectious focus (i.e., OM) is found. Even with normal laboratory parameters, however, these patients are at risk for bacteremia and subsequent focal infection, including meningitis. Therefore, it has been our tendency to admit infants who are aged 2 to 3 months with fever and marked irritability, even when CBC and CSF are initially normal.

The febrile child between 3 and 36 months of age with signs of focal infection (e.g., irritability, meningismus, tachypnea, flank tenderness) should be evaluated with the appropriate diagnostic tests and treated for the identified infection. However, if the child with a temperature of 39°C (102°F) or higher does not have localizing symptoms or laboratory/radiograph results indicative of definitive focal infection, he or she should be evaluated with a blood culture for occult bacteremia. Highly febrile children with OM on examination and those who have had a simple febrile seizure (discussed in the following) are also considered to be at risk for occult bacteremia. Teach et al. have analyzed data from a large multicenter trial and indicate that the Yale Observation Scale is not clinically useful in identifying patients with occult bacteremia. Therefore, all such children, despite "well" clinical appearance, are at risk for occult bacteremia.

There has been considerable controversy in the literature about the appropriate initial treatment of children at risk of such occult bacteremia (see Chapter 84). One large, prospective controlled study (Jaffe et al., 1987) found no benefit of oral amoxicillin versus placebo in preventing "serious bacterial infection" (SBI), although the number of randomized children with this outcome were few, and the power to detect a significant difference was small. In 1994, Fleisher et al. compared oral amoxicillin with intramuscular ceftriaxone in preventing SBI and found the latter to be protective. Several recent meta-analyses have attempted to further determine whether antibiotics decrease the risk of meningitis in children evaluated for occult bacteremia. Rothrock et al. determined the risk of SBI in children with occult *S. pneumonia* bacteremia to be decreased in those treated with oral antibiotics (3.3%) compared with those not receiving any antibiotic treatment (9.7%). They also found a reduction from 2.7% to 0.8% for meningitis in children treated with oral antibiotics compared with untreated children. Although Rothrock et al. reported that this difference in risk of meningitis was not statistically significant using their method of analysis, it should be noted that performance of a chi square test on their data yields a *P*-value of less than .05 and also that they eliminated children who underwent a lumbar puncture from the analysis. Bulloch et al. recently performed a meta-analysis that concluded that the use of antibiotics in children at risk for occult bacteremia did not statistically reduce serious sequelae (OR = 0.60; 95% CI 0.10 to 3.49), whereas ceftriaxone compared with oral antibiotics did significantly prevent SBI in children with proven bacteremia (OR = 0.25; 95% CI 0.07 to 0.89). The recent epidemiologic changes due to the *H. influenzae* type B vaccine are responsible for *S. pneumoniae* currently being the most common causative organism of occult bacteremia. *S. pneumoniae* bacteremia spontaneously resolves in most cases. Recent studies indicate that the current incidence of meningitis in children with occult bacteremia is much lower than reported when *H. influenzae* was a significant causative organism of occult bacteremia. Therefore, the role of treatment with empiric antibiotics has been further questioned.

Currently, several options are available to physicians facing this common but challenging situation in the ED (see Chapter 84). One approach is that of obtaining a blood culture and providing expectant therapy without antibiotics, including antipyretics, close observation for progression of symptoms, and reevaluation for any positive culture. Many authorities, including a panel of experts in pediatric emergency medicine and infectious diseases convened to recommend clinical practice guidelines, whose recommendations were published simultaneously in *Annals of Emergency Medicine* and *Pediatrics* (Baraff et al., 1993), suggest presumptive antibiotic treatment with intramuscular ceftriaxone for children at highest risk for occult bacteremia. Jaffe and Fleisher found the sensitivity of a WBC count of 10,000/mm ^{3} or more to be 92%, with a positive predictive value of 4.6% for occult bacteremia. Recently, Kuppermann et al. derived a multivariable model to predict occult pneumococcal bacteremia in children 3 to 36 months of age with temperatures of 39°C or higher and without focal infection. Age less than 24 months, high temperature, and absolute neutrophil count of 10,000/mm^3 or more (WBC 15,000/mm^3 or more) were identified as independent predictors of occult pneumococcal bacteremia and therefore may help delineate patients at highest risk. Others have differed with the published clinical practice guidelines regarding empiric antibiotic therapy, and even to the necessity of obtaining blood cultures in children who do not appear toxic (Kramer and Shapiro, 1997). The optimal management of children at risk for bacteremia may evolve as more emergent techniques to detect bacteremia at the time of the initial ED visit are developed. For example, currently available PCR assays for pneumococcal bacteria currently do not have the sensitivity or specificity needed to screen for occult bacteremia. However, further investigation may yield rapid, predictive tests for occult bacteremia in the form of pneumococcal PCR, plasma tumor necrosis factor, or interleukin 1b levels. Finally, it is likely that the introduction of a conjugated pneumococcal vaccine in the near future will almost completely eliminate the problem of occult bacteremia.

An additional consideration revolves around children with risk factors for bacteremia and whose initial presentation was notable for such irritability that a lumbar puncture was deemed necessary. Although clinical appearance may not predict occurrence of bacteremia per se, lumbar puncture itself has been reported to be associated with the occurrence of meningitis in bacteremic children. Thus, it might be particularly desirable to empirically treat infants with antibiotics who have undergone this procedure but whose cerebrospinal fluid was normal and were thus being considered for an ambulatory disposition.

Recent studies by Shaw et al. and Hoberman et al. have established the overall prevalence of occult UTI in young children without an identified source of infection to be between 3 and 5%. The risk is highest in febrile Caucasian girls less than 2 years of age and in uncircumcised boys less than 1 year of age. Laboratory testing is indicated for young febrile children without an identifiable focus of infection. One approach is to obtain a urinalysis and urine culture in febrile boys less than 6 months of age as well as in any age febrile boy who is uncircumcised and not yet toilet-trained. Febrile girls less than 2 years of age should have urine studies obtained if any two of the following characteristics are present: fever of 39°C (102.2°F) or higher, 1 year of age or younger; Caucasian, fever lasting 2 days or longer; or no identifiable source of infection. Aseptic urethral catheterization or suprapubic aspiration is an appropriate method to obtain urine for the diagnosis of urinary tract infections. Positive dipstick urinalysis with microscopic evidence of pyuria (5 WBC/HPF or greater) or any bacteria/HPF has a sensitivity between 65 and 83%; therefore, urinalysis alone is not adequate to diagnose UTI. Urine dipstick and culture should be performed for all children at significant risk for occult UTI (see Chapter 84).

Simple febrile seizures occur in 3 to 5% of all children (see Chapter 70). They are defined as generalized tonic-clonic seizures without focal neurologic findings, occurring only once per febrile illness (usually in the first 12 hours of onset of fever), in children 6 months to 5 years of age, lasting less than 15 to 20 minutes in duration. By definition, they are seizures accompanied by fever in children without CNS infection. The dilemma that faces the emergency physician is to decide whether a febrile seizure is truly such or if a child presenting with a fever and seizure requires a lumbar puncture to rule out meningitis. Green et al. reviewed 503 cases of meningitis in children 2 to 15 years of age and noted that no cases of bacterial meningitis presented solely as a seizure without any other neurologic signs or symptoms (nuchal rigidity, irritability, prolonged seizure activity, or multiple seizures). Nonetheless, each case of a child presenting with what appears to be a simple febrile seizure, even with a history of such episodes, needs to be evaluated individually. Whether a lumbar puncture is performed or not, the physician should evaluate each child for the underlying source of fever. The threshold to perform a lumbar puncture should be extremely low in children younger than 12 months of age (because of the difficulty in recognizing signs and symptoms of meningitis in very young infants) and in children pretreated with antibiotics (because symptoms of partially treated meningitis may be minimal or absent). Children with atypical febrile seizures should be closely evaluated for CNS infection and a lumbar puncture strongly considered.

Children older than 36 months of age usually can be managed on the basis of degree of irritability, evidence of meningeal signs, and/or other foci of infection found on history and physical examination. These children need not be screened routinely for occult bacteremia. After excluding meningitis, there are several important infections that may be present in ill-appearing, febrile children in this age group, without obvious initial focus. These include meningococcemia, Rocky Mountain spotted fever, salmonellosis or shigellosis, and pyelonephritis (see Chapter 84). Early institution of presumptive therapy may be lifesaving in some of these situations, so their possibility must be borne in mind with toxic, febrile children at any age. Obviously, if certain high-risk features are missed during the initial triage (or evolve after triage) and are encountered early during the physician's careful clinical assessment (e.g., a significant hemorrhagic rash), the child must be managed with extreme urgency (as befits the reclassification into the high-risk category).

Other causes of acute febrile episodes should be kept in mind, including intoxications, environmental exposure, and immunization reactions. Poisonous exposures to aspirin, atropinics, amphetamines, antihistamines, and cocaine present with hyperpyrexia (see Chapter 88). Additional uncommon febrile drug reactions include the serotonin syndrome occurring with the combined use of monoamine oxidase inhibitors and analgesic, antitussive, or psychotropic serotonergic medications (e.g., meperidine, dextromethorphan, fluoxetine), and the neuroleptic malignant syndrome (see Chapter 88). History of environmental exposure in the face of severe hyperpyrexia may represent heat stroke rather than an infectious cause for the increased temperature (see Chapter 89). The diphtheria–pertussis–tetanus immunization is associated with fever that occurs within 48 hours (occurring less often with DTaP vaccine administration). Fever, at times accompanied by a faint rash, may occur 7 to 10 days after immunization with the live-attenuated measles vaccine or the measles–mumps–rubella vaccine.

Fevers of unknown origin (FUO) are defined as daily temperatures of 38.5°C (101.3°F) or higher for at least 2 weeks without discernible cause. Infections commonly causing prolonged fever in children include Epstein-Barr virus infections, osteomyelitis, *Bartonella henselae* infections (cat-scratch disease), UTIs, Lyme disease, and HIV. Noninfectious causes of prolonged fever include neoplasms, collagen vascular diseases, and inflammatory disorders (ulcerative colitis or Crohn's disease).

Symptomatic Treatment

Over the years, there has been considerable debate about the theoretical benefits of fever in the mechanism of host defense, but few definite benefits have been demonstrated in humans. For example, the exanthem of varicella lasts about 1 day longer in children treated aggressively with antipyretics than in control patients. Nevertheless, most clinicians find that febrile children, especially those with high fevers (temperatures of 39.5°C [103°F] or higher), feel better if the temperature is brought down with antipyretic medication. Furthermore, antipyresis may allow the physician to see the child at his or her “best” and, in particular, can help in the decision to perform a lumbar puncture. However, antipyresis will not aid the clinician in discriminating bacteremic children from those with viral syndrome. Both groups typically respond with an equivalent pronounced drop in temperature.

In general, antipyretic therapy should parallel the pathophysiologic basis of the fever. When the fever is caused by altered hypothalamic set point, as in infection, antigen–antibody reactions, and malignancy, attempts to reset the “thermostat” with antipyretic medications are most likely to enhance patient comfort. Antipyretics in common use today work via the inhibition of hypothalamic prostaglandin synthesis. If fever is caused by imbalance of heat production and heat loss mechanisms, such as in heat stroke, urgent cooling by physical removal of heat is necessary, such as with ice water baths, and antipyretics will not help (see Chapter 89). Rarely, a patient with infection will have extreme hyperpyrexia (temperature of 41.1°C [106°F] or higher) and will require urgent temperature reduction with both antipyretics and external cooling. Patients with ongoing febrile seizures also warrant rapid treatment with antipyretics and external cooling, although tepid to cool water sponging usually is sufficient. On the other hand, children at risk for recurrent febrile seizures do not, unfortunately, tend to be protected by rapid use of “prophylactic” antipyresis at first sign of fever.

Most authorities consider acetaminophen to be the pediatric antipyretic of choice. The current dosage recommendation for acetaminophen is 10 to 15 mg/kg given every 4 to 6 hours, with a maximum of five doses per day, resulting in 50 to 75 mg/kg per day. Several recent reports have stressed that, although very rare, repetitive dosing of acetaminophen at the upper limit of, or just slightly above, recommended dosages may result in severe or fatal fulminant hepatic failure. This is particularly the case for children who were fasting (e.g., because of vomiting, diarrhea with febrile illness), under age 2 years, treated for several days, or treated with adult-intended preparations (see Kearns et al., 1998, and Heubi et al., 1998). Ibuprofen is now available in pediatric formulations and is typically dosed at 5 to 10 mg/kg per dose, given every 6 to 8 hours, with a maximum of four doses per day (e.g., 30 to 40 mg/kg per day). There has been concern that ibuprofen might manifest increased incidence of serious gastrointestinal bleeding, renal failure, or allergic reactions relative to acetaminophen, but this has not been borne out in large, prospective studies (see Lesko and Mitchell, 1995).

Nevertheless, in our view, ibuprofen does not confer any significant advantage over acetaminophen. Its proponents note greater antipyretic efficacy, faster onset of action, and longer duration than acetaminophen on a milligram-for-milligram basis, but this is hardly clinically significant, especially at safe, equi-antipyretic doses of both medications (e.g., 10 mg/kg of ibuprofen and 12 to 15 mg/kg of acetaminophen). Recently, some concern has been raised over undesirable effects of ibuprofen's anti-inflammatory activity in the treatment of routine febrile illnesses, particularly varicella, with a potential for exacerbating invasive streptococcal disease. Although not proven to be causally related, it might be prudent to particularly avoid ibuprofen in such cases of suspected or at-risk streptococcal disease (see Curtis, 1996). Aspirin is no longer recommended for routine antipyretic use in children because of its potential to cause gastrointestinal bleeding and its implication as a possible risk factor for Reye syndrome (see Chapter 83 and Chapter 93). The practice of combining or alternating ibuprofen and acetaminophen for “really bad fevers” is bound to confuse parents, result in increased risk of drug toxicity, and add to unreasonable “fever phobia.” It is important to remember that many parents greatly fear even moderately high fever in their children and require reassurance that the fever itself, in its usual range of severity, does not cause damage. They need education about appropriate indications for antipyretic treatment, particularly seeking to reduce fever-associated discomfort, rather than a modestly elevated temperature itself. They need further education about appropriate, safe antipyretic dosing regimens, the lack of urgency in treating fever (unless temperature goes above 41.1°C [106°F] or there are prolonged febrile seizures), and most important, the concept that the overall well-being of the child, in context with age, usually is far more important than the temperature per se.

Selected Readings

Al-Eissa YA. Lumbar puncture in the clinical evaluation of children with seizures associated with fever. Pediatr Emerg Care 1995;11(6):347–350.

Alpern ER, Alessandrini EA, Shaw KN, McGowan KL. Prevalence, time to detection, and outcome of occult bacteremia in an urban pediatric emergency department (Abstract). New Orleans, LA: Ambulatory Pediatric Association Program and Abstracts, 1998:118.

American Academy of Pediatrics. Practice parameter: the neurodiagnostic evaluation of the child with a first simple febrile seizure. Pediatrics 1996;97(5):769–771.

Baker MD, Avner JR, Bell LM. Failure of infant observation scales in detecting serious illness in febrile 4- to 8-week-old infants. Pediatrics 1990;85(6):1040–1043.

Baker MD, Bell LM, Avner JR. Outpatient management without antibiotics of fever in selected infants. N Engl J Med 1993;329(20): 1437–1441.

Baker MD, Fosarelli PD, Carpenter RO. Childhood fever: correlation of diagnosis with temperature response to acetaminophen. Pediatrics 1987;80(3):315–318.

Baker RC, Sequin JH, Leslie N, et al. Fever and petechiae in children. Pediatrics 1989;84(6):1051–1055.

Baraff LJ, Bass J W, Fleisher GF, et al. Practice guideline for the management of infants and children 0 to 36 months of age with fever without source. Pediatrics 1993;92:1–12.

Baraff LJ, Osland S, Prather M. Effect of antibiotic therapy and etiologic microorganism on the risk of bacterial meningitis in children with occult bacteremia. Pediatrics 1993;92:140–143.

Baskin M, O'Rourke EJ, Fleisher GR. Outpatient treatment of febrile infants 28 to 89 days of age with intramuscular administration of ceftriaxone. J Pediatr 1992;120(1):22–27.

Bass JW, Steele RW, Wittler RR, et al. Antimicrobial treatment of occult bacteremia: a multicenter cooperative study. Pediatr Infect Dis J 1993;12(6):466–473.

Bonadio WA, Hegenbarth M, Zachariason M. Correlating reported fever in young infants with subsequent temperature patterns and rate of serious bacterial infections. Pediatr Infect Dis J 1990;9:158–160.

Bonadio WA, Smith DS, Sabnis S. The clinical characteristics and infectious outcomes of febrile infants aged 8 to 12 weeks. Clin Pediatr 1994;33(2):95–99.

Bramson RT, Meyer TL, Silbiger ML, et al. The futility of the chest radiograph in the febrile infant without respiratory symptoms. Pediatrics 1993;92(4):524–526.

Bulloch B, Craig WR, Klassen TP. The use of antibiotics to prevent serious sequelae in children at risk for occult bacteremia: a meta-analysis. Acad Emerg Med 1997;4:679–683.

Curtis N. Non–steroidal anti-inflammatory drugs may predispose to invasive group A streptococcal infections. Arch Dis Child 1996;75:547.

Dagan R, Powell KR, Hall CB, Menegus MA. Identification of infants unlikely to have serious bacterial infection although hospitalized for suspected sepsis. J Pediatr 1985;107(6):855–860.

Doctor A, Harper MB, Fleisher GR. Group A beta-hemolytic streptococcal bacteremia: historical overview, changing incidence, and recent association with varicella. Pediatrics 1995;96(3):428–433.

Ferrera PC, Bartfield JM, Snyder HS. Neonatal fever: utility of the Rochester Criteria in determining low risk for serious bacterial infections. Am J Emerg Med 1997;15(3):299–302.

Fleisher GR, Rosenberg N, Vinci R, et al. Intramuscular versus oral antibiotic therapy for the prevention of meningitis and other bacterial sequelae in young, febrile children at risk for occult bacteremia. J Pediatr 1994;124(4):504–512.

Green SM, Rothrock SG, Clem KJ, et al. Can seizures be the sole manifestation of meningitis in febrile children? Pediatrics 1993;92(4):527–534.

Hoberman A, Wald ER. Urinary tract infections in young febrile children. Pediatr Infect Dis J 1997;16(1):11–17.

Huebi JE, Barbacci MB, Zimmerman HJ. Therapeutic misadventures with acetaminophen: hepatotoxicity after multiple doses in children. J Pediatr 1998;132:22–27.

Huttenlocher A, Newman TB. Evaluation of the erythrocyte sedimentation rate in children presenting with limp, fever, or abdominal pain. Clin Pediatr 1997;36(6):339–344.

Isaacman DJ, Zhang Y, Reynolds EA, Ehrlich GD. Accuracy of a polymerase chain reaction-based assay for detection of pneumococcal

bacteremia in children. Pediatrics 1998;101(5):813–816.

Jacobs RF, Schutze GE. *Bartonella henselae* as a cause of prolonged fever and fever of unknown origin in children. Clin Infect Dis 1998;26:80–84.

Jaffe DM, Fleisher GR. Temperature and total white blood cell count as indicators of bacteremia. Pediatrics 1991;87(5):670–674.

Jaffe DM, Tanz RR, Davis AT, et al. Antibiotic administration to treat possible occult bacteremia in febrile children. N Engl J Med 1987;317:1175–1180.

Jaskiewicz JA, McCarthy CA, Richardson AC, et al. Febrile infants at low risk for serious risk for serious bacterial infection: an appraisal of the Rochester Criteria and implications for management. Pediatrics 1994;94(3):390–396.

Kearns GL, Leeder JS, Wasserman GS. Acetaminophen overdose with therapeutic intent. J Pediatr 1998;132:5–8.

Kramer MS, Shapiro ED. Management of the young febrile child: a commentary on recent practice guidelines. Pediatrics 1997;100(1):128–134.

Kuppermann N, Fleisher GR, Jaffe DM. Predictors of occult pneumococcal bacteremia in young febrile children. Ann Emerg Med 1998;31(6): 679–687.

Lee GM, Harper MB. Risk of bacteremia for febrile young children in the post- *Haemophilus influenzae* type b era. Arch Pediatr Adolesc Med 1998;152(5):624–628.

Lesko SM, Mitchell AA. An assessment of the safety of pediatric ibuprofen. JAMA 1995;273:929–933.

Lieu TA, Baskin MN, Schwartz JS, Fleisher GR. Clinical and cost-effectiveness of outpatient strategies for management of febrile infants. Pediatrics 1992;89(8):1135–1144.

Maller JS, Gorelick MH. Use of monitoring devices. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:33–37.

Mandl KD, Stack AM, Fleisher, GR. Incidence of bacteremia in infants and children with fever and petechiae. J Pediatr 1997;131(3):398–404.

Press S. Association of hyperpyrexia with serious disease in children. Clin Pediatr 1994;33(1):19–25.

Rothrock SG, Harper MB, Green SM, et al. Do oral antibiotics prevent meningitis and serious bacterial infections in children with *Streptococcus pneumoniae* occult bacteremia? A meta-analysis. Pediatrics 1997;99(3):438–444.

Saper CB, Breeder CD. The neurologic basis of fever. N Engl J Med 1994;330:1880–1886.

Schuchat A, Robinson K, Wenger J, et al. Bacterial meningitis in the United States in 1995. N Engl J Med 1997;337(14):970–976.

Shaw KN, Gorelick M, McGowan KL, et al. Prevalence of urinary tract infection in febrile young children in the emergency department. Pediatrics 1998;102(2):e16.

Shaw KN, McGowan KL, Gorelick MH, Schwartz S. Screening for urinary tract infection in Infants in the emergency department: which test is best? Pediatrics 1998;101(6):e1.

Strait RT, Ruddy RM, Friedland LR, Duncan KM. A pilot study of the predictive value of plasma tumor necrosis factor Alpha and interleukin 1 beta for *Streptococcus pneumoniae* bacteremia in febrile children. Acad Emerg Med 1997;4(1):44–51.

Teach SJ, Fleisher GR, Occult Bacteremia Study Group (sup A). Efficacy of an observation scale in detecting bacteremia in febrile children three to thirty-six months of age, treated as outpatients. J Pediatrics 1995;126(6):177–181.

Torrey SB, Henretig F, Fleisher G, et al. Temperature response to antipyretic therapy in children: relationship to occult bacteremia. Am J Emerg Med 1985;3(3):190–192.

CHAPTER 29

Foreign Body—Ingestion/Aspiration

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Through play, experimentation, and normal daily activities, children are likely to place foreign bodies just about anywhere. Once an object or foodstuff is in a child's mouth, it can lodge in the respiratory tree or be ingested. Young age (6 months to 4 years), a tendency to hold objects in the mouth, easy distractibility, inappropriate-for-age foods, and inappropriate playthings place the child at risk for foreign body aspiration or ingestion. Often, the “choking episode” completely clears the foreign body; however, the sequelae of an aspirated object can range from an immediate life-threatening event to a slowly evolving pneumonia. The seriousness of the foreign body ingestion is determined by the nature of the object (e.g., round, long, sharp, corrosive) and the potential level of lodgment in the gastrointestinal (GI) tract. Fortunately, children typically swallow round rather than sharp objects. Generally, most ingested foreign material is well tolerated, and many ingestions go unnoticed by the family as well as by the child.

PATHOPHYSIOLOGY

There are three main pathophysiologic considerations for aspirated and ingested foreign bodies: the anatomic determinants of lodgment site, the physical properties of the foreign body (size, shape, and composition), and the local tissue reaction to the foreign body.

The respiratory tract, once distal to the larynx, gradually narrows with each airway generation, whereas the GI tract has several sites of anatomic or functional narrowing that occur throughout. An ingested foreign body may lodge in three distinct esophageal sites—thoracic inlet, level of the aortic arch, or gastroesophageal junction—may be unable to pass through the pylorus, or may become impacted in the duodenum, cecum, appendix, rectum, or any other location of congenital or acquired narrowing.

The nature of the foreign body (size, shape, and composition) determines the site of lodgment and the potential for local tissue interaction. The widest diameter of the aspirated or ingested foreign body and the distensibility of the tissue determines, in part, where it lodges within the respiratory or GI tract. A sharp or long object may become impacted even where there is no anatomic narrowing. The aspirated object may affect air movement minimally, until the “fit” with the airway is sufficient to completely or intermittently impede air flow.

The composition of the foreign body also determines the local tissue reaction and the evolution of complications. A disc battery may erode through the esophageal wall much more rapidly than a coin. In the bronchial tree, the fatty oils in some aspirated foods (e.g., peanuts) create a more severe pneumonitis than a similarly sized plastic or metal object. Less commonly, the patient may absorb compounds from the foreign body that cause systemic toxicity.

DIFFERENTIAL DIAGNOSIS

Gastrointestinal Foreign Body

Esophagus

Impaction in the esophagus is the most common and most serious consequence of a foreign body ingestion. Most childhood esophageal foreign bodies are round or cuboidal objects. Coins account for 50 to 75% of childhood esophageal foreign bodies, with pennies predominating (Fig. 29.1). This contrasts with adults, whose impacted esophageal foreign bodies tend to be foodstuffs (meat) and sharp objects (bones). Esophageal foreign bodies in adults are often associated with underlying conditions that affect the esophagus (e.g., intrinsic strictures, neuromuscular conditions, extrinsic pressure), whereas most children with esophageal impactions have a structurally and functionally normal esophagus. Naturally, children with esophageal strictures either acquired (e.g., secondary to caustic ingestions) or from congenital conditions and their repair (e.g., esophageal atresia, tracheoesophageal fistula) are at increased risk for recurrent esophageal impactions, even with foodstuffs (e.g., hot dogs, chicken).

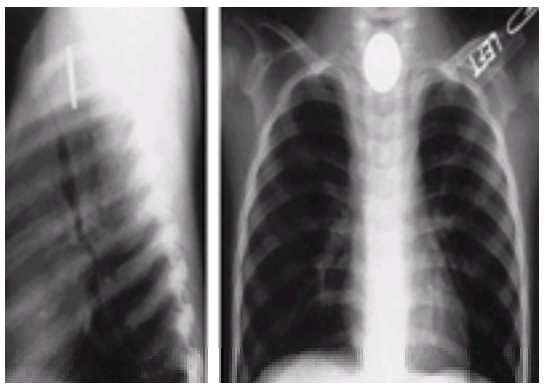

FIGURE 29.1. Two-view chest radiograph demonstrating impacted esophageal coin located at the thoracic inlet.

Foreign bodies of the esophagus tend to lodge at three sites. The most proximal location, at the thoracic inlet (Fig. 29.1), accounts for 60 to 80% of esophageal foreign bodies. The next most common level of lodgment is at the gastroesophageal junction, accounting for 10 to 20%, and last, at the level of the aortic arch, accounting for 5 to 20%. The level of lodgment in children with underlying esophageal conditions or strictures depends on the nature and location of the constricting lesion.

Foreign bodies that remain lodged in the esophagus may lead to potentially serious complications. For example, coins can cause respiratory distress with upper airway compromise, esophageal perforation, mediastinitis, and aortic and tracheal fistula formation. Therefore, it is imperative that the physician be alert to the possibility of esophageal foreign bodies, especially in the susceptible 6-month-old to 4-year-old age group.

Stomach and Lower Gastrointestinal Tract

Objects that can pass safely into the stomach generally traverse the remainder of the GI tract without complication. Safe passage has been documented in hundreds of cases involving various foreign objects (Fig. 29.2). This may not be true of some long (greater than 5 cm) objects that are unable to negotiate the turns of the duodenum and some other tight bends in the lower GI tract. This also may not be true of some very sharp objects (sewing needles are particularly high risk), which may perforate the hollow viscera. Bowel perforation from sharp objects has resulted in peritonitis, abscess formation, inflammatory tumors, hemorrhage, and death.

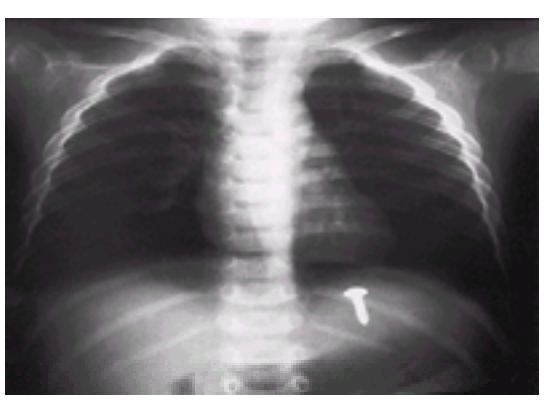

FIGURE 29.2. Abdominal radiograph demonstrating gastric radiopaque foreign body—a screw—that passed without complications.

Respiratory Foreign Body

Upper Airway

Foreign bodies that lodge in the upper airway can be immediately life-threatening. Such occurrences are responsible for more than 300 childhood deaths in the United States annually. Of fatalities caused by food aspiration, 65% occur in children who are less than 2 years old. The most common foods responsible include hot dogs, candy, nuts, and grapes. Childhood fatalities from man-made objects tend to be conforming objects, with balloons accounting for nearly one-third; children less than 3 years of age account for 65% of these fatalities. Children with foreign bodies in their upper airways present with acute respiratory distress, stridor, increased respiratory effort, or complete obstruction of their upper airway. In patients with complete airway obstruction, emergency treatment depends on proper application of basic life support (BLS) skills. Back blows and chest compressions are used in infants, and the Heimlich maneuver is used in toddlers, children, and adolescents. If these methods fail to dislodge the foreign body, rapid progression to direct visualization and manual extraction is necessary (see Chapter 1 and Chapter 5).

Lower Respiratory Tract

Because of the ubiquitous nature of the presenting symptoms, the frequency of the asymptomatic presentation, and the potential for false-negative and false-positive screening radiographs, childhood foreign bodies of the lower

tracheobronchial tree present a diagnostic challenge to all who treat children.

Foreign bodies of the lower respiratory tract are seen more commonly in the younger pediatric age groups. Approximately 60 to 80% of pediatric tracheobronchial foreign bodies occur in children less than 3 years old. In children, aspirated foreign bodies show only a slight propensity to lodge in the right lung. The nature of the aspirated objects is fairly consistent throughout studies in several countries. Organic matter accounts for most aspirations, with nuts (mainly peanuts) and seeds (sunflower and watermelon) accounting for 30 to 70% of cases, followed by other food products, including apples, carrots, and plants and grasses (Table 29.1). Plastics and metals make up a minority of aspirated objects (Fig. 29.3), and coin aspiration has been reported only rarely.

Foreign Body	Percent
Peanuts	38
Other nuts	10
Other organic (food) material	16
Seeds, weeds, or twigs	7
Plastics	6
Popcorn	6
Pins, screws, tacks, or nails	6
Crayons	2
Rocks or stones	1
Miscellaneous[b]	8

[a]Included a total of 440 foreign bodies removed. (Adapted from Black RE, Johnson DG, Mattak ME. Bronchoscopic removal of aspirated foreign bodies in children. J Pediatr Surg 1994;29:662–664.)
[b]Included cotton/lint, earrings, bullet shell casings, tooth, staple, shirt label, pellet, spring, aluminum foil, seashell, pencil lead, screw/silver, chalk, chain, coin, chicken bone, plaster, Styrofoam cup fragment, and others.

Table 29.1. Aspirated Foreign Bodies in Children Recovered at Bronchoscopy[a]

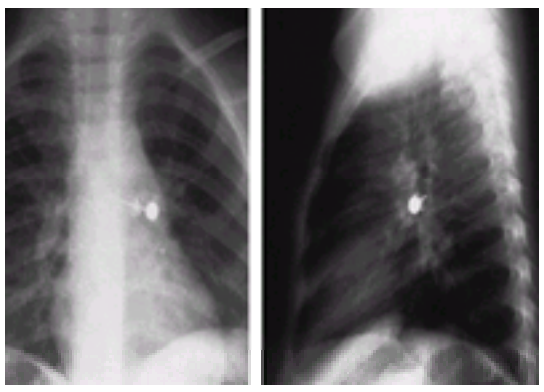

FIGURE 29.3. Two-view chest radiograph demonstrating aspirated radiopaque foreign body—an earring—located in the left bronchus.

The diagnosis of foreign body aspiration is often delayed. Previously, diagnosis on the day of aspiration occurred in fewer than half the cases, and the diagnosis was made a week or more after the aspiration in 20 to 30% of cases. A more recent report suggests diagnosis within a day in 70% of patients. Symptoms at diagnosis include cough in 75 to 90%, wheezing in 50 to 75%, and respiratory distress in 25 to 60%. The classic clinical triad of an aspirated foreign body (wheeze, cough, and decreased breath sounds) is present in only about one-third of all cases of pediatric foreign body aspiration. It has been demonstrated that this triad is more likely to occur the more delayed the evaluation from the aspiration event. Approximately 20% of patients with aspirated foreign bodies are asymptomatic. It should be noted, however, that in most surveys of pediatric foreign body aspiration, a history of aspiration, if sought, is present in more than 75% of patients.

EVALUATION AND DECISION

Unknown Location

Generally, the symptom complex and history that surround the event provide the clues necessary to decide whether to evaluate the respiratory tract or GI tract. Symptoms of cough, respiratory distress with tachypnea or retractions, stridor, wheezing, or asymmetric aeration suggest a foreign body in the airway. Symptoms of gagging, vomiting, drooling, dysphagia, pain, or localization suggest esophageal impaction. However, impacted esophageal foreign bodies may induce secondary airway symptoms, either location may induce coughing, vomiting, or gagging initially, and both types may be asymptomatic. If the history and physical examination do not provide the necessary clues, initial evaluation with a chest radiograph (to include the upper abdomen and oropharynx) will suffice as the first screen for a radiopaque esophageal or gastric foreign body. Coupling this with an expiratory chest radiograph (as outlined later) screens for an aspirated foreign body (Fig. 29.4). Alternatively, the combination of a soft-tissue lateral neck and a “wide” chest radiograph that includes oropharynx and abdomen is also used as an initial foreign body search.

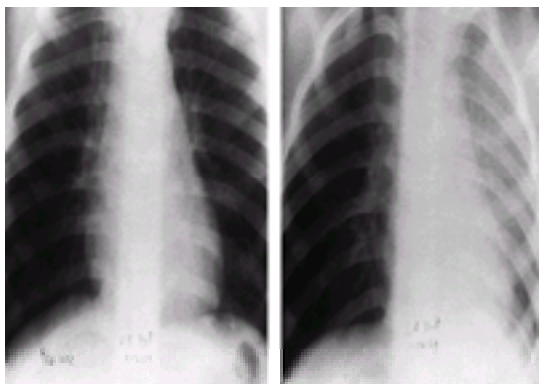

FIGURE 29.4. Inspiratory and expiratory chest radiographs demonstrating air trapping in the right lung during expiration, indicating likely right-sided foreign body. A peanut was removed at bronchoscopy.

Gastrointestinal Foreign Body

Esophageal Foreign Body: Diagnosis

Children with esophageal foreign bodies often have a history of having swallowed the foreign body. Symptoms associated with esophageal impaction include pain with swallowing, refusal to eat, foreign body sensation or localization, drooling, and vomiting. When these symptoms are associated with a history of foreign body ingestion, the diagnosis is straightforward. In the absence of an ingestion history, the diagnosis may be more subtle because these same symptoms occur with such common childhood ailments as acute gastroenteritis, pharyngitis, and gingivostomatitis. Any patient with swallowing difficulty requires a thorough examination, including mouth, oropharynx, neck, chest, and abdomen, as well as a radiologic evaluation in some cases (see Chapter 53).

The approach to a child with a foreign body ingestion is outlined in Figure 29.5. Children may be asymptomatic with an esophageal foreign body. Studies have demonstrated that 30 to 40% of children who showed coins in the esophagus were asymptomatic in the emergency department (ED). Therefore, it is suggested that all children with a history of ingested foreign bodies undergo radiographic evaluation, with the exception of the asymptomatic patient who has ingested a small (less than 1 cm in maximum diameter), nonsharp object (see Fig. 29.5). If the foreign body is not radiopaque (yet is large enough to become impacted) and the patient's symptoms suggest esophageal impaction, it is necessary to use contrast esophagrams to rule out an esophageal foreign body. Fortunately, childhood esophageal foreign bodies tend to be radiopaque (e.g., coins), so diagnosis with plain radiographs is not difficult (Fig. 29.1). Children with a predisposing condition (tracheoesophageal fistula repair, esophageal stricture) who have symptoms of an esophageal foreign body after eating should have contrast esophagrams; plain radiographs are not necessary. Similarly, with nonradiopaque ingestions and symptoms suggestive of impaction in children without underlying conditions, contrast esophagrams or esophagoscopy should be performed initially.

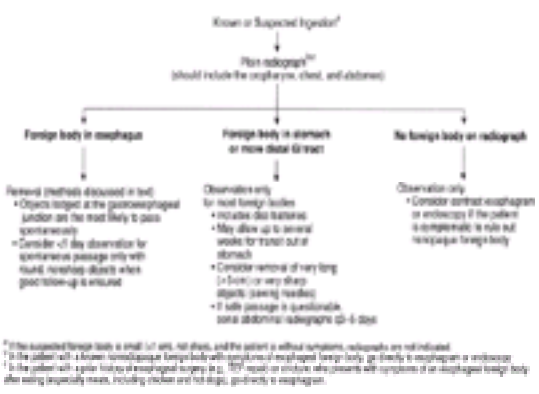

FIGURE 29.5. Management of ingested foreign body.

Handheld metal detectors may provide an alternative to conventional radiography as an initial screen when a coin ingestion is suspected. In study situations, these devices compare favorably with radiography in determining presence or absence of a coin and determining coin location (esophagus, or more distal GI tract). Users should gain some metal detector experience using x-ray confirmation before abandoning radiography, and patient follow-up is suggested because esophageal coins may be missed (this may be especially true in obese children).

Esophageal Foreign Body: Removal

In general, once an esophageal foreign body is detected, it should be removed promptly. This is especially true of sharp esophageal foreign bodies and disc batteries. It has been noted that some impacted esophageal foreign bodies pass spontaneously, regardless of location. Spontaneous passage is most likely to occur when the object is lodged at the gastroesophageal junction. It may be prudent to allow a period of less than 1 day for spontaneous passage of round, noncorrosive objects in the asymptomatic patient. This should be done only if good follow-up can be ensured and in consultation with the physician who will be involved in the removal. Handheld metal detectors may have a role in following such patients with coin ingestions. Serious complications have not resulted from an esophageal coin that was impacted for less than a few days; however, the esophageal mucosa may grow around a coin after several days, which

could hinder removal attempts.

Removal techniques for impacted esophageal foreign bodies vary regionally and may depend on the duration of impaction and the nature of the foreign body. Removal methods currently used include rigid esophagoscopy under general anesthesia, flexible endoscopy (with or without conscious sedation), and for round objects, a balloon-tipped catheter under fluoroscopic guidance or bougienage to advance the object into the stomach. All of these methods have a high success rate; provincial opinion and local referral patterns will dictate which method is used. Esophagoscopy under general anesthesia has been the method of choice for many years. This technique has proved to be safe and efficacious. The balloon-tipped catheter technique has been criticized because of lack of airway control, poor control of the foreign body during extraction, and inadequate visualization of the esophagus. Bougie dilators may be used to push the coin into the stomach, where spontaneous passage should occur. Both Foley catheter and Bougie dilator methods have been used successfully with low complication rates at several institutions and are less expensive than esophagoscopy with general anesthesia. These methods should be attempted only by clinicians familiar with the techniques and are generally reserved for rounded esophageal foreign bodies that have been impacted for less than a few days. Use of medications (e.g., glucagon, diazepam) to reduce muscular tone, to enhance esophageal motility, or to relax the lower esophageal sphincter has been suggested to facilitate passage from the esophagus. This has not been investigated thoroughly enough to offer guidelines for routine clinical application and dosing.

Stomach and Lower Gastrointestinal Tract

As mentioned, most foreign bodies of the stomach and lower GI tract can be managed expectantly (Fig. 29.2). Management recommendations for sharp objects are varied but conservative—watchful waiting usually is safe. Sewing needles seem to have increased propensity for perforation, however, and should probably be removed. Long objects (greater than 5 cm) should also be removed from the stomach. If the long or very sharp object has passed out of the stomach at the time of evaluation or, in any instance, to a place where a safe journey through the remainder of the GI tract is questionable, serial abdominal radiographs every 3 to 5 days and serial examinations may be necessary to document continued passage. Most round objects (e.g., coins) will traverse the GI tract in 3 to 8 days without any complication. Some physicians advocate parental examination of the stool for the foreign body, although in practice, this may not be very useful. It is an unpleasant task that is commonly, and understandably, abandoned. Furthermore, inability to retrieve the foreign body after 1 week or more of stool examination often heightens parental concern that some untoward complication has developed. Occasionally, some objects remain in the stomach for a long duration. A prolonged time, up to weeks, should be allowed for passage of inert objects out of the stomach before surgical or endoscopic removal is attempted.

Disc Battery Ingestion

Disc batteries are used as a power source for many household items, ranging from watches, cameras, and calculators to hearing aids. Therefore, these intriguing, bite-sized, “slippery when wet” batteries are often within reach of children. Most of these batteries fall into one of three varieties—a manganese dioxide system, a silver oxide system, or a mercuric oxide system. These systems may cause corrosive injury to the hollow viscera. Early reports of disc battery ingestions emphasize serious hemorrhagic sequelae, so disc battery ingestion often raises the level of concern for the physician caring for the child. Subsequent studies of large series of ingested disc batteries have emphasized the benign nature of most ingestions. A disc battery that reaches the stomach safely is likely to pass through the remainder of the GI tract without complication, and no operative or endoscopic intervention is indicated unless symptoms suggest serious sequelae. Only sporadic cases of systemic absorption of battery contents have been suggested in the literature, and no serious toxicities have been reported. Disc batteries that lodge in the esophagus should be removed promptly because of the potential to erode rapidly through the esophagus.

Respiratory Foreign Body

Lower Respiratory Tract: Diagnosis

A high clinical index of suspicion is necessary to diagnose foreign body aspiration accurately and promptly. Symptoms seen in pediatric foreign body ingestions also are present in other common diseases such as upper respiratory tract infection, bronchiolitis, pneumonia, and asthma. A few radiographic techniques serve as the most important diagnostic aids. Yet, when the clinical suspicion of foreign body aspiration is high (good history for aspiration, new onset of symptoms with focal physical findings), the lack of confirmatory radiographic studies should not dissuade the clinician from pursuing bronchoscopy for diagnosis and treatment. In patients diagnosed early, as many as one-third have normal chest radiographs. The abnormal findings seen on a chest radiograph include air trapping, atelectasis, and consolidation. The more time that has elapsed since the aspiration event, the more likely the chest radiograph will be abnormal and the greater the percentage of patients who exhibit consolidation and atelectasis. Inspiratory and expiratory films comparing the relative deflation of the two lungs may demonstrate unilateral air trapping indicative of a foreign body (see Fig. 29.4). In some series, up to 80% of the foreign bodies demonstrated abnormalities using inspiratory and expiratory chest radiographs. In the young or uncooperative child, in whom obtaining an adequate expiratory film may be difficult, lateral decubitus chest radiographs (both obtained during inspiration) that compare the relative deflation of the dependent lung may be a useful adjunct. In equivocal cases, chest fluoroscopy may show mediastinal shift during respiration; unfortunately, even this technique does not have 100% sensitivity.

The approach to diagnosing foreign body aspiration is outlined in Figure 29.6. In instances in which a respiratory foreign body is being considered, the patient should be kept on a nil per os (NPO) basis until the diagnosis is confirmed or the decision for bronchoscopy is made. The first step in the patient suspected of foreign body aspiration is inspiratory and expiratory chest radiographs. If these studies are normal, the aspiration history is poor, the material uncommonly aspirated, and the patient has mild or no symptoms without focal findings on physical examination, then discharge with follow-up in a few days usually is adequate. If diagnosis is still unclear after plain films and there is a historical or clinical suspicion of aspiration, fluoroscopy may be obtained, looking for air trapping and evidence of mediastinal shift away from

the foreign body. In some instances in which there are focal physical findings (unilateral wheeze, decreased aeration) and a good aspiration history, the patient might go directly to bronchoscopy. Similarly, in some cases, despite normal radiographic evaluation (including fluoroscopy), when there is a high clinical index of suspicion, bronchoscopy is necessary to confirm the presence of a pulmonary foreign object.

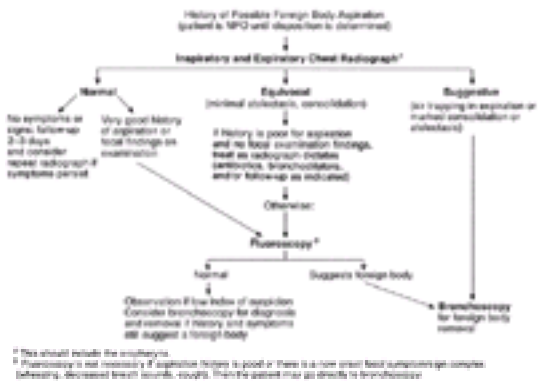

FIGURE 29.6. Guidelines for management of the child with suspected foreign body aspiration.

A history of aspirated foreign body should be sought in all cases of new-onset respiratory distress, wheezing, or cough, with special consideration to the high-risk children from 6 months to 4 years of age. History taking should include questions about recent choking episodes, especially when eating nuts (peanuts), seeds, apples, and carrots. The differential diagnosis of foreign body aspiration includes many common childhood diseases, including upper respiratory infection (URI), bronchiolitis, viral and bacterial pneumonitis, and reactive airway disease; specific questioning concerning aspiration events should be explored.

Lower Respiratory Tract: Removal

Once a foreign body of the lower respiratory tract has been identified, bronchoscopic removal is performed under general anesthesia. This technique is successful in more than 98% of cases and only rarely is a thoracotomy required. The procedure often can be done on an outpatient surgery basis, although any preoperative or postoperative concerns about the patient's respiratory status mandate in-hospital observation. Potential postoperative complications after removal of an aspirated foreign body include atelectasis, pneumonia, stridor, bronchospasm or laryngospasm, and retained foreign body.

Suggested Readings

Biehler JL, Tuggle D, Stacy T. Use of the transmitted-receiver metal detector in evaluation of pediatric coin ingestions. Pediatr Emerg Care 1993; 9:208–210.

Black RE, Johnson DG, Matlak ME. Bronchoscopic removal of aspirated foreign bodies in children. J Pediatr Surg 1994;29:682–684.

Conners GP. A literature based comparison of three methods of pediatric esophageal coin removal. Pediatr Emerg Care 1997;13:154–157.

Conners GP, Chamberlain JM, Ochsenschlager DW. Symptoms and spontaneous passage of esophageal coins. Arch Pediatr Adolesc Med 1995;149:36–39.

Conners GP, Chamberlain JM, Weiner PR. Pediatric coin ingestion: a home based survey. Am J Emerg Med 1995;14:638–640.

Harned RK, Strain JD, Hay TC, Douglas, MR. Esophageal foreign bodies: safety and efficacy of Foley catheter extraction of coins. Am J Radiol 1997;168:443–446.

Hodge D, Tecklenburg F, Fleisher G. Coin ingestion: does every child need a radiograph? Ann Emerg Med 1985;14:443–446.

Kelly JE, Leech MH, Carr MG. A safe and cost-effective protocol for the management of esophageal coins in children. J Pediatr Surg 1993;28:898–900.

Puhakka H, Svedstrom E, Kero P, et al. Tracheobronchial foreign bodies: a persistent problem in pediatric patients. Am J Dis Child 1989;143: 543–545.

Rimell FL, Thome A, Stool S, et al. Characteristics of objects that cause choking in children. JAMA 1995;274:1763–1766.

Sacchetti A, Carraccio C, Lichenstein R. Hand-held metal detector identification of ingested foreign bodies. Pediatr Emerg Care 1994;10:204–207.

Schunk JE, Corneli H, Bolte R. Pediatric coin ingestions: a prospective study of coin location and symptoms. Am J Dis Child 1989;143:546–548.

Schunk JE, Harrison M, Corneli HM, Nixon GW. Fluoroscopic Foley catheter removal of esophageal foreign bodies in children: experience with 415 episodes. Pediatrics 1994;94:709–714.

Stallings HC, Baker SP, Smith GA, et al. Childhood asphyxiation by food: a national analysis and overview. JAMA 1984;251:2231–2235.

Swischuk LE. Foreign body in the lower airway. In: Emergency Imaging of the Acutely Ill or Injured Child. 3rd ed. Baltimore: Williams & Wilkins, 1994:113–122.

Wiseman NE. The diagnosis of foreign body aspiration in childhood. J Pediatr Surg 1984;19:531–535.

Textbook of Pediatric Emergency Medicine

CHAPTER 30

Gastrointestinal Bleeding

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Gastrointestinal (GI) bleeding is a relatively common problem in pediatrics. Most infants and children who arrive in the emergency department (ED) with what appears to be GI bleeding have an acute, self-limited GI hemorrhage and are hemodynamically stable. In such patients, three important questions must be asked: 1) Is the patient really bleeding? 2) Is the blood coming from the GI tract? and 3) Is there more than a trivial amount of blood? Children with only a few drops or flecks of blood in the vomit or stool should not be considered “GI bleeders” if their history and physical examinations are otherwise unremarkable. Likewise, many substances ingested by children may simulate fresh or chemically altered blood. Red food coloring (as in some cereals, antibiotic and cough syrups, Jell-O, and Kool-Aid), as well as fruit juices and beets, may resemble blood if vomited. Melena may be confused with dark or black stools due to iron supplementation, dark chocolate, bismuth, spinach, cranberries, blueberries, grapes, or licorice. In these cases, confirmation of the absence of blood with Gastroccult (vomitus) and Hematest or Hemoccult (stool) tests will allay parental anxiety and prevent unnecessary concern and workup. Gastroccult is a specific and sensitive assay, stable in an acid environment, which can detect as little as 300 µg/dL of hemoglobin. A careful search for other causes of presumed GI bleeding, such as recent epistaxis, dental work, and sore throat, should be sought.

In most cases of upper and lower GI bleeding, the source of the bleeding is inflamed mucosa (infection, allergy, drug-induced, stress-related, or idiopathic). The emergency physician must be vigilant in differentiating inflammatory conditions that are often self-limited from causes that may require emergent surgical or endoscopic intervention, such as ischemic bowel (intussusception, volvulus), structural abnormalities (Meckel's diverticulum, angiodysplasia), and portal hypertension (esophageal varices).

INITIAL ASSESSMENT

The differential diagnosis of GI bleeding is broad. A systematic approach to all patients includes the following:

1. Assessment of the severity of the bleeding and institution of appropriate resuscitative measures
2. Establishing the level of bleeding within the GI tract
3. Pertinent history, physical examination, and laboratory tests based on knowledge of age-related causes
4. Emergency treatment based on general categories of causes

Estimation of blood loss (a few drops, a spoonful, a cupful, or more) should be obtained initially. Vomiting of coffee ground material does not necessarily signify a specific quantity of blood, nor does the vomiting of bright red blood mean that major bleeding is taking place. Hemoglobin and hematocrit are unreliable estimates of acute blood loss because of the time required for hemodilution to occur. The estimated volume of blood loss should be correlated with the patient's clinical status. The presence of resting tachycardia, pallor, prolonged capillary refill time, and metabolic acidosis points to significant blood loss. An orthostatic decrease in systolic blood pressure of 10 mm Hg or more or an increase of 20 beats/minute in pulse suggests a 10 to 20% loss of intravascular volume. Hypotension is a late finding in young children and demands immediate resuscitative measures.

ESTABLISHING THE LEVEL OF BLEEDING

There are two general categories of GI bleeding: upper and lower. *Upper GI bleeding* refers to bleeding proximal to the ligament of Treitz. *Lower GI bleeding* is distal to the ligament. In most cases, the clinical findings along with nasogastric lavage will delineate the cause of bleeding within the GI tract. *Hematemesis* is defined as vomiting of blood, which is either fresh and bright red or old with the appearance of coffee grounds. Melena, the passage of stool that is shiny, black, and sticky as a result of enzymatic or bacterial action on intraluminal blood, reflects bleeding from either the upper GI tract or the proximal small bowel. In general, the darker the blood in the stool, the higher it originates in the GI tract. Hematochezia, or the passage of bright red blood per rectum, is usually a manifestation of lower GI bleeding. Currant jellylike stools indicate vascular congestion and hyperemia as seen with intussusception. Maroon-colored stools occur

with a voluminous bleed anywhere proximal to the rectosigmoid area, such as seen with a Meckel's diverticulum.

All patients with a significant bleeding episode should have a nasogastric tube placed for diagnostic purposes (Fig. 30.1). In patients with hematemesis or melena, a positive examination of a nasogastric aspirate confirms an upper source of GI bleeding, whereas a negative result almost always excludes an active upper GI bleed. Occasionally, a postpyloric upper GI lesion, such as a duodenal ulcer, bleeds massively without reflux into the stomach, resulting in a negative examination of stomach aspirate. An upper GI endoscopic study is the best method to detect such a lesion. Patients with hematochezia and massive rectal bleeding should likewise have a nasogastric tube placed. Because blood exerts a cathartic action, brisk bleeding from an upper GI lesion may induce rapid transit through the gut, thus preventing blood from becoming melanotic. In patients with hematochezia manifested as bloody diarrhea or minimally blood-streaked stools, a lower GI source should be investigated.

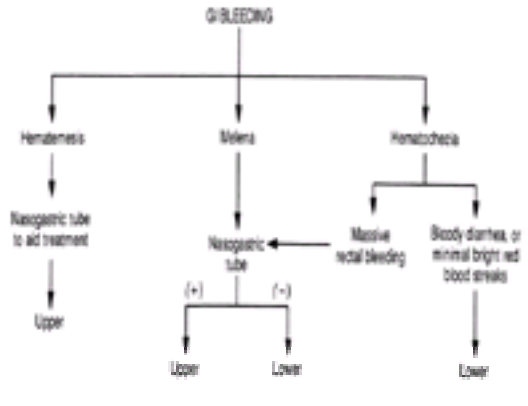

FIGURE 30.1. Establishing level of GI bleeding.

UPPER GASTROINTESTINAL BLEEDING

Differential Diagnosis

As seen in Table 30.1, Table 30.2 and Table 30.3, there is considerable overlap between age groups and causes of upper GI bleeding. Mucosal lesions, including esophagitis, gastritis, stress ulcers, peptic ulceration, and Mallory-Weiss tears, are the most common sources of GI bleeding in all age groups (see Chapter 93). Of all cases of upper GI bleeding in children, 95% are related to mucosal lesions and esophageal varices.

Neonatal Period (<1 mo)	Infancy (1-2 yr)	Preschool Age (2-5 yr)	School Age (5-18 yr)
Swallowed maternal blood	Gastritis	Epistaxis	Gastritis
Hemorrhagic gastritis	Esophagitis	Gastritis	Mallory-Weiss tear
Stress ulcer	Mallory-Weiss tear	Esophagitis	Peptic ulcer
Idiopathic	Stress ulcer	Mallory-Weiss tear	Stress ulcer
Bleeding diathesis	Pyloric stenosis	Toxic ingestion	Toxic ingestion
Esophagitis	Vascular malformation	Stress ulcer	Esophagitis
Duplication	Toxic ingestion	Foreign body	Inflammatory bowel disease
Vascular malformations	Duplication	Vascular malformation	Esophageal varices
		Esophageal varices	Vascular malformation
		Hemobilia	Hemobilia

Table 30.1. Etiology of Upper Gastrointestinal Bleeding Based on Age (in order of frequency of occurrence)

Neonatal Period	Gastritis
Swallowed maternal blood	Mallory-Weiss tear
Infancy	*School Age*
Gastritis	Gastritis
Esophagitis	Mallory-Weiss tear
Mallory-Weiss tear	Peptic ulcer
Preschool Age	
Epistaxis	

Table 30.2. Common Causes of Upper Gastrointestinal Bleeding Based on Age

Ulcer	Duplication
Vascular malformation	Esophageal varices

Table 30.3. Life-Threatening Causes of Upper Gastrointestinal Bleeding

Hematemesis in a healthy newborn most likely results from swallowed maternal blood either at delivery or during breast-feeding (i.e., cracked nipples). The Apt test can differentiate neonatal from maternal hemoglobin based on the conversion of oxyhemoglobin to hematin when mixed with alkali. To perform the Apt test, the physician should mix one part bloody stool or vomitus with five parts water, centrifuge at 2000 rpm for 2 minutes, and mix the supernatant with 1 mL of 0.1 N sodium hydroxide. Fetal hemoglobin is more resistant to denaturing than adult hemoglobin and remains pink, whereas maternal hemoglobin becomes brown.

Although rare, hemorrhagic disease of the newborn should be considered with prolongation of the prothrombin time, if vitamin K has not been administered.

Significant and sometimes massive upper GI hemorrhage in a newborn infant may occur with no demonstrative anatomic lesion or only “hemorrhagic gastritis” at endoscopy. This is usually a single, self-limited event that is benign if treated with appropriate blood replacement and supportive measures.

Critically ill children of any age are at risk for developing stress-related peptic ulcer disease. Such ulcers occur with life-threatening illnesses, including shock, respiratory failure, hypoglycemia, dehydration, burns (Curling ulcer), intracranial lesions or trauma (Cushing ulcer), renal failure, and vasculitis. These ulcers may develop within minutes to hours after the initial insult and primarily result from ischemia. Hematemesis, melena, and perforation of a viscus may accompany stress-associated ulcers. Hematemesis secondary to gastroesophageal reflux and esophagitis is uncommon but should be considered in patients who are severely symptomatic with vomiting or aspiration. Hematemesis following the acute onset of vigorous vomiting or retching at any age suggests a Mallory-Weiss tear.

Idiopathic peptic ulcer disease is a common cause of GI bleeding in preschool and older children. Most preschool children with idiopathic ulcers develop GI bleeding (hematemesis or melena). Complications, including obstruction and perforation, may occur. Younger children have less characteristic symptoms, often localize abdominal pain poorly, and may have vomiting as a predominant symptom. Older children and adolescents describe epigastric pain in a pattern typical of adults. *Helicobacter pylori* has emerged as a leading cause of secondary gastritis, particularly in older children. Similar to adults, pediatric patients with evidence of *H. pylori* infection are often treated with antibiotics, bismuth preparations, and H_2 antagonists or omeprazole (Prilosec).

In older children, the possibility of bleeding esophageal varices must be considered in the differential diagnosis of upper GI bleeding. Although variceal bleeding is rare in infancy, esophageal and gastric varices associated with portal hypertension are the most common cause of severe upper GI hemorrhage in older children. One-half to two-thirds of these children have an extrahepatic presinusoidal obstruction, often resulting from portal vein thrombosis, as the cause of portal hypertension. Omphalitis with or without a history of umbilical vein cannulation, dehydration, and a number of other factors may contribute. Other children with portal hypertension have hepatic parenchymal disorders such as neonatal hepatitis, congenital hepatic fibrosis, cystic fibrosis, or biliary cirrhosis associated with biliary atresia. Two-thirds of patients with portal hypertension develop bleeding before 5 years of age, and 85% do so by 10 years of age.

Evaluation and Decision

History and Physical Examination

Pertinent historical elements to be sought include a history of umbilical catheterization or sepsis in the neonatal period, previous episodes of bleeding from the GI tract or other sites, and past hematologic disorders and liver disease. A family history of peptic ulcer disease can be found in up to 30% of patients with idiopathic ulcers. Ingestions should be sought as a possible cause. These include theophylline, aspirin, iron, nonsteroidal anti-inflammatory drugs (NSAIDs), alcohol, and steroids. Massive hemorrhage associated with right upper quadrant pain and jaundice in the posttrauma patient indicates bleeding into the biliary tract (hemobilia).

The physical examination should include visualization of the posterior nose and pharynx to eliminate epistaxis as a source of bleeding. Signs of liver disease or portal hypertension may be subtle in children. Icterus, abdominal distension, prominent abdominal venous pattern, hepatosplenomegaly, cutaneous spider nevi, and ascites suggest liver disease and/or portal hypertension with esophageal varices.

Laboratory Evaluation

Laboratory tests are not useful for identifying a precise cause of upper GI bleeding. Mucosal lesions are more likely than

esophageal varices to be associated with prior occult bleeding. A low mean corpuscular volume (MCV) and hypochromic, microcytic anemia suggest chronic mucosal bleeding. Initial low white blood cell and platelet counts may be seen in either hypersplenism from portal hypertension or sepsis with associated stress mucosal ulceration. Abnormal hepatic studies, including an elevation of serum bilirubin, transaminase, prothrombin time, and a low serum albumin, are suggestive of esophageal varices as a cause of bleeding. A blood urea nitrogen (BUN):creatinine ratio greater than 30 may indicate blood resorption and an upper GI source of bleeding.

Diagnostic Approach

Once it has been determined that a significant upper GI bleed has occurred and hemodynamic stability is restored, identification of the specific disorder is the next step (Fig. 30.2). If the bleeding is mild and self-limited or the gastric aspirate negative, a minor mucosal lesion is likely. Although mucosal lesions such as esophagitis, gastritis, or peptic ulcer disease can present with severe bleeding, most often bleeding from mucosal lesions is self-limiting and will respond to conservative medical management. In patients with persistent or recurrent hemorrhage, emergent endoscopy may be necessary if the bleeding is considered life-threatening (continued transfusion requirement, hemodynamic instability). In a small percentage of patients in whom bleeding is massive, making endoscopic visualization impossible, angiography or radionuclide studies (Technetium-sulfur colloid/Tc-labeled red blood cells) may be indicated. Treatment of specific mucosal conditions and esophageal varices is discussed in Chapter 93.

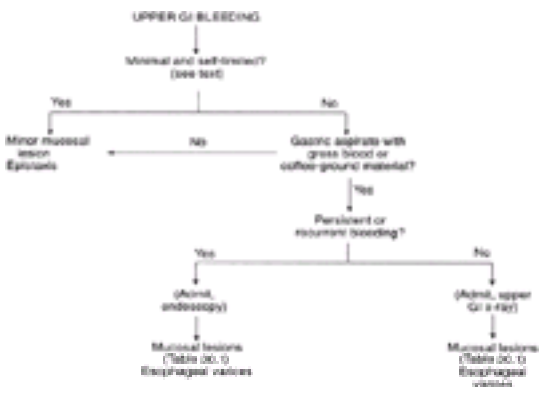

FIGURE 30.2. Diagnostic approach to upper GI bleeding.

Eighty to eighty-five percent of upper GI bleeding stops spontaneously, regardless of the source, before or early in the hospital course. In stable patients who have stopped bleeding, double-contrast barium examination of the upper GI tract and endoscopy provide valuable and often complementary information. In this group of patients, endoscopy need not be performed on an emergent basis and may be done electively in the first 12 to 24 hours after admission. Elective endoscopy should be performed in patients who stop bleeding spontaneously but who have required transfusion and/or have a history of previously unexplained upper GI bleeding episodes.

LOWER GASTROINTESTINAL BLEEDING

Differential Diagnosis

Neonatal Period (0 to 1 Month)

As is true for upper GI bleeding, a common cause of blood in the stool in well infants is the passage of maternal blood swallowed either at delivery or during breast-feeding from a fissured maternal breast. Although hemorrhagic disease of the newborn is uncommon after prophylactic administration of vitamin K at delivery, maternal drugs that cross the placenta, including aspirin, phenytoin, cephalothin, and phenobarbital, may interfere with clotting factors and cause hemorrhage. Infectious diarrhea can occur in very young infants, and stools may contain blood or mucus. Common bacterial pathogens in this age group include *Campylobacter jejuni* and *Salmonella*.

In ill-appearing infants with lower GI bleeding, midgut volvulus, necrotizing enterocolitis, and Hirschsprung's disease should be considered (Table 30.4). Malrotation with midgut volvulus is most common during this period. Initially, bilious vomiting, abdominal distension, and pain are present. Melena is seen in 10 to 20% of patients and signifies vascular compromise. Of all cases of necrotizing enterocolitis, 10% occur in term infants. These patients can present with nonspecific signs of sepsis (temperature instability or apnea and bradycardia) as well as with specific GI tract findings, such as abdominal distension, pain, and abdominal wall erythema. GI bleeding can be in the form of guaiac-positive or grossly bloody stools. Hirschsprung's disease with enterocolitis may present with GI bleeding in the newborn period. Enterocolitis has recently been shown to occur in up to 25% of children with Hirschsprung's disease. The risk of enterocolitis remains high until about 6 months of age. The diagnosis should be considered in any newborn who does not pass meconium in the first 24 to 48 hours of life.

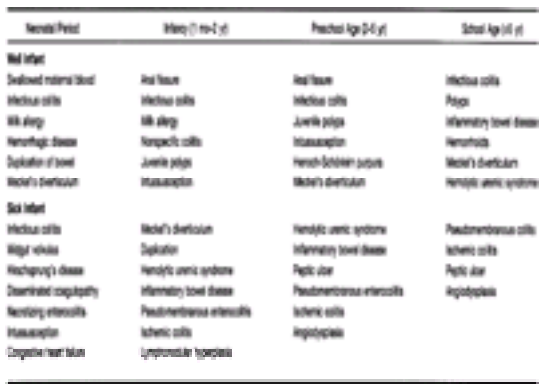

Table 30.4. Etiology of Lower Gastrointestinal Bleeding Based on Age (in order of frequency of occurrence)

Other common causes of lower GI bleeding are listed in Table 30.5. Life-threatening causes are listed in Table 30.6.

Neonatal Period	*Preschool Age*
Swallowed maternal blood	Anal fissure
Infectious colitis	Infectious colitis
Milk allergy	*School Age*
Infancy	Infectious colitis
Anal fissure	
Infectious colitis	
Milk allergy	

Table 30.5. Common Causes of Lower Gastrointestinal Bleeding Based on Age

Midgut volvulus	Pseudomembranous colitis
Intussusception	Ischemic colitis
Meckel's diverticulum	Peptic ulcer
Hemolytic uremic syndrome	

Table 30.6. Life-Threatening Causes of Gastrointestinal Bleeding

Infancy (1 Month to 2 Years)

In the first 2 years of life, anal fissures are the most common cause of rectal bleeding and are usually associated with hard stools or constipation. Treatment with stool softeners and sitz baths will often resolve the problem spontaneously in most patients. Milk or soy enterocolitis usually occurs during the first month of life or shortly thereafter. These infants can present with chronic diarrhea, stools containing blood or mucus, or less commonly, fulminant colitis and shock. Milk-protein allergy responds to a change in formula from cow's milk or soy protein to a casein hydrolysate (Nutramigen, Alimentum, Pregestimil). Breast-fed infants whose mothers drink cow's milk may develop an allergic colitis that responds to removal of cow's milk from the mother's diet. Infectious enterocolitis as a cause of bloody diarrhea is common in all age groups. Bacterial causes (*Salmonella, Shigella, Campylobacter*, pathogenic *Escherichia coli,* and *Yersinia enterocolitica*) should be identified with stool cultures. In symptomatic infants and children, the presence of leukocytes in a stool smear for white cells may aid in preliminary diagnosis. Pseudomembranous colitis should be considered in any infant or child with bloody stools and a history of recent antibiotic therapy. “Nonspecific colitis” has been demonstrated to be a common cause of hematochezia in infants less than 6 months of age. Although the cause of nonspecific colitis is unknown, it may represent a variation in the colonic response to viral invasion.

Meckel's diverticulum should be suspected in infants or young children who pass bright or dark red blood per rectum. Intermittent painless bleeding or massive GI hemorrhage can occur. Sixty percent of complications from Meckel's diverticulum (hemorrhage and intestinal obstruction) occur in patients less than 2 years of age.

Idiopathic intussusception may occur in children 3 months to 1 year of age, with 80% occurring before 2 years of age. In children older than 3 years, a lead point (polyp, Meckel's diverticulum, or hypertrophied lymphoid patch) is often found. Paroxysmal pain may be associated with guaiac-positive stools or hematochezia. Lethargy alone (without pain) has been increasingly recognized as a presenting symptom of intussusception in young children. Lymphonodular hyperplasia is a

common cause of rectal bleeding in this age group and may cause mild, painless hematochezia. The nodular lymphoid response is self-limited and does not require any specific therapy. Intestinal duplications are an uncommon cause of lower GI bleeding and, when diagnosed, are usually found in children less than 2 years of age. Duplications can be found anywhere in the GI tract but are most common in the distal ileum. These usually present with obstruction and lower GI bleeding.

Preschool (2 to 5 Years)

The two conditions most likely to cause bleeding in children 2 to 5 years of age are juvenile polyps and infectious enterocolitis. Most polyps in childhood are inflammatory without significant malignant potential and are often multiple. Thirty to forty percent are palpable on rectal examination. Polyps may cause painless rectal bleeding in this age group. Significant bleeding is unusual. Infectious causes of colitis are similar to those discussed in younger age groups. Hematochezia is often a manifestation of systemic disease in infancy and throughout childhood. Hemolytic uremic syndrome is the most prevalent of these conditions reported in infants and children up to 3 years of age. Bloody diarrhea may precede the development of renal and hematologic abnormalities. GI manifestations of Henoch-Schönlein purpura (HSP) occur in 50% of patients and include colicky abdominal pain, melena, and bloody diarrhea. These symptoms precede the characteristic rash in 20% of patients. GI complications of HSP include hemorrhage (5%), intussusception (3%), and rarely, intestinal perforation.

Angiodysplasia is a rare cause of GI bleeding but can be associated with massive hemorrhage. Vascular lesions of the GI tract probably have a congenital basis. Several recognized syndromes, including Rendu-Osler-Weber syndrome, blue rubber bleb syndrome, and Turner's syndrome may be associated with intestinal telangiectasia.

School Age (5 Years through Adolescence)

For the most part, the diagnostic considerations relevant to the preschool child apply to school-age and adolescent children with the addition of inflammatory bowel disease. Although inflammatory bowel disease occurs in younger children, it is rare before the age of 10 years. Rectal bleeding is a common presentation of both ulcerative colitis and Crohn's disease. Massive lower GI bleeding occurs in 2 to 5% of children with Crohn's disease. Toxic megacolon is a life-threatening presentation of both ulcerative colitis and Crohn's disease.

Evaluation and Decision

History and Physical Examination

Symptoms of an acute abdominal process, including abdominal pain, distension, and vomiting, should be elicited. A history of bloody diarrhea may indicate infectious colitis, intussusception, or hemolytic uremic syndrome. Extraintestinal manifestations of inflammatory bowel disease, including weight loss, anorexia, and arthralgias, may be predominant symptoms in school-aged children. The dietary history may suggest features of a protein intolerance (cow's milk). Firm stool streaked with red blood characterizes anal fissures. A detailed family history (bleeding diathesis, familial polyposis) and drug history (NSAIDs, salicylates, iron) or antibiotics (pseudomembranous colitis) is important in patients with lower GI bleeding. Long-standing constipation with acute onset of bloody diarrhea suggests enterocolitis associated with Hirschsprung's disease.

Physical examination to detect abdominal obstruction (abdominal tenderness, distension, palpable mass, peritoneal signs, hyperactive bowel sounds) is the most urgent task of the evaluating physician. Careful separation of the buttocks with eversion of the anal mucosa may reveal a fissure. Prominent or multiple perianal skin tags may raise suspicion of Crohn's disease. Rectal polyps may be palpable. Cutaneous lesions may provide important diagnostic clues in patients with GI bleeding. Eczema may be associated with milk allergy, whereas erythema nodosum is the most common skin manifestation of inflammatory bowel disease. Mucocutaneous pigmentation (Peutz-Jegher syndrome) and cutaneous or subcutaneous tumors (Gardner's syndrome) indicate intestinal polyposis.

Diagnostic Approach

Rectal bleeding is a common complaint in the pediatric age group (Fig. 30.3). The causes of lower GI bleeding vary significantly with age and often are transient and benign. Occasionally, lower GI bleeding reflects a life-threatening pathologic condition, and establishment of a specific diagnosis becomes urgent.

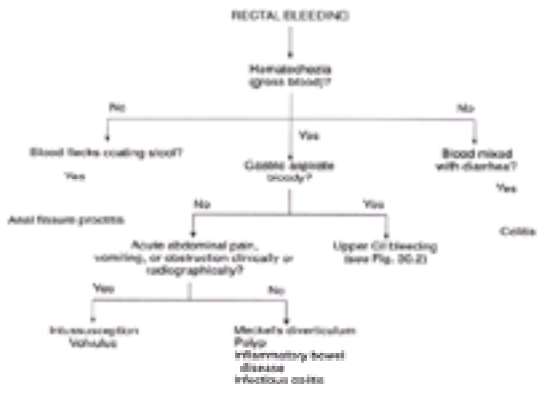

FIGURE 30.3. Diagnostic approach to lower GI bleeding.

The priority of the emergency physician in evaluating the patient with lower GI bleeding is to identify lower tract bleeding associated with intestinal obstruction. Intussusception and a late presentation of midgut volvulus secondary to malrotation are the major types of intestinal obstruction associated with lower GI hemorrhage. All causes of abdominal obstruction (e.g., adhesions, incarcerated hernia, appendicitis) eventually cause bleeding, however, if diagnosis is delayed and vascular compromise occurs.

Severe lower GI bleeding leading to hemodynamic instability or requiring transfusion is rare in pediatrics, and gastric lavage is essential in these cases to rule out a possible source in upper GI tract. Meckel's diverticulum is the most common cause of severe lower GI bleeding in all age groups. Following Meckel's diverticulum, Crohn's disease and arteriovenous malformation are prominent causes of massive lower GI bleeding in adolescents.

The urgency and extent of evaluation of patients with lower GI bleeding will depend on the amount of bleeding, the patient's age, and associated physical findings. In a healthy infant with a few streaks of blood in the stool and a normal examination, stool culture and observation are reasonable. If hematochezia is found and nasogastric aspirate is negative, significant pathology must be sought. Flat and upright abdominal radiographs should be performed if an obstructive process (e.g., intussusception, volvulus) is suspected by history or physical examination. The absence of radiographic findings should not, however, deter the physician from pursuing further diagnostic evaluation. A barium or air enema examination is diagnostic and often therapeutic in children with intussusception. If obstruction is not considered likely, the decision to perform contrast enema examination or colonoscopy will depend on the diagnosis suspected. Air-contrast barium enema is extremely valuable in the detection of polyps or inflammatory bowel disease. Indications for colonoscopy include severe bleeding, moderate but persistent bleeding with a negative double-contrast barium enema, or a lesion of unknown nature seen on barium enema. If undefined bleeding persists, radionuclide studies or angiography should be considered. A technetium scan may detect ectopic gastric mucosa as seen in Meckel's diverticulum, whereas angiography will help identify bleeding vascular malformations in the GI tract. Ongoing, undiagnosed GI hemorrhage accounts for fewer than 10% of cases in infants and children. Exploratory laparotomy may be necessary and lifesaving in these circumstances.

SUMMARY

Management of acute GI bleeding often requires a team approach, including the emergency physician, surgeon, and gastroenterologist. The foremost goals of ED evaluation of patients with GI bleeding are establishment of hemodynamic stability and determination of level of bleeding. All patients with nontrivial upper GI bleeding should be admitted for observation and further evaluation. If an acute abdominal process is suspected, surgical consultation and diagnostic workup should be instituted. If rectal bleeding is mild and self-limited and the history and physical are unremarkable, further investigation with a gastroenterologist is recommended.

Suggested Readings

Gryboski JD. Peptic ulcer disease in children. Pediatr Rev 1990;12:15–21.

Kirks DR, Caron KH. Gastrointestinal tract. In: Kirks DR, ed. Practical Pediatric Imaging: Diagnostic Radiology of Infants and Children. 2nd ed. Boston: Little, Brown, 1991.

Latt TT, Nicholl R, Domizio P, et al. Rectal bleeding and polyps. Arch Dis Child 1993;69:144–147.

Luks FI, Yzabeck S, Perreault G, et al. Changes in presentation of intussusception. Am J Emerg Med 1992;10:574–576.

McRury JM, Barry RC. A modified Apt test: a new look at an old test. Pediatr Emerg Care 1994;10:189–191.

Mondrgon LT, Gomez MA, Tiscarreno MA, et al. Angiodysplasia of the colon in children. J Pediatr Surg 1995;30:72–75.

O'Hara S. Acute gastrointestinal bleeding. Radiol Clin North Am 1997;35(4):897–895.

Silber G. Lower gastrointestinal bleeding. Pediatr Rev 1990;12:85–92.

Teach SJ, Fleisher GR. Rectal bleeding in the pediatric emergency department. Ann Emerg Med 1994;23(6):1252–1258.

Vinton NE. Gastrointestinal bleeding in infancy and childhood. Gastroenterol Clin North Am 1994;23:93–122.

CHAPTER 31

Groin Masses

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Children occasionally present to the emergency department (ED) with an inguinal mass. It may be noticed during a diaper change, or the older child may bring it to the parent's attention; sometimes an adolescent arrives alone seeking help. There are many different causes, ranging from inconsequential to serious (Table 31.1). One generally can ascertain the correct diagnosis based on the age and sex of the child, the location of the mass, how painful it is, how rapidly it has evolved, and whether there are any associated symptoms or signs (Fig. 31.1). Admittedly, such categorization is imperfect; however, we believe it is useful nonetheless. Lymph node enlargement and retractile, undescended, and traumatically dislocated testes are discussed in this section. Hernia and hydrocele (as well as scrotal masses) are addressed in Chapter 58 and Chapter 118.

Painful
Torsion of an undescended testicle[a]
Trauma (e.g., dislocated testicle)[a]
Incarceration or strangulation of an indirect inguinal hernia[a]
Lymphadenitis
Usually or Comparatively Painless
Hernia
Hydrocele
Lymphadenopathy
Retractile or undescended testicle

[a]Urgent or emergent condition.

Table 31.1. Causes of Inguinal Masses

FIGURE 31.1. A. Groin masses in boys. **B.** Groin masses in girls.

DIFFERENTIAL DIAGNOSIS

Lymphadenopathy and Lymphadenitis

There are two groups of inguinal nodes: superficial and deep. The superficial ones can be subdivided into a horizontal group that runs parallel to the inguinal ligament and a vertical group located lateral to it. The horizontal group drains lymph from 1) the skin of the lower abdominal wall, perineum, and gluteal region; 2) the skin of the penis and scrotum; 3) the mucosa of the vagina; and 4) the lower anal canal. The vertical group drains lymph from 1) the gluteal region; 2) the

penis and deep structures of the scrotum; 3) the anterior and lateral areas of the thigh and leg; and 4) the middle and medial portions of the foot. The deep inguinal nodes, which lie beneath the fascia lata medial to the femoral vein, drain 1) all of the superficial nodes; 2) the clitoris or glans of the penis; 3) the medial areas of the thigh and leg; and 4) the lateral portion of the foot.

A healthy child can have a few small nodes normally. These tend to be less than half a centimeter long, and they are oval, firm, slightly moveable, and nontender. If the nodes are enlarged (especially unilaterally) or tender, erythematous, or suppurating, further evaluation is necessary (see Chapter 44).

Inguinal adenopathy—nodes that are enlarged but nontender—often is part of a more generalized lymphadenopathy. The list of causes of generalized lymphadenopathy is extensive and includes collagen vascular diseases (e.g., juvenile rheumatoid arthritis, serum sickness), immunologic disorders (e.g., chronic granulomatous disease), metabolic diseases (e.g., Gaucher's disease, Niemann-Pick disease), and certain anemias (e.g., sickle cell disease, thalassemia). Inguinal nodes may be enlarged because of malignancy (e.g., acute lymphocytic leukemia), but this is rarely the sole presentation of a malignant tumor. Of note, some local tumors, such as testicular tumors, metastasize to the inguinal nodes. Although many infections, particularly viral ones (e.g., human immunodeficiency virus, Epstein-Barr virus), produce inguinal adenopathy, these usually cause generalized lymphadenopathy as well as hepatosplenomegaly and other abnormalities.

Inflammation or infection of the gluteal region, perineum, genitalia, or ipsilateral lower extremity is the most common cause of isolated inguinal adenopathy or adenitis. These areas must be examined carefully. Chronic eczema, tinea cruris, or an innocuous inflammation (an insect bite or diaper rash) may produce lymphadenopathy. In such cases, treatment of the underlying condition suffices. If lymphadenitis—enlargement with tenderness, erythema, or suppuration—is detected, the node itself is probably infected. Group A b-hemolytic streptococcus, *Staphylococcus aureus,* or an enteric organism is the usual pathogen, depending on the site of the primary infection. A culture from the primary site or node aspirate helps identify the organism. Most children can be treated as outpatients with oral antibiotics (e.g., cephalexin or amoxicillin–clavulanate), but children with severe symptoms should be admitted and treated with intravenous antibiotics (e.g., oxacillin, cefazolin). Abscesses caused by these pathogens should be incised and drained (see Chapter 84).

Venereal diseases can result in inguinal adenopathy or adenitis in adolescents (see Chapter 84). Herpes is a common cause of genital ulcerations and bilaterally enlarged, painful lymph nodes. Enlarged lymph glands may precede the appearance of vesicles occasionally. Oral acyclovir shortens the duration of symptoms and viral shedding in primary disease but is less effective in recurrences.

The chancre of primary syphilis is painless and indurated and has raised, firm borders, a shallow, smooth base, and no exudate. Bilateral (70%) and unilateral, nontender inguinal adenopathy is common. A positive rapid plasma reagin (RPR) confirms the diagnosis, but this test is nonreactive in up to 30% of patients with primary syphilis. Recommended treatment for primary syphilis is benzathine penicillin G 2.4 million units intramuscularly.

Chancroid is more common in developing countries than in the United States. It is caused by *Haemophilus ducreyi,* which is hard to isolate and requires selective media. Unlike syphilis, the chancroid ulcer is painful and soft and has ragged edges and a deep, friable base covered with a dirty yellow exudate. About half of patients develop painful adenitis, usually unilaterally. The node or nodes often suppurate and drain spontaneously. The preferred treatment is azithromycin 1 g orally or ceftriaxone 250 mg intramuscularly.

Lymphogranuloma venereum, which occurs mostly in tropical and subtropical countries, is caused by *Chlamydia trachomatis.* The genital papule, vesicle, or ulcer often is missed because it is painless, inconspicuous, and transitory. One or more unilaterally enlarged, moderately tender, fluctuant nodes are characteristic. If left untreated, these nodes drain and form fistulae. Three weeks of treatment with doxycycline, erythromycin, or a sulfonamide is necessary.

Granuloma inguinale is caused by the Gram-negative bacillus *Calymmatobacterium granulomatis.* Granuloma inguinale is rare in the United States but if untreated results in extensive subcutaneous granulomas (pseudobuboes), which mimic suppurative lymph glands. The initial small, relatively painful, red nodule or vesicle progresses to a red mass of granulomatous tissue, which ulcerates and coalesces. Both tetracycline and trimethoprim–sulfamethoxazole are somewhat effective.

An enlarged, tender inguinal node can be caused by plague, brucellosis, tularemia, or cat-scratch disease if the portal of entry for the infective organism is the lower extremity. *Yersinia pestis,* which causes plague, is extremely rare in the United States and is commonly transmitted by flea bites. The buboes are firm and nonfluctuant but extremely tender. The overlying skin is often warm and edematous. Mortality can be as high as 80%. Streptomycin and gentamycin are the drugs of choice for children.

Inguinal lymphadenopathy can be seen in tularemia when it is caused by an infected tick bite. Enlarged tender lymph nodes precede the appearance of a small papule that later ulcerates. Streptomycin and gentamycin are the preferred treatments in children.

Infection with *Bartonella henselae* (cat-scratch disease) results in regional lymphadenopathy that is usually red, indurated, and warm. Usually, the lymphadenopathy resolves spontaneously within 2 to 4 months. Antibiotic treatment is of questionable value.

Filariasis, which is found in the tropics, can produce adenopathy or adenitis associated with lower extremity lymphedema and scrotal pathology.

Retractile, Undescended, or Traumatically Dislocated Testes

If the inguinal mass is firm, oval, and nontender and is associated with an empty scrotum, it probably is a retractile or undescended testis. A retractile testis, in contrast to a truly undescended one, is pulled into its abnormally high position by a hyperactive cremasteric reflex and can be milked back into the scrotum by the examiner. The scrotum appears fully developed when the testicle is retractile. Although it may retract again, it will ultimately assume a normal position; therefore, no treatment is needed.

An undescended testicle is found in 2 to 4% of term boys. As expected, cryptorchidism is about 10 times more common in premature boys. Its incidence correlates inversely with gestational age; for example, practically all babies who weigh less than 900 g are cryptorchid. The incidence falls to 0.7% by 1 year of age. Because this is similar to the incidence in adult men, it seems that spontaneous descent occurs rarely after 1 year of age. The testis can lodge anywhere along its natural line of descent—for example, intra-abdominally, in the inguinal canal, or just outside the external inguinal ring. It also may be discovered in an ectopic location—for example, in a superficial pouch near the external ring or, less commonly, in the abdominal, suprapubic, perineal, or femoral areas. Cryptorchidism is right-sided in 50% of patients, left-sided in 25%, and bilateral in 25%. There is a right-sided predominance because the right testicle descends later than the left one during embryologic development. Bilaterally undescended testicles occur more often in premature boys and in conjunction with some anatomic, enzymatic, and chromosomal disorders that are encountered in the delivery suite or shortly thereafter. There is an increased occurrence of cryptorchidism among family members.

If left untreated, various complications, including testicular atrophy, infertility, malignancy, torsion, injury related to trauma, and development of a hernia, can ensue. Germ cell depletion may be visible by as early as the first birthday; other degenerative changes, such as Leydig cell atrophy, smaller seminiferous tubules, and peritubular fibrosis, follow. These manifestations are worse in testicles located more proximally. Interestingly, these histologic abnormalities also develop in the normally descended testicle, although later and less severely. Adults with untreated bilateral cryptorchidism generally are sterile, but those with untreated unilateral cryptorchidism are somewhat infertile as well. Compared with men who have normally descended testicles, the incidence of malignancy is increased 22-fold. The malignancy usually is a seminoma, presenting in the third decade of life. Although intra-abdominal testes are most at risk, cancer also occurs more often in the contralateral descended testis. An undescended testicle is particularly prone to torsion and more likely to be injured from trauma. In such cases, if it is located in the inguinal region, it presents as a painful groin mass. Finally, 90% of undescended testicles are associated with a patent processus vaginalis, increasing the chance that a hernia will develop.

Early referral to a urologist is warranted. Currently, most urologists recommend orchiopexy between 1 and 2 years of age. At surgery, the testis, spermatic cord, and vascular structures are mobilized and brought down into the scrotum, where the testis is either pexed or placed in a dartos pouch; in addition, the processus vaginalis is ligated if it is patent. Although it is hoped that earlier surgery will lessen the incidences of infertility and malignancy, this has not yet been proved conclusively. Orchiopexy facilitates diagnosis of a malignancy because the testis can be palpated more easily in the scrotal sac. Hormonal therapy (e.g., human chorionic gonadotropin [HCG]) may be beneficial in a few select cases; however, studies from the United States have demonstrated that it is not particularly effective, especially when retractile testes are excluded. Some clinicians report that it does not work as well in younger boys, and it also is ineffective if the testis is located ectopically. Many pediatric endocrinologists and urologists use it mainly to differentiate retractile from undescended testes.

Finally, a traumatically dislocated testicle may be discovered in the groin. Testicular dislocation occurs primarily in the older adolescent and young adult but is uncommon even then. It usually follows major trauma—in particular, a motor vehicle accident. An associated injury, such as a pelvis or femur fracture, often is found. Despite swelling, ecchymosis, and tenderness, the scrotum feels empty. As mentioned, sometimes the testis is palpated in an abnormal location, most often in the groin in the superficial pouch anterior to the external oblique aponeurosis. At surgery, the testis is reduced into the scrotum and pexed to the wall.

EVALUATION AND DECISION

In evaluating groin masses, one should consider the sex of the child, presence or absence of pain, status of the testes (in boys), response to attempted reduction, history of trauma, and findings of local infection.

Boys (Fig. 31.1A)

Focusing first on boys, pain often heralds a potentially emergent condition, including torsion of an undescended testis, an incarcerated or strangulated hernia, or a significant injury. One begins the evaluation by carefully palpating the scrotum. An empty scrotum points to a dislocated testis after trauma or spontaneous torsion of an undescended testis. In a boy with bilaterally descended testes, an isolated, painful groin mass may represent an incarcerated or strangulated inguinal hernia. The finding of penile lesions, such as those of herpes or syphilis, or obvious signs of inflammation (erythema, fluctuance) suggest inguinal lymphadenitis.

Painless groin masses in boys are usually not urgent. If the mass is reducible, it is an inguinal hernia, which calls for elective surgical repair. The absence of a testis in the scrotum on the side of the mass suggests a retractile or undescended testis. A retractile testis is more likely in the boy presenting with new-onset swelling; the diagnosis can be confirmed in most cases by “milking” the testis into the scrotum. When both testes are descended, a painless mass is likely to be either a hydrocele or an enlarged lymph node. One must keep in mind that a recently incarcerated hernia may be painless and is easily confused with a solitary, enlarged lymph node.

Girls (Fig. 31.1B)

As for girls, one can approach the diagnosis of groin masses by first ascertaining the presence or absence of pain. The highest priority in girls with painful masses is to identify an incarcerated or strangulated hernia. Local lesions or signs of

inflammation point to lymphadenitis.

Although hernias occur less often in girls than in boys, they are still relatively common. The ability to reduce a mass with gentle pressure confirms this diagnosis. When a painless mass is irreducible, a recently incarcerated hernia (particularly involving an ovary) or an enlarged lymph node represent the most likely causes.

Suggested Readings

LYMPHADENOPATHY AND LYMPHADENITIS

Darville T, Jacobs RF. Lymphadenopathy, lymphadenitis, and lymphangitis. In: Jenson H, Baltimore RS, eds. Pediatric Infectious Diseases: Principles and Practice. East Norwalk, CT: Appleton & Lange, 1995:751.

McAllister K, Kazragis R. Cutaneous lesions of the male genitalia. In: Harwood-Nuss A, Linden CH, Luten RC, et al, eds. The Clinical Practice of Emergency Medicine. 2nd ed. Philadelphia: Lippincott Raven, 1996:263.

Peter G, ed. 1997 Red Book: Report of the Committee on Infectious Diseases. 24th ed. Elk Grove Village, IL: American Academy of Pediatrics, 1997.

Tunnesen WW Jr. Lymphadenopathy. In: Signs and Symptoms in Pediatrics. Philadelphia: JB Lippincott, 1983:45.

RETRACTILE AND UNDESCENDED TESTICLE

Bloom DA, Wan J, Key D. Disorders of the male external genitalia and inguinal canal. In: Kelalis PP, King L, Belman B, eds. Clinical Pediatric Urology. Philadelphia: WB Saunders, 1992:1015.

Colodny AH. Undescended testes: is surgery necessary? N Engl J Med 1986;314:510.

Grosfeld JL. Current concepts in inguinal hernia in infants and children. World J Surg 1989;13:506.

Kogan S. Cryptorchidism. In: Kelalis PP, King L, Belman B, eds. Clinical Pediatric Urology. Philadelphia: WB Saunders, 1992:1050.

Neely EK, Rosenfeld RG. The undescended testicle: when and how to intervene. Contemp Pediatr 1990;7:21.

Rajfer J, Handelsman DJ, Swerdloff RS, et al. Hormonal therapy of cryptorchidism. N Engl J Med 1986;314:466.

Rozanski TA, Bloom DA. The undescended testis: theory and management. Urol Clin North Am 1995;22:107.

SCROTAL TRAUMA

Munter DW, Faleski EJ. Blunt scrotal trauma: emergency department evaluation and management. Am J Emerg Med 1989;7:227.

CHAPTER 32

Hearing Loss

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Normal hearing plays a critical role in the proper development of speech and language. Any persistent hearing loss will distort a child's perception of expressive speech and language and will compromise the ability to attain normal language. Failure to recognize hearing impairment can negatively impact school performance, socialization, and emotional development. Acute hearing loss is of two main types: conductive and sensorineural. Several serious—even life-threatening—disorders can accompany acute hearing loss. Therefore, prompt clinical evaluation is mandated when hearing loss is suspected.

Hearing loss can occur as an isolated symptom or in association with auditory or central nervous system (CNS) dysfunction. The differential diagnosis of hearing loss includes congenital and acquired causes. It may be produced by the abnormal transmission of sound waves to the inner ear (conductive hearing loss) or by the defective processing of sound waves (sensorineural hearing loss) in the inner ear (Table 32.1). In young children, the possibility of acute hearing loss may be suspected by parents when the child does not respond to noise or to simple commands. Abnormal or delayed language development may be a sign of a more chronic process. Older children and adolescents may complain directly of hearing difficulty.

Table 32.1. Differential Diagnosis of Hearing Loss

PATHOPHYSIOLOGY

An intricate series of properly aligned anatomic and physiologic connections is responsible for the precise functioning of the auditory system. When sound reaches the tympanic membrane, vibrations of the membrane are transmitted to the ossicles within the middle ear and from there to the cochlea, which is located in the inner ear. Within the cochlea, specialized receptors in the form of hair cells in the spiral organ of Corti convert this mechanical energy to nerve impulses that are transmitted to the CNS by way of the cochlear (acoustic) nerve, the auditory portion of the eighth cranial nerve. Anatomically, the acoustic apparatus is closely related to the vestibular system, which is concerned with the proprioceptive senses of posture and equilibrium. The three semicircular canals of the vestibular system are connected to the cochlear system; therefore, abnormalities in the inner ear may cause auditory and vestibular symptoms.

DIFFERENTIAL DIAGNOSIS

Conductive Hearing Loss

In children, conductive hearing loss occurs when there is a decrease in the transmission of sound waves to the cochlea from an external source. Commonly, middle ear effusion (acute or chronic), impacted cerumen, foreign body in the external ear canal, and ossification of the middle ear ossicles produce conductive hearing loss (Table 32.1). In children with chronic recurrent otitis media (OM), a cholesteatoma—an epidermal inclusion cyst of the middle ear—may develop and cause a slowly progressive conductive hearing loss. Acute head injury, especially in association with a basilar skull

fracture, may produce a conductive hearing loss secondary to hemotympanum, rupture of the tympanic membrane, or disruption of the inner ear ossicles. Rarely, the conductive hearing loss may be secondary to malformations of the external or middle ears, such as absence of the external ear canal.

Congenital Sensorineural Hearing Loss

Approximately 1 of every 750 infants is born with congenital hearing loss. Diagnostic possibilities include genetic disorders, chromosomal abnormalities, metabolic and storage diseases, and abnormal development of the auditory apparatus (Table 32.1). Congenital hearing loss secondary to aplasia of the inner ear (Michel's aplasia) and abnormal cochlear development (Mondini's aplasia) or absence of parts of the cochlear apparatus (Scheibe's aplasia, Alexander's aplasia) are reported in children. Sensorineural hearing loss has been described in more than 70 syndromes, including Waardenburg syndrome (facial dysmorphism, white forelock), Jervell and Lange-Nielsen syndrome (prolonged Q-T syndrome), Usher's syndrome (retinitis pigmentosa and sensorineural hearing loss), and Alport's syndrome (nephritis, optic abnormalities, and hearing loss). The chromosomal disorders caused by trisomies (especially trisomies 13, 14, 15, 18, and 21) are associated with defects in hearing. Many of these patients are diagnosed because of anatomic features associated with each of these disorders, although the hearing loss that occurs may be present at birth or develop over time. Individual patients with presumed idiopathic sensorineural hearing loss have a recurrence rate in siblings of 10%, much closer to the 25 to 50% rate of an inherited disorder than the overall risk of 1 in 750 for the general population. These data support the theory that the etiology of presumed isolated causes of sensorineural hearing loss may be genetic in origin.

Acquired Sensorineural Hearing Loss

Although acquired sensorineural hearing loss occurs less commonly than congenital hearing loss, the absence of associated symptoms may make it a more difficult diagnosis. An array of clinical problems can produce sensorineural hearing loss during childhood.

Acute Infection

Hearing loss secondary to bacterial meningitis is the most common cause of acquired sensorineural hearing loss. Reported in 15 to 20% of patients with meningitis, the hearing loss is usually profound and often bilateral. It is most commonly associated with infections caused by *Haemophilus influenzae*, *Streptococcus pneumoniae*, or *Neisseria meningitidis* and can develop despite appropriate antimicrobial therapy. Vaccine programs that have led to a decrease in the incidence of *H. influenzae* infections have been instrumental in decreasing the occurrence of this complication. Hearing loss from meningitis is thought to result from direct toxicity of the cochlear hair cells, and recent studies suggest adjunctive therapy with dexamethasone may decrease the incidence of this complication.

Untreated congenital syphilis is another infection associated with acquired sensorineural hearing loss. Hearing loss has also been described with congenital infections caused by rubella, toxoplasmosis, cytomegalovirus (CMV), and perinatally acquired herpes simplex infections.

Viral infection of the labyrinth (also called viral cochleitis) secondary to mumps, parainfluenzae, adenovirus, herpes simplex, CMV, and rubeola have been described and confirmed by serologic studies. Labyrinthitis usually has symptoms related to inflammation of the inner ear and involvement of the vestibular apparatus, and patients may complain of vomiting, tinnitus, and vertigo.

Vascular Insufficiency

Sudden hearing loss secondary to vascular insufficiency has been described in the pediatric patient. Vascular insufficiency may compromise blood flow to the cochlea, producing a hypoxic insult to the sensitive nerve cells in the organ of Corti. Once injured, these nerve cells may not regenerate and profound sensorineural hearing loss can develop. In children, sickle cell disease, long-standing diabetes mellitus, and hyperviscosity states associated with polycythemia can compromise cochlear blood flow and produce sudden hearing loss.

Perilymphatic Fistula

Anatomic defects in the bony or membranous enclosure that normally surrounds the perilymphatic space can produce a perilymphatic fistula. These defects may produce an anomalous communication between the middle and inner ear compartment and should be considered in the differential diagnosis of the pediatric patient with acute sensorineural hearing loss. A perilymphatic fistula can occur at any age. The sudden ingress of air into the inner ear is thought to produce the symptom complex of hearing loss, tinnitus, vertigo, dizziness, and nystagmus. Trauma usually underlies the development of a fistula. This is especially true in any patient who has a recent history of vigorous exercise or changes in barometric pressure associated with airplane travel or scuba diving. Although unilateral hearing loss is most common, bilateral hearing deficits have been described. Occasionally, a perilymphatic fistula will develop in a child with previously abnormal hearing. Therefore, this diagnosis needs to be considered in any patient who has sudden onset of hearing loss, fluctuation in hearing, or complaints of progressive hearing loss, regardless of baseline hearing function. Patients with a perilymphatic fistula generally have a normal otoscopic examination. If a tympanogram is performed, middle ear effusion usually is absent. Emergent referral to an otolaryngologist is warranted because surgery may be required for closure of the anatomic defect.

Head Trauma

Both the vestibular and cochlear nerves can be injured with fractures of the temporal bone. Assessment of audiologic function should be considered in any child with major head trauma; computed tomography (CT) scan may be required to

diagnose these injuries.

Acoustic Trauma

Immediate, severe, and permanent hearing loss can follow even a short period of exposure to sounds greater than 140 dB. Exposure to sounds in the 80- to 100-dB range can produce hearing loss with chronic exposure and is most commonly diagnosed in adolescents. Rock concerts, stereo headphones, machinery, and explosive devices are capable of producing sound at the intensity required to produce this condition.

Chronic/Recurrent Otitis Media

A history of chronic/recurrent OM may predispose patients to the development of sensorineural hearing loss. This hearing loss is thought to be related to inflammatory changes in the inner ear that are produced by the diapedesis of toxins through the round window membrane. Such involvement of the inner ear has been confirmed pathologically by the presence of labyrinthitis in patients with acute OM. Because middle ear effusions produce a conductive hearing loss, the clinician must be aware of the possibility of OM producing a mixed picture.

Miscellaneous

Acoustic neuroma, CNS tumors, and leukemic infiltrates also are associated with sensorineural hearing loss. Other considerations in pediatric patients include Kawasaki disease, hypothyroidism, lightning injury, hyperlipidemia, and the use of ototoxic drugs in the neonatal period. Finally, some children will have no demonstrable cause for their hearing loss.

EVALUATION AND DECISION

Any complaint of hearing loss requires prompt evaluation. Common causes of acute hearing loss are listed in Table 32.2. Life-threatening causes of acute hearing loss are rare in pediatric patients (Table 32.3). The initial step in evaluation is to confirm the presence of acute hearing loss (Fig. 32.1). Although sophisticated hearing tests are best performed by an audiologist, the emergency physician should attempt bedside testing of gross hearing function. In young children, behavioral responses to loud stimuli can be assessed. Without attracting visual attention, an auditory stimulus (e.g., vigorous hand clapping or ringing a bell) can be presented to the child. Eye blinking or turning toward the stimulus represents a positive response and suggests some degree of intact hearing. In older children or adolescents, hearing can be assessed by asking the patients whether they hear a low-intensity sound such as a watch ticking or fingers rubbing together. Because hearing dysfunction can be subtle and can occur over the entire range of auditory frequencies, these bedside tests may underestimate the degree of hearing impairment. Therefore, an abnormal test should be considered a confirmation of hearing impairment; a negative test needs to be interpreted in the context of the chief complaint of the patient. If the history remains strongly suggestive of hearing loss, the physician should assume some degree of hearing loss despite the results of bedside testing.

FIGURE 32.1. Evaluation of hearing loss.

Conductive Hearing Loss	Sensorineural Hearing Loss
Middle ear effusion	TORCH infections
Impacted cerumen	Birth asphyxia
Foreign body of external ear canal	Acute otitis media
	Viral labyrinthitis
	Bacterial meningitis
	Perilymphatic fistula
	Trauma
	Acoustic neuroma

Table 32.2. Common Causes of Acute Hearing Loss

Acute head injury	Leukemic infiltrate
Brain tumor	Vascular insufficiency

Table 32.3. Life-Threatening Causes of Acute Hearing Loss

Critical elements of the medical history should include the onset of the hearing loss and the duration of symptoms. Family history of hearing loss may suggest the diagnosis of a genetic disorder with delayed presentation of hearing loss. A history of birth asphyxia, hyperbilirubinemia, or maternal infection points to a neonatal cause. A recent history of head trauma or barotrauma (e.g., scuba diving) may suggest the diagnosis of perilymphatic fistula. Fever and otalgia suggest a diagnosis of acute OM. Associated neurologic symptoms such as tinnitus, vertigo, and dizziness suggest inner ear disease or CNS involvement. Headache can be a marker for tumor of the CNS or extension of middle ear infection (Fig. 32.1).

On physical examination, the presence of fever may suggest an infection such as OM or viral labyrinthitis. A detailed otoscopic examination to detect the presence of a middle ear effusion, impacted cerumen, perforated tympanic membrane, foreign body, or other abnormality of the tympanic membrane is a priority. Tympanometry can be used to supplement the physical examination, especially if the otoscopic examination suggests the presence of middle ear disease. Because of the intricate relationship between the cranial nerves, a careful neurologic examination is required. Within the petrous bone, the cochlear nerve is closely related to the seventh cranial nerve and the vestibular branch of the eighth cranial nerve. An ipsilateral facial nerve palsy in a patient with hearing impairment suggests an intracranial process. Vestibular function should be tested looking for the presence of nystagmus at rest as well as with directed gaze. Patients with vestibular dysfunction often fall to one side with Romberg testing or have difficulty with rapid alternating movements or finger-to-nose testing.

Once hearing loss is established, the next step in the emergency department (ED) is to differentiate conductive from sensorineural hearing loss with the use of tuning fork tests (Fig. 32.1). Conductive hearing loss can be confirmed by the Weber test. In the Weber test, a vibrating, 512-Hz tuning fork is placed in the midline of the patient's forehead. The patient will describe the sound as being more prominent on the side of the conductive hearing loss or the side opposite the sensorineural hearing loss. For the Rinne test, the vibrating tuning fork is placed against the mastoid process. When the patient signals that the vibration has ceased, the tuning fork is placed adjacent to that ear to determine whether the patient can hear the sound of the still-vibrating tuning fork. Patients with conductive hearing loss will not be able to hear the tuning fork. This is a negative Rinne test (bone conduction greater than air conduction). Sensorineural hearing loss shows up as a positive Rinne test (air conduction greater than bone conduction). Finally a test by confrontation is done by placing a tuning fork at a point equidistant from both ears. Regardless of the type of hearing loss, the patient will report the sound to be higher on the side with normal hearing.

Laboratory evaluation is seldom necessary in the ED; when needed, it should focus on diagnosis that may be contemplated after obtaining a detailed history and physical examination. Complete blood count (CBC) and peripheral blood smear, renal function tests, serologic tests for syphilis, TORCH titers, and bacteriologic cultures should be performed only if the history and physical suggest an associated diagnosis. Thyroid function tests, lipid profile, and serum calcium levels should be individualized in the context of clinical findings.

In children with the clinical suspicion of intracranial pathology, a radiologic evaluation assists in the diagnosis. Patients with known or suspected congenital malformation of the middle and inner ears should be evaluated with a CT scan because bony detail is essential for diagnosis. Inner ear abnormalities have been demonstrated in 8 to 20% of patients with sensorineural hearing loss. A CT scan also should be performed in patients with suspected fracture of the temporal bone. Magnetic resonance imaging (MRI) is replacing the CT scan for the diagnosis of acoustic neuroma. With the addition of paramagnetic contrast, high-resolution scanning, and thin-section techniques, excellent detail of the internal auditory canal can be achieved.

Most children with decreased hearing in the ED have a conductive hearing loss. If impacted cerumen is seen on examination, it must be removed because it may be the cause of the decreased hearing and it prevents further evaluation. Children whose hearing improves after disimpaction and who have a normal otoscopic examination need no further treatment. Patients without impacted cerumen and those who fail to improve after removal of cerumen may have a foreign body in the ear canal. Only large objects that completely obstruct the external auditory canal should impair hearing; thus, this diagnosis is easily established during otoscopic examination.

The next step in the evaluation is a careful examination of the tympanic membrane, including pneumatic otoscopy. Many patients will show evidence of a middle ear effusion, the most common cause of hearing loss seen in the ED. Rarely, a cholesteatoma may been seen through the translucent tympanic membrane. Sensorineural hearing loss is seen less often in the ED. Most children with sensorineural hearing loss have congenital problems that are diagnosed during the course of routine care, although occasional cases will be brought to the attention of the emergency physician. Among the acquired causes, those that must be diagnosed urgently include CNS tumors, vascular accidents, and perilymphatic fistula. In cases of acquired sensorineural hearing loss, a history of trauma should be sought. Direct blows to the head

may cause temporal bone fractures or a perilymphatic fistula, and sudden changes in pressure may injure the cochlea. If there is no preceding trauma, a careful neurologic examination should be performed, looking for evidence of CNS tumors. A CT scan is indicated if these lesions are suspected. The most common cause of acquired sensorineural hearing loss seen in children in the ED without a history of trauma is viral labyrinthitis. These patients usually have associated tinnitus, vertigo, and vomiting but no focal neurologic abnormalities. Most of the remaining causes of hearing loss are idiopathic. Vascular insufficiency merits consideration in children with sickle cell anemia, diabetes mellitus, and collagen vascular disease. If the cause of the hearing loss remains uncertain, otolaryngologic consult and evaluation should be considered.

Suggested Readings

Brookhouser PE. Sensorineural hearing loss in children. Pediatr Clin North Am 1996;43:1195–1216.

Brookhouser PE, Worthington DW. Middle ear disease in young children with sensorineural hearing loss. Laryngoscope 1993;103:371–378.

Chan KH. Sensorineural hearing loss in children: classification and evaluation. Otolaryngol Clin North Am 1994;27:473–486.

Council on Scientific Affairs. Magnetic resonance imaging of the head and neck region. JAMA 1988;260:3313–3326.

Cox LC. Otoacoustic emissions as a screening tool for sensorineural hearing loss (Editorial). J Pediatr 1997;130:685–686.

Dennis JM, Neely JG. Basic hearing tests. Otolaryngol Clin North Am 1991;24:253–276.

Early Identification of Hearing Impairment in Infants and Young Children. NIH Consensus Statement 1993;11:1–24.

Elwany S, Kamel T. Sensorineural hearing loss in sickle cell crisis. Laryngoscope 1988;98:386–389.

McIntyre PB, Berkey CS, King SM, et al. Dexamethasone as adjunctive therapy in bacterial meningitis: a meta-analysis of randomized clinical trials since 1988. JAMA 1997;278:925–931.

Meyerhoff WL, Cass S, Schwaber MK, et al. Progressive sensorineural hearing loss in children. Otolaryngol Head Neck Surg 1994;110:569–579.

Pappas DG, Simpson LC, Godwin GH. Perilymphatic fistula in children with preexisting sensorineural hearing loss. Laryngoscope 1988;98:507–510.

CHAPTER 33

Patients with Heart Murmurs

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In a “first encounter” examination of a child, the emergency physician will often hear a cardiac murmur—one previously 33 known to the family or one freshly discovered—and then he or she must determine how to fit this finding into the sometimes complicated evaluation of the patient's current illness. Is the murmur an incidental finding of no relevance? Is it suggestive of heart disease? If so, are there cardiac-related symptoms? Is it related to life-threatening illness, not necessarily of cardiac origin?

Certain priorities prevail that should precede the further definition of the murmur per se. It is not important to pinpoint a primary cardiac diagnosis immediately, but it is essential to determine whether the patient is in cardiac decompensation, whether life is at risk, and whether there is incipient need for evaluation by a cardiologist or cardiac surgeon.

A murmur is a noise created by the turbulence of blood flow, under varying pressures, through chambers and vessels and across valves that are of unequal sizes and shapes. The murmur itself is not abnormal. Most murmurs, in fact, are sounds created by normal turbulence and, therefore, are best referred to as “normal” rather than the less appropriate adjectives, “innocent” or “functional.” Some loud murmurs are created by clinically inconsequential defects (e.g., small ventricular septal defects), whereas other, barely discernible, murmurs may be associated with far more serious defects or illnesses, such as acute myocarditis or transposition of the great vessels.

The ultimate goal of the chapter is to provide criteria for determining whether a patient's murmur is irrelevant or associated with life-threatening illness of cardiac or extracardiac origin, and whether the patient needs cardiac consultation now, eventually, or not at all.

DIFFERENTIAL DIAGNOSIS

History

Unless the patient is in extremis, a careful, focused history is always the starting place. For the patient in whom a murmur is heard, in addition to usual history related to the present complaint, relevant questions might include the following:

- Is the murmur known to have been present? Since the neonatal period? If not, since when? Has it been evaluated at any previous examination? If so, what were the findings and conclusion? Has there been cardiac surgery?
- Was there an antecedent illness before discovery of the murmur? Sore throat? Viral infection?
- Are there or have there been associated signs or symptoms? Edema? Cyanosis? Difficult breathing? Hypertension? Chest pain? Joint symptoms? Feeding problems? Weight gain or loss? Congenital defects?
- Does the child tire easily? How far can he or she walk? Does the child squat after walking? Climb stairs? Have cyanotic “spells”?

Examination

The examination should consist of as complete a physical as possible under the prevailing circumstances.

Murmur Characteristics

Murmur characteristics can be defined in print only qualitatively or, at best, in a crude quantitative manner. Most physicians are not put in the position often enough of having to dissect a murmur in intricate detail to achieve the precision of the cardiologist. Therefore, although the characteristics are briefly defined here, the subsequent course of this chapter makes only minimal use of them. More detailed descriptions of murmur characteristics are given in the references at the end of this chapter.

- *Timing and duration:* Systolic (between the first and second heart sounds) or diastolic (between the second and first sounds)? Early, mid, late, or throughout systole (holosystolic)? Beginning in systole and persisting into diastole

(continuous)?

- *Intensity (loudness):* Usually graded from barely discernible (grade I) through accompanied by a palpable thrill (grade IV) to audible without making contact with the chest (grade VI).
- *Shape:* Terms such as *diamond-shaped, plateau, crescendo,* and *decrescendo* are easily understood when learned in conjunction with a phonocardiogram; interpreting them by ear requires much practice and training.
- *Quality: Musical, blowing, rumbling, wood-sawing, harsh, vibratory, twang, soft, rough, grating,* and *click* are among the subjective terms uses. One's own interpretation of “quality” is useful mostly in distinguishing the possibility that there are two separate murmurs.
- *Frequency (pitch):* Described qualitatively as low, medium, or high.
- *Location and transmission:* Location is the point of maximum intensity of the murmur (upper, lower, or mid left or right sternal margin; apex; midclavicular or axillary line; at which rib interspace?). Transmission refers to the areas of maximal spread of the sound (to the back; to the neck).

Precordial Examination

The remainder of the precordial examination should be completed; the physician should inspect for chest bulge, palpate for thrills and clicks and points of maximal impulse, and listen carefully for the heart sounds and adventitial sounds. The physician must interpret the first and second sound for intensity and splitting. The third and fourth sounds, opening snaps, clicks, rubs, and some unusual rhythms may be confused for murmurs at times and should be kept in mind as possible confounding factors.

Associated Signs and Symptoms

Vital Signs

Normal vital signs for age are listed in Appendix D.

- *Heart rate and rhythm:* Check the heart rate and listen and palpate for rhythm disturbances (see Chapter 74 and Chapter 82). The normally rapid rate of the infant makes evaluation of the murmur even more difficult. Palpate the femoral artery pulsations.
- *Blood pressure:* Lower extremity pressures are normally measured 10 to 40 mm Hg higher than upper pressures.
- *Respirations:* Count the rate and observe for retraction.
- *Body temperature:* Infective endocarditis must be thought of in any child with heart disease and fever.

Color

Central cyanosis (see Chapter 16) is diffuse and is best differentiated from peripheral cyanosis by involvement of the tongue. It may be subtle and sometimes difficult for the unpracticed eye to appreciate. If accompanied by clubbing of the distal fingers in the older child, cyanosis is probably persistent and chronic. Severe pallor related to marked anemia may be associated with high-output cardiac failure.

Other Signs Associated with Cardiac Failure (see Chapter 82)

- *Edema:* More likely to be dependent and pitting in cardiac disease. In the preambulant child, dependent edema may be appreciated best along the back, rather than the lower extremities, and also may be prominent in the periorbital area.
- *Neck veins:* Look for distended jugular veins in the neck of the patient when lying or propped at a 45-degree angle.
- *Respiratory effort:* Look for tachypnea, grunting, difficult breathing (particularly subcostal retractions), or a preferred upright position. Listen to the lung fields for crackles and wheeze.
- *Organ enlargement:* Palpate for a soft, engorged liver, mostly involving the left lower lobe. Check for splenomegaly.

Remaining Examination

- *Skin:* Look for petechiae on the surface of the skin, in the conjunctivae, and under the fingernails. Also search for erythema marginatum and subcutaneous nodules. Look for thoracotomy scars.
- *Joints:* Check for tenderness, redness, heat, and swelling (see Chapter 57).
- *Neurologic:* Cranial nerve deficiencies? Paresis? Papilledema?
- *Nutritional evaluation:* Are the child's height and weight within a reasonable percentile compatible with the parents'? Is the weight percentile significantly lesser or greater than that for height?

Ancillary Diagnostic Aids

None of the following need be used routinely, but most should be obtainable and interpretable for the emergency department (ED) setting. They should be ordered selectively if clinical assessment of the child does not allow a satisfactory conclusion regarding the significance of the murmur.

Electrocardiography

A lead II rhythm strip can be used for monitoring the patient in distress. A full 14-lead electrocardiogram (ECG), using age- and size-appropriate electrodes, should be readily obtainable for screening and evaluation purposes. The emergency physician should have a working knowledge of the criteria for determining normality, which may vary with the age group of the child (see Chapter 82 and Appendix D).

Echocardiography

Echocardiography allows definitive diagnosis for many congenital cardiac lesions, determination of the severity of cardiac failure, and differentiation of myocarditis from pericardial effusion. In evaluation of many lesions, echocardiography has replaced cardiac catheterization for preoperative diagnosis. Because it is expensive and the techniques vary significantly, the procedure is best reserved for patients for whom cardiologic consultation is to be obtained. The cardiologist will be in the best position to guide the technician as to optimal methodology.

Procedures such as angiocardiography, electrophysiologic mapping, and cardiac flow determinations, although they may prove essential for eventual diagnosis, require cardiac consultation and should not be part of the ED evaluation.

Chest Radiograph

Films should be taken in both posteroanterior (PA) and lateral views. The physician should look for gross cardiac enlargement in the PA views, which may be determined in older children by a transverse diameter greater than 50% of the width of the thoracic cage. In infants, the diameter normally may be considerably wider than that ratio. Thymic shadows and less than full inspiration may be confounding factors. The lung fields should be evaluated for increased or diminished pulmonary flow.

Pulse Oximetry

Pulse oximetry is used to evaluate the presence and degree of oxygen desaturation.

Blood Studies

Tests that might be of ancillary value under specific circumstances include a complete blood count, sedimentation rate, arterial blood gas measurements, blood culture, antistreptolysin titer, sickle cell screening, and antinuclear antibody.

EVALUATION AND DECISION

Neonates and older infants usually have different clinical presentations than older children. Therefore, in our first encounter evaluation, we divide murmur patients by two age groupings: from birth to 3 years of age (Fig. 33.1 A and Fig. 33.1 B) and over 3 years of age (Fig. 33.2A,Fig. 33.2B and Fig. 33.2C). Table 33.1 lists conditions that may be associated with murmurs.

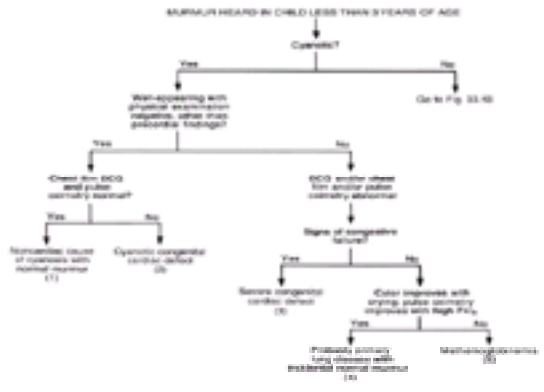

FIGURE 33.1A. Assessment of a noncyanotic child less than 3 years of age in whom a murmur is heard. (Numbers in parentheses refer to specific citations in text.)

FIGURE 33.1B. Assessment of a noncyanotic child less than 3 years of age in whom a murmur is heard. (Numbers in parentheses refer to specific citations in text.) *VH,* ventricular hypertrophy.

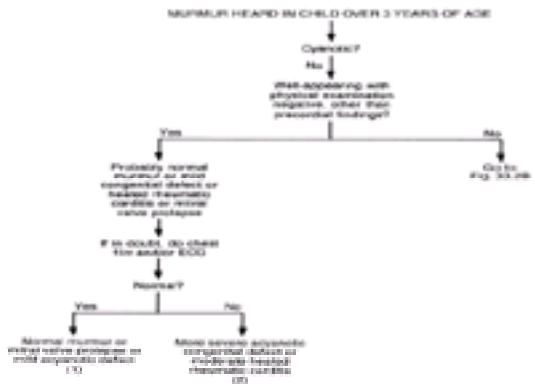

FIGURE 33.2A. Assessment of a noncyanotic, well-appearing child 3 years of age and over in whom a murmur is heard. (Numbers in parentheses refer to specific citations in text.)

FIGURE 33.2B. Assessment of a noncyanotic, sick child 3 years or over in whom a murmur is heard. (Numbers in parentheses refer to specific citations in text.)

FIGURE 33.2C. Assessment of a cyanotic child 3 years of age or over. (Numbers in parentheses refer to specific citations in text.)

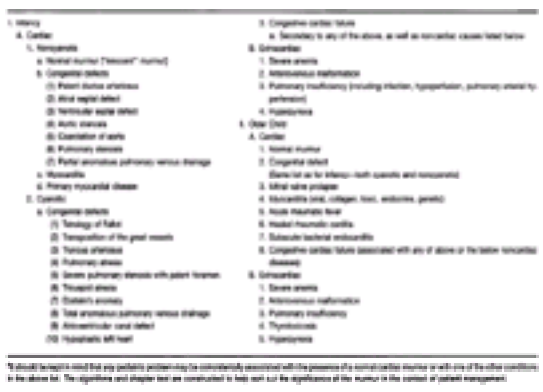

Table 33.1. Conditions That May Be Associated with Presence of a Cardiac Murmur[a]

Infants Less Than 3 Years of Age

Neonates or children less than 3 years of age in whom a murmur is heard require extremely careful assessment but are not necessarily in serious difficulty. The lead point in the evaluation is the presence or absence of cyanosis, preferably confirmed by pulse oximetry.

Infants Less Than 3 Years of Age Who Are Cyanotic (Fig. 33.1A)

Any baby who has a murmur and appears cyanotic should have a thorough physical examination and also should have an ECG, chest film, pulse oximetry, and possibly arterial gases.

If the physical examination is normal, except for the cyanosis and the murmur, and the ECG, film, and pulse oximetry are normal, the infant probably has a normal murmur and a noncardiac cause of only apparent cyanosis (peripheral acrocyanosis, polycythemia) (Fig. 33.1 A [1]).

A cyanotic infant who appears well but has an abnormal ECG and chest film and has diminished arterial saturation by oximetry or blood gas evaluation probably has cyanotic heart disease of a type not likely to get into early trouble (Fig. 33.1A[2]). This could include tetralogy of Fallot, transposition of the great vessels with single ventricle, truncus arteriosus, Ebstein's anomaly, tricuspid atresia, anomalous pulmonary venous drainage, or moderately severe pulmonary stenosis with shunting through an atrial or ventricular septal defect. These babies should be discussed with a cardiologist and referred for evaluation and echocardiogram. Neonates should be admitted for observation in the hospital, but children more than 4 weeks old do not necessarily need to be admitted.

If the cyanotic infant appears acutely ill, the likelihood is that the chest film and/or the ECG and pulse oximetry will be abnormal. If the findings on physical examination suggest congestive heart failure (CHF), the baby probably has severe cyanotic congenital heart disease or an extremely severe acyanotic defect with the cyanosis related to poor perfusion and failure (Fig. 33.1A [3]). Defects in the neonate could include extreme aortic or pulmonary stenosis, pulmonary atresia with intact ventricular septum, and hypoplastic left heart. In the somewhat older infant, considerations include tricuspid atresia, Ebstein's anomaly, large arteriovenous malformation, atrioventricular canal defect, large ventricular septal defect, and total anomalous pulmonary venous drainage. These babies should be admitted to the hospital or transferred immediately to a center for diagnostic cardiac workup and therapy.

If the evaluation of the sick cyanotic baby does not suggest CHF and the saturation improves somewhat with crying and in oxygen, the baby probably has primary lung disease caused by infection, hypoperfusion, or pulmonary arteriolar hypertension (Fig. 33.1A [4]). These babies should be admitted for further evaluation and therapy and the murmur followed closely during therapy, although it is most likely a normal murmur. If the saturation by oximetry of an infant not in failure does not improve with oxygen, the patient may have methemoglobinemia (Fig. 33.1A [5]), either on a congenital basis or secondary to toxin. Co-oximetry studies of blood gases should be done (see Chapter 88).

Infants Less Than 3 Years of Age Who Are Not Cyanotic (Fig. 33.1B)

Infants younger than 3 years old who are not cyanotic should be evaluated carefully, looking for the abnormalities as outlined previously under “Examination.”

If the baby has a negative physical examination and appears well, except for the murmur (Fig. 33.1B [1]), the murmur may represent a congenital cardiac defect that is physiologically insignificant at the time (small patent ductus, atrial or ventricular septal defect, mild aortic or pulmonary stenosis, partial anomalous pulmonary venous drainage) or a normal murmur. These children can be followed by the primary care physician. If doubt exists about the infant's status because of the intensity or transmission of the murmur, an ECG and chest film should be ordered; if normal, they confirm the above impression. Peripheral pulmonary stenosis, related to angulation of the distal pulmonary arteries, is a common cause of normal murmurs in neonates; this murmur transmits well to the back, may be quite loud, and in time, will not be discernible.

If the ECG shows abnormal atrial or ventricular hypertrophy and the chest film shows cardiac enlargement or abnormal pulmonary vasculature, a more serious degree of the same acyanotic defects, or possibly an acyanotic Tetralogy of Fallot, is likely, and nonemergent referral to a cardiologist is warranted (Fig. 33.1B [2]).

If the acyanotic baby with a murmur appears ill, an ECG and chest film are obtained. If these are normal, the murmur is most likely inconsequential, and the baby should be evaluated for an underlying medical or surgical illness related to the presenting complaints at this visit (Fig. 33.1B [3]). It is important to think of noncardiac conditions, such as severe anemia or hyperpyrexia, as causes of the murmur.

If the infant has signs suggesting cardiac failure, ECG findings of marked ventricular hypertrophy or tall, pointed P waves might be indicative of a severe acyanotic congenital cardiac defect (large ventricular septal defect, large patent ductus, severe aortic or pulmonary stenosis) (Fig. 33.1B [4]).

A baby in failure with only T-wave or ST-segment changes on ECG may have viral myocarditis, primary myocardial disease, or an extracardiac problem that causes high cardiac output (severe anemia, large arteriovenous malformation). Some of these babies, although not cyanotic on the basis of their underlying lesion, may show a degree of cyanosis with diminished saturation on pulse oximetry because of the hypoperfusion related to cardiac failure. All such babies need admission or transfer to a tertiary pediatric center for emergency evaluation and treatment (Fig. 33.1B[5]).

If the chest film or ECG is not normal and the baby appears ill but does not have signs that suggest CHF, a primary pulmonary disease should be considered, with the murmur being either a normal one or representing a milder acyanotic defect (Fig.33.1B[6]). Admission for therapy and further evaluation is appropriate.

Children 3 Years and Older

The assessment and disposition of an older child in whom a murmur is discovered is somewhat less of a challenge to the emergency physician. The child is more cooperative and generally more interactive than the infant, and as a result, the sometimes subtle evaluation of the child's state of well-being is less uncertain. The heart rate normally is slower, allowing

easier dissection of the murmur.

By the time children who live in geographic areas in which modern medical care is available reach 3 years of age, most congenital lesions have been discovered and many have been surgically repaired. Acquired cardiac and noncardiac illnesses, therefore, play a more prominent role in assessment. Still, some congenital lesions go unrecognized, some do not require surgery, and others that require surgery have not had access because of economic or social reasons.

Normal murmurs are by far the most common ones discovered at a first encounter with an older child. Table 33.2 describes the characteristics generally associated with a normal murmur. If the examining physician is satisfied that the murmur is a normal one and the child has no other symptoms referable to the cardiovascular system, cardiac consultation is neither necessary nor advisable. Still, it often is difficult to distinguish a normal murmur from the murmur of such intracardiac lesions as small atrial or ventricular defects or mild aortic or pulmonic stenosis; the management guidelines outlined in this chapter obviate the need for the emergency physician to be concerned about such a differential. For the purposes of the ED evaluation, it is important to determine only whether the patient is in difficulty or needs further evaluation. Again, the presence or absence of cyanosis is the lead finding.

Timing: midsystole
Intensity: grades I through IV
Location of maximal intensity: midsternal border
Radiation: possibly to the precordium and neck, but not the back
Quality: "twangy" or "vibratory"
Heart Sounds: readily definable, including splitting of S_2

Table 33.2. Characteristics Usually Associated with a “Normal” Murmur

Children 3 Years of Age and Over Who Are Not Cyanotic (Fig. 33.2A and Fig. 33.2B)

The acyanotic child with a murmur who seems essentially well (Fig. 33.2A [1]) and has a negative physical examination other than the precordial finding most likely has a normal murmur but may have a mild acyanotic congenital defect, a healed rheumatic carditis, or a mitral valve prolapse. The femoral artery pulsations should always be palpated for the possibility of coarctation of the distal aorta. In mitral valve prolapse, a midsystolic “click” is a more constant finding than the murmur, which follows the click and is quite variable. These children should be followed by the primary care physician. If in doubt because of the intensity or transmission of the murmur, an ECG and chest film should be ordered. If these are normal, one has further assurance that primary care follow-up is all that is necessary. If these are abnormal, there is concern about the possibility of a more severe acyanotic defect or moderately severe healed rheumatic carditis (Fig. 33.2A [2]). Referral should be made to a cardiologist on a nonemergency basis for further differentiation.

In evaluating the acyanotic child who appears acutely ill and has a murmur (Fig. 33.2B [1]), a careful history should be taken regarding prior or recent antecedent illness, including a significant sore throat or “viral” illness. If there is such a history, the possibility of swollen, red, and tender joints should be sought. If these are present, the child is febrile, and an ECG is abnormal, the child should be hospitalized to be evaluated for the possibility of acute rheumatic fever (see Chapter 82). On the other hand, the acyanotic ill child with objective joint findings but a normal ECG is more likely to have a normal murmur with a concurrent arthriticlike illness, such as Henoch-Schönlein purpura (HSP), juvenile rheumatoid arthritis, or septic arthritis (Fig. 33.2B [2]). These children need diagnostic evaluation of their acute illness and follow-up of the murmur.

The ill-appearing acyanotic child who has a murmur and a history of known chronic or recent antecedent illness but has no objective joint findings should have an ECG and a chest film taken. If the ECG shows ST-segment and T-wave abnormality and if the heart is enlarged, the patient probably has myocarditis, with the murmur resulting from turbulence created by ventricular dilation and atrioventricular valve insufficiency (Fig.33.2B [3]). The cause could be viral, collagen vascular, toxic, or endocrine (see Chapter 82, Chapter 97 and Chapter 101). These children must be admitted for evaluation and therapy.

The ill-appearing acyanotic child who has a murmur but no chronic or recent antecedent illness and who shows signs of CHF may have severe acyanotic heart disease, myocarditis, or high-output failure secondary to severe anemia, large arteriovenous malformation, or thyrotoxicosis (Fig. 33.2B [4]).

Whether in congestive failure or not, the ill-appearing child with a murmur and fever should be examined carefully for splenomegaly and petechiae on the skin surface, on the conjunctivae, and under the nailbeds. If these are found, although the murmur could represent the entire list of cardiac diseases, the important immediate concern is that of infectious endocarditis (Fig. 33.2B [5]). The child should be admitted for evaluation, cardiac consultation, and echocardiography (see Chapter 82).

If the patient with a murmur and petechiae is not showing signs of failure, the murmur may be normal or represent a mild acyanotic defect. In addition to infectious endocarditis, consideration must be given to other conditions manifested with petechiae, such as meningococcemia, idiopathic thrombocytopenic purpura (ITP), HSP, and rickettsial infection (Fig. 33.2B [6]). Blood cultures should be drawn, appropriate emergency treatment initiated, and the child admitted for further

evaluation and treatment.

If the acyanotic ill-appearing child with a murmur is not in failure, has no splenomegaly or petechiae, and had a normal ECG, the murmur is most likely normal or associated with the high cardiac output of hyperpyrexia or anemia (Fig. 33.2B [7]). These children should be evaluated for their underlying condition.

Children 3 Years and Older Who Are Cyanotic (Fig. 33.2C)

As with infants, cyanotic older children with murmurs should have an ECG, chest film, pulse oximetry, and possibly, arterial gases after a careful history and complete physical examination. If a patient is tested with these and the results are normal, except for the cyanosis and murmur, the child probably has a noncardiac cause of the cyanosis (polycythemia) (Fig. 33.2C [1]). The murmur may be normal or associated with a coincidental acyanotic congenital defect. The primary condition should be investigated.

The cyanotic child who is not acutely ill but has an abnormal ECG, chest film, and pulse oximetry has cyanotic heart disease that possibly could be improved surgically (Fig.33.2C [2]). The child should be referred to a cardiologist on an nonemergent basis for further evaluation, including echocardiography.

If the cyanotic child appears acutely ill and has signs of CHF, severe cardiac disease is present (Fig. 33.2C [3]). The causes could include a decompensating congenital cyanotic cardiac defect, in which case the cyanosis would be intense (Table 33.1), or failure secondary to acquired disease, in which the cyanosis is related to hypoperfusion and usually less intense. These children need to be admitted for therapy and evaluation.

Whether or not there are signs of congestive failure in the cyanotic child with a murmur, if there is fever, splenomegaly, and/or petechiae on the skin, on the conjunctivae, or under the nailbeds, blood cultures should be drawn for the likelihood of infective endocarditis (Fig. 33.2C [4]). If the child is not in failure and petechiae are found, infective endocarditis is still a possibility but other noncardiac causes of petechial presentations must be considered (meningococcemia, Valsalva maneuvers, ITP, HSP). Blood cultures should be drawn and the child admitted.

A careful neurologic examination should be part of the evaluation of every ill child with cyanotic heart disease. If findings are abnormal, consideration has to be given to the complications of hypoxemic “spells,” cerebrovascular accident, or if febrile, brain abscess (Fig. 33.2C [5]).

If the ill cyanotic child with a murmur and abnormal chest film shows significant improvement of oxygen saturation with supplemental oxygen, the child most likely has primary pulmonary disease. The ECG abnormality, if there is one, would most likely consist of tall, pointed P waves and evidence of right ventricular hypertrophy (Fig. 33.2C [6]). These children need admission for evaluation and therapy.

In the cyanotic child with a murmur who has a normal ECG and chest film, abnormal pulse oximetry, but normal arterial PO_2, the possibility must be considered of acute toxin-induced methemoglobinemia, with the murmur being normal or representing a mild acyanotic defect (Fig. 33.2C [1,7]).

SUMMARY

This chapter has provided clinical guidelines for the initial assessment and disposition of infants and children in whom a murmur is discovered during the first visit. Although diagnoses have been listed and suggested for most of the legs of the decision paths, it has also been shown that definitive diagnosis of the underlying lesion is not the primary aim of ED evaluation; careful assessment of the patient and safe disposition are required at first encounter. Particularly in older children, many readers probably can do considerably more dissection of the characteristics of the murmur and heart sounds and can arrive at more specific cardiac diagnoses than have been used in the evaluation approach described in this chapter. This process can be augmented, if desired, by supervised practice, by careful use of the references, and by judicious cardiac consultation.

Suggested Readings

Athreya B, Silverman BK. Pediatric Physical Diagnosis. East Norwalk, CT: Appleton-Century-Crofts, 1985.

Bisno AL, Gerber MA, Gwaltney JM, et al. Diagnosis and management of group A streptococcal pharyngitis: a practical guideline. Clin Infect Dis 1997;25:574–583.

Danford DA, Nasir A, Gumbiner C, et al. Cost assessment of the evaluation of heart murmurs in children. Pediatrics 1993;91:365–370.

Fuerst RS. Use of pulse oximetry. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:823–828.

Hoffmann JIE. Cardiology. In: Rudolph AM, Hoffmann JIE, Rudolph CD, eds. Pediatrics. 20th ed. Stamford, CT: Appleton & Lange, 1996:1409–1428.

Mangione S, Nieman LZ. Cardiac auscultatory skills of internal medicine and family practice trainees. A comparison of diagnostic proficiency. JAMA 1997;278:717–722.

Newburger JW, Rosenthal A, Williams RG, et al. Non-invasive tests in the initial evaluation of heart murmurs in children. N Engl J Med 1983;308:61–64.

Park MK, Guntheroth WG. How to Read Pediatric ECGs. 3rd ed. St Louis: Mosby-Year Book, 1992.

Rice MJ, McDonald RW, Reller MD, Sahn DJ. Pediatric echocardiography: current role and a review of technical advances. J Pediatr

1996;128:1–14.

Wadsworth MR, Silverman BK. The electrocardiogram in infants and children. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:759–769.

CHAPTER 34

Hematuria

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Hematuria, the presence of red blood cells (RBCs) in the urine, is a common presenting complaint in the emergency department (ED). Disease processes manifested by gross hematuria or other symptoms such as acute onset of edema, headache, and hypertension are the context in which hematuria requires urgent/emergent evaluation in the ED. Microscopic hematuria may be accompanied by other signs and symptoms or may be completely asymptomatic; it can usually be evaluated in the outpatient setting. Red or brown urine does not always indicate hematuria. Several foods, substances, and drugs may color the urine; therefore, it is important to document the presence of blood in the urine. Reagent strips (based on the peroxidase reaction with hemoglobin) can be used as the initial screening test for hematuria. Heme-positive reagent strips must be confirmed by microscopic examination for the presence of RBCs because both hemoglobinuria and myoglobinuria can cause a positive reaction in the absence of RBCs. The presence of 5 to 10 or more RBCs per high-power field (RBC/HPF) is abnormal and warrants further workup. The evaluation of a child with hematuria must take into consideration the clinical presentation, patient and family histories, physical examination, and complete urinalysis so that a logical, orderly, and cost-effective approach can be undertaken.

PATHOPHYSIOLOGY

Red cells can be added to the urine at any point along the urinary tract—from the glomerulus, through the tubule, or through the collecting system, the ureter, the bladder, or the urethra. The pathophysiology of hematuria can be explained by categorizing it as either glomerular or nonglomerular. Immune-mediated inflammatory damage to the glomerular filtration surface, as seen in postinfectious nephritis, causes disruption of the glomerular basement membrane with subsequent leakage of red cells and protein. Glomerular bleeding that results in gross hematuria may be brown, smoky, or cola- or tea-colored as a result of the acidic urine changing the hemoglobin to hematin. Red cells may become enmeshed in the protein matrix to form RBC casts, a sensitive indicator of glomerular hematuria. The renal papillae are sites of nonglomerular bleeding that are susceptible to microthrombi and anoxia in patients with sickle cell disease or trait. Inflammation of the tubules and interstitium caused by antibiotics can result in hematuria, proteinuria, and eosinophiluria. Nonsteroidal agents can produce hematuria from both tubulointerstitial nephritis and inhibition of prostaglandin synthesis. Grossly bloody urine that is bright red or pink with or without clots is more likely to be originating from the lower urinary tract, usually the bladder or urethra. Hematuria from trauma to the kidney or bladder is caused by contusions, hematomas, or lacerations anywhere along the tract. Increased vascularity from infection or chemical irritation can lead to leakage of RBCs into the urine. Exercise-related hematuria results from ischemic injury as well as direct trauma. Benign familial hematuria, a principal cause of asymptomatic hematuria, is caused by leakage of RBCs through a thin glomerular basement membrane and rarely comes to the attention of the emergency physician except as an incidental finding.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of hematuria is vast and can be categorized based on whether the site of bleeding is within the urinary system (glomerular, extraglomerular) or secondary to a systemic process (Table 34.1). The most common causes of hematuria (Table 34.2) are urinary tract infection (either cystitis or pyelonephritis), acute poststreptococcal glomerulonephritis, and trauma, the latter two also being the most common of the potentially life-threatening causes. Other potentially serious causes of hematuria (Table 34.3) include hematologic disorders, renal stones with obstruction, tumors, and hemolytic uremic syndrome (HUS). Other glomerular causes of hematuria that are primary renal diseases include nonstreptococcal postinfectious glomerulonephritides, membranous glomerulonephritis, immunoglobulin A (IgA) nephropathy, and Alport's syndrome (hereditary nephritis). Hematuria as a manifestation of a systemic condition is most commonly seen in children with vasculitides such as Henoch-Schöonlein purpura, systemic lupus erythematosus (SLE), and polyarteritis nodosa.

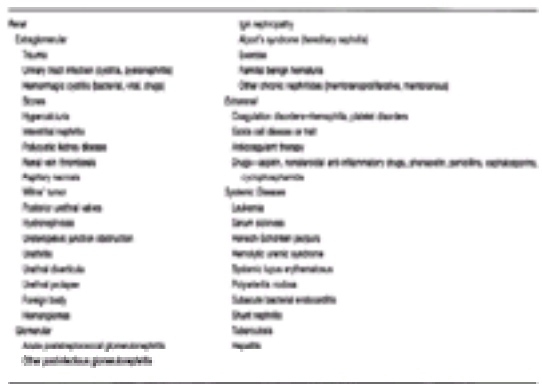

Table 34.1. Principal Causes of Hematuria in Children

Urinary tract infection—cystitis, pyelonephritis	Interstitial nephritis
Trauma (kidney, bladder, urethra)	Benign hematuria
Acute poststreptococcal glomerulonephritis	Urethritis
Sickle cell disease or trait	

Table 34.2. Common Causes of Hematuria

Trauma (kidney, bladder)	Tumor
Acute glomerulonephritis	Hematologic disorders
Hemolytic uremic syndrome	Toxins
Renal stones with obstruction	

Table 34.3. Life-Threatening Causes of Hematuria

Extraglomerular causes of hematuria include congenital anomalies such as diverticula of the urethra and bladder; hemangiomas in the bladder; cysts of the kidneys, as in polycystic or multicystic kidney; and obstruction of the ureteropelvic junction. In addition to congenital anomalies, renal vein thrombosis secondary to a coagulation disorder or to placement of an umbilical catheter is a cause of hematuria in the neonate. Wilms' tumor is a common childhood solid tumor associated with hematuria in 12 to 25% of the cases. Nephrolithiasis should be considered if there is a family history or a predisposing condition such as recurrent infection, bladder dysfunction (seen in myelomeningocele), or chronic diuretic therapy (as seen in infants with bronchopulmonary dysplasia). Hypercalciuria and cystinuria are metabolic diseases that also predispose patients to renal stones and hematuria. Finally, urethral prolapse, seen most commonly in girls 2 to 4 years of age, may present with vaginal bleeding that can contaminate a collected urine specimen and be misinterpreted as hematuria.

EVALUATION AND DECISION

The initial evaluation of hematuria must begin with confirmation of blood in the urine. Further investigation of the cause and treatment includes detailed patient and family histories, careful physical examination, and microscopic urinalysis (which lends information to probable causes and helps determine the site of bleeding within the urinary tract system). A specific diagnosis may or may not be made in the ED, and the patient may require further diagnostic testing. The most important role for the emergency physician in evaluating a child with hematuria is to identify serious, treatable, and progressive conditions such as trauma, nephritis associated with hypertension, bleeding disorders, and infection.

Blood in the urine may come from sources outside the urinary tract. Vaginal hemorrhage in the female secondary to infection, foreign body, or trauma (sometimes secondary to abuse) may contaminate the urine. In addition, parents may report finding blood in the urine when, in fact, a rectal fissure has caused a small hemorrhage, producing a mixture of blood and urine in the diaper or underwear. In prepubertal girls, urethral prolapse may present with vaginal bleeding, which may be confused with hematuria.

Urine dipsticks positive for blood require microscopic examination of the urine. Hemoglobinuria from hemolysis and

myoglobinuria from rhabdomyolysis will cause a positive dipstick reaction for blood and an absence of RBCs on urine microscopy. Many dyes, drugs, and pigments will change the urine color to pink, red, brown, or black but will not result in a positive dipstick test for blood. A partial list includes beets, blackberries, urates, aniline dyes, bile pigments, porphyrin, diphenylhydantoin, phenazopyridine (Pyridium), rifampin, deferoxamine, phenolphthalein, ibuprofen, methyldopa, chloroquine, homogentisic acid, and *Serratia marcescens* infection.

The history taking for infants and neonates with hematuria should include questions about umbilical vessel catheters (renal venous or arterial thrombosis), passage of clots on voiding (hemorrhagic disorders), abdominal swelling or palpable mass (tumor, polycystic disease, ureteropelvic junction obstruction, posterior urethral valves), and significant birth asphyxia (corticomedullary necrosis). Dysuria or frequency in children and adolescents suggests cystitis, whereas flank, abdominal, or back pain suggests trauma, genitourinary infection, or stones as the cause. Sore throat, upper respiratory infection, or pyoderma (preceding or appearing concurrently with the onset of hematuria) points to acute postinfectious glomerulonephritis, streptococcus being the most common bacterial cause. A history of gross hematuria with a concomitant viral upper respiratory or gastrointestinal infection may also suggest IgA nephropathy. Hematuria associated with systemic disorders may be uncovered by eliciting a history of skin rashes and arthralgia or arthritis as seen in Henoch-Schönlein purpura and SLE. Both sickle cell anemia and sickle cell trait are associated with chronic, asymptomatic gross hematuria. Finally, a history of drug use, especially use of the penicillins and cephalosporins, may point to interstitial nephritis as the cause. Antibiotic-associated tubulointerstitial nephritis is associated with high-dose, long-term antibiotic therapy and is characterized clinically by fever, rash, eosinophilia with pyuria, eosinophiluria, hematuria, proteinuria, and nonoliguric renal failure. Family history of renal stones, deafness, nephritis, renal anomalies, or hematologic disease may suggest a diagnosis in the child such as Alport's syndrome (hereditary nephritis), sickle cell anemia, or hemophilia.

Physical examination of a child with hematuria should always include a blood pressure measurement. Hypertension may accompany glomerulonephritis, obstructive uropathy, Wilms' tumor, polycystic kidney, or vascular disease. Periorbital edema and facial swelling may be the first physical sign of nephritis. Urethral prolapse presents as a doughnut-shaped mass at the site of the urethral meatus, which is usually hyperemic and friable with scant bloody drainage.

Bruising of the abdomen, flank, or back should raise suspicion of trauma, including child abuse, as a cause of hematuria. Tenderness of the flank or lower abdomen may signal pyelonephritis, obstructed kidney, or lower urinary tract infection. Flank or abdominal masses suggest Wilms' tumor or hydronephrosis, hydroureter, or polycystic kidney. Petechial or purpuric lesions on the skin and arthritis may accompany hematuria seen in vasculitic syndromes such as Henoch-Schönlein purpura, HUS, and SLE. Pallor may be a sign of anemia from chronic renal insufficiency, HUS, hemoglobinopathy, leukemia, or tumors.

A careful, detailed urinalysis plays an essential role in the evaluation of the child with hematuria. Several clues in the urinalysis can help localize the site of hematuria. RBC casts, cellular casts, tubular cells, tea-colored or smoky-brown urine, and proteinuria 2+ or greater by dipstick all point to glomerular bleeding. In addition, the presence of dysmorphic RBCs and/or acanthocytes (ring-formed RBCs with one or more protrusions of different shapes and sizes) as well as measurement of the mean volume of urine erythrocytes using a Coulter counter have been used as markers of glomerular bleeding (erythrocyte volume less than 50 μm^3). In contrast, nonglomerular bleeding is suggested by red or pink urine, blood clots, no proteinuria (or less than 2+ in the absence of gross hematuria), and normal morphology of erythrocytes. Calcium oxalate crystals may be seen in the urine of patients with renal stones.

Other blood studies may be useful in selected cases and include a complete blood count (CBC), prothrombin time (PT), partial thromboplastin time (PTT), erythrocyte sedimentation rate (ESR), blood urea nitrogen (BUN), serum creatinine, complement levels (C3 and C4), and streptococcal serologies (antistreptolysin O, anti-DNase-B, and antihyaluronidase titers). The history and physical examination should direct the emergency physician to those additional tests that are needed, if any. Most patients, other than those with isolated microscopic hematuria or lower urinary tract infection, will require at least a CBC as part of their evaluation.

A clinical algorithm for evaluating hematuria in the ED is shown in Figure 34.1. The first step is to confirm the presence of true hematuria. If a traumatic cause for the hematuria is suspected based on history or physical findings, emergent evaluation for serious anatomic lesions must be initiated. Parenchymal contusions, lacerations, renal transections, and pedicle disruptions are possible injuries. Hematuria is the cardinal marker of renal injury, with the severity paralleling the magnitude of the injury (except for renal pedicle injuries, which may have no associated hematuria). Microscopic hematuria greater than 20 to 50 RBC/HPF, gross hematuria, or history of significant mechanism of injury to the flank or abdomen necessitates emergent imaging. Hematuria disproportionate to the injury may indicate a congenital renal anomaly or tumor.

FIGURE 34.1. Approach to hematuria in the emergency department.

If there is no history of trauma, coagulopathies should be considered as a cause. However, the medical history alone usually will point to this cause because the sudden occurrence of isolated hematuria in a previously healthy child is unlikely with either a congenital or acquired bleeding disorder. Hematuria in a child known to have hemophilia or a related disorder often requires minimal investigation and is managed in accordance with standard protocols. If an acquired coagulopathy is suspected, a CBC with platelet count, PT, and PTT is warranted.

If trauma and coagulopathies are considered unlikely, identifying the site of bleeding as either glomerular or nonglomerular (based on urinalysis and other signs or symptoms) can direct further evaluation and diagnosis. Acute glomerulonephritis characterized by hypertension, edema, RBC casts, proteinuria, and tea-colored urine most often follows a streptococcal infection and merits serious consideration in the ED because it may cause significant hypertension and pulmonary edema requiring immediate intervention. HUS is a serious disorder that may present with glomerular-induced hematuria and proteinuria as well as a characteristic microangiopathic hemolytic anemia, thrombocytopenia, and renal failure. Laboratory studies useful in children suspected of having nephritis include a CBC, ESR, BUN, serum creatinine, complement levels, and antistreptococcal antibodies. Other nephritides may be associated with vasculitis (Henoch-Schönlein purpura, SLE, periarteritis nodosa, Wegener's granulomatosis) and may require further diagnostic evaluation before a specific diagnosis is made.

Most children without a history of trauma who are evaluated for gross and/or microscopic hematuria in the ED have a urinary tract infection (UTI). The infection may either be in the upper tract (e.g., pyelonephritis, characterized by fever, chills, flank pain, vomiting, and dysuria) or lower tract (e.g., cystitis, characterized by dysuria, frequency, and occasionally, abdominal pain and fever). The cause of UTI is either bacterial or viral. Acute hemorrhagic cystitis is often associated with adenovirus. The findings of pyuria and bacteriuria on urinalysis suggest an infectious cause, although their absence does not exclude either pyelonephritis or cystitis; thus, a urine culture is essential if no other cause has been uncovered. If the clinical suspicion is high for a bacterial UTI, presumptive antimicrobial treatment should be initiated.

Severe flank pain radiating to the groin is characteristic of renal colic from calculi, which may present with either gross or microscopic hematuria. Stones may occur in children with metabolic abnormalities or stasis secondary to obstruction and in premature infants taking furosemide, especially those with bronchopulmonary dysplasia. Crystals may be seen on urinalysis; further investigation with intravenous pyelography, renal ultrasound, or spiral computed tomography (CT) usually confirms stones if a plain abdominal radiograph does not reveal the presence of radiopaque material. Hypercalciuria is an important cause of hematuria in children and may be idiopathic or secondary to another disease and can lead to nephrocalcinosis.

Hematuria that persists after the previously mentioned causes have been ruled out or deemed unlikely based on history and physical examination usually does not require further evaluation in the ED and should be pursued by the primary health care provider, possibly in collaboration with a pediatric nephrologist. These additional causes are listed in Figure 34.1 and Table 34.1 and may require more extensive imaging, intervention such as renal biopsy, metabolic studies, or serial urinalyses (benign hematuria, exercise-induced hematuria).

Suggested Readings

Baker MD, Baldassano RN. Povidone -iodine as a cause of factitious hematuria and abnormal urine coloration in the pediatric emergency department. Pediatr Emerg Care 1989;5:240.

Fogazzi GB, Ponticelli C. Microscopic hematuria diagnosis and management. Nephron 1996;72:125.

Hockberger RS, Schwartz B, Connor J. Hematuria induced by urethral catheterization. Ann Emerg Med 1987;16:550.

Inglefinger JR, Davis AE, Grupe WE. Frequency and etiology of gross hematuria in a general pediatric setting. Pediatrics 1977;59:557.

Jones CL, Eddy AA. Tubulointerstitial nephritis. Pediatr Nephrol 1992;6:572.

Lettgen B, Wohlmuth A. Validity of G1-cells in the differentiation between glomerular and non-glomerular haematuria in children. Pediatr Nephrol 1995;9:435.

Lieu TA, Grasmeder HM, Kaplan BS. An approach to the evaluation and treatment of microscopic hematuria. Pediatr Clin North Am 1991;38:579.

Roy S. Hematuria. Pediatr Ann 1996;25:284.

Stapleton FB, Roy S, Noe HN, Jerkins G. Hypercalciuria in children with hematuria. N Engl J Med 1984;310:1345.

Trachtman H, Weiss RA, Bennett B, et al. Isolated hematuria in children: indications for renal biopsy. Kidney Int 1984;25:94.

White RH. The investigation of hematuria. Arch Dis Child 1989;64:159.

CHAPTER 35

Hypertension

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Until recently, hypertension was considered predominantly a disorder of adulthood. Although there are currently no universally accepted standards for determining whether a child should be classified as hypertensive, the most commonly used definition of hypertension is a systolic, diastolic, or mean arterial pressure that is above the 95th percentile for age. Children with excessive height or lean body mass for age may not be included in this definition. Beyond this specific definition of hypertension, however, the nomenclature is often confusing because several other terms have been used with varying meanings, including *hypertensive crisis, malignant hypertension,* and *accelerated hypertension.* The Second Task Force on Blood Pressure Control in Children divided hypertension into two distinct classes: *significant (moderate) hypertension* was defined as blood pressure measurements persistently between the 95th and 99th percentiles for age and sex, and *severe hypertension* was defined as blood pressure measurements persistently at or above the 99th percentile for age and sex (Table 35.1). For the sake of the present discussion, the classification system of The Second Task Force can be expanded to include two additional terms. *Hypertensive urgency* is a severely elevated blood pressure that may be potentially harmful but is without evidence of end-organ damage or dysfunction. For the purposes of evaluation and treatment, blood pressure measurements at or above “severe” levels should be considered a hypertensive urgency even in the absence of symptoms. *Hypertensive emergency* describes a situation in which elevated blood pressure is associated with evidence of secondary organ damage such as hypertensive encephalopathy or acute left ventricular failure. It is important to note that these definitions do not rely solely on a particular level of blood pressure, but they also rely on the effect of an elevated blood pressure (usually markedly elevated) on end organs. Hypertensive urgencies ordinarily develop over days to weeks, whereas hypertensive emergencies generally develop within hours. Hypertensive emergencies are more commonly seen in individuals with poorly controlled chronic hypertension. In previously normotensive individuals, however, a hypertensive emergency may be the result of such factors as head trauma, drug ingestion, acute glomerulonephritis, toxemia, or pheochromocytoma.

Age Group	Significant Hypertension (mm Hg)*	Severe Hypertension (mm Hg)*
Newborn–7 days	Systolic BP ≥96	Systolic BP ≥106
8–30 days	Systolic BP ≥104	Systolic BP ≥110
Infant (<2 yr)	Systolic BP ≥110 Diastolic BP ≥63	Systolic BP ≥118 Diastolic BP ≥82
Children (2–5 yr)	Systolic BP ≥116 Diastolic BP ≥74	Systolic BP ≥124 Diastolic BP ≥84
Children (6–9 yr)	Systolic BP ≥121 Diastolic BP ≥81	Systolic BP ≥130 Diastolic BP ≥86
Children (10–12 yr)	Systolic BP ≥127 Diastolic BP ≥83	Systolic BP ≥134 Diastolic BP ≥90
Adolescents (13–15 yr)	Systolic BP ≥135 Diastolic BP ≥86	Systolic BP ≥144 Diastolic BP ≥92
Adolescents (16–18 yr)	Systolic BP ≥142 Diastolic BP ≥92	Systolic BP ≥150 Diastolic BP ≥98

Adapted with permission from the Task Force on Blood Pressure Control in Children. Report of the second task force on blood pressure control in children—1987. Pediatrics 1987;79:1–25.
BP, blood pressure.

Table 35.1. Classification of Hypertension by Age Group

Appropriate blood pressure cuff size is essential for accurate measurement of blood pressure. The inflatable rubber bladder should be long enough to completely encircle the circumference of the arm (overlap is acceptable). Bladder width should be approximately 40% of arm circumference at a point halfway between the acromion and the olecranon. A narrow cuff can produce falsely elevated readings. Sphygmomanometer measurements are difficult to perform in neonates, and blood pressure determinations are more reliably performed with oscillometric or Doppler devices. In older children, however, abnormal oscillometric readings should be verified manually with a sphygmomanometer. Current guidelines recommend that the fifth Korotkoff sound be used to define diastolic blood pressure in children, adolescents, and adults.

Ideally, repeated measures should be recorded over a series of visits before a diagnosis of hypertension is made. However, in the emergency department (ED) setting, the child with an initial moderate or severe hypertensive reading should have blood pressure measurements redone after a brief period of quiet rest. If the second reading remains mildly or moderately elevated and the child is asymptomatic with no evidence of end-organ compromise, outpatient follow-up is required. The child with evidence of hypertensive urgency or emergency, on the other hand, demands immediate attention.

PATHOPHYSIOLOGY/DIFFERENTIAL DIAGNOSIS

Hypertension is not a disease itself but rather the result of one or more pathologic processes. In many instances, the nature of the pathologic processes can be determined. Often, however, a specific cause for childhood hypertension cannot be identified, even after a comprehensive workup. In that case, the elevated blood pressure is called essential or primary hypertension. In fact, it has been suggested that for the general pediatric population, the incidence of essential hypertension is similar to that of the adult population. Although most cases of hypertension are considered essential, a number of other causes largely depend on age (Table 35.2). In newborn infants, for example, the most common causes of hypertension are renal artery thrombosis or stenosis, coarctation of the aorta, congenital renal malformations, and bronchopulmonary dysplasia. In children under the age of 6 years, coarctation of the aorta, renal parenchymal diseases, and renal artery stenosis remain the most common causes. Children between 6 and 10 years of age may have renal artery stenosis and renal parenchymal disease as the source of severe hypertension, although essential hypertension is the leading cause of moderate hypertension in this age group. In older children, although essential hypertension is the more common diagnosis, hypertension may be the result of renal parenchymal disease (Table 35.3). In adolescent girls, oral contraceptive pills may be a cause of hypertension, and in all age groups, drug- or toxin-induced hypertension should be considered. Life-threatening causes of hypertension (Table 35.4) are relatively uncommon in the pediatric age group and most often represent a severe presentation of the common causes.

Table 35.2. Complete Differential Diagnosis of Hypertension

Age Group	Cause
Newborn infants	Renal artery thrombosis, renal artery stenosis, congenital renal malformations, coarctation of the aorta, bronchopulmonary dysplasia
Infancy–6 yr	Renal parenchymal diseases,* coarctation of the aorta, renal artery stenosis
6–10 yr	Essential hypertension (including obesity), renal artery stenosis, renal parenchymal diseases
Adolescence	Essential hypertension (including obesity), renal parenchymal diseases

Adapted with permission from the Task Force on Blood Pressure Control in Children. Report of the second task force on blood pressure control in children—1987. Pediatrics 1987;79:1–25.
*Includes renal structural and inflammatory lesions, as well as tumors.

Table 35.3. Common Causes of Hypertension

Age Group	Cause
Infancy	Coarctation of the aorta, valvular insufficiency, congenital adrenal hyperplasia, renal vascular disease, renal parenchymal disease
Childhood	Renal parenchymal disease, renal vascular disease, coarctation of the aorta, pheochromocytoma, increased intracranial pressure, bacterial endocarditis, drug-induced/toxicologic
Adolescence	Renal parenchymal disease, pheochromocytoma, toxemia of pregnancy, drug-induced/toxicologic

*Any increase in blood pressure that is sufficiently acute or elevated to cause systemic symptoms should be considered life-threatening.

Table 35.4. Selected Life-Threatening Causes of Hypertension [a]

EVALUATION AND DECISION

The extent of ED assessment of the child with hypertension depends on the level of blood pressure and the clinical signs and symptoms of the child at presentation. The asymptomatic child with incidental, newly diagnosed mild or moderate hypertension (“significant” in Table 35.1) generally requires a brief but thorough history and physical examination with emphasis on detecting pathophysiologic risk factors for hypertension and signs and symptoms of the complications of chronic hypertension. Barring the discovery of any of these, the child should be referred for follow-up and further

evaluation with his or her primary care physician. The emergency physician in this circumstance should not institute outpatient treatment. In fact, although it is appropriate to initiate the process of patient education in the ED regarding such factors as weight loss, salt reduction, and exercise, a definitive diagnosis of hypertension should not be offered until the child has had repeated measurements of his or her blood pressure on several occasions.

The workup of a child with severe hypertension requires careful evaluation for the presence of clinical findings that may represent either the primary cause of the elevated blood pressure or the secondary, systemic effects. Relevant history includes frequent urinary tract infections, unexplained fevers, hematuria, dysuria, frequency, and edema—all suggestive of possible renal disease. A history of umbilical artery catheterization as a neonate may indicate risk of renal artery stenosis. Ingestion of prescription, over-the-counter, or illicit drugs may support the diagnosis of drug-induced hypertension. Alternatively, a history of sweating, flushing, palpitations, fever, and weight loss may indicate a pheochromocytoma.

Physical examination should concentrate on identifying involved organ systems. Thus, the cardiac examination should inspect for evidence of congestive heart failure (CHF) and pulmonary edema. Femoral pulses should be palpated because their absence suggests aortic coarctation. Neurologic evaluation should include funduscopic examination for such hypertensive changes as hemorrhages, infarcts, and disc edema. In addition, testing of the pupillary light reflex and visual acuity, and observation for sensorimotor symmetry should be included. Abdominal examination may reveal the presence of a bruit or palpable kidneys, implicating a renovascular or renal cause for the hypertension.

Ancillary investigations depend on the severity of the child's hypertension. In the child with severe hypertension, a number of laboratory studies are appropriate. Complete blood count, electrolytes, blood urea nitrogen, serum creatinine, and uric acid and urinalysis are warranted in the asymptomatic child with severe hypertension. In addition, urine culture should be obtained in all girls and in boys with known renal pathologic conditions. In the child with hypertensive emergency, early intravenous access should also be established.

In symptomatic children with hypertension, further evaluation includes an electrocardiogram and chest radiograph. These tests help evaluate myocardial function and the extent of damage from hypertension, as well as the presence of CHF. Although several additional sophisticated and/or invasive studies exist for the evaluation of hypertension, these are rarely part of the routine ED assessment.

In some instances, the cause of hypertension is already known, as in a child with end-stage renal disease, or is immediately apparent, as in an adolescent who has used cocaine. When the cause is not readily identified, a systematic approach is indicated (Fig. 35.1). Because diseases of the genitourinary tract are among the more common sources for hypertension in children, a logical starting point is to ascertain whether the history suggests prior urinary infection, whether the physical examination identifies signs of renal disease (e.g., edema or an abdominal bruit of renal artery stenosis), or whether the urinalysis is abnormal. Of the cardiovascular causes, coarctation of the aorta is the most likely to manifest in a toddler or older child with previously undiagnosed hypertension. Physical examination in patients with this lesion is notable for femoral pulses that are absent or diminished compared with those in the upper extremities. When a careful evaluation of the renal and cardiovascular systems is not revealing, a detailed neurologic examination is in order because any condition accompanied by increased intracranial pressure may elevate the systemic blood pressure. The history and signs of head trauma (see Chapter 105) are usually obvious but may be occult when injuries in young children are not witnessed or are intentionally inflicted. The emergency physician should specifically examine the fundi for papilledema and ascertain the presence of ataxia or other focal neurologic findings. Although the endocrine disorders are relatively rare, careful history taking and a directed physical examination may identify temperature sensitivity, obesity, a goiter, abdominal striae, or abnormal pigmentation suggestive of hyperthyroidism or Cushing's syndrome. Intermittent headaches and flushing occur with pheochromocytoma. Finally, specific questioning about the ingestion of illicit drugs or other medications and a toxicologic screen are appropriate when no other cause for hypertension is apparent. A negative evaluation for a source of an elevation of blood pressure in the ED is compatible with, but not sufficient for, the diagnosis of essential hypertension. Follow-up is always indicated, usually accompanied by further testing in patients under the age of 10 years.

FIGURE 35.1. Diagnostic approach to acute hypertension in a previously healthy child. (See Fig. 35.2 for initial triage and stabilization for significant or severe hypertension.)

FIGURE 35.2. Approach to the initial ED triage and stabilization of the hypertensive child.

MANAGEMENT

The decision to treat a child with hypertension in the ED setting depends on the acuteness of the rise in blood pressure, the presence of symptoms, preexisting medical problems, and the extent of end-organ damage (Fig. 35.2).

For children with hypertensive emergencies (i.e., blood pressure associated with evidence of end-organ damage or dysfunction), treatment must be rapid and, at the same time, cautious. There is a compelling reason for this conservative approach. Under normal conditions, the cerebral vasculature autoregulates cerebral blood flow, maintaining flow at a relatively constant level despite variations in the blood pressure. Thus, as blood pressure rises, the cerebral vasculature constricts, and as blood pressure falls, the vasculature normally dilates. However, when blood pressure rises significantly higher than normal, the range of pressures over which cerebral autoregulation occurs shifts to higher values, particularly in those with chronic hypertension. Overaggressive lowering of these hypertensive values may bring the blood pressure below the “new” lower limit of autoregulation. At this point, vessels are maximally dilated and ischemia may occur, leading to neurologic deficits, including infarction of the optic nerve; thus, attention to vision and the pupillary reflex provides a means of monitoring for this problem. In most emergent cases, it is recommended that blood pressure be reduced gradually—generally by about 25% over several minutes to several hours, depending on the severity of the emergency. Management of hypertensive emergencies is discussed further in Chapter 86.

The choice of which drug to use in the ED treatment of a child with hypertensive crisis depends on the severity of the patient's hypertension, the patient's current medications, the suspected cause of the hypertension, and the organs involved. Thus, hypertension caused by a catecholamine-secreting tumor (pheochromocytoma) might best be controlled with an a-blocking agent such as phentolamine. On the other hand, if hypertension is associated with an intracerebral bleed, medications that cause an increase in cerebral blood flow, such as nifedipine and nicardipine, are best avoided.

In addition to treating the elevation in blood pressure, the child with complications of hypertension also may require treatment for the specific complications. Thus, the child with seizures or CHF often requires the standard treatment for those problems in addition to antihypertensive therapy. However, when other complications are thought to be secondary to severe hypertension, treatment of the hypertension should take precedence. Of course, attention to airway, breathing, and circulation are to be given priority over all other therapeutic interventions.

Specific Therapy

Generally, only children presenting with hypertension that falls into the categories of hypertensive emergency or hypertensive urgency require pharmacologic intercession in the ED. As mentioned, the medication used depends on a number of factors, including the treating physician's familiarity with the antihypertensive agents.

Hypertensive Emergencies

Children with hypertension and major end-organ abnormalities such as CHF, encephalopathy, or seizures require immediate blood pressure reduction. Careful attention to airway, breathing, and circulation is followed by establishment of vascular access and treatment with one of the following parenteral medications (Table 35.5).

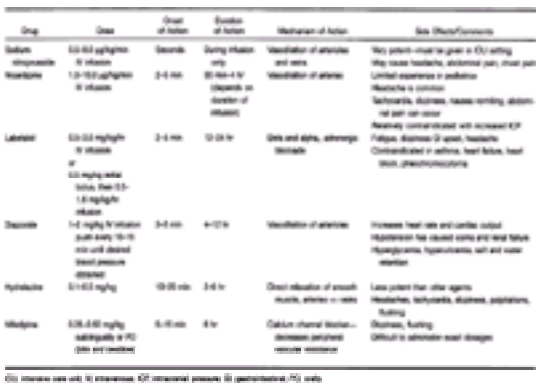

Table 35.5. Drugs Commonly Used for the Treatment of Hypertensive Emergencies in Children

Sodium Nitroprusside

Nitroprusside is a powerful vasodilator, affecting the smooth muscle of both the resistance and capacitance vessels. Its onset of action is almost immediate, and its duration of action is extremely short. Because of its venous dilatory effects, nitroprusside reduces preload, thus often improving cardiac output if CHF is present. Nitroprusside is metabolized to thiocyanate, and cyanide is an intermediary in its metabolism. Consequently, cyanide and thiocyanate toxicity must be considered a risk of its use, although the circumstances under which these develop have been poorly characterized.

Nitroprusside is given as an intravenous infusion, starting at a dosage of 0.5 mg/kg/min and increasing as needed to 8.0 µg/kg/min. The patient should be kept in the recumbent position because of the frequency of orthostatic hypotension. The degree of drop in blood pressure is dose-related; thus, the infusion should be started at the low end of the dosage range and titrated to achieve the desired blood pressure levels. The average dosage required for control of hypertension is approximately 3.0 µg/kg/min. Because nitroprusside has an extremely short half-life, blood pressure returns to pretreatment levels within 1 to 10 minutes of cessation of the infusion. Patients treated with nitroprusside should be admitted to an intensive care unit for blood pressure monitoring.

Nicardipine

Nicardipine is a calcium channel blocker that is effective in controlling elevated blood pressure when administered intravenously. It acts by reducing peripheral vascular resistance without reducing cardiac output. It has also been shown to diminish both cardiac and cerebral ischemia. Although there is relatively little literature on nicardipine dosing in children, recent studies suggest that an infusion rate of 1 to 10 µg/kg/min is effective. Because its onset of action is 2 to 5 minutes, the infusion can be started at the low end of this range and titrated upward. The half-life of nicardipine is approximately 40 minutes, although its duration of action after cessation of administration increases with duration of the infusion. Thus, nicardipine's effect on blood pressure generally diminishes within 30 minutes of discontinuing an infusion of less than 2 hours' duration, but its effect remains for up to 4 hours after discontinuing a 12- to 20-hour infusion. Excessive reductions in blood pressure, although uncommon, can be reversed by administration of calcium. *Experience with nicardipine in children is extremely limited; thus, this drug should be used with foremost caution.* Because it elevates intracranial pressure, nicardipine should generally be avoided in patients with increased intracranial pressure.

Labetalol

Labetalol is a combined a- and b-adrenergic blocking agent with a rapid onset of action (usually 5 to 10 minutes) when given intravenously. Because marked orthostatic hypotension can occur with its use, the patient should be kept in the supine position during and for some time after administration.

When administered for hypertensive emergencies, labetalol may be given as an initial minibolus of 0.5 mg/kg followed by a constant infusion of 0.5 to 1.5 mg/kg/hr, titrated to achieve the desired blood pressure. Alternatively, some prefer to give successive miniboluses of 0.3 to 1.0 mg/kg as often as every 10 to 15 minutes as needed. Others start with an infusion of 0.5 mg/kg/hr titrated slowly to a maximum of 3 mg/kg/hr. Labetalol can be somewhat difficult to titrate to effect; thus, it should be used with foremost caution. Because labetalol is a b blocker, it should not be used in patients with asthma, heart block, or CHF. It is also contraindicated for the treatment of hypertension secondary to pheochromocytoma.

Diazoxide

Diazoxide has a structural similarity to the thiazide diuretics but exerts its primary effect on arterial smooth muscle, causing vasodilation of mainly the resistance vessels. Although its rapid intravenous infusion rarely causes the blood pressure to drop below the normal range, there have been reported cases of coma and renal failure associated with hypotension after diazoxide infusion. Consequently, its use should be accompanied by careful monitoring.

Diazoxide has traditionally been given as a rapid bolus injection of 5 mg/kg per dose. However, less hypotension has been noted when the drug is given as frequent, small boluses of 1 to 2 mg/kg over 5 to 10 seconds every 10 to 15 minutes until the desired blood pressure is obtained. The effect of diazoxide is noted within 3 to 5 minutes, and its duration of action is generally between 4 and 12 hours.

Hypertensive Urgencies

The following drugs have less potency and/or a slower onset of action than those just discussed. As a result, they are more appropriate for hypertensive crises not accompanied by life-threatening manifestations.

Nifedipine

Nifedipine is a calcium channel blocker that can be given by the sublingual, oral, or rectal route. Recently, it has been shown to be effective for the treatment of hypertensive urgencies in children. It acts by decreasing peripheral vascular resistance.

Nifedipine is administered sublingually in a dose of 0.25 to 0.5 mg/kg (up to a maximum of 20 mg). Alternatively—perhaps preferably—the capsule can be chewed and swallowed. Onset of action is within 5 to 15 minutes; duration of action is approximately 6 hours. Facial flushing is a common side effect of nifedipine administration.

Hydralazine

Hydralazine is an arteriolar vasodilator that can be given intramuscularly but has even faster onset of action when given intravenously (10 to 20 minutes). Because hydralazine is less potent than several of the other parenteral antihypertensives, it may not be the drug of choice when life-threatening signs or symptoms are evident.

Hydralazine is given at a starting dose of 0.1 mg/kg intravenously and may be increased to 0.5 mg/kg. Its duration of action is 3 to 6 hours. Reflex tachycardia is a relatively common side effect and often necessitates the addition of a b-adrenergic blocking agent.

Other Agents

Several other agents have been used in the treatment of hypertension in children, including trimethaphan, minoxidil, methyldopa, reserpine, and clonidine. Because these agents often take considerably more time to lower blood pressure than those previously discussed and have a more serious profile of side effects, they are generally not considered the best choice for management of hypertensive emergencies.

SUMMARY

It is not unusual that a child presenting to the ED will be found to have an elevation in blood pressure. In many such cases, the blood pressure will normalize with rest or acclimation to the environment. Occasionally, however, the elevation in blood pressure will be sustained. In children with asymptomatic hypertension and no target organ involvement, the emergency physician must ensure adequate follow-up. Hypertension that affects or threatens to affect end organs, on the other hand, requires evaluation and initiation of treatment in the ED (Fig. 35.2). Indeed, in severe or life-threatening cases, blood pressure reduction will need to be instituted before the cause of the hypertension is known.

Suggested Readings

Buchi KF, Siegler RL. Hypertension in the first month of life. J Hypertension 1986;4:525–528.

Daniels SR. The diagnosis of hypertension in children: an update. Pediatr Rev 1997;18:131–135.

Farine M, Arbus GS. Management of hypertensive emergencies in children. Pediatr Emerg Care 1989;5:51–55.

Fivush F, Neu A, Furth S. Acute hypertensive crises in children: emergencies and urgencies. Curr Opinion Pediatr 1997;9:233–236.

Groshong T. Hypertensive crisis in children. Pediatr Ann 1996;25:368, 370–371, 375–376.

National High Blood Pressure Education Program Working Group on Hypertension Control in Children and Adolescents. Update on the 1987 task force report on high blood pressure in children and adolescents: a working group report from the National High Blood Pressure Education Program. Pediatrics 1997;98:649–658.

Task Force on Blood Pressure Control in Children. Report of the second task force on blood pressure control in children--1987. Pediatrics 1987;79:1–25.

Varon J, Fromm RE. Hypertensive crises. The need for urgent management. Postgraduate Med 1996;99:189–191, 195–203.

CHAPTER 36

Immobile Arm

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An infant or child brought for evaluation of an “immobile arm” is not moving the limb because of pain or weakness. The evaluation is often challenging because most of these children are preverbal; therefore, the history is second- or third-hand if available at all. The patient is unable to report symptoms or localize pain, and the physical examination often is difficult because of the child's fear of strangers. These children can be considered as having an upper extremity equivalent of “limp”; by using historical information, physical findings, selective radiologic studies, and laboratory tests, the physician can evaluate and treat children with this complaint.

DIFFERENTIAL DIAGNOSIS

Table 36.1 lists most conditions that cause decreased use of the arm. Trauma is the most common cause of decreased arm movement in children. Any injury from the clavicle to the fingertips can cause pain in a child and lead to diminished use of the limb; these injuries range from serious (fracture or dislocation with neurovascular compromise) to a simple contusion. Most young children with diminished arm use have a radial head subluxation (“nursemaid's elbow”), fracture, or soft-tissue injury. Although one can often elicit a history of trauma, the diagnosis must be considered even in its absence because of unwitnessed events in preverbal children or, less commonly, intentional injuries inflicted by caretakers who are not forthcoming. With musculoskeletal injuries, the child may have an obvious abnormality such as a deformity or a contusion or more subtle findings of localized tenderness or decreased arm movement. Children with hemophilia may have hemarthrosis or hematoma with minimal trauma. Radiographs are useful for demonstrating most fractures or dislocations but may appear normal with Salter type I fractures and nursemaid's elbow, as well as with contusions and other minor soft-tissue injuries.

Table 36.1. Differential Diagnosis of the Immobile Arm

Although much less common than trauma, infection may also cause decreased use of an arm. There may be a history of fever, and onset of arm disuse often is less abrupt than with trauma. The infection can be located at any point from the shoulder to finger and may be superficial (e.g., cellulitis, paronychia) or deep. Arthritis and osteomyelitis often have associated localized swelling, warmth, and tenderness; infected joints usually have limited, painful range of motion. With more severe infections, the child may be febrile and appear ill (especially if bacteremic). Laboratory findings may include elevated white blood cell count and sedimentation rate (ESR) or C-reactive protein (CRP), and blood cultures may yield the offending organism. Acutely, radiographs often are nondiagnostic; if arthritis or osteomyelitis is suspected, arthrocentesis and technetium bone scan should be considered.

Other inflammatory causes of arm pain include noninfectious arthritis and myositis. In addition to a swollen, tender joint, children with arthritis caused by postinfectious, Lyme, and rheumatologic diseases may have multiple joint involvement, rash, fever, adenopathy, heart murmur, hematuria, or bloody stools. If the examination suggests an inflammatory arthritis but cannot exclude a septic process, arthrocentesis is necessary for definitive diagnosis.

Tumors are an uncommon cause of diminished arm use. The tumors can be benign or malignant and of bone, cartilage, or muscle, or they may represent neoplastic infiltration of bone marrow (e.g., leukemia, neuroblastoma). Tumors usually are less acute in onset; cardinal symptoms may include pain and, perhaps, increasing mass or joint swelling, although the lesions may be asymptomatic. Occasionally, tumors lead to a pathologic fracture. Systemic complaints, including fever, malaise, and weight loss, may be present. Physical examination may reveal localized tenderness, joint swelling, or a mass of the soft tissue or bone. With leukemia or neuroblastoma, fever, abdominal mass, hepatosplenomegaly, or pathologic adenopathy also may be found. Plain radiographs are of obvious importance; lesion location and radiologic appearance (density and peripheral margin) can be diagnostically significant. Complete blood count (CBC) and ESR or CRP are helpful in screening for possible infection or bone marrow neoplasm.

Children with neurologic abnormalities will have diminished use of an arm because of weakness, with or without pain. An isolated monoplegia may be caused by a radiculopathy, plexopathy, or neuropathy that results from compression,

inflammation, or injury. Trauma, particularly traction on the arm, commonly leads to neurologic abnormalities (e.g., brachial plexus injury from birth); however, nontraumatic conditions may have an abrupt onset with no apparent antecedent illness. The child will have diminished arm movement and weakness, and he or she may even experience pain; unlike the previously discussed causes of arm disuse, however, the pain (if present) usually is not reproducible with palpation and is not accompanied by swelling or redness. Reflexes may be diminished or absent. It is important to identify any associated neurologic abnormalities because facial or leg weakness may be subtle but would point to a lesion in the central nervous system.

Children with hemoglobinopathies (most commonly sickle cell disease) may present with decreased arm use because of vaso-occlusive crisis, causing ischemia or infarction of bone marrow with acute bone pain. Long bones are commonly affected; however, young children often have involvement of the small bones of the hands and feet (dactylitis). Usually, no precipitating events are identified. The child is in pain and has localized tenderness and swelling of the involved areas; associated warmth and erythema may be present. Acutely, no bony abnormalities show on radiographs. Because the “hand–foot syndrome” may be the first clinical manifestation of sickle cell disease, all African-American children with unexplained limb pain or swelling must be screened for hemoglobinopathy if not tested previously. It also is particularly important to consider septic arthritis and osteomyelitis in children with sickle cell disease because they are susceptible to infection and the clinical findings may overlap with bone infarction, particularly if fever and leukocytosis are present.

Several other much less common processes can cause decreased upper limb use. These include avascular necrosis of the humeral head or capitellum in otherwise healthy children and reflex sympathetic dystrophy. Table 36.2 lists common causes of diminished arm use. Table 36.3 lists life- and limb-threatening causes of diminished arm use.

Table 36.2. Common Causes of Diminished Arm Use

Table 36.3. Life- and Limb-Threatening Causes of Diminished Arm Use

EVALUATION AND DECISION

The evaluation of the child who has diminished arm movement consists of a complete history, a thorough physical examination, radiographic studies, and selected laboratory tests, when indicated. Based on these findings, appropriate management can be undertaken.

A history of any trauma should be ascertained. Details of the event may provide clues to the type of injury incurred; a fall onto an outstretched hand may cause a wrist, forearm, or elbow injury, whereas a sudden arm pull by a caretaker can cause dislocation or subluxation of the radial head (nursemaid's elbow). Some children with radial head subluxation may have a mechanism of injury other than a pull. If an immediate causative traumatic event is not elicited, the duration, course, and pattern of diminished arm use should be clarified. Fever, malaise, rash, or weight loss may give clues to a systemic illness. If the patient is an infant, it should be determined whether the arm disuse was from birth: a difficult delivery may lead to clavicular fractures or brachial plexus injuries. Infants do not always mount a febrile response to infection and may have only nonspecific symptoms of diminished feeding, increased sleeping, lethargy, or irritability. General medical history should include any history of inflammatory process, hemophilia, or sickle cell disease.

After a careful history, a complete physical examination should be performed. Fever should be noted and may indicate infection or, less likely, inflammatory or neoplastic processes. Observation and inspection, sometimes from a distance of several paces, can provide information that might otherwise not be obtainable (because many children cry when approached or touched by a stranger). The position of the arm should be noted. A child with nursemaid's elbow often holds the arm pronated and slightly flexed with obvious diminished movement, although often without apparent discomfort. A child with neurologic abnormality may hold the arm limply at the side of the body. Close inspection for areas of deformity, redness, swelling, or bruising should be noted. Observation of the child's reach and grasp for an interesting object can provide information about the active range of motion and neurologic function. While distracting the child, the physician should palpate the limb systematically from clavicle to fingertips to identify areas of warmth, swelling, or tenderness. Joints should be assessed for warmth, swelling, tenderness, and range of motion; however, if a history of trauma is present, manipulation can be deferred until an acute fracture has been excluded. Neurovascular integrity of the arm should be assessed carefully. A thorough general examination that notes rash, other joint abnormalities, hepatosplenomegaly, adenopathy, abnormal mass, and neurologic status should be performed.

Plain radiographs are one of the most useful studies for evaluating the child with diminished arm use. They may reveal a fracture or dislocation, joint effusion, or lytic bone lesion. If a discrete area of tenderness is identified, radiographs of that location, including the joint above and below, should be obtained. If the focus of pain is not apparent, it may be necessary to obtain radiographs of the entire limb from clavicle and shoulder to fingers.

A CBC may help in the diagnosis of infection, inflammation, malignancy, or hemoglobinopathy. Although nonspecific, an

ESR or CRP may be useful in differentiating inflammatory or infectious processes from other causes.

Other tests helpful in selected cases include blood culture (if an infectious process is suspected), hemoglobin electrophoresis (if sickle cell disease is a possibility), and technetium bone scan (for osteomyelitis, septic arthritis, aseptic necrosis). Arthrocentesis is imperative if septic arthritis is a possibility, and if osteomyelitis is suspected, evaluation and treatment should proceed urgently.

When a child is brought for evaluation of diminished arm use, the physician should first determine whether this decrease in mobility resulted from a specific traumatic event (Fig. 36.1). If the history is classic for radial head subluxation, the patient is holding the arm pronated and slightly flexed, and there is no localized tenderness or swelling, the physician may attempt reduction. If the child does not regain full use of the arm quickly, as in most other cases of trauma, radiographs should be obtained. For many conditions, the radiographic studies make the diagnosis (e.g., fracture, dislocation). Normal radiographs in the setting of acute trauma usually imply soft-tissue injury, and the patient should be treated symptomatically with close follow-up, provided that neurovascular integrity is established. If radiographs appear normal but the child has reproducible tenderness localized to the epiphyseal plate, the patient should be treated for a Salter type I fracture. Occasionally, a child with a nursemaid's elbow may have an atypical history (e.g., “fell onto arm”); if radiographs exclude a fracture but the patient is holding the arm in a characteristic position, a reduction should be attempted.

FIGURE 36.1. Approach to the child with diminished arm use.

Children with neurologic abnormalities should be evaluated urgently to localize the site and cause of the impairment; the appropriate subspecialist (neurologist, neurosurgeon) should be involved.

If the child has no clear history of trauma but is afebrile and has no obvious localizing findings of infection, the limb should be evaluated radiographically. Abnormalities revealed might include fracture, dislocation, tumor, or effusion. If radiographs are normal in these children, one could consider obtaining a CBC, ESR or CRP, blood culture, or hemoglobin electrophoresis to evaluate for occult infectious or inflammatory processes.

Children who are febrile, have signs of localized inflammation (e.g., warm, swollen joint), or have evidence of systemic illness should have CBC, ESR or CRP, and blood culture obtained in addition to radiographs. Based on specific findings, further evaluation might include arthrocentesis, bone scan, or rheumatologic tests. When the initial history, physical examination, laboratory tests, and radiographs localize site and cause of the pathology, the physician can begin specific treatment.

A few children with no history of trauma in whom a thorough initial evaluation is unrevealing will have a nursemaid's elbow. Therefore, an attempt at reduction is warranted in selected cases. Children with persistently diminished arm movement who are afebrile and nontoxic and who have no localizing findings, normal neurovascular function, and normal laboratory tests likely have an occult soft-tissue injury and can be treated as outpatients. A few of these children may have indolent pathologic processes or occult fractures; therefore, close follow-up must be ensured. These patients should be reevaluated every few days until normal arm use is regained or until evidence of a pathologic process develops. If arm disuse persists, a more extensive evaluation to diagnose or exclude occult fracture, infection, tumor, or collagen vascular disease is in order.

Suggested Readings

Bora FW. The Pediatric Upper Extremity. Philadelphia: WB Saunders, 1986.

Cassidy JT, Petty RE. Textbook of Pediatric Rheumatology. 2nd ed. New York: Churchill Livingstone, 1990.

Dich VQ, Nelson JD, Haltalin KC. Osteomyelitis in infants and children. Am J Dis Child 1975;129:1273–1278.

Fenichel GM. Clinical Pediatric Neurology: A Signs and Symptoms Approach. Philadelphia: WB Saunders, 1988.

McCarthy PL, Wasserman D, Spiesel SZ, et al. Evaluation of arthritis and arthralgia in the pediatric patient. Clin Pediatr 1980;19:183–190.

Schaller JG. Arthritis and infections of bones and joints in children. Pediatr Clin Am 1977;24:775–790.

Schunk JE. Radial head subluxation: epidemiology and treatment of 87 episodes. Ann Emerg Med 1990;19:1019–1023.

Schutzman SA, Teach S. Upper-extremity impairment in young children. Ann Emerg Med 1995;26:474–479.

Watson JR, Burko H, Megas H, Robinson M. The hand-foot syndrome in sickle-cell disease in young children. Pediatrics 1963;31:975–982.

Weissberg ED, Smith AL, Smith DH. Clinical features of neonatal osteomyelitis. Pediatrics 1974;53:505–510.

CHAPTER 37

Injury—Ankle

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Young children with ankle injuries may complain of pain anywhere from their midcalf to their toes because it is often difficult for children to localize pain. Conversely, disease in the lower leg and foot can cause referred pain to the ankle.

The ankle joint is composed of three bones: the tibia, the fibula, and the talus. The bony prominence of the distal fibula constitutes the lateral malleolus, whereas the prominence of the distal tibia forms the medial malleolus. The physes are located one to two fingerbreadths above the distal ends of the tibia and fibula.

The ankle ligaments are attached to the physes. The distal tibial physis is the most commonly injured growth plate in the lower extremities. It is second only to the distal radius in the incidence of physeal injuries.

Growth plates and bone are weaker than ligaments. Consequently, ankle trauma in children under 14 to 15 years of age is much more likely to cause fractures of the physis and the adjacent epiphysis and/or metaphysis than ligamentous injuries or sprains.

DIFFERENTIAL DIAGNOSIS

A number of traumatic injuries may cause ankle pain (Table 37.1). Although trauma is the most common cause of ankle pain in children, infectious, rheumatologic, inflammatory, neoplastic, and hematologic abnormalities also should be considered (Table 37.2). Again, a complaint of ankle pain may result from a lesion anywhere between the knee and the toe, particularly in the preverbal child. The most common injuries vary according to age (Table 37.3).

Leg	Sprains
Tibial fractures	Contusions
(toddler's fracture)	Osteochondritis dissecans
Fibular fractures	Hemarthrosis
Contusions	*Foot*
Compartment syndrome	Fractures
of the calf	Talar
Ankle	Navicular
Fractures	Fifth metatarsal (Jones fracture)
Distal tibial	Calcaneal
Distal fibular	Sprains
Physeal	Contusions

Table 37.1. Differential Diagnosis of Traumatic Injuries That Cause Ankle Pain

Trauma	Brodie's abscess (subacute osteomyelitis of the distal tibia)
Fractures	*Rheumatologic*
Sprains	Juvenile rheumatoid arthritis
Contusions	Rheumatic fever
Osteochondritis dissecans	Reiter's syndrome
Hemarthrosis	*Hematologic*
Inflammatory	Sickle cell disease (pain crisis)
Tendonitis	Hemophilia (hemarthrosis)
Synovitis	*Osteochondroses (avascular necrosis)*
Periostitis	Köhler's disease (navicular)
Sever's disease (calcaneal apophysitis)	Freiberg's disease (second metatarsal)
Infectious	*Tumors*
Osteomyelitis	Ewing's sarcoma
Soft-tissue abscess	Osteoid osteoma
Septic joint	

Table 37.2. Differential Diagnosis of Ankle Pain

Toddler	Child	Adolescent
Spiral fracture of tibia	Salter I fracture of distal fibula	Ankle sprain
Soft-tissue contusion	Soft-tissue contusion	Soft-tissue contusion

Table 37.3. Common Injuries Associated with Ankle Pain According to Age

Ankle Fractures

Fractures of the ankle account for 5.5% of all fractures in pediatrics. The system used to classify ankle fractures in children differs from the one used in adults because of the presence of growth plates and the possible implications of physeal injuries. The Salter-Harris classification is most commonly applied, as described in Chapter 115.

Inversion ankle injuries in the preadolescent most commonly cause a Salter type I fracture of the distal fibula (Fig. 37.1). Clinically, the patient has swelling about the lateral malleolus and tenderness at the distal fibular physis. Fractures confined to the physes may not be visible on a routine radiograph.

FIGURE 37.1. Inversion injury.

In severe inversion injuries, the distal fibular fracture just described may be accompanied by a fracture of the medial malleolus (Fig. 37.2). This medial malleolus fracture is usually a Salter type III or IV fracture of the distal tibia. These patients will have tenderness at the medial malleolus as well as the distal fibular physis.

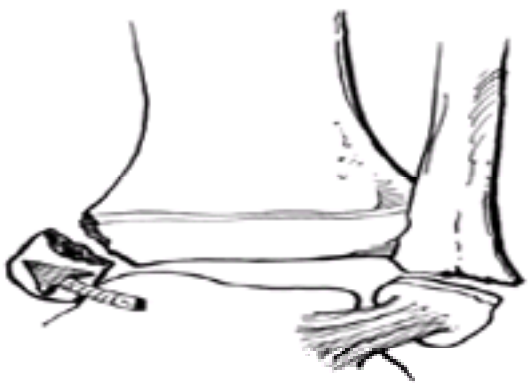

FIGURE 37.2. Severe inversion injury.

Fractures resulting from eversion of the ankle are usually a combination of a Salter type II fracture of the lateral tibia and a transverse fracture of the fibula (Fig. 37.3). The fibular fracture is relatively high (4 to 7 cm above the fibular physis).

Therefore, it is important to examine the full length of the fibula in patients with ankle injuries.

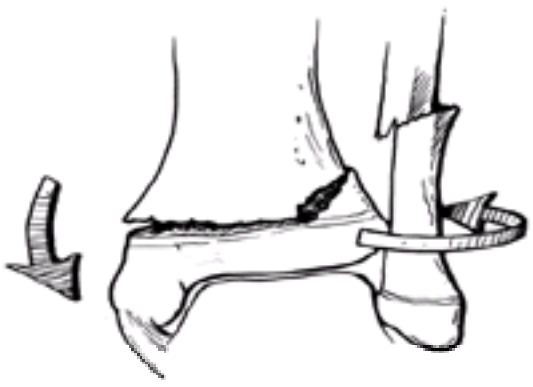

FIGURE 37.3. Eversion injury.

Direct axial compression of the ankle is uncommon but can cause a Salter type V injury to the distal tibia.

External rotation injuries are responsible for lesions known as transitional fractures. Transitional fractures occur during adolescence when closure of the growth plates is beginning. Closure of the distal tibial physis starts in the center of the bone and then spreads medially, posteriorly, and finally, laterally. The distal tibial physis closes before the distal fibular physis.

As skeletal maturity (and physeal closure) progresses, the relative strengths of various parts of the tibia change. As a result, the same mechanism of injury may cause very different fracture patterns depending on the patient's age. The juvenile fracture of Tillaux and the triplane fractures are examples of transitional fractures.

A juvenile Tillaux fracture, in which a fragment of bone is torn off the lateral border of the tibia by the anterior tibiofibular ligament (Fig. 37.4 and Fig. 115.63), is a Salter type III injury of the distal tibia. This fracture is seen almost exclusively in patients between the ages of 12 and 14 years because closure of the medial aspect of the distal tibial physis begins around this time, whereas the lateral aspect remains open and, consequently, less stable for an additional 18 months. The greater the skeletal maturity of the patient, the more lateral the epiphyseal fracture line occurs.

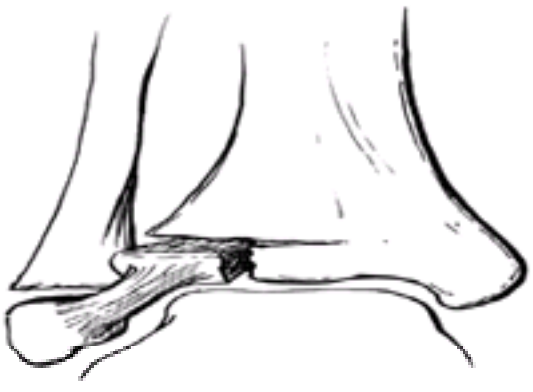

FIGURE 37.4. Juvenile Tillaux, Salter-Harris type III fracture of the distal tibial physis; the medial part of the tibial physis is fused.

Diagnosis of these fractures may be difficult because routine radiographs may not show the fracture line well. If displacement is minimal, the only radiographic sign may be a slight widening of the lateral tibial physis or a faint vertical fracture line through the epiphysis on anteroposterior (AP) or oblique views. In some cases, the only finding may be local tenderness in the area of the lateral tibial physis. Multiple oblique views, computed tomography (CT), or conventional tomography may be needed to adequately delineate the extent of the fracture.

Growth arrest and angular deformity are rare because these fractures occur at the time of physeal closure. However, ankle joint arthritis may complicate the long-term outcome if the diagnosis is missed or the reduction is inadequate.

Triplanar fractures are characterized by a fracture line that runs in three planes: coronal, sagittal, and transverse. They are a combination of a juvenile Tillaux and a Salter type II fracture of the distal tibia. Two types of triplane fractures have been reported. The first is a three-fragment fracture (Fig. 37.5). The first fragment is the same as the one found in the juvenile Tillaux fracture—a fragment of epiphysis torn off the anterolateral quadrant of the tibia. The second fragment is the remaining medial part of the epiphysis, which is attached to a posterior spike of metaphyseal bone. The third fragment is the tibial shaft.

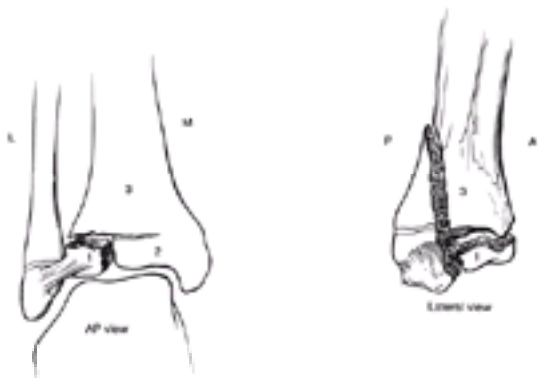

FIGURE 37.5. Three-fragment triplanar fracture. *L,* lateral; *M,* medial; *P,* posterior; *A,* anterior.

A two-fragment fracture has also been reported. The first fragment is again the lateral tibial epiphysis, but it is attached to a posterior spike of metaphyseal bone. The second fragment is the remaining medial epiphysis and is attached to the tibial shaft (Fig. 37.6).

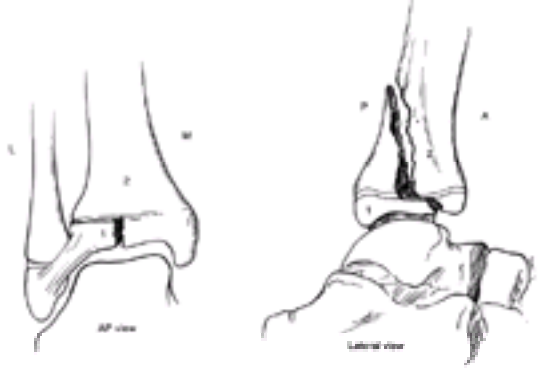

FIGURE 37.6. Two-fragment triplanar fracture. *L,* lateral; *M,* medial; *P,* posterior; *A,* anterior.

Radiographically, triplanar fractures have the appearance of a Salter type III fracture on the AP view and a Salter type II fracture on lateral view. If only the AP view is obtained, it may be difficult to distinguish these fractures from the juvenile fracture of Tillaux. The key to diagnosis is the posterior metaphyseal spike seen on the lateral film.

Ankle Sprains

Ankle sprains in the child or preadolescent are less common than fractures because the ligaments in this age group are much stronger than growth plates or even bone. If a ligamentous injury occurs in a child with an open growth plate, an associated avulsion fracture is almost always present. However, once skeletal maturity is reached, ankle sprains become the most common of sports injuries.

Inversion injuries cause 85% of ankle sprains. The most commonly injured structures are the lateral ligaments. Three lateral ligaments support the ankle joint: the anterior talofibular (ATFL), the calcaneofibular (CFL), and the posterior talofibular (PTFL) (Fig. 37.7). The ATFL is the weakest and most commonly injured of the three. The CFL is intermediate in strength and rarely is injured without an associated tear of the ATFL. The PTFL is the strongest and least injured of the lateral ligaments. Because its fibers run horizontally, only extreme dorsiflexion will stress the PTFL. The peroneus brevis tendon also traverses the lateral aspect of the ankle joint and can be injured by inversion stress. It inserts at the base of the fifth metatarsal.

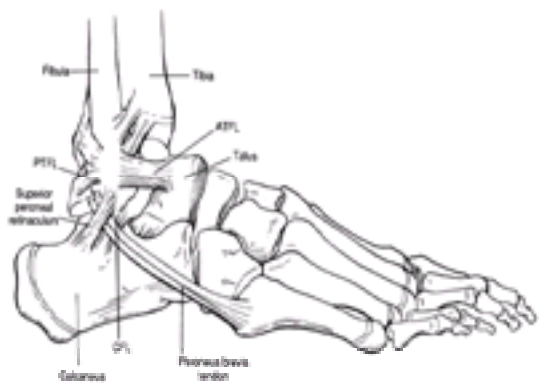

FIGURE 37.7. Lateral view of the ankle. *ATFL,* anterior talofibular ligament; *PTFL,* posterior talofibular ligament; *CFL,* calcaneofibular ligament.

Eversion injuries account for 15% of ankle sprains. The deltoid ligament, which supports the medial aspect of the ankle, is most commonly affected by this mechanism (Fig. 37.8). It is made up of deep and superficial fibers. Eversion may also cause disruption of the tibiofibular syndesmosis, which connects the distal tibia and fibula.

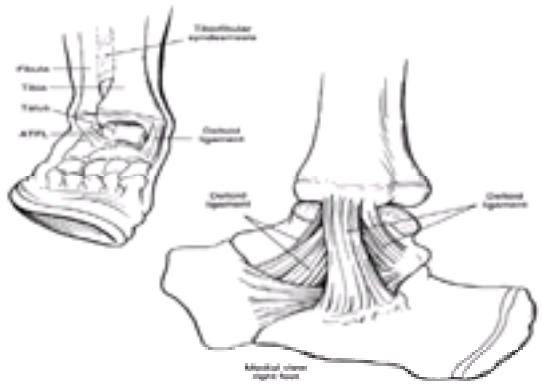

FIGURE 37.8. Ankle eversion injury. *ATFL,* anterior talofibular ligament.

Classification of Ankle Sprains

There are many systems of classification for ankle sprains. Table 37.4 provides guidelines that can be used in grading injuries to the lateral ligaments.

[illegible]	[illegible]	[illegible]	[illegible]
[illegible]	[illegible]	[illegible]	[illegible]
[illegible]	[illegible]	[illegible]	[illegible]
[illegible]	[illegible]	[illegible]	[illegible]

Table 37.4. Classification of Ankle Sprains

Injuries Associated with Ankle Sprains

Approximately 7% of ankle sprains are accompanied by osteochondral fractures of the talus. The medial dome is more commonly fractured than the lateraldome. Avulsions of the peroneus brevis tendon from the base of the fifth metatarsal have been observed in up to 14% of patients with ankle ligament ruptures. If this injury occurs in a child younger than 15 years of age, the avulsed fragment is usually an apophysis and is considered a Salter type I injury. In the older patient, the displaced portion represents a bony fragment and is known as a Jones fracture.

EVALUATION AND DECISION

History

Attempting to obtain a reliable history from patients with ankle injuries can be frustrating. It is a rare occasion when a patient says, “I sustained an inversion injury while playing basketball!” More commonly, the description is, “I twisted it, and it hurts.” Nevertheless, the mechanism of injury, if obtained, can provide a clue to the diagnosis. Other questions include: When did the injury occur? Did swelling occur immediately or gradually? Is there a history of any previous injury to that limb? and Does the patient have a history of any other medical problems—osseous, neurologic, or muscular disease?

A history of fever, rash, or other joint involvement combined with a history of minimal or no trauma suggests nontraumatic diagnoses such as septic joint, arthritis, or collagen vascular disease.

Physical Examination

General Inspection

The physician should look for obvious deformities, open wounds, loss of anatomic landmarks, local swelling, and ecchymosis. If an obvious deformity is present, manipulation of the extremity should be kept to a minimum and neurovascular status assessed promptly. Any break in the skin may communicate with the joint space or constitute an

open fracture. The need for antibiotic coverage must be evaluated immediately.

Neurovascular Evaluation

The physician should palpate the dorsalis pedis and posterior tibial arteries. Skin temperature, color, and capillary refill should be noted. The absence of pulses or the presence of pallor requires immediate attention. A Doppler device may help identify pulses.

Vascular compromise is usually caused by a posterior dislocation. Traction reduction of the deformity should be attempted as rapidly as is feasible by performing the following steps: 1) sedate the patient; 2) apply longitudinal traction to the foot; 3) if relocation is not accomplished in step 2, apply longitudinal traction and pull the foot in a posterior-to-anterior direction; and 4) immobilize the ankle and obtain radiographic studies. If the vascular status has not been compromised, the physician should continue with the examination and evaluate the nerves that cross the ankle. Soft touch and pain sensation of the foot should be tested.

Bony Palpation

All three bones of the ankle joint (the tibia, the fibula, and the talus) should be traced, searching for areas of point tenderness. It is important to palpate the distal tibial and fibular physes because fractures in these areas may not be evident on a radiograph. Any tenderness found along a physis should be considered a Salter type I fracture at the least, even if radiographic studies are negative. Also, the only clue to a juvenile fracture of Tillaux may be tenderness at the lateral tibial physis. The physician should remember to palpate the fibula proximal to the ankle joint. External rotation and triplanar injuries may be associated with high fibular fractures.

Finally, the foot should be examined. This should include palpation of the dome of the talus. This is performed most easily with the foot in plantar flexion. The base of the fifth metatarsal should be palpated. Tenderness here suggests an avulsion of the peroneus brevis tendon.

Once one area of point tenderness is found, examination of the entire joint should continue. A single injury may cause many abnormalities.

Ligament Palpation

The physician should palpate for tenderness along all three lateral ligaments, remembering that each one arises from the distal fibula. The ATFL can be further tested by inverting and plantar flexing the foot. This will increase pain if injury to this ligament is present. More than 4 cm of swelling in an area of lateral ligament tenderness is highly suggestive of significant ligament injury.

The superficial fibers of deltoid ligament on the medial aspect of the joint should be examined. The deep fibers are intra-articular and nonpalpable; therefore, rupture may be present without much medial tenderness. Isolated injuries to the deltoid are rare because of the great strength of this ligament. If the deltoid ligament has been damaged, the tibiofibular syndesmosis is usually disrupted along with it.

Injuries to the tibiofibular syndesmosis may be explored by squeezing the midshafts of the tibia and fibula together, externally rotating the foot, or forcefully dorsiflexing the ankle with the patient supine. Exacerbation of pain with these maneuvers suggests syndesmonic disruption.

Stability Testing

An attempt should be made to assess the stability of the ankle joint. However, stability testing in the immediate postinjury period may be limited significantly by pain, swelling, and/or muscle spasm. Several maneuvers are useful, but they are generally not performed if an ankle fracture is present.

- *Anterior drawer test*: The anterior talofibular ligament is the only structure that prevents forward subluxation of the talus. The anterior drawer test is performed to assess the anterior stability of the ankle joint and the integrity of the ATFL (Fig. 37.9). The test is positive if the foot can be pulled forward by more than 4 mm or if there is a significant difference in the degree of anterior movement in the injured ankle compared with the normal ankle.

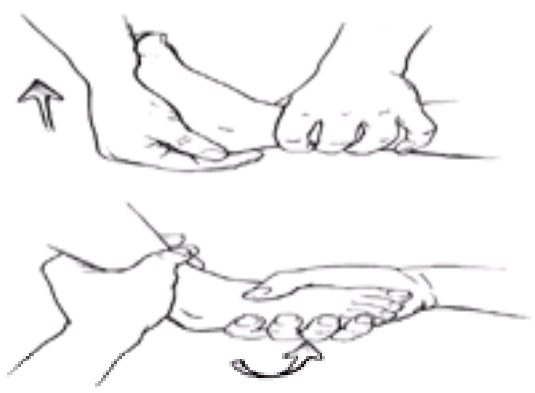

FIGURE 37.9. *Top,* The anterior drawer test is performed by placing the patient's heel in the palm of the examiner's hand with the ankle at a 90-degree angle to the long axis of the leg. The examiner gently, but firmly, moves the heel and foot forward *(arrow). Bottom,* In the talar tilt maneuver, the heel is firmly adducted *(arrow)* and assessed for increased laxity or instability compared with the noninjured side.

- *Talar tilt test:* This test is used to examine the lateral stability of the ankle joint. It is performed by firmly adducting the heel and looking for increased laxity compared with the noninjured joint (Fig. 37.9). Both the anterior talofibular and calcaneofibular ligaments must be torn to cause gross lateral ankle instability.

Radiographic Imaging

With the advent of cost containment, a number of recent studies have evaluated the use of specific physical findings to predict which patients should have radiographs of the ankle taken. The Ottawa Ankle Rules suggest that radiographs need be obtained only if there is pain near the malleoli and one or both of the following: 1) inability to bear weight (four steps) immediately after the injury and in the emergency department; or 2) bone tenderness at the posterior edge or tip of either malleolus. These criteria apparently predict ankle fractures in adults with 100% sensitivity. Another study evaluated the use of the same criteria to predict ankle fracture in children and found the same results. However, the sample size was small, and there was a paucity of children under 10 years of age; therefore, further investigation is required before these criteria can be adopted safely in the pediatric population.

Radiographic evaluation of the ankle should include at least three views: AP, lateral, and mortise. If tenderness of the proximal fibula is noted, full-length views of the fibula are essential. Tenderness at the base of the fifth metatarsal mandates visualization of this area on the lateral film. If radiographic findings are questionable, consideration should be given to obtaining comparison views of the noninjured ankle.

Areas of soft-tissue swelling should be noted. This may be the only clue to a Salter type I fracture of the distal fibula. Stress films to evaluate growth plate injuries are rarely necessary and may cause further damage. Ultrasound has also been used to evaluate children who have tenderness over the lateral malleolus but normal ankle radiographs; this can help assess the possibility of an occult Salter-Harris type I fracture of the distal fibular physis. However, this practice is not currently the standard of routine care.

The value of stress films to assess ligament damage is also questionable. Severe pain and muscle spasms often prohibit stress maneuvers. Arthrography may be more helpful but is seldom indicated in the acute setting. This method uses the location of extravasated contrast material in the ankle joint to identify ligamentous ruptures.

CT of the ankle is often necessary to fully evaluate triplane fractures.

Approach to Diagnosis

The approach (Fig. 37.10) to the evaluation and diagnosis of traumatic ankle injuries relies primarily on physical findings and the results of radiographic evaluation. Initially, pulses and sensation are assessed. Loss of pulses or sensation suggests a fracture or dislocation and the need for a rapid reduction; when available without delay, orthopedic consultation is advisable. After immobilization to prevent further compromise and the provision of analgesia, a radiograph should be obtained immediately. If neurovascular status is adequate and the general inspection reveals no obvious abnormalities, the remainder of the physical examination should be continued as described previously.

FIGURE 37.10. Evaluation and diagnosis of traumatic ankle injuries.

Next, the area should be examined for open wounds. If open wounds are present, the physician should fashion a sterile saline dressing and immobilize the extremity before obtaining a radiograph. In addition, the administration of intravenous antibiotic therapy and tetanus prophylaxis should be considered.

If radiographic studies indicate a fracture or dislocation, the specific injury should be treated (see Chapter 115). Analgesia should be administered as needed.

If no fracture is evident on radiography but tenderness is elicited over a physis, the diagnosis of a Salter type I injury is made and appropriate immobilization performed. A negative radiograph in the absence of bony tenderness suggests the diagnosis of contusion or ligamentous injury. The diagnosis of a grade II or III sprain is rendered in the patient with joint instability. If the ankle is stable but pain is elicited with ligamentous stress or palpation, a grade I sprain is diagnosed.

TREATMENT

Fractures

Fracture reduction is usually accomplished by reversing the mechanism of injury. Closed reduction and a short leg cast are usually adequate for Salter-Harris type I and II fractures of the distal tibia and fibula. Often, younger children require placement of a long leg cast, even for minor fractures, because of their ability to accidentally (or intentionally) slip out of short leg casts.

One recent study recommends the use of a tubular bandage (Tubigrip, Seton Health Care PLC) and crutches—in lieu of a short leg plaster cast—in patients with clinically suspected but nonradiographically evident Salter-Harris type I fractures of the distal fibula. The group treated with the tubular bandage had a shorter time to recovery; however, further study is required to determine whether this method is adequate.

Some displacement can be accepted in younger patients because of their ability to remodel. Salter type III and IV injuries involve the articular surface and are therefore less stable. They require anatomic realignment, often by open reduction. A long leg cast is commonly applied in any rotational injury.

Sprains

A common approach to the treatment of ankle sprains is described by the mnemonic *RICE* (rest, ice, compression, elevation). This treatment approach should be initiated within 36 hours of the injury.

- *Rest* The patient is allowed to ambulate or exercise only if the activity causes no pain or swelling during the activity or within 24 hours. Otherwise, crutches and light weight bearing are recommended until ambulation without pain is possible.
- *Ice* The patient should apply ice directly to the ankle for 20 minutes every 2 hours, if possible, for the first 48 hours after injury.
- *Compression* The goal of compression is to keep (and/or push) fluid out of the area of the ankle joint. This can be accomplished using an elastic bandage starting at the foot and wrapping proximally toward the ankle. For additional compression, any bulky padding can be applied to the malleoli and then secured with an elastic wrap.
- *Elevation* To help decrease or prevent swelling, the patient should elevate the ankle as often as possible.

Splinting

If swelling or pain is severe, a stirrup and/or posterior splint should be applied to the ankle. Air splints can also be used; they allow dorsiflexion and plantar flexion while maintaining medial and lateral stability.

Rehabilitation

Early rehabilitation shortens the period of disability considerably. Plantar flexion and dorsiflexion exercises are initiated as soon as possible, followed by toe raises and inversion/eversion exercises.

Orthopedic Referral

Absolute indications for orthopedic referral include 1) obvious deformity with growth plate involvement; 2) neurovascular compromise; 3) suspected syndesmotic injury; 4) a grade III sprain; and 5) locking of the ankle.

Suggested Readings

Chande VT. Decision rules for roentgenography of children with acute ankle injuries. Arch Pediatr Adolesc Med 1995;149:255.

Devalentine SJ. Epiphyseal injuries to the foot and ankle. Clin Podiatr Med Surg 1987;4:279.

Dias LS, Glegerich CR. Fractures of the distal tibial epiphysis in adolescence. J Bone Joint Surg 1983;65:438.

Gleeson AP, Stuart MJ, Wilson B, et al. Ultrasound assessment and conservative management of inversion injuries of the ankle in children. J Bone Joint Surg 1996;78B:483.

Gregg JR, Das M. Foot and ankle problems in the preadolescent athlete. Clin Sports Med 1982;1:131.

Gross RH. Foot pain in children. Pediatr Clin North Am 1986;33:1395.

Gross RH. Ankle fractures in children. Bull NY Acad Med 1987;63:739.

Hergenroeder AC. Diagnosis and treatment of ankle sprains. Am J Dis Child 1990;144:809.

Hergenroeder AC, Garrick JG. Prophylactic ankle bracing. Pediatr Clin North Am 1990;37:1175.

Lassiter TE, Malone TR, Garrett WE. Injury to the lateral ligaments of the ankle. Orthop Clin North Am 1989;20:629.

Lucchesi GM, Jackson RE, Peacock WF, et al. Sensitivity of the Ottawa rules. Ann Emerg Med 1995;26:1.

Stefanich R, Lozman J. The juvenile fracture of Tillaux. Clin Orthop Relat Res 1986;210:219.

Stiell IG, Greenberg GH, McKnight RD, et al. Decision rules for the use of radiography in acute ankle injuries: refinement and prospective validation. JAMA 1993;269:1127.

Trott AW. Fractures of the foot in children. Orthop Clin North Am 1976;7:677.

Volger HW. Unusual juvenile ankle fracture: explanation and surgical repair. J Foot Surg 1990;29:516.

CHAPTER 38

Injury—Head

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PEDIATRIC HEAD TRAUMA

Head injuries in children are common, accounting for 600,000 emergency department (ED) visits annually. Although most of these injuries are minor, head trauma causes significant pediatric morbidity and mortality. Trauma is the leading cause of deaths in children older than 1 year of age, and central nervous system (CNS) trauma is the most common cause of death among injured children. In 1986, approximately 7,000 children died from head injuries, and it is estimated that each year an additional 29,000 children under 19 years of age suffer permanent disability.

The most common mechanisms of injury for the entire pediatric group are falls (37%), motor vehicle (18%) and pedestrian (17%) accidents, and bicycle injuries (10%); most fatal injuries occur because of motor vehicle–related injuries. The mechanism of pediatric head injury varies with age; younger children are more likely to fall or be abused, whereas older children often are injured in sporting or motor vehicle accidents (in addition to falls).

Many of the serious neurologic complications of head injury are evident soon after the traumatic event; however, some life-threatening injuries can appear initially as trivial head trauma. To manage head injuries best, the physician must approach the child in a systematic manner to address all injuries (because global resuscitation is the first priority of cerebral resuscitation), identify and treat any neurologic complications, and prevent ongoing cerebral insult.

PATHOPHYSIOLOGY

Neurologic injury after head trauma is related to the unique physiology and pathophysiology of the brain and the intracranial environment. The brain is a semisolid structure bathed in cerebrospinal fluid (CSF) and covered by the fine inner pia-arachnoid membrane and the outer thick fibrous layer of dura, all of which are encased in the skull, which is covered by the five-layer structure of the scalp. After infancy (when the skull sutures fuse), the cranial vault becomes a stiff and poorly compliant housing for the brain. Because the intracranial volume is relatively fixed, any change in the volume of one of the intracranial components (blood, brain, and CSF) must occur at the expense of the others; if the other components do not decrease proportionally, intracranial pressure (ICP) will increase.

Brain injury occurs in two phases: primary and secondary. The primary injury is the mechanical damage sustained at the time of trauma and can be caused by direct impact of the brain against the internal calvarial structures, by bone or foreign bodies projected into the brain, and by shear forces delivered to the white matter tracts. Secondary brain injury is further neuronal damage sustained after the traumatic event to cells not initially injured. This results from numerous causes, including hypoxia, hypoperfusion, and metabolic derangements, and may result from sequelae of the primary injury (e.g., cerebral edema, expanding intracranial mass) or be caused by extracranial injuries (e.g., hypotension from excessive blood loss, hypoxia from pulmonary contusion). The clinician's goal is to identify and treat any complications of primary brain injury to limit further neuronal damage by secondary brain injury.

One of the most common causes of secondary brain injury is cerebral ischemia resulting from diminished cerebral blood flow. Cerebral perfusion pressure (CPP) is related to the difference between the mean arterial pressure (MAP) of blood flowing to the brain and the ICP. In the healthy child, blood flow to the brain is maintained at a constant rate over a wide range of systemic blood pressures by means of autoregulatory changes in the cerebrovascular resistance so that the brain does not suffer ischemia or excessive blood flow during periods of relative hypotension or hypertension, respectively. With severe injuries, this autoregulatory control may be lost and the cerebral blood flow can become directly dependent on the CPP; with low MAP or increased ICP, inadequate blood flow and cerebral ischemia results. In addition to potential for causing decreased cerebral blood flow and ischemia, increased ICP, if left unchecked, can lead to brain herniation and compression. This may be caused by a number of posttraumatic conditions, including cerebral edema and expanding intracranial mass.

Clinical symptoms of increased ICP or herniation include headache, vomiting, irritability, lethargy, visual abnormalities (double vision, depressed acuity), and gait abnormalities or weakness. Signs include depressed level of consciousness, abnormal vital signs (bradycardia, hypertension, respiratory irregularity), cranial nerve palsies, hemiparesis, and

decerebrate posturing. The classic findings in transtentorial herniation are headache, decreasing level of consciousness followed by ipsilateral pupillary dilation (third cranial nerve [CN] palsy), and contralateral hemiparesis or posturing. If the process continues unchecked, dilation of the opposite pupil, alteration in respirations, and ultimately, bradycardia and arrest ensue. For a more detailed description of the anatomy, pathophysiology, and treatment of specific head injuries, see Chapter 105.

DIFFERENTIAL DIAGNOSIS

Head trauma may cause injuries of the scalp, skull, and intracranial contents. Although each is discussed here separately, the clinician must remember that these injuries may occur alone or in combination, and all potential injuries must be kept in mind when dealing with each.

Scalp

The scalp consists of five layers of soft tissue that cover the skull; contusions and lacerations of this structure are common results of head trauma. The outermost layers of the scalp are skin and the subcutaneous tissue; edema and hemorrhage here may produce a mobile swelling. The third layer, the galea aponeurotica, is a strong membranous sheet that connects the frontal and occipital bellies of the occipitofrontalis muscle. The remaining two layers, deep to the galea, are the loose areolar tissue and pericranium. Subgaleal hematomas may result from more forceful blows if vessels in the fourth layer bleed and dissect the galea from the periosteum, or they may be signs of an underlying skull fracture. In subperiosteal hematomas, or cephalohematomas, the swelling is localized to the underlying cranial bone and most often occurs with birth trauma. Scalp lacerations may occur with or without underlying contusions or fractures and often require suturing.

Skull

Linear, diastatic, depressed, compound, and basilar skull fractures may result from head trauma. Linear fractures account for 75 to 90% of skull fractures in children and often manifest with localized swelling and tenderness. The significance of linear fractures is debated, but their potential importance is twofold. Their presence reflects the degree of traumatic impact; because the force required to fracture a child's skull is significant, risk of an intracranial injury (ICI) is significantly more likely (as much as 10 to 20 times) with a fracture. The second feature important in linear skull fractures derives from their location; those that cross the path of a major vascular structure (middle meningeal artery, large dural sinuses) signal potential for intracranial hemorrhage from these vessels.

Diastatic fractures are traumatic separations of cranial bones at one or more suture sites. A depressed skull fracture is present when the inner table of the skull is displaced by more than the thickness of the entire bone. These may be palpable and are diagnosed with tangential skull radiographs or computed tomography (CT). Compound fractures are those that communicate with lacerations.

Basilar skull fractures often are difficult to detect on routine radiographs or CT scans; however, their location produces clinical signs that lead to the diagnosis. Fractures of the petrous portion of the temporal bone may cause hemotympanum, hemorrhagic or CSF otorrhea, or Battle's sign (bleeding into mastoid air cells with postauricular swelling and ecchymosis). Fracture of the anterior skull base may cause a dural laceration with subsequent drainage of CSF into paranasal sinuses and rhinorrhea. Anterior venous sinus drainage may cause blood leakage into the periorbital tissues (“raccoon's eyes”). Given the location of basilar skull fractures, associated CN palsies may occur. CT scan is indicated to try to localize the area of fracture and to identify any associated intracranial pathology (which occurs in approximately 20% of children with basilar skull fractures who have a Glasgow Coma Scale [GCS] score of 15 and a nonfocal neurologic examination).

Intracranial Injury

Insults to intracranial contents include functional derangements without demonstrable lesions on CT scan (concussion, posttraumatic seizures), hemorrhage (cerebral contusion, epidural hematoma, subdural hematoma, subarachnoid hemorrhage, and intracerebral hemorrhage), and acute brain swelling. Penetrating brain injuries rarely occur in children.

Concussion

Concussion is the most minor brain injury and is characterized by posttraumatic alteration in mental status that may or may not involve loss of consciousness. No consistent associated pathologic lesion in the brain has been identified. The child may have a depressed level of consciousness, pallor, vomiting, amnesia, and confusion; however, the clinical picture usually normalizes within several hours without specific therapy.

Posttraumatic Seizures

Posttraumatic seizures can be divided temporally into immediate, early, and late, and they occur in 5 to 10% of children hospitalized for head trauma.

Immediate seizures occur within seconds of the trauma and probably represent traumatic depolarization of the cortex. They usually are generalized and rarely recur.

Early seizures occur within 1 week of the trauma (most occur within 24 hours) and often are the result of focal injuries (contusion, laceration, ischemia, edema). Skull fractures, intracranial hemorrhage, and focal signs all are associated with increased risk of early posttraumatic seizures; therefore, an early seizure should prompt investigation of these

possibilities.

Late seizures occur more than 1 week after the traumatic event and may be attributed to scarring associated with local vascular compromise, distortion, and mechanical irritation of the brain. These seizures are more likely to occur in children with severe head injuries, dural lacerations, and intracranial hemorrhages. A substantial number of patients will have subsequent seizures.

Cerebral Contusion

Cerebral contusion is a bruising or crushing of brain and often results from blunt head trauma. The site of contusion may be a “coup” lesion, with the injured cerebral cortex directly beneath the site of impact (with or without skull fracture), or a “contrecoup” lesion, with damage opposite the site of impact; the contusion is demonstrable by CT. Children with cerebral contusion may have had loss of consciousness (not imperative), may show a depressed level of consciousness or symptoms of vomiting or headache, and may have focal neurologic signs or seizures.

Epidural Hematoma

Epidural hematoma (EDH) is a collection of blood between the skull and dura. An overlying fracture is present in 60 to 80% of cases, and depending on the location and vascular structure involved, the hemorrhage may be of arterial or venous origin; injury to the middle meningeal artery often is responsible for temporal EDH. The classic pattern of a lucid interval between initial loss of consciousness and subsequent neurologic deterioration occurs in only a minority of children with EDH. Furthermore, patients occasionally may develop EDH after relatively minor trauma with no history of loss of consciousness. Although many children present with significant lethargy, focal neurologic signs, or a clinical pattern consistent with temporal lobe herniation as the hematoma expands, some children with EDH are alert, have a nonfocal neurologic examination, and have only symptoms of headache or persistent vomiting; nevertheless, rapid deterioration can ensue.

Subdural Hematomas

Subdural hematomas (SDH) occur as a result of bleeding between the dura and the arachnoid membranes covering the brain parenchyma. They may result from direct trauma or from shaking injuries and result from tearing of the cortical bridging veins or from bleeding from the cortex itself. Subdural hematomas may be bilateral, and often, there is an associated underlying brain injury. Skull fractures occur in only a minority of cases. Children with SDH often have seizures, may present with evidence of acutely elevated ICP, or may have more nonspecific signs of vomiting, irritability, or low-grade fever. Physical examination often reveals an irritable or lethargic infant or child with a bulging fontanel who may or may not have neurologic abnormalities. CT scan commonly demonstrates crescent-shaped subdural collections.

Intracerebral Hematoma

Posttraumatic intracerebral hematomas are unusual in children. Blood within the parenchyma usually is the result of severe focal injury or penetrating trauma, usually manifests with severe neurologic compromise, and often portends a poor prognosis.

Subarachnoid Hemorrhage

Subarachnoid hemorrhage may occur after head trauma (including shaking injuries in infants) and may cause headache, neck stiffness, and lethargy in the child.

Acute Brain Swelling

The most common diagnosis in children with severe head injury (GCS score of less than 8) is diffuse cerebral swelling; in one study, this occurred in 50% of cases, whereas intracranial hemorrhages were less common at 20% of cases. These children have a depressed level of consciousness and may have focal neurologic signs or symptoms of herniation.

Penetrating Injuries

Penetrating head injuries are uncommon in children and may be caused by bullets, teeth (e.g., dog bites), or other objects (e.g., dart, pencil, pellet) penetrating the skull. These injuries have obvious potential for extensive damage to the brain and intracranial vessels.

EVALUATION AND DECISION

The clinical spectrum of head injury in children varies from a small contusion of the scalp with no neurologic sequelae to severe intracranial trauma that causes death. The general approach is essentially the same as with any child who presents with trauma, paying particular attention to potential CNS damage. Following the ABCs (airway, breathing, and circulation) of resuscitation, the physician must systematically evaluate and stabilize the child with head trauma. The goals of management are to identify complications of the head trauma and to prevent secondary brain injury. Because some complications of head trauma may not manifest immediately, the assessment period includes the initial evaluation in the ED and a more extended observation period, either in the hospital or as an outpatient, as clinically indicated. Specific therapy will vary, based on specific diagnosis in each case, and may include supportive care and possible neurosurgical intervention. Although complications are more common in children with severe head injury, they also occur in children with apparently minor head trauma; thus, all patients merit some degree of scrutiny.

The immediate management of the child varies with the degree of compromise. A brief initial assessment is done to

determine immediate stability. In the older child, verbal response to a question often establishes the adequacy of the airway, ventilation, and cognitive function. If the child is unconscious or has unstable vital signs, immediate resuscitation is initiated to ensure a patent airway (with cervical spine immobilization), effective ventilation, and adequate tissue perfusion (see Chapter 1); efforts to decrease possible increased ICP may be indicated depending on the degree of neurologic compromise (see Chapter 105). The child with airway and hemodynamic stability and with only mild to moderate depression of mental status can undergo a more timely evaluation to identify subtle or occult abnormalities.

Clinical Assessment

History

The history should be obtained from the patient (if age and level of consciousness permit) and from any witnesses to determine the nature and severity of the impact as well as the prehospital course. Specifics of the traumatic event should include how, when, and where the trauma occurred as well as details such as height of a fall, type of impact surface, and type and velocity of striking objects. Occurrence of loss of consciousness should be determined as well as duration. If the event was not witnessed and the patient is amnestic, the clinician should assume that loss of consciousness occurred. Occurrence of seizure activity (including details of time of onset posttrauma, duration, and focality) as well as the child's level of alertness since the injury should be noted; in addition, the presence of vomiting, irritability, ataxia, and abnormal behavior are all signs of possible brain injury. Vomiting after a head injury is common; however, more than several hours of unremitting vomiting may signal intracranial abnormalities. If the child is verbal, he or she should be questioned about presence of headache or neck pain, amnesia, weakness, visual disturbances, or paresthesias. In many cases, elicited symptoms may be the only evidence of underlying CNS injury. In infants, symptoms of ICI may be subtle or absent; therefore, the clinician should pay particular attention to any alteration in behavior in this age group. Progression or resolution of any symptoms, neurologic signs, and level of consciousness since the traumatic episode must be defined clearly. One should also inquire about the patient's medical history and factors predisposing to head trauma (e.g., seizure disorder, gait disturbance, bleeding diathesis, alcohol abuse, illicit drug use). When there are discrepancies in the history, when the history does not fit the physical findings, or when there is a skull fracture or ICI in a young child without a history of significant trauma, one should suspect nonaccidental injury.

Physical Examination

After a primary survey with appropriate resuscitation, a thorough physical examination should be performed with special emphasis on the vital signs, head and neck, and the neurologic examination. Bradycardia may be a sign of increased ICP, even in the alert or minimally drowsy child; it is of particular concern when associated with hypertension, abnormal breathing pattern, depressed level of consciousness, or neurologic abnormality. Bradycardia also may be seen with spinal cord injuries caused by unopposed parasympathetic tone; in those cases, it often is associated with hypotension, flaccidity, a sensory level, and absent deep tendon reflexes. Tachycardia may reflect hypovolemia (especially if associated with hypotension), hypoxia, or anxiety. Isolated head injuries rarely cause hypovolemia (except in infants with large subgaleal or intracranial hematomas); therefore, hypotension should alert one to an extracranial source of hemorrhage.

The head should be inspected and palpated carefully for scalp swelling, lacerations, irregularities of the underlying bony structure, and fontanel fullness (in infants). Signs of basilar skull fracture (periorbital or postauricular hemorrhage in the absence of direct trauma, hemotympanum, CSF otorrhea, or rhinorrhea) and retinal abnormalities (hemorrhage or papilledema) should be noted. All children with depressed mental status, a mechanism of injury suggesting a significant force, or neck pain should have cervical spine immobilization at least until its integrity has been confirmed radiographically; one should note cervical abrasions, deformity, or tenderness—findings that may indicate underlying cervical spine injuries.

Neurologic examination encompasses assessment of the child's mental status as well as cranial nerve, motor, sensory, cerebellar, and reflex functions. Serial examinations are important in the child with head trauma to document improvement or deterioration. The GCS is a convenient way to quantify level of consciousness and monitor neurologic progression. The GCS scores patient performance in three areas: eye opening, verbal ability, and motor ability. It also assesses level of alertness, mentation, and major CNS pathways (Table 38.1); an individual's score may range from a low of 3 to a high of 15. The scale has been modified for more age-appropriate behaviors in infants (Table 38.2). Although ICIs are more common in a child with a low GCS score, even a child with a score of 13 to 15 may harbor life-threatening complications of head trauma (e.g., EDH), especially if neurologic abnormalities are present. Further evaluation of mental status includes assessing orientation and memory. Subtle signs (irritability and high-pitched cry) may indicate underlying abnormalities in infants.

Activity	Best Response	Score
Eye Opening	Spontaneous	4
	To verbal stimuli	3
	To pain	2
	None	1
Verbal	Oriented	5
	Confused	4
	Inappropriate words	3
	Nonspecific sounds	2
	None	1
Motor	Normal spontaneous movements	6
	Localizes pain	5
	Withdraws to pain	4
	Abnormal flexion (decorticate rigidity)	3
	Abnormal extension (decerebrate rigidity)	2
	None	1

Table 38.1. Glasgow Coma Scale Score

Activity	Best Response	Score
Eye Opening	Spontaneous	4
	To speech	3
	To pain	2
	None	1
Verbal	Coos, babbles	5
	Irritable, cries	4
	Cries to pain	3
	Moans to pain	2
	None	1
Motor	Normal spontaneous movements	6
	Withdraws to touch	5
	Withdraws to pain	4
	Abnormal flexion (decorticate rigidity)	3
	Abnormal extension (decerebrate rigidity)	2
	None	1

Table 38.2. Modified Coma Scale for Infants

CN function is assessed by checking for facial symmetry, corneal reflexes, presence of a gag reflex, full extraocular movements, pupillary size, and pupillary reactivity. In the comatose patient or in the child with possible neck injury who is uncooperative, lateral gaze may be tested by caloric stimulation of the vestibular apparatus (but not the “doll's eye” maneuver) once tympanic membrane integrity has been established.

Examination of the motor system to evaluate both CNS and spinal cord function varies with age and level of consciousness. The alert patient should have individual muscle groups tested and gait evaluated. The child with a depressed level of consciousness may have motor responses elicited by noxious stimuli (e.g., sternal rub, nailbed pressure). Deep tendon reflexes and Babinski's sign also should be evaluated. Obviously, a complete physical examination with attention to possible thoracic, abdominal, pelvic, and extremity injuries should be performed.

Radiographic Investigation

Complications of head trauma may be identified with radiographic studies, which include plain radiographs of the skull and cervical spine and CT scans of the head; specific studies are indicated based on the child's history and physical findings. Although magnetic resonance imaging (MRI) is an additional imaging modality for the cranial contents, limited availability and prolonged study time limit its use for evaluation of acute trauma at this time. All children with significant head trauma should be evaluated for associated cervical spine injuries; this evaluation will be clinical, with or without radiographic studies, based on the specific circumstances (see Chapter 106).

CT provides excellent images of the intracranial contents and therefore is the diagnostic modality of choice when intracranial pathology is suspected. Indications include history of penetrating trauma, loss of consciousness (particularly if more than brief) or seizure, altered level of consciousness, focal neurologic abnormalities, presence of full fontanel, skull fracture, and persistent vomiting or progressive headache. Many authors now recommend performing a CT scan in patients with any loss of consciousness because the incidence of ICIs in alert children with nonfocal neurologic examinations who lost consciousness and underwent CT is approximately 3 to 6%; however, only a small number of these children require neurosurgical intervention. Because children younger than 24 months of age are difficult to assess, cannot report symptoms, have impaired skull integrity, are at risk for abuse, and often are minimally symptomatic with ICI, the clinician should favor a particularly low threshold for obtaining CT scans for patients in this age group who have head trauma and any symptoms. The younger the age, the more difficult to assess and the higher the incidence of ICIs (and particularly of occult or asymptomatic intracranial injuries); although this is a continuum, the clinician should have a very low threshold for obtaining CT scans for infants younger than 2 to 3 months of age who have trauma, unless trivial, even without symptoms. In all cases, the patient's condition must be stabilized before transfer to the neuroradiologic suite; the patient should be monitored appropriately and accompanied by a health care professional with medication and equipment necessary for resuscitation.

Over the years, one of the most controversial issues in the management of head trauma has been indications for skull radiographs. Inherently, skull radiographs are of limited value in that they reveal primarily bony abnormalities, giving little or no information about ICIs. The presence of a skull fracture, however, may imply significant impact with a higher likelihood for ICI, particularly if the fracture crosses the path of a major vessel. Although clearly any child for whom there is significant concern for ICI should undergo CT scan, there may still be a small role for skull radiographs in certain select circumstances when immediate CT is not warranted, yet significant chance of fracture exists to justify the test. Data suggest that a substantial number of children younger than 12 months of age who have ICIs are minimally symptomatic; however, most of the asymptomatic children will have an associated skull fracture. Aside from being less costly, one advantage of obtaining skull radiographs is that children do not require sedation, which is necessary for many young children who undergo CT (especially those 4 to 24 months of age). One reasonable approach to imaging is to use skull radiographs as a screening tool in children with scalp findings (because most fractures are associated with swelling or scalp abnormalities) or with a history of significant trauma (e.g., a fall of more than 3 feet) who do not have other symptoms of ICI. Children with fractures identified on skull radiographs should then undergo CT scans because they are at increased risk for associated ICI. Other indications for skull radiographs include a suspicion of depressed fracture or penetrating trauma and the possibility of a foreign body.

Approach

The goals of management are to define specific anatomic lesions (e.g., skull fracture, ICI) and to prevent secondary brain injury. Pediatricians and emergency physicians will be the initial clinicians to evaluate and manage most children with head trauma. Neurosurgical consultation should be considered for all children with penetrating trauma, prolonged loss of

consciousness, abnormal mental status or neurologic examination, skull fractures, and intracranial complications. The urgency of neurosurgical involvement varies with the acuity of the patient's clinical condition.

One approach (Fig. 38.1) to diagnosing complications of head trauma involves determining whether a penetrating injury has occurred. If so, brain or vascular injury is likely, and emergent CT scanning and neurosurgical consultation are mandated in addition to stabilization.

FIGURE 38.1. Approach to the child with head trauma. *GCS*, Glasgow Coma Scale; *ICI,* intracranial injury; *CT,* computed tomography.

If the head injury has resulted from blunt trauma, it must be determined whether an ICI is present. Suggestive history includes loss of consciousness (especially if more than brief); prolonged, focal, or early seizure; and definite bony abnormality or underlying condition that predisposes to ICI (e.g., coagulopathy). Physical findings indicative of possible intracranial abnormalities include a GCS score of less than 15, focal neurologic abnormalities, a definite palpable bony depression, and signs of a basilar skull fracture. If any of these findings is present, then, in addition to supportive therapy, CT scan and possible neurosurgical consultation are indicated. Abnormalities on the CT scan might include intracranial hemorrhage, diffuse cerebral swelling, or skull fracture; if the CT scan is normal, concussion or extracranial injury has likely occurred.

If these findings are not present, the child will be alert with a nonfocal neurologic examination. The drowsy child who quickly becomes alert or the child with a history of momentary loss of consciousness who is alert with a nonfocal examination may show no evidence of intracranial complications; however, the child may have had more than a trivial head injury. If these patients do not undergo a CT scan, they should be observed in the ED for at least 4 to 6 hours after the injury for signs and symptoms of complications, including neurologic abnormalities, mental status depression, persistent vomiting, and progressively severe headache. A CT scan should be obtained if these signs or symptoms develop. As previously stated, discrete abnormalities may be identified on CT scan; however, if the scan is normal, the child has experienced a concussion or extracranial injury.

Children likely to display abnormalities on skull radiographs but in whom immediate imaging of intracranial contents is not indicated include those who are awake and alert but in whom there is a history of a foreign body, possible penetration of the bony calvarium, or question of a palpable bony irregularity. Another relative indication for skull radiographs is a significant subgaleal hematoma, especially if it overlies the middle meningeal groove. One should also consider obtaining skull radiographs in infants under the age of 1 year with a history of a fall of more than 3 feet onto a hard surface or with a palpable hematoma (if they have not undergone CT) because an infant's skull has limited ability to withstand trauma.

Skull radiographs may show an extracranial or intracranial foreign body, may reveal a fracture, or may be normal. Obviously, immediate CT scan (with neurosurgical consultation) is mandated for patients with an intracranial foreign body because neuronal or vascular damage is likely. CT is also indicated if skull radiographs demonstrate a skull fracture because this significantly increases the likelihood of an associated ICI. The child with normal skull radiographs and examination who is alert has likely experienced an extracranial injury or a concussion.

The remaining children who sustained impact of minimal force, had no loss of consciousness, and are alert and asymptomatic with normal examinations likely have only minor head trauma with or without extracranial injuries, including contusions and lacerations. Home observation is appropriate management for most of these patients. Rarely, intracranial complications develop in these children, causing symptoms hours after the traumatic event; therefore, caretakers should be given a printed list of signs and symptoms indicative of increased ICP with instructions to check the child at regular intervals and to return to the ED if symptoms occur. The caretakers must be reliable and able to return with the child if necessary, and there must be no suspicion of abuse or neglect; otherwise, admission for observation in the hospital should be considered (Table 38.3).

Traumatic force not life-threatening	No intracranial abnormalities on CT (if obtained)
Glasgow Coma Scale score of 15	Reliable caretakers who are able to return if necessary
Nonfocal neurologic examination	No suspicion of abuse or neglect
No significant symptoms	
No history of prolonged loss of consciousness (or normal CT if it did occur)	

CT, computed tomography.

Table 38.3. Criteria for Discharge with Home Observation

Suggested Readings

Bruce DA, Raphaely RC, Goldberg AI, et al. Pathophysiology, treatment and outcome following severe head injury in children. Child's Brain 1979;5:174–191.

Davis RL, Mullen N, Makela M, et al. Cranial computed tomography scans in children after minimal head injury with loss of consciousness. Ann Emerg Med 1994;24:640–644.

Dietrich AM, Bowman MJ, Ginn-Pease MD, et al. Pediatric head injuries: can clinical factors reliably predict an abnormality on computed tomography? Ann Emerg Med 1993;22:1535–1540.

Greenes DS, Schutzman SA. Infants with isolated skull fracture: what are their clinical characteristics, and do they require hospitalization? Ann Emerg Med 1997;30:253–259.

Greenes DS, Schutzman SA. Occult intracranial injury in infants. Ann Emerg Med 1998;32, in press.

Gruskin K, Schutzman SA. Head trauma in children less than two years of age: are there predictors for complications? Arch Pediatr Adolesc Med, in press.

Hahn YS, McLone DG. Risk factors in the outcome of children with minor head injury. Pediatr Neurosurg 1993;19:135–142.

James HE. Neurologic evaluation and support in the child with an acute brain insult. Pediatr Ann 1986;15:16–22.

Kadish HA, Schunk JE. Pediatric basilar skull fractures: do children with normal neurologic findings and no intracranial injury require hospitalization? Ann Emerg Med 1995;26:37–41.

Krauss JF, Rock A, Hemyari P. Brain injuries among infants, children, adolescents, and young adults. Am J Dis Child 1990;144:684–691.

Lloyd DA, Carty H, Patterson M, et al. Predictive value of skull radiography for intracranial injury in children with blunt head injury. Lancet 1997;349:139–143.

Masters SJ, McClean PM, Argarese JS, et al. Skull x-ray examinations after head trauma. N Engl J Med 1987;316:84–91.

Quayle KS, Jaffe DM, Kuppermann N, et al. Diagnostic testing for acute head injury in children: when are head computed tomography and skull radiographs indicated? Pediatrics 1997;99:1–8.

Rivara F, Tanagucki D, Parish RA, et al. Poor prediction of positive computed tomographic scans by clinical criteria in symptomatic pediatric head trauma. Pediatrics 1987;80:579–584.

Schunk JE, Rodgerson JD, Woodward GA. The utility of head computed tomographic scanning in pediatric patients with normal neurologic examination in the emergency department. Pediatr Emerg Care 1996;12:160–165.

Shackford SR, Wald SL, Ross SE. The clinical utility of computed tomographic scanning and neurologic examination in the management of patients with minor head injuries. J Trauma 1992;33:385–394.

Tecklenburg FW, Wright MS. Minor head trauma in the pediatric patient. Pediatr Emerg Care 1991;7:40–47.

Tepas JJ, DiScala C, Ramenofsky ML, Barlow B. Mortality and head injury: the pediatric perspective. J Pediatr Surg 1990;25:92–96.

Zuckerman GB, Conway EE. Accidental head injury. Pediatr Ann 1997;26:621–632.

CHAPTER 39

Injury—Knee

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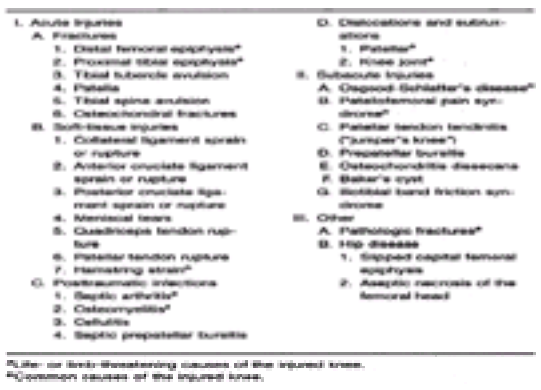
Acute pain or injury to the knee is a common complaint in the emergency department (ED). Many injuries are minor and require only limited therapy; others, however, require consultation with an orthopedist, either in the ED or for subsequent evaluation after pain and inflammation subside. The emergency physician can provide appropriate therapy or determine the need for consultation, based on a comprehensive history, physical examination, and an appropriate radiographic evaluation.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of acute and chronic knee injuries is summarized in Table 39.1. The pertinent anatomy is illustrated in Figure 39.1 and Figure 39.2.



I. Acute Injuries
- A. Fractures
  1. Distal femoral epiphysis*
  2. Proximal tibial epiphysis*
  3. Tibial tubercle avulsion
  4. Patella
  5. Tibial spine avulsion
  6. Osteochondral fractures
- B. Soft-tissue injuries
  1. Collateral ligament sprain or rupture
  2. Anterior cruciate ligament sprain or rupture
  3. Posterior cruciate ligament sprain or rupture
  4. Meniscal tears
  5. Quadriceps tendon rupture
  6. Patellar tendon rupture
  7. Hamstring strain*
- C. Posttraumatic infections
  1. Septic arthritis*
  2. Osteomyelitis*
  3. Cellulitis
  4. Septic prepatellar bursitis
- D. Dislocations and subluxations
  1. Patella*
  2. Knee joint*

II. Subacute Injuries
- A. Osgood-Schlatter's disease*
- B. Patellofemoral pain syndrome*
- C. Patellar tendon tendinitis ("jumper's knee")
- D. Prepatellar bursitis
- E. Osteochondritis dissecans
- F. Baker's cyst
- G. Iliotibial band friction syndrome

III. Other
- A. Pathologic fractures*
- B. Hip disease
  1. Slipped capital femoral epiphysis
  2. Aseptic necrosis of the femoral head

*Life- or limb-threatening causes of the injured knee.
†Common causes of the injured knee.

Table 39.1. Differential Diagnosis of the Injured Knee

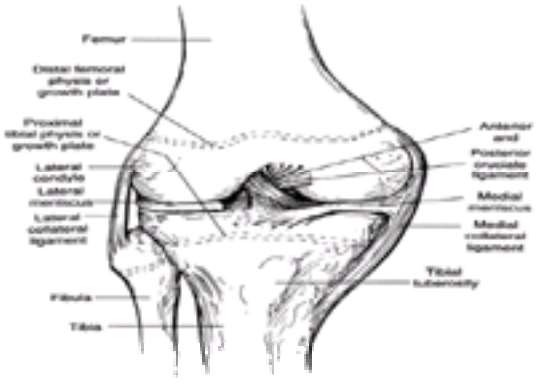


FIGURE 39.1. Anatomy of the knee—anterior view (patella removed).

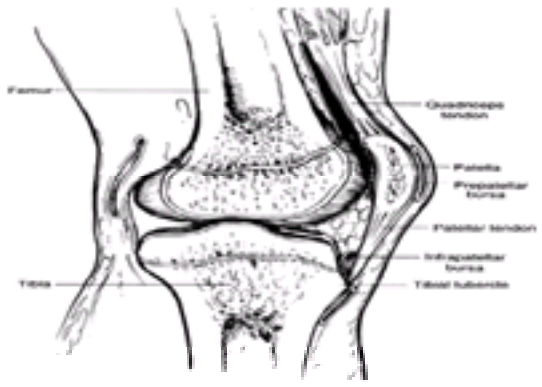


FIGURE 39.2. Anatomy of the knee—sagittal section.

Acute Injuries

Fractures

When first described in the 1800s, a separation fracture of the distal femoral epiphysis (wagon-wheel injury) usually occurred when a child's leg was caught in a wagon wheel, the child's thigh or torso stopped against the wagon, and the knee hyperextended. Now, this injury occurs most commonly during contact sports or in car accidents. It is classified by the Salter-Harris pattern (Chapter 115) and by the displacement of the epiphysis (usually lateral or medial). The injury usually follows significant direct (e.g., being run over by a car's wheels) or indirect (e.g., being hit during contact sports from the lateral side with the foot fixed by cleats) trauma. The patient has severe pain, refuses to bear weight, and experiences joint and soft-tissue swelling and possibly deformity. Distal neurovascular status should be assessed because compromise of the popliteal artery occurs in 1% of cases and peroneal nerve injury occurs in 3% of cases. Usually, radiographs are diagnostic but may be normal if the injury is a nondisplaced Salter-Harris type I fracture. Lateral and anteroposterior (AP) stress views may be necessary.

Separations of the proximal tibial epiphysis are more rare than those of the distal femoral epiphysis and also are likely to involve vascular compromise because of the proximity of the popliteal artery to the posterior aspect of the tibial epiphysis. The patient will have severe pain, limited range of motion (ROM), and commonly, a hemarthrosis. If displaced, the knee will be deformed. Distal neurovascular status should be assessed. Usually, radiographs are diagnostic but may be normal if the injury is a nondisplaced Salter-Harris type I fracture. Lateral and AP stress views may be necessary.

Acute traumatic avulsion of the tibial tubercle is caused by acute stress on the knee's extensor mechanism. The quadriceps muscle group extends the knee by way of the patella and the patellar ligament. The patellar ligament inserts on the tibial tuberosity and may avulse it during sudden acceleration (e.g., beginning a jump) or deceleration (e.g., landing after a jump). The patient will have tenderness and swelling over the tibial tubercle and is unable to extend the knee fully. A lateral radiograph is diagnostic.

Fractures of the patella are more common in adults because the child's patella is surrounded by cartilage, protecting it from direct trauma. A medial avulsion fracture suggests that the mechanism was a patellar dislocation that spontaneously reduced. The patient's knee will be swollen and tender and will resist full extension. A radiograph is diagnostic, although a bipartite patella may be confused with an acute fracture. The curved radiolucent line associated with a bipartite patella usually is in the superior lateral quadrant, a rare area for a fracture, and should not be associated with the soft-tissue swelling and effusion seen with a fracture.

Analogous to an adolescent who ruptures the anterior cruciate ligament, children sustain avulsion fractures of the tibial spine at the point where the anterior cruciate ligament inserts. The tibial spine is incompletely ossified and may avulse before the ligament ruptures. The patient will have a hemarthrosis and will be unable to bear weight. If the patient tolerates an examination, the Lachman test (Fig. 39.3) may be positive because the injury is similar mechanically to an anterior cruciate ligament tear. AP, lateral, and intercondylar or tunnel view radiographs will show the avulsed fragment. The visible ossified fragment may be small because the tibial spine is mostly radiolucent cartilage.

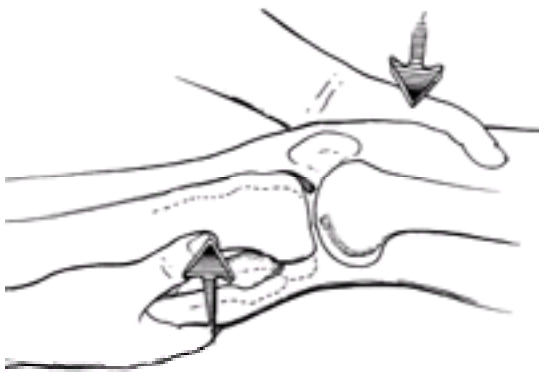


FIGURE 39.3. Testing for anterior cruciate ligament injury with the Lachman test. Flex the knee 20 to 30 degrees, support the thigh with one hand, and grasp the calf with the other hand. Move the tibia forward on the femur. Observe the tibial tubercle for movement and feel for excessive forward movement of the tibia in relation to the femur.

Osteochondral fractures are fractures of articular cartilage and underlying bone not associated with ligamentous attachments. These fractures usually involve the patella and occur during patellar dislocations or may involve the femoral condyle. Occasionally, a patient sustains a direct blow to the knee, but more commonly, the knee is injured during a twisting injury. The patient has severe pain, holds the knee flexed, and refuses to bear weight. Often, the patient has immediate swelling of the knee consistent with a hemarthrosis. Because the fragment may be in the intercondylar notch, radiographs need to include an intercondylar or tunnel view.

Dislocations

Patellar dislocation occurs as the quadriceps muscles pull along the patellar tendon to extend the knee. If the vastus medialis fibers do not keep the patella in the intercondylar groove, the patella may dislocate laterally. This often recurrent injury rarely occurs from direct force but rather happens more often during dancing or gymnastics. The patient may feel or hear a ripping or popping sensation or sound. The patient complains of intense pain and holds the knee flexed. The patella is visible more laterally than normal. The dislocation may be reduced before radiographs are taken. If the history is consistent with dislocation but the patient is no longer in pain and has a normal examination, he or she may have subluxated the patella. The patient is observed carefully for a high-riding or laterally displaced patella. Measurement is made of the quadriceps or Q angle formed by a line drawn from the anterior superior iliac spine to the midpatella and one drawn from the midpatella to the tibial tubercle. For men, the average Q angle is 14 degrees; for women it is 17 degrees. An angle above 20 degrees is abnormal. A patellar apprehension test is performed by gently attempting to move the patella laterally. If the patient becomes apprehensive or grabs the examiner's hand, this suggests that he or she has subluxated the patella. Radiographs are always obtained to rule out an associated avulsion fracture of the patella or femoral condyle.

In a child, the knee joint itself rarely dislocates; usually, the distal femoral or proximal tibial epiphysis separates first. Dislocation occurs only with trauma that involves significant force, such as a high-speed motor vehicle accident. The knee appears obviously deformed with the tibia or femoral condyles abnormally prominent in an anterior or posterior dislocation, respectively. Disruption of the popliteal artery may occur with the dislocation, and the resulting hypoperfusion may be limb-threatening. Posterior tibial and dorsalis pedis pulses and peroneal nerve function must be documented; radiographs will confirm the diagnosis.

Soft-Tissue Injuries

Medial or lateral collateral ligament injuries are rare when the epiphysis is open because the involved ligaments are stronger than the growth plate. The lateral collateral ligament inserts on the fibular head proximal to the physis, and the medial collateral ligament (MCL) inserts on the tibia distal to the physis. In older patients, the MCL may be damaged by a blow to the lateral side of the knee during contact sports or stress during a skiing accident, when the athlete “catches an edge” and falls forward with the leg rotated externally. Severe collateral ligament injury may be associated with anterior cruciate ligament (ACL) or meniscal damage. On examination, the knee may be swollen only minimally but will be tender over the involved ligament. The knee then should be tested for lateral laxity in full extension (associated with more severe injuries) and in 30 degrees of flexion (associated with less severe injuries), as shown in Figure 39.4. Orthopedic referral may be indicated if the examination reveals lateral or medial laxity.

FIGURE 39.4. Testing for collateral ligament injury. Test the knee in full extension and in 30 degrees of flexion. To test for medial collateral ligament injury, hold and apply force to the medial side of the ankle with one hand and apply pressure over the fibular head with the other hand. To test for lateral collateral ligament injury, hold and apply force to the lateral side of the ankle with one hand and apply pressure just below the medial side of the knee with the other hand. If the knee “opens up” laterally or medially more than the uninjured knee, the collateral ligament is injured.

Anterior cruciate ligament injuries occur in many scenarios but usually involve rotational forces on a fixed foot. The patient often reports the sensation of a “pop.” The joint usually swells rapidly as a result of hemarthrosis and has a marked decrease in ROM. The Lachman test (Fig. 39.3) is sensitive in detecting ACL injuries but may be falsely negative soon after the injury when the knee is swollen and painful. Examining the uninjured knee can be helpful for comparison. Arthroscopy or magnetic resonance imaging (MRI) is often needed for definitive diagnosis. ACL injuries are rare before adolescence because in a child, the ACL's insertion point, the tibial spine, is incompletely ossified. Therefore, the same force that would produce an ACL injury in an adolescent will cause an avulsion fracture of the tibial spine in a child.

Posterior cruciate ligament injuries are extremely rare and usually result from direct force on the tibial tubercle, pushing the tibia posteriorly on the femur. The posterior drawer sign will be present in most cases (Fig. 39.5).

FIGURE 39.5. Testing for posterior cruciate ligament injury with the posterior drawer test. With the patient supine and the knee flexed to 90 degrees, sit on the patient's foot to stabilize it. Attempt to force the tibia posteriorly. Posterior movement greater on the injured side than on the uninjured side is abnormal and suggests a posterior cruciate ligament injury.

The menisci are tough fibrocartilage pads that help distribute the body's weight over the femoral and tibial condyles. They can be injured when the knee is twisted during weight bearing. The patient may report a popping sensation and the feeling of the knee “giving out.” More chronically, the patient may report that the knee suddenly refuses to extend fully, “locking up,” and then suddenly “unlocking.” Joint-line tenderness is almost always present but must be differentiated from the tenderness associated with collateral ligament injuries. An effusion is commonly detected. Acutely, the injury may be difficult to diagnose because the patient has significantly reduced ROM, making the classic McMurray's sign difficult to elicit (Fig. 39.6). The Apley compression test (Fig. 39.7) requires less knee flexion and may be easier for the patient to tolerate. Radiographs generally are normal. A subluxating patella, ACL injury, or osteochondral fracture also may cause a popping sensation, and the patellofemoral pain syndrome may be associated with “giving way” of the knee.

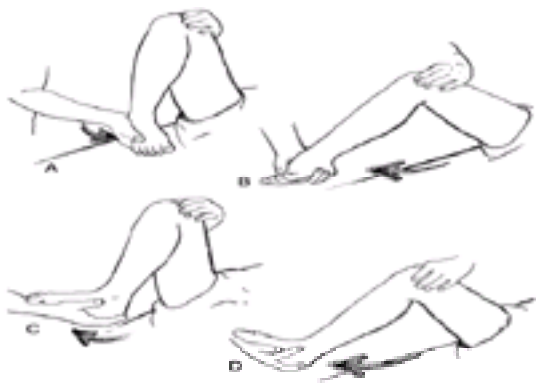


FIGURE 39.6. A–D. Testing for meniscal injury with the McMurray test. Grasp the patient's foot with one hand and place the other hand over the joint lines. Fully flex and extend the knee while alternately internally and externally rotating the tibia. The injured meniscus may be felt as a grinding or snapping sensation as the knee is manipulated.

FIGURE 39.7. Testing for meniscal injury with the Apley compression test. With the patient prone and the knee flexed to 90 degrees, apply pressure to the heel while the tibia is rotated. If this produces pain that resolves when the tibia is distracted from the femur while rotated, a meniscal injury should be suspected.

The quadriceps or patellar tendon can rupture acutely, especially in an older athlete who jumps or falls a great distance. The tendon will be tender directly over the rupture, and the patella may be positioned abnormally. A hemarthrosis may be present, and radiographs may show the abnormally positioned patella.

The three hamstring muscles (semitendinosus, semimembranosus, and the biceps femoris) flex the knee and may be strained in young athletes. The semitendinosus and semimembranosus insert along the medial popliteal space, and the biceps femoris tendon runs laterally. The patient may describe an acute pain or even a pop in the back of the thigh. Often, this injury presents subacutely with posterior thigh and popliteal knee pain when the hamstrings are strained by

repetitive use.

Posttraumatic Infection

Although not considered injuries, acute infections may present after a vague history of trauma. Physical findings of acute infection are present. The most common disorders are septic arthritis, osteomyelitis, cellulitis, and septic prepatellar bursitis.

Subacute Injuries

Many subacute knee problems manifest acutely in the ED. Osgood-Schlatter's disease of the tibial tubercle may lead to similar symptoms as a traumatic avulsion of the tubercle; however, with Osgood-Schlatter's disease, the symptoms have been noted for days or weeks. The symptoms of Osgood-Schlatter's disease are exacerbated by squatting or jumping, but they do not cause the same disability as an acute avulsion. The disease is usually seen in patients between 10 and 15 years of age. It may be caused by recurrent contractions of the patellar tendon during knee extension, traumatizing the tendon's insertion on the tibial tubercle during the child's growth spurt. The patients have localized tenderness and occasionally swelling over the tibial tubercle. The patient will refuse to extend the knee against force (e.g., perform a deep-knee bend) and have difficulty going up or down stairs, although they may have a normal gait on a level surface. To eliminate the possibility of a neoplasm, the physician should always obtain radiographs. They will either be normal or show irregularity of the tubercle.

Patellofemoral pain syndrome (PFPS) or chondromalacia patella may be caused by malalignment of the extensor mechanism of the knee. The patella transmits the force of the quadriceps muscles to the patellar tendon to extend the knee. The vastus lateralis, vastus intermedius, and rectus femoris pull the patella slightly laterally and need to be balanced perfectly by the vastus medialis to keep the patella tracking across the articular cartilage correctly. Medially squinting patellae and patella alta predispose patients to PFPS. Some of these patients have chondromalacia patella, with softening of the cartilage. The patient with PFPS has patellar pain with running and especially going down inclines or stairs. The patient also may have the sensation of the knee giving out when descending, although an actual fall usually does not occur. The patient has pain when sitting for a prolonged time with the knee flexed at 90 degrees. The pain disappears once the patient is ambulatory. This is called the movie sign. On examination, the patient may have the malalignment just mentioned, tenderness of the articular surface of the patella, and a positive patellar stress test. This test is performed with the patient in the supine position with the knee fully extended. The patient is asked to relax the quadriceps so that the physician can move the patella. With the patella pulled inferiorly, the physician should gently press down on it and ask the patient to tighten the quadriceps. (A younger patient should be asked to “push the knee into the examination table.”) This will move the patella superiorly as the physician continues to press down. A patient with PFPS will have acute pain with this maneuver; radiographs, however, are normal.

Patellar tendon tendinitis, or “jumper's knee,” occurs in patients during their growth spurt, especially those involved in jumping (knee extension) sports. The knee is tender on the inferior pole of the patella and the adjacent patellar tendon, but not on the tibial tubercle; radiographs generally are normal.

Prepatellar bursitis occurs after acute or chronic trauma to this bursa, which overlies the patella. The patient will have swelling over the anterior aspect of the knee, especially over the patella. A septic bursitis may need to be ruled out by needle aspiration.

Osteochondritis dissecans is the separation of a small portion of the femoral condyle with the overlying cartilage. The patient usually is an adolescent with a 1- to 4-week history of nonspecific knee pain. The physical examination may be normal or the femoral condyle may be tender. Because AP and lateral radiographs may not show the lesion, a tunnel or intercondylar view should be obtained.

Iliotibial band syndrome usually occurs in older runners who complain of pain over the lateral aspect of the knee. The iliotibial band moves in an anterior or posterior direction across the lateral femoral condyle as the knee flexes and extends. This repetitive movement may cause the pain. When examined, the patient is tender over the lateral femoral epicondyle, palpable 2 cm above the joint line. Radiographs are normal.

The Baker's cyst is a herniation of the synovium of the knee joint or a separate synovial cyst located in the popliteal fossa. The patient complains of popliteal pain and swelling only if the cyst enlarges. The sac can be palpated in the posterior medial aspect of the popliteal space and may be transilluminated. For the most part, radiographs will be normal or show soft-tissue swelling.

In any patient with knee pain, with or without a history of trauma, benign (e.g., osteochondroma and nonossifying fibroma) and malignant tumors (e.g., osteosarcoma or Ewing's sarcoma), the various causes of monoarticular arthritis (see Chapter 57), and hip disease that may present with knee pain (e.g., slipped capital femoral epiphysis or aseptic necrosis of the femoral head) must be considered.

EVALUATION AND DECISION

Four points are critical in the patient's history: 1) the activity and forces that led to the injury (e.g., direct or indirect force, direction of the force, and whether the foot was planted); 2) any sensations or noises (e.g., “locking,” “pops,” “snaps,” “rips,” or “tears”); 3) the initial location of the pain; and 4) the timing of any swelling.

The possibility of abuse in young children must always be considered, especially if the injury is unexplained, the history is implausible, or the seeking of medical care was delayed unreasonably.

Most severe injuries (ACL or meniscal injuries) occur when the patient is involved in high-velocity weight-bearing activities, especially running and making sharp cuts or being subjected to direct valgus stress. Non–weight-bearing injuries (e.g., diving) usually result in patellar dislocation or subluxation only. Direct trauma to the front of the knee may cause posterior cruciate ligament injuries or patellar fractures, whereas lateral to medial (valgus) forces may cause collateral or cruciate ligament damage or fractures. Distinct popping noises or tearing sensations are reported in ACL injuries and patellar subluxation. Locking of the knee often may be reported in meniscal injuries, but not immediately after the injury. Although the knee may “hurt all over” when seen in the ED, the patient may be able to localize the initial pain. Meniscal or collateral ligament injuries cause pain on the lateral or medial aspect of the knee, whereas ACL injuries hurt just inferior to the patella, and Osgood-Schlatter's disease is painful a few centimeters inferior to the patella over the tibial tubercle. Swelling within 2 hours strongly suggests hemarthrosis and an associated ACL injury, meniscal injury, or osteochondral fracture. Swelling after 4 to 12 hours is more likely to be an isolated effusion without an associated fracture or ligamentous injury.

In subacute injuries, inquiry is made about hip or groin pain because the hip and knee share sensory nerves. Legg-CalvéPerthes disease or a slipped capital femoral epiphysis may cause knee pain. The patient is asked about changes in physical activities or footwear. A sensation of the knee giving way without actually falling when going down stairs or inclines may be elicited in patellofemoral pain syndrome (i.e., chondromalacia patella), whereas exacerbation doing deep-knee bends suggests Osgood-Schlatter's disease.

Examination of the patient should include walking and standing to check for medially deviated “squinting” patellae. The knees are inspected and palpated in two positions, sitting relaxed with the knees at 90 degrees and supine. When sitting, the knees are inspected for swelling, bony changes (e.g., swelling over the tibial tubercle in Osgood-Schlatter's disease), joint-line tenderness (meniscal injuries), and quadriceps atrophy. The knee should be extended and flexed while inspecting for patellar tracking and palpating for crepitations.

With the patient supine, inspection and palpation are repeated over the joint line, collateral ligaments, patella, and tibial tuberosity. If the knee appears swollen, an effusion is sought. Normally, synovial fluid coats the patellar surface but does not separate the patella and femur. When fluid separates the two bones, a sharp pat on the patella results in the sensation of a tap as the two bones meet. If the joint contains a large amount of fluid, the patella will not touch the femur but will feel as if it is sitting on a cushion. The ROM of the knee and quadriceps strength are documented.

The physician should test for collateral and cruciate ligament damage, meniscal injuries, patellar subluxation, and patellofemoral pain syndrome, using the appropriate maneuvers (Table 39.2). Each test is described in the appropriate differential diagnosis section.

Maneuver	Diagnosis
Collateral laxity test (Fig. 39.4)	Collateral ligament injury
Lachman test (Fig. 39.3)	Anterior cruciate ligament injury
Posterior drawer test (Fig. 39.5)	Posterior cruciate ligament injury
McMurray test (Fig. 39.6)	Meniscal injury
Apley compression test (Fig. 39.7)	Meniscal injury
Patellar apprehension test	Patellar subluxation
Patellar stress test	Patellofemoral pain syndrome

Table 39.2. Summary of Diagnostic Maneuvers for the Injured Knee

Distal pulses, the posterior tibial, and the dorsalis pedis should be palpated, and the peroneal nerve function should be assessed. The deep peroneal nerve enervates the ankle dorsiflexors. The extensor hallucis longus can be tested by opposing dorsiflexion of the great toe. The deep peroneal nerve supplies sensation to the web space between the great and second toes. Patients with knee symptoms should have a careful hip examination because aseptic necrosis of the femoral head or a slipped capital femoral epiphysis may present with knee pain.

All patients with acute knee injuries should have AP and lateral radiographs, and if indicated, a patellar (or skyline view) radiograph should be taken. If the injury is more chronic, an intercondylar or tunnel view should also be taken to evaluate for osteochondritis dissecans.

Figure 39.8 summarizes an approach to the child with an acutely injured knee. If the initial evaluation suggests vascular compromise, traction and reduction of the knee should be attempted and an emergency orthopedics consultation should be obtained. If the patella is obviously dislocated, it may be reduced before obtaining radiographs. Postreduction radiographs and a careful examination for physeal tenderness then can exclude the diagnosis of a fracture. If the patient's knee is too painful or swollen to allow a complete examination or if the patient has a hemarthrosis, ligament or meniscal damage should be suspected, and the knee should be immobilized for a maximum of three days until seen by an orthopedic surgeon.

FIGURE 39.8. Approach to the patient with an acute knee injury.

If the patient tolerates an examination, a series of maneuvers may suggest collateral ligament injury, cruciate ligament injury, meniscal injury, or patellar subluxation as the diagnosis (Table 39.2 summarizes the diagnostic maneuvers for the knee). Next an assessment for popliteal tenderness to exclude a Baker's cyst or hamstring strain is performed. Finally, if no signs of infection or hip disease exist, the patient may have a tendon rupture or a mild sprain, strain, or contusion.

Often, a patient may come to the ED with a history of trauma and knee pain that has been present for more than 1 or 2 days (Fig. 39.9). In addition to the standard AP, lateral, and patellar views, a tunnel or intercondylar view should be taken to exclude a fracture, tumor, and osteochondritis dissecans. If the initial knee and hip examinations do not suggest a diagnosis and no signs of infection exist, the diagnostic maneuvers in Table 39.2 should be completed. The patient may have an old collateral ligament, cruciate ligament, or meniscal injury and may require an orthopedic referral.

FIGURE 39.9. Approach to the patient with a subacute knee injury.

Suggested Readings

Chung S. Handbook of Pediatric Orthopedics. New York: Van Nostrand Reinhold, 1986.

Garrick JG. Knee problems in adolescents. Pediatr Rev 1983;4:235–243.

Harries M, Williams C, Stanish WD, Micheli LJ, eds. Oxford Textbook of Sports Medicine. New York: Oxford University Press, 1994.

Hoppenfield S. Physical Examination of the Spine and Extremities. Norwalk, CT: Appleton-Century-Crofts, 1976.

Rockwood CA, Wilkins KE, King RE, eds. Fractures in Children. 4th ed. Philadelphia: JB Lippincott, 1996.

Roy S, Irvin R. Sports Medicine: Prevention, Evaluation, Management, and Rehabilitation. Englewood Cliffs, NJ: Prentice-Hall, 1983.

Smith JB. Knee problems in children. Pediatr Clin North Am 1986;33: 1439–1456.

Tachjian MO. Clinical Pediatric Orthopedics. Stamford, CT: Appleton & Lange, 1997.

CHAPTER 40

Injury—Shoulder

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This chapter focuses on the diagnosis of the child with an overtly painful, injured shoulder (for the preverbal child with a possible shoulder injury presenting with an immobile arm, see Chapter 36). Children have different causes for their shoulder injuries than do adults because of their open growth plates, and young patients may be more difficult to examine because of anxiety and limited verbal skills. Figure 40.1 shows the important bony anatomy.

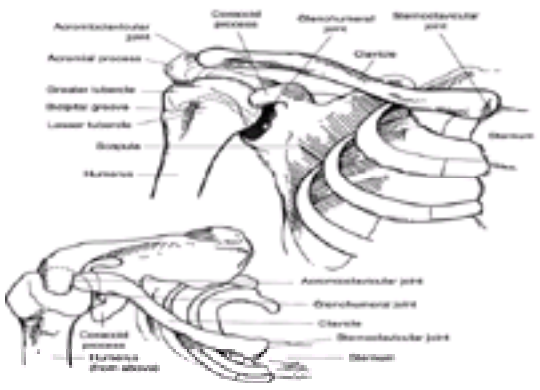

FIGURE 40.1. Anatomy of the shoulder.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis depends mainly on exactly where the patient has pain. In this chapter, injuries are described anatomically, from the sternoclavicular joint to the humeral shaft (Table 40.1 and Table 40.2).

Sternoclavicular joint	Rotator cuff
Dislocation*	Impingement
Sprain	Tear
Clavicle	Humerus
Physeal separation of medial end of the clavicle	Fracture of proximal humeral physis
Fracture	Stress fracture of proximal humeral physis ("Little League shoulder")
Contusion ("shoulder pointer")	Fracture of shaft
Osteolysis	Biceps tendon tendinitis
Acromioclavicular joint dislocation or sprain ("shoulder separation")	Pathologic fracture*
Scapula fracture	Referred pain (from)
Glenohumeral joint	Myocardium*
Dislocation ("shoulder dislocation")	Diaphragm*
Subluxation	Neck
	Thoracic outlet syndrome

*Potentially life-threatening conditions.

Table 40.1. Differential diagnosis of the injured Shoulder

Clavicle fracture	Humerus
Glenohumeral joint	Fracture of proximal humeral physis
Dislocation ("shoulder dislocation")	Fracture of shaft
Subluxation	

Table 40.2. Common Causes of the Injured Shoulder

Physeal (growth plate) separations of the medial clavicle are caused by indirect trauma that forces the shoulder medially and separates the growth plate. This injury mimics the sternoclavicular dislocations seen in adults. Before 18 years of age, injury to the sternoclavicular joint causes physeal separations because the epiphysis of the medial clavicle begins to ossify between 13 and 19 years of age and fuses between 22 and 25 years of age. At that time, a similar injury will cause sternoclavicular joint dislocations. Most separations are anterior, and the patient has swelling and tenderness over the sternoclavicular joint. Anteroposterior (AP) and superiorly projected lordotic radiographs comparing both clavicles may not visualize the lesion. Computed tomography (CT) or magnetic resonance imaging (MRI) is usually necessary to delineate the lesion. If the dislocation is posterior, the aorta or trachea may be injured; the child may remain asymptomatic or complain of choking or difficulty breathing. If the growth plate and ligaments are not disrupted by the injury, a simple sprain has occurred.

The clavicle is a commonly fractured bone in children, most often in the middle or lateral third of the bone. The clavicle is subject to any medially directed force on the upper limb (e.g., a fall on an outstretched hand) but is most commonly fractured by a direct blow. Although subclavian vessels and the brachial plexus are just beneath the clavicle, they are rarely injured because the subclavius muscle is interposed between the bone and vessels and the thick periosteum of the clavicle rarely splinters. A neonate's birthing injury or an infant's greenstick fracture of the clavicle may go unnoticed until the focal swelling of the developing callus is noted. In the older child, the arm droops down and forward, and the head may be tilted toward the affected side because of sternocleidomastoid muscle spasm. Localized swelling, tenderness, and crepitations may be noted. A radiograph will confirm the diagnosis. Rarely, a radiograph obtained because of clavicular trauma will show a congenital pseudarthrosis, or false joint of the clavicle.

Older children with a clavicular contusion or “shoulder pointer” may have swelling and tenderness over the distal clavicle. This injury can be differentiated from an acromioclavicular (AC) joint sprain by careful physical examination (the tenderness should be localized to the distal clavicle and not over the AC joint) and radiographs (see the section on AC joint separation).

Osteolysis of the distal clavicle with resorption of the bone may develop after minor injuries to the clavicle. Patients experience chronic pain and mild swelling 2 to 3 weeks after the initial injury. Radiographs are diagnostic.

AC joint injuries usually cause fractures of the distal clavicle in patients less than 13 years of age. Older children may injure the AC joint (“shoulder separations”) either by a direct blow to the shoulder or by transmitted force from a fall on an outstretched hand. The child will have pain with any motion of the shoulder and tenderness over the AC joint. In a first-degree sprain, the clavicle is not elevated above the acromion. In second- and third-degree sprains, swelling and elevation should be present. Bilateral “stress view” radiographs of the AC joint may be obtained to compare the separation on the normal and affected sides. A shoulder pointer contusion of the distal clavicle can be differentiated by tenderness limited to the distal clavicle and a normal radiograph. Cosmetic deformities and degenerative changes of the distal clavicle may complicate these injuries, even with appropriate therapy.

Scapula fractures are rare in pediatrics and usually occur only after major direct trauma such as a motor vehicle accident or a fall from a height. The child will have tenderness over the scapula. The patient often sustains other more life-threatening injuries (e.g., head injuries, rib fractures, pneumothoraces).

Shoulder or glenohumeral joint dislocations are rare in children less than 12 years old. These injuries become common in adolescence as the skeleton matures. The glenohumeral joint is shallow, allowing a wide range of motion, but increasing the risk of dislocation. The patient is injured when an already abducted and externally rotated arm is forcibly extended posteriorly. This action leverages the humeral head out of the glenoid fossa. The trauma can damage the axillary nerve or fracture the humeral head. More than 95% of all dislocations are anterior, and less than 5% are posterior. The patient will be in severe pain, supporting the affected arm internally rotated and slightly abducted (i.e., the patient cannot bring the elbow to his or her side). The shoulder contour is sharp, unlike the smooth contour of the opposite shoulder, and the acromion is prominent (Fig. 40.2). Sensation over the shoulder, especially laterally (axillary nerve distribution), and distal pulses should be documented. Radiographs should always be obtained because a humeral head or even a clavicular fracture may mimic a shoulder dislocation. An AP, transcapular lateral (Neer's), and an axillary view will show the location of the dislocation and the presence of any fractures.

FIGURE 40.2. An older adolescent patient with left anterior glenohumeral joint dislocation. Notice the sharp contour of the shoulder, the fullness below the glenoid fossa, and the prominent acromion.

If the patient has a history consistent with dislocation but has more range of motion than expected and the radiograph is normal, the patient may have spontaneously reduced a dislocated shoulder or only sprained the ligaments overlying the glenoid fossa and subluxated the glenohumeral joint. An apprehension test should be performed (Fig. 40.3).

FIGURE 40.3. The apprehension test to evaluate for shoulder subluxation. The patient's shoulder should be abducted passively and rotated externally. If this elicits apprehension or pain, the test is positive. If not, the examiner then should apply anteriorly directed pressure to the posterior aspect of the humeral head. If this elicits pain, then the test also is positive, and the patient's shoulder may have subluxed.

Actual tears of the rotator cuff are rare before 21 years of age. However, if the rotator cuff muscles are weak, the humeral head is displaced upward during overhead motion and may impinge the tendon of the supraspinatus muscle as it runs below the acromion. Impingement symptoms usually occur with repetitive overhead motions (e.g., throwing a ball). The pain is poorly localized. The patient may have mild tenderness under the acromion when the arm is abducted and may have increasing pain when the arm is abducted passively between 80 and 120 degrees. A test for impingement is illustrated in Figure 40.4. If the impingement test produces pain below the acromion in an adolescent athlete, the patient may have a lax glenohumeral joint. Plain radiographs are usually normal, and an MRI may be necessary.

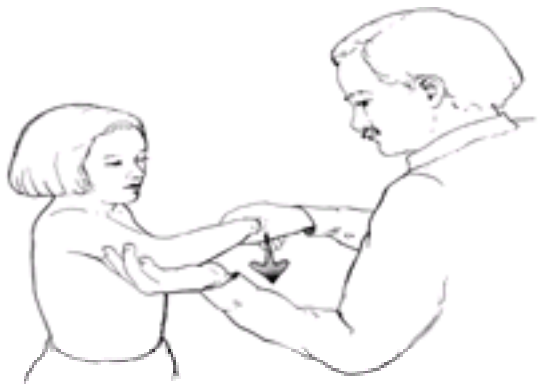

FIGURE 40.4. The impingement test to evaluate for impingement of the supraspinatus tendon. The patient's arm should be held in internal rotation and 90 degrees of forward flexion. Force the forearm downward by bringing the humeral head up against the acromion. Pain localized below the acromion suggests impingement.

Because the ligamentous attachments are stronger than the growth plate, fracture separations of proximal humeral epiphysis occur until the patient's epiphysis closes between 16 and 19 years of age. The injury occurs because of direct or indirect trauma, such as a backward fall or an attempt to break a fall with a hand. The patient usually has mild swelling and local tenderness. AP and lateral radiographs confirm the diagnosis, although in patients under the age of 6 years, the injury may be difficult to visualize because the epiphysis is mainly cartilaginous. Even in older patients, slight widening of the epiphysis may be difficult to see, and comparison views of the uninjured shoulder may be necessary.

Stress fractures of the proximal humeral epiphysis, or “Little League shoulder,” are caused by repetitive internal rotation of an abducted, externally rotated shoulder during the throwing motion. The child has diffuse shoulder pain that worsens after throwing. The proximal humerus may be tender, and radiographs show widening of the proximal humeral epiphysis.

Transverse or comminuted fractures of the humeral shaft may occur from direct trauma, whereas spiral fractures usually occur from indirect trauma (e.g., a fall on a hand). If the history is implausible, inconsistent from one caretaker to another, or the patient is less than 2 years of age, the child should be evaluated for physical abuse. The patient will have obvious pain, tenderness, and local deformity. Care must be taken not to miss an associated neurovascular injury because the radial nerve runs along the humeral shaft. Radial nerve damage results in weakness of wrist extension and anesthesia of the skin between the first and second metacarpals. Radiographs help rule out a pathologic fracture (e.g., through a unicameral bone cyst or tumor).

Shoulder pain is rarely related to tendinitis of the tendon of the long head of the biceps. This tendon runs through the bicipital groove just anterior and medial to the greater humeral tuberosity. The tendon moves within the groove during

internal and external rotation. The patient often has chronic pain and tenderness over the bicipital groove.

A painful shoulder or fracture that follows minimal trauma may be caused by a benign or malignant tumor or by nonneoplastic bone lesions. Osteochondromas (exostoses) are outgrowths of benign cartilage from the bone adjacent to the epiphysis and present with a mass adjacent to a joint. The nonossifying fibromas (called fibrous cortical defects if smaller than 0.5 cm) are common asymptomatic lesions that may lead to pathologic fractures.

The malignant chondroblastoma is a rare tumor, but its most common location is the proximal humerus. The patient often has joint pain from effusion associated with this tumor. Osteogenic sarcomas and Ewing's sarcoma are more common but involve the humerus in only 10% of cases

Unicameral and aneurysmal bone cysts are asymptomatic until the bone fractures, but they are not neoplastic lesions. Unicameral cysts are more likely to affect the humerus.

Shoulder pain also may be referred from the neck (e.g., cervical disc herniation), myocardium, or diaphragm (e.g., a splenic hematoma) after trauma to those areas.

Thoracic outlet syndrome, with compression of the medial cord of the brachial plexus (C8–T1) may rarely present as shoulder pain. The pain and anesthesia most commonly follow the dermatome of the ulnar nerve. The pain may be reproduced by 180 degrees of forward flexion; radiographs should include the neck to exclude a cervical rib.

EVALUATION AND DECISION

Initially, the patient's neurovascular status is assessed and fracture stabilization provided, if necessary (Fig. 40.5).

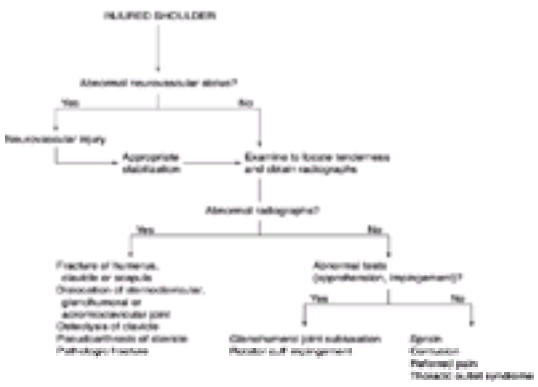

FIGURE 40.5. Approach to the patient with an injured shoulder.

For an isolated shoulder injury, the pain should be localized as specifically as possible and the mechanism of injury determined. An attempt is made to determine whether the trauma was direct or indirect and to ascertain what position the shoulder was in when the injury occurred. If the pain is chronic, the position or motion that most exacerbates the pain is determined (e.g., throwing a ball). The patient is asked about any distal paresthesias, associated pain, and trauma (e.g., neck, chest, abdomen). The possibility of abuse in young children should always be considered, especially if the injury is unexplained, the history is implausible, or the seeking of medical care was delayed unreasonably.

Initially, the patient is observed without clothes over the shoulder for positioning of the arm, swelling, deformity, or any asymmetry. Then, one asks the patient to point with one finger to the most painful area. This observation period before the formal physical examination is especially important in a young, anxious child and helps prioritize the rest of the evaluation.

If the child seems anxious, the uninjured side is examined first. The examiner carefully palpates the entire shoulder from sternoclavicular joint to the shaft of the humerus. Swelling and tenderness at the sternoclavicular joint suggests a physeal separation or dislocation at this site. The clavicle is covered only by a thin platysma muscle and a fracture is easily seen and palpated. Just lateral to the clavicle is the AC joint. Elevation of the clavicle above the acromion or tenderness of the articulation suggests AC joint dislocation (“shoulder separation”) or contusion. With the shoulder in external rotation, palpation just lateral to the acromion will locate the greater tuberosity of the humerus. Just in front of the greater tuberosity is the tendon of the long head of the biceps within the bicipital groove. Pressure may produce exquisite tenderness in this area, so palpation should be gentle; if uncertainty about a finding of tenderness exists, a comparison with the examination of the uninjured side is helpful. Finally, the proximal humeral shaft and the scapula are palpated.

During the neurologic evaluation, it is important that sensation is tested over the deltoid muscle because the axillary nerve may be damaged during a shoulder separation

Next follows an examination of the patient's active and passive range of motion (Fig. 40.6), checking for abduction, adduction, forward flexion, backward extension, internal rotation, and external rotation. Internal and external rotation can be observed easily in a child by asking the patient to touch behind the neck (external rotation) and lower back (internal rotation).

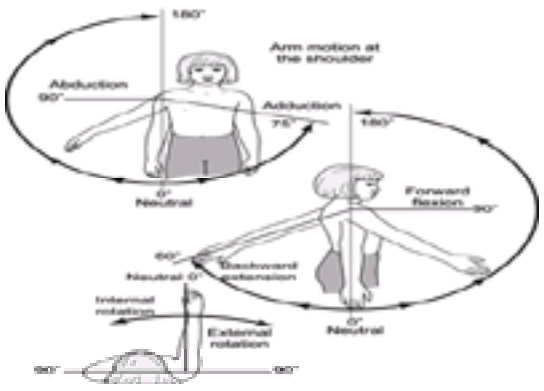

FIGURE 40.6. Range of motion of the shoulder joint.

Once the pain has been localized, appropriate radiographs are obtained, and when indicated, two additional specific tests are performed: the apprehension test for shoulder subluxation and the impingement test for rotator cuff impingement (Fig 40.3 and Fig 40.4).

As previously discussed, patients with normal radiographs and negative maneuvers are most likely to have sprains or contusions, but occasionally, they may be experiencing referred pain.

Suggested Readings

Chung S. Handbook of Pediatric Orthopedics. New York: Van Nostrand Reinhold, 1986.

Harries M, Williams C, Stanish WD, et al, eds. Oxford Textbook of Sports Medicine. New York: Oxford University Press, 1994.

Hoppenfield S. Physical Examination of the Spine and Extremities. Norwalk, CT: Appleton-Century-Crofts, 1976.

Ireland ML, Andrews JR. Shoulder and elbow injuries in the young athlete. Clin Sports Med 1988;7:473–486.

Rockwood CA, Wilkins KE, King RE, eds. Fractures in Children. 4th ed. Philadelphia: JB Lippincott, 1996.

Roy S, Irvin R. Sports Medicine: Prevention, Evaluation, Management, and Rehabilitation. Englewood Cliffs: Prentice-Hall, 1983.

Tachjian MO. Clinical Pediatric Orthopedics. Stamford, CT: Appleton & Lange, 1997.

Tibone J. Shoulder problems of adolescents. Clin Sports Med 1983;2:423–427.

CHAPTER 41

Jaundice—Unconjugated Hyperbilirubinemia

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Jaundice—a yellowish discoloration of the skin, tissues, and bodily fluids—indicates an elevated serum bilirubin concentration. Hyperbilirubinemia results from increased production or impaired excretion of bilirubin. Both physiologic and pathologic etiologies may give rise to unconjugated (or indirect) hyperbilirubinemia; to distinguish, the clinician must consider the patient's age, the rate of increase and peak level of serum bilirubin, and the historical features of the case. Prompt evaluation and therapy for unconjugated hyperbilirubinemia are particularly important for infants less than 1 week old. If bilirubin levels rise too high, these young infants risk developing neurodevelopmental deficits as well as kernicterus.

PATHOPHYSIOLOGY

Bilirubin is formed from the degradation of hemoglobin and other heme-containing proteins. Heme protoporphyrin becomes oxidized, producing biliverdin, which then is reduced to form bilirubin. In the blood, bilirubin is bound to albumin. The liver uptakes bilirubin for conjugation by a glucuronyl transferase, and then excretes the conjugated form into bile. The net balance of its entry into and removal from the circulation determines serum bilirubin concentration. In the newborn infant, multiple factors contribute to jaundice, including high bilirubin load from increased red blood cell (RBC) volume and decreased red cell survival, as well as impaired plasma binding, liver uptake, conjugation, and excretion of bilirubin.

Older children and adults normally have serum bilirubin concentrations below 1 mg/dL and appear jaundiced with a bilirubin level above 2 mg/dL. Newborn infants becomes visibly jaundiced when their serum bilirubin concentration is above 5 mg/dL. In conjugated, or direct, hyperbilirubinemia, the conjugated (or direct reacting) portion of the serum bilirubin exceeds 2.0 mg/dL and is more than 15 to 30% of the total. Otherwise, the hyperbilirubinemia is unconjugated, or indirect (see Chapter 42).

In neonates, hyperbilirubinemia may be associated with neurotoxicity, encephalopathy, and kernicterus. Kernicterus is a rare but devastating neurologic disorder with characteristic yellow staining in the basal ganglia and other brain regions.

DIFFERENTIAL DIAGNOSIS

For patients with primarily unconjugated hyperbilirubinemia who are beyond the neonatal period, the most common causes are hemolytic processes resulting in overproduction of bilirubin (Table 41.1). Extravascular blood in a concealed hemorrhage may also be metabolized into supraphysiologic concentrations of bilirubin. Infants and children with jaundice may have enzyme deficiencies or other conditions that impair the hepatic uptake or conjugation of bilirubin. Upper gastrointestinal (GI) obstruction generally causes conjugated hyperbilirubinemia but may occasionally be associated with an unconjugated hyperbilirubinemia. In the neonatal period, physiologic and breast milk jaundice are common. The potential for maternal–fetal blood group incompatibility is critical to recognize.

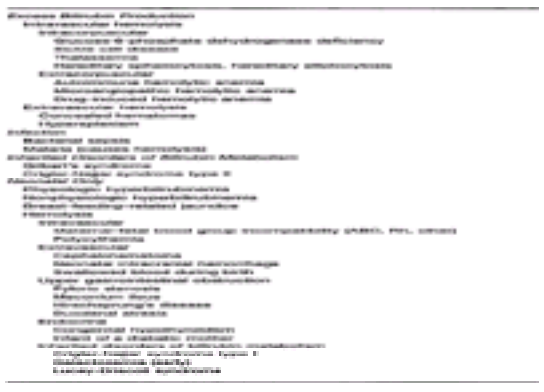

Table 41.1. Causes of Primarily Unconjugated Hyperbilirubinemia

Excess Bilirubin Production

The numerous causes of hemolysis may be classified as intravascular or extravascular. Intravascular hemolysis may be further divided into intracorpuscular and extracorpuscular defects (Table 41.1). Inborn errors of metabolism, such as glucose-6-phosphate dehydrogenase (G6PD) deficiency, may result in destruction of RBCs. This disorder is common in African-American and Asian children as well as those of Mediterranean origin. Patients with G6PD deficiency who are exposed to oxidant stress (e.g., fava beans, sulfa drugs) may have acute rapid hemolysis. Neonates with G6PD deficiency have an increased risk of jaundice that is only partially attributable to hemolysis.

Hemoglobinopathies, including sickle cell disease, can result in hemolysis, as can the impairments in hemoglobin chain synthesis that occur in the thalassemias. Defects in the RBC membrane found in hereditary spherocytosis and hereditary elliptocytosis increase the fragility of the corpuscles. Extracorpuscular causes of RBC destruction include the autoimmune, microangiopathic, and drug-induced hemolytic anemias.

Hematomas, pulmonary hemorrhages, and other collections of extravasated blood undergo hemolysis and, if sufficiently large, can elevate serum levels of unconjugated bilirubin. Various hypersplenic states, including splenic sequestration crisis in sickle cell disease, may result in anemia with accompanying hemolysis and hyperbilirubinemia.

Infection

Jaundice may be a harbinger of serious infection. Bacterial endotoxins reduce bile flow and can cause hyperbilirubinemia. The neonate with jaundice as well as poor feeding, lethargy, or fever should be evaluated for sepsis and urinary tract infection. However, sepsis is exceedingly rare among well-appearing jaundiced neonates who have no additional signs or symptoms, occurring at a rate considerably below 1%. Malaria, caused by *Plasmodium* species, is endemic in tropical regions. In patients with malaria, a high degree of parasitemia may result in massive hemolysis presenting with jaundice.

Inherited Disorders of Bilirubin Metabolism

Gilbert's syndrome is a common cause of mild, intermittent, unconjugated hyperbilirubinemia that occurs in as many as 6% of the population. Patients with Gilbert's syndrome have a partial deficiency of glucuronyl transferase. They generally do not present until late childhood or early adolescence, when they may develop nonspecific abdominal pain, nausea, and mild jaundice during an intercurrent illness. Other liver function studies are normal, and there is no evidence of hemolysis or hepatosplenomegaly. The serum bilirubin rarely exceeds 5 mg/dL.

Crigler-Najjar syndrome is characterized by the absence or deficiency of the enzyme bilirubin glucuronyl transferase. Type I, the more severe form, manifests soon after birth and is associated with high morbidity and mortality. Type II, the milder form, caused by an incomplete enzyme deficiency, typically presents in infancy or later in childhood but has been reported to first appear as late as adolescence. The type II form is generally treatable with phenobarbital.

Special Considerations in the Neonate

Physiologic Neonatal Hyperbilirubinemia

Most newborns develop a mild hyperbilirubinemia; approximately 60% manifest clinical signs of physiologic jaundice. Physiologic jaundice peaks between 3 and 5 days of life in the term infant and requires no treatment. Because at high levels bilirubin may be associated with neurotoxic effects, careful attention should be paid to distinguishing physiologic from nonphysiologic jaundice.

Nonphysiologic Neonatal Hyperbilirubinemia

One to two percent of newborns require readmission within the first week of life, and up to 85% of these readmissions are for nonphysiologic neonatal hyperbilirubinemia. Jaundice in the term newborn is nonphysiologic if it is conjugated or appears within the first 24 hours of life. Other indications that jaundice is not physiologic are a peak serum total bilirubin concentration of 17 mg/dL or higher in the breast-fed infant and 15 mg/dL or higher in the formula-fed infant. Also, infants with persistence of jaundice beyond the first week of life, or whose serum bilirubin level increases more than 5 mg/dL per day, should be followed closely for nonphysiologic jaundice. Risk factors for nonphysiologic neonatal hyperbilirubinemia include history of a sibling with hyperbilirubinemia, breast-feeding, lower gestational age, maternal diabetes, bruising (from birth trauma), and Asian race. Dehydrated neonates may develop unconjugated hyperbilirubinemia.

Breast-Feeding and Jaundice

Breast-fed newborns develop a greater degree of hyperbilirubinemia more often than do formula-fed newborns. Breast-milk jaundice, occurring in 1% of newborns, is associated with the breast milk itself and may be hormonally mediated or related to intestinal excretion and resorption of bile. Many cases of jaundice in breast-fed infants are caused by nonoptimal breast-feeding practices that result in dehydration.

Hemolysis

Birth trauma, when associated with a cephalohematoma, extensive bruising, or swallowed maternal blood, can result in hyperbilirubinemia. Intracranial, pulmonary, or other concealed hemorrhage also can lead to extravascular hemolysis. Similarly, polycythemia, caused by delayed clamping of the cord or maternal–fetal or fetal–fetal transfusion (in multiple gestations), increases the RBC mass and causes jaundice in neonates.

When maternal antibodies are produced against fetal red cell antigens, the neonate can develop a Coombs' positive isoimmune hemolytic anemia. The risk of kernicterus in infants with hyperbilirubinemia as a result of isoimmune hemolytic anemia is much greater than the risk in infants with nonhemolytic causes of jaundice. Fetal Rh and A and B blood group antigens are most commonly etiologic in the hemolysis syndrome, although dozens of antigens have been implicated. Rh-negative mothers may become sensitized to an Rh-positive fetus during pregnancy and mount an antibody response to a fetus during a subsequent pregnancy. Administration of Rhogam to Rh-negative mothers who have not yet developed anti-Rh antibodies can prevent Rh isoimmunization. ABO hemolytic disease of the newborn generally occurs in infants with A or B blood groups whose mothers have type O blood. Maternal anti-A and anti-B antibodies are produced and can result in hemolysis with a positive direct Coombs' test.

Upper Gastrointestinal Obstruction

Pyloric stenosis, meconium ileus, Hirschsprung's disease, duodenal atresia, and other causes of upper GI obstruction may present with jaundice and clinical signs of obstruction. In neonates, obstruction can increase enterohepatic circulation or decrease the enzyme activity responsible for bilirubin uptake, resulting in unconjugated hyperbilirubinemia. In contrast, older children and adults with upper GI obstruction and jaundice generally have a conjugated hyperbilirubinemia.

Endocrine Disorders

Unconjugated hyperbilirubinemia may be the presenting sign of congenital hypothyroidism, preceding other manifestations by several weeks. The mechanism probably relates to reduced bile flow. Other signs that may be present include persistent poor feeding, prolonged jaundice, constipation, and hypotonia. Infants of diabetic mothers are also at increased risk of jaundice, with as many as 19% developing nonphysiologic hyperbilirubinemia.

Inherited Disorders of Bilirubin Metabolism

Very high, rapidly rising levels of bilirubin not responsive to phototherapy raise concern for Crigler-Najjar syndrome type I or Lucey-Driscoll syndrome. Lucey-Driscoll syndrome is probably caused by an inhibition of glucuronyl transferase. Infants with galactosemia may exhibit an unconjugated hyperbilirubinemia during the first week of life. Older infants with galactosemia tend to have a conjugated hyperbilirubinemia. Infants with galactosemia usually also present with vomiting, failure to thrive, poor feeding, abdominal distension, and hypoglycemia.

EVALUATION AND DECISION

An approach to the patient with unconjugated hyperbilirubinemia is outlined in Figure 41.1. Hemolysis and Gilbert's syndrome are the most common cause of jaundice in the patient beyond the neonatal period (Table 41.2). During the neonatal period, physiologic jaundice and breast-feeding–related jaundice are the most likely causes. The differential diagnosis is broad, and evaluation should always begin with a detailed history and physical examination.

FIGURE 41.1. Evaluation of the pediatric patient with unconjugated hyperbilirubinemia.

Excess Bilirubin Production
Glucose-6-phosphate dehydrogenase deficiency
Sickle cell disease
Infection
Malaria (causes hemolysis)
Inherited Disorders of Bilirubin Metabolism
Gilbert's syndrome
Neonatal Only
Physiologic hyperbilirubinemia
Nonphysiologic hyperbilirubinemia
Breast-feeding–related jaundice
Overproduction of hemoglobin
Hemolysis
Maternal-fetal blood group incompatibility (ABO, Rh, other)
Cephalohematoma

Table 41.2. Common Causes of Primarily Unconjugated Hyperbilirubinemia

Acute Hemolysis
Infection
Bacterial sepsis
Malaria (causes hemolysis)
Neonatal Only
Nonphysiologic hyperbilirubinemia
Hemolysis
Maternal-fetal blood group incompatibility (Rh, ABO, other)
Polycythemia
Upper gastrointestinal obstruction
Endocrine
Congenital hypothyroidism
Inherited disorders of bilirubin metabolism
Crigler-Najjar syndrome type I
Galactosemia (early)
Lucey-Driscoll syndrome

Table 41.3. Life-Threatening Causes of Primarily Unconjugated Hyperbilirubinemia

History

A general clinical history may help guide the workup. An infant who has been lethargic or apneic or a child who has been ill and febrile may require evaluation for serious bacterial infections (Table 41.3). A neonate with persistent or bilious emesis may have an upper GI obstruction.

The clinician should ascertain whether there are factors predisposing a patient to jaundice. One such factor is a family history of jaundice or anemia or a racial or ethnic origin associated with hemolytic anemias. African-American race and Mediterranean ancestry are associated with G6PD deficiency. Additional causes of jaundice run in families or have racial predisposition. African-American patients are much more likely to have sickle cell disease. Mediterranean and Asian children have a higher incidence of thalassemia. East Asian neonates are more likely to develop nonphysiologic hyperbilirubinemia. A history of drug ingestion may lead to the diagnosis of drug-induced hemolysis. A dietary history may identify an agent such as the fava bean that induces hemolysis in patients with G6PD deficiency. Residence in or travel to sub-Saharan Africa, South East Asia, or parts of Central and South America carries a risk of exposure to malaria.

Special Historical Considerations in the Neonate

For the newborn, timing of the onset of icterus is critical. Most jaundice that appears before the first 24 hours of life is pathologic. Maternal blood type and Rhogam status should be ascertained to establish risk factors for isoimmune hemolytic anemia. Previous bilirubin levels and the results of Coombs' testing should be ascertained. Feeding practices influence development of jaundice; a breast-fed infant is at risk for breast-feeding–related jaundice, including jaundice resulting primarily from dehydration. Knowledge of weight gain or loss may help judge hydration status.

Physical Examination

The general appearance of the patient will help guide the clinician as to the likelihood of a serious underlying condition such as bacterial sepsis. In the neonate, poor feeding, lethargy, apnea, tachypnea, and temperature instability are highly concerning.

The sclera and skin should be examined closely under adequate light. In a patient with dark skin, palms and soles may be less pigmented and easier to assess for icterus. Gentle pressure with one finger to blanch the skin facilitates inspection of skin color. In neonates, jaundice progresses in a cephalocaudad direction. Newborns with jaundice below the knees and on the palms have the highest levels of serum bilirubin. Pallor may indicate anemia from hemolysis or bleeding.

Presence of a cephalohematoma or large areas of ecchymosis may point to an extravascular hemolysis as the cause of hyperbilirubinemia. Hepatomegaly may indicate underlying liver dysfunction. Splenomegaly can indicate a hypersplenic state such as splenic sequestration in sickle cell disease. Splenomegaly may also be present in lupus, which is associated with an autoimmune hemolysis. In a neonate with jaundice and vomiting, an abdominal mass suggests upper

GI obstruction.

Laboratory Testing

The serum bilirubin level should always be determined. Jaundiced patients beyond the neonatal period should be evaluated for anemia with a complete blood count (CBC) and reticulocyte count. Those with evidence of anemia and/or hemolysis should have a peripheral blood smear examined microscopically. Characteristic abnormal morphology such as sickle cells, spherocytes, or elliptocytes may be identified. Helmet and fragmented cells are diagnostic of a microangiopathic hemolytic anemia, such as that occurring in hemolytic uremic syndrome. Malarial ring forms may be apparent. Nucleated RBCs and Howell-Jolly bodies indicate a sustained hemolysis. Patients with anemia or hemolysis should also have a Coombs' test performed to look for evidence of autoimmune hemolysis. Testing for G6PD should be performed if the patient has risk factors or a consistent clinical presentation. Hemoglobin electrophoresis may be used to diagnose hemoglobinopathies such as sickle cell disease and thalassemia. If hepatomegaly is present or if there is no evidence of anemia, liver function studies should be performed. Patients with no laboratory abnormalities other than serum unconjugated bilirubin below 5 mg/dL have Gilbert's syndrome, a benign condition.

Special Laboratory Considerations in the Neonate

In neonates, it may be important to determine the rate of rise of serum bilirubin with serial measurements. The clinician must know whether a newborn has a set-up for maternal–fetal isoimmune anemia. Therefore, either the mother's blood and Rh type and antibody status should be obtained, or the infant's blood type should be determined and Coombs' testing performed. A neonate with probable physiologic jaundice does not need to undergo an anemia or hemolysis workup if he or she has no family history of hemolytic disease, no maternal–fetal blood group incompatibility, and no physical stigmata of anemia.

If clinical signs of obstruction are present, the patient should undergo appropriate laboratory testing such as abdominal radiographs, ultrasound, or upper GI series with contrast. The neonate with fever or ill appearance should be evaluated for serious bacterial infection, with peripheral white blood cell count, urine analysis, and cerebrospinal fluid analysis as well as blood, urine, and cerebrospinal fluid cultures. Results of the newborn screen for congenital hypothyroidism may be available. The newborn with symptoms of congenital hypothyroidism needs to have a determination of T_4 level. A newborn with poor feeding, vomiting, or failure to gain weight should be evaluated for galactosemia. If the newborn with galactosemia has already started feeds, the urine will contain reducing substances (Clinitest positive) but no glucose.

Approach

Beyond the Neonatal Period

In all children with jaundice, a total bilirubin level with fractionation and CBC should be performed. If a patient appears acutely ill, the physician should proceed with the appropriate evaluation and treatment for sepsis. Among well-appearing patients, the hematocrit determines the likely diagnostic possibilities and appropriate studies.

Anemia

Anemic children are suspect for having hemolytic processes, including autoimmune hemolytic anemia, hemoglobinopathies (e.g., thalassemia or sickle cell anemia), enzyme deficiencies (e.g., G6PD), red cell membrane defects (e.g., spherocytosis), hypersplenism, drug reactions, hemolytic uremic syndrome, and malaria. Extravascular hemolysis and resultant jaundice occur occasionally in children with concealed blood loss or large hematomas. A family history of hemolytic anemia and severe jaundice, particularly among children of Mediterranean descent, suggests G6PD deficiency, whereas abnormal RBC morphology points to sickle cell anemia, hereditary spherocytosis, or heredity elliptocytosis. Additional clues on the peripheral smear include helmet and fragmented cells in hemolytic uremic syndrome and ring forms in malaria.

A positive Coombs' test is seen with autoimmune hemolytic anemia. In patients with splenomegaly, hemolysis may lead to jaundice. Finally, drug reactions and unusual hemolytic anemias should be considered.

Normal Hematocrit

When unconjugated hyperbilirubinemia occurs without anemia, abnormal liver function studies (transaminases, prothrombin time, and partial thromboplastin time) differentiate hepatic disease from inherited disorders of bilirubin metabolism. Among these latter disorders, only Gilbert's syndrome, which produces a mild elevation in the serum bilirubin level, is at all common.

Neonatal Period

As for older children, an ill appearance and/or fever suggest sepsis. Other disorders likely to cause lethargy include bowel obstruction, hypothyroidism, and inborn errors of metabolism, such as galactosemia. Among well-appearing neonates, the presence of anemia serves as an important point in the differential diagnosis.

Anemia

The foremost consideration is the infant with indirect hyperbilirubinemia and anemia is isoimmune hemolytic disease, caused by blood group incompatibility, because this disorder may lead to kernicterus. The diagnosis can be established by determining the blood group and Rh status of the maternal–infant dyad in combination with a Coombs' test in the infant. Other disorders that produce jaundice include enzymatic and structural disorders of the red cells (e.g., G6PD,

hereditary spherocytosis) and the poorly understood occurrence of jaundice in infants of diabetic mothers.

Normal Hematocrit

In the absence of anemia, extravascular hemolysis is a common cause of jaundice in the newborn, with the breakdown of hemoglobin occurring in a cephalohematoma, large ecchymosis, or from swallowed blood. In addition, polycythemic infants are prone to jaundice. Also, hemolysis of even small amounts of hemoglobin may markedly elevate the serum bilirubin level in the infant with an immature liver. Thus, the same disorders that are diagnosed in anemic infants may occur in the jaundiced neonate with a normal hematocrit.

Most infants with indirect hyperbilirubinemia will have a negative evaluation for the disorders previously listed. If the bilirubin level is less than 12 mg/dl, rises slowly, and resolves before 8 days of age, one can diagnose physiologic hyperbilirubinemia without further laboratory studies. When these conditions are not met, the most likely cause for the jaundice is the hormonal impairment of bilirubin conjugation. Other possibilities, either alone or in combination with breast milk, include Crigler-Najjar and Lucey-Driscoll syndromes.

MANAGEMENT

For patients beyond the neonatal period, the management of hyperbilirubinemia is primarily directed at identification and treatment of the underlying cause. In some cases of severe hyperbilirubinemia, such as those caused by Crigler-Najjar syndrome type II, phenobarbital may be indicated. In contrast, newborns with jaundice require careful monitoring and sometimes specific therapies for hyperbilirubinemia because the neonatal central nervous system is susceptible to the toxic effects of bilirubin. The emergency physician may initiate the management of term newborn infants with jaundice and arrange hospitalization or subsequent follow-up after discharge. Infants discharged home from their birth hospitalization on the first or second day of life are at increased risk of readmission for hyperbilirubinemia. The management of premature infants with hyperbilirubinemia is highly specialized and not discussed here.

The goal of neonatal hyperbilirubinemia management is to prevent neurotoxicity, encephalopathy, and kernicterus. The jaundiced newborn needs to be kept well hydrated, and enteral feeding should be encouraged to promote bilirubin excretion. When bilirubin levels rise significantly, phototherapy and exchange transfusion may be indicated.

Phototherapy can be initiated with either an overhead bank of lights or a fiberoptic light source in a blanket. The optimal wavelength is 425 to 475 nm. Phototherapy primarily promotes 1) photoisomerization of unconjugated bilirubin to a less toxic isomer, which is excreted in the bile, and 2) structural isomerization of bilirubin to lumirubin, which is excreted in the bile and urine.

Indications for phototherapy and exchange transfusion vary according to the age of neonate. For the term neonate who develops jaundice after the first day of life and has no evidence of hemolysis, indications for phototherapy and exchange transfusion are shown in Table 41.4. When there is evidence of isoimmune hemolysis, phototherapy should be started immediately and a neonatologist should be consulted regardless of bilirubin level.

	Total Serum Bilirubin Level (mg/dL)			
Age (h)	Consider Phototherapy[b]	Phototherapy	Exchange Transfusion if Intensive Phototherapy[c] Fails	Exchange Transfusion and Intensive Phototherapy
<24	—	—	—	—
24–48	≥12	≥15	≥20	≥25
49–72	≥15	≥18	≥25	≥30
>72	≥17	≥20	≥25	≥30

Adapted with permission from the American Academy of Pediatrics practice parameter for management of hyperbilirubinemia in the healthy term newborn.
[b]Term infants who are clinically jaundiced at <24 hours old are not considered healthy and require further evaluation.
[b]Phototherapy at these total serum bilirubin levels is a clinical option, meaning that the intervention is available and may be used on the basis of individual clinical judgment.
[c]Intensive phototherapy should produce a decline of total serum bilirubin of 1 to 2 mg/dL within 4 to 6 hours and the total serum bilirubin level should continue to fall and remain below the threshold level for exchange transfusion. If this does not occur, it is considered a failure of phototherapy.

Table 41.4. Management of Hyperbilirubinemia in the Healthy Term Newborn[a]

During phototherapy, the baby should be undressed to maximize the exposed surface area of the skin. Intensive phototherapy with two banks of lights or two fiberoptic blankets will improve efficacy. When using overhead lights, the infant's eyes must be shielded and maintenance fluid requirements are increased. Phototherapy is relatively contraindicated in patients with conjugated hyperbilirubinemia because it can cause the “bronze baby syndrome.”

When bilirubin levels are toxic, exchange transfusion may be necessary. Exchange transfusion is most commonly indicated in infants with hemolytic disease. Generally, fresh irradiated reconstituted whole blood is pushed in through an umbilical vein catheter while blood is pulled out through an umbilical artery catheter. Careful monitoring is necessary. Complications include electrolyte and acid-base disturbances, hemolysis, and infection.

For jaundiced, breast-fed infants, the interruption or discontinuation of breast-feeding should be discouraged. Any of several management strategies, however, are accepted: 1) the infant may be observed while normal breast-feeding continues; 2) if bilirubin levels are high (Table 41.4), the infant may continue to breast-feed while receiving phototherapy; 3) breast-feeding may be supplemented with or without administration of phototherapy; and 4) breast-feeding may be interrupted and formula may be substituted with or without administration of phototherapy.

Suggested Readings

Gartner LM. Neonatal jaundice. Pediatr Rev 1994;15:422–432.

Hinkes MT, Cloherty JP. Neonatal hyperbilirubinemia. In: Cloherty JP, Stark AR, eds. Manual of Neonatal Care. Philadelphia: Lippincott-Raven, 1998:175–209.

Martinez JC, Maisels MJ, Otheguy L, et al. Hyperbilirubinemia in the breast-fed newborn: a controlled trial of four interventions. Pediatrics 1993;91:470–473.

Nathan DG, Orkin SH. Nathan and Oski's Hematology of Infancy and Childhood. 5th ed. Philadelphia: WB Saunders, 1998.

Provisional Committee for Quality Improvement and Subcommittee on Hyperbilirubinemia. Practice parameter: management of hyperbilirubinemia in the healthy term newborn. American Academy of Pediatrics. Pediatrics 1994;94:558–565.

Watchko JF, Oski FA. Bilirubin 20 mg/dL = vigintiphobia. Pediatrics 1983;71:660-3.

CHAPTER 42

Jaundice—Conjugated Hyperbilirubinemia

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The presence of jaundice in a child can be a useful indicator of occult pathology. The finding of icterus should set in motion a careful diagnostic search to elucidate the cause. The ultimate goal, to identify precisely the cause of the clinical syndrome, may rest in some cases with the longitudinal caretaker. In all cases, however, the emergency physician at first visit must separate patients whose admission can be temporized from those who require urgent intervention and/or immediate hospitalization.

PATHOPHYSIOLOGY

Unconjugated bilirubin is largely a product of converted heme from senescent red blood cells. Unconjugated bilirubin is transported from extrahepatic reticuloendothelial cells to the liver, bound to albumin. Albumin is detached as the bilirubin gains entry into the hepatocyte. In the liver cell, bilirubin is conjugated with glucuronide by the action of uridine diphosphate glucuronyl transferase. The soluble conjugated diglucuronide then is secreted across the canalicular membrane into the bile. In the intestine, as a result of the activity of bacterial flora, bilirubin is converted to urobilinogen. A portion of urobilinogen is reabsorbed into the portal circulation and is taken up by the liver cells, only to be reexcreted into the bile. A small percentage of urobilinogen escapes into the systemic circulation and is excreted in the urine. The unabsorbed urobilinogen is excreted in the stool as fecal urobilinogen.

In hepatocellular disease, the damaged liver may be unable to excrete the conjugated bilirubin produced in normal amounts. Or, in the absence of hepatic damage, regurgitation into the plasma of conjugated bilirubin may result from functional cholestasis, disruption of the hepatic architecture, or extrahepatic biliary obstruction. In most instances of jaundice primarily related to hepatic disease, the plasma exhibits elevated concentrations of unconjugated and conjugated bilirubin. Overt mechanical obstruction of bile excretion leads to raised plasma levels of conjugated bilirubin, and only as secondary liver damage occurs do unconjugated bilirubin levels rise.

DIFFERENTIAL DIAGNOSIS

Conjugated hyperbilirubinemia, defined as an elevated total bilirubin with greater than 30% direct reacting, always is pathologic. The differential diagnosis includes a variety of structural defects, infections, hepatotoxins, inborn errors of metabolism, and familial syndromes (Table 42.1).

Table 42.1. Causes of Conjugated Hyperbilirubinemia in Infants and Children

Although only a few diseases commonly cause conjugated hyperbilirubinemia (Table 42.2), all are serious. In addition, several less common conditions are important considerations because they are life-threatening (Table 42.3).

Infancy	Childhood
Idiopathic cholestasis	Viral hepatitis
Biliary atresia	Hepatotoxins
Perinatal infections (TORCH)	
Sepsis/urinary tract infection	

Table 42.2. Common Causes of Conjugated Hyperbilirubinemia

Fulminant hepatic failure	Abdominal crisis, sickle hemoglobinopathy
Septicemia	Hepatorenal syndrome
Intra-abdominal sepsis	Reye syndrome (usually anicteric)
Pyogenic liver abscess	
Suppurative cholangitis	
Peritonitis	

Table 42.3. Life-Threatening Causes of Conjugated Hyperbilirubinemia

EVALUATION AND DECISION

It is convenient to divide the approach to patients with conjugated hyperbilirubinemia by age, focusing first on those less than 8 weeks old and then on those who are older. A suitable framework for the evaluation of the infant in the first 8 weeks of life focuses on duration of symptoms and mode of presentation. The most important considerations in the evaluation of the older patient are the presence of chronic disease, predisposition to biliary tract disturbances, and exposure to contagion and hepatotoxins (Fig. 42.1).

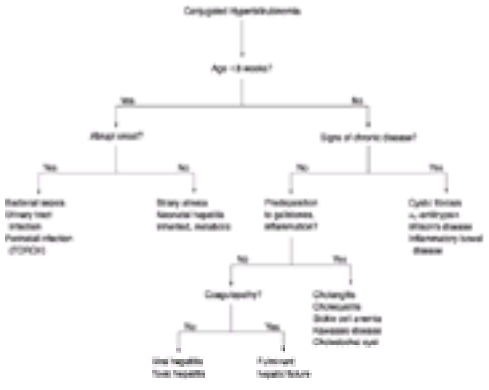

FIGURE 42.1. Approach to the patient with conjugated hyperbilirubinemia.

Infants Less Than 8 Weeks Old

The onset of conjugated hyperbilirubinemia in the first 2 months of life may be abrupt or insidious. Infants with generalized viral infections acquired in utero and during the birthing process are more likely to present shortly after birth. Perinatal cytomegalovirus (CMV) infection, rubella, toxoplasmosis, herpes simplex, and syphilis may account for irritability, jitteriness, seizures, microcephaly, hepatomegaly, icterus, and petechiae.

The diagnosis of sepsis or urinary tract infection should be considered in any infant who abruptly develops icterus. Hyperbilirubinemia may occur antecedent to blood cultures becoming positive and may precede findings of anorexia, vomiting, abdominal distension, fever, hepatomegaly, or alterations in respiratory pattern or sensorium. The precise mechanism of jaundice that complicates these infections is not completely understood. Sepsis, if accompanied by hepatic hypoperfusion, can be associated with prolonged biochemical and histologic changes within the liver. Similarly, children with prolonged congestive heart failure or postoperative repair of congenital heart disease may develop jaundice and abnormalities in biochemical tests of liver function. This is called ischemic hepatitis.

Infants with intrahepatic and extrahepatic biliary atresia have a failure of bile secretion associated with a significant hyperbilirubinemia. Although symptoms vary in onset, they usually are less acute than those seen in the infectious states. Infants affected with atresia generally appear well, with the exception of jaundice and hepatomegaly. Infants with

complete obstruction pass clay-colored stools. Failure to thrive may be a delayed manifestation.

The characteristic clinical pattern with idiopathic cholestasis consists of onset of jaundice in the second or third week of life. Initially, stool color is normal, but the stools may become acholic after several weeks. The presence of acholic stools may make it difficult to differentiate between obstructive jaundice caused by hepatocellular disease and that caused by obstruction of the biliary tree.

The inherited and metabolic conditions (e.g., galactosemia, tyrosinemia, cystic fibrosis; a_1-antitrypsin deficiency) associated with conjugated hyperbilirubinemia are typically insidious in onset. Although of a diverse nature, these diseases largely are characterized by inconstant jaundice, failure to thrive, developmental delay, and metabolic derangements. Unexplained fatality in the sibship or unexplained pulmonary, gastrointestinal, neurologic, or psychiatric disturbance in other family members may provoke diagnostic consideration.

The priorities for the emergency physician are to diagnose medically treatable infections, to identify metabolic disorders for which effective therapy is available, and to detect extrahepatic obstructive lesions that are amenable to surgical correction. The evaluation begins with cultures of cerebrospinal fluid, blood, urine, and stool. Infants also should have complete blood and platelet counts and should be tested for prothrombin time, hepatic enzymes (AST, ALT, and GGT), ammonia, albumin, total protein and protein electrophoresis, alkaline phosphatase, electrolytes, blood urea nitrogen, creatinine, and blood sugar. Urine should be tested for reducing substances. Additional studies useful for the longitudinal care physician include sweat iontophoresis, a_1-antitrypsin, TORCH and HBV serology, immunoglobulin M (IgM), urine examination for CMV, red blood cell galactose1-phosphate uridyltransferase activity, stool examinations, abdominal ultrasonography, and hepatobiliary scintigraphy.

Inpatient observation is appropriate in this age group because the diagnosis rarely can be established in the emergency department (ED). Empiric therapy for sepsis or urinary infection often is warranted, pending culture results.

Children More Than 8 Weeks Old

In the evaluation of conjugated hyperbilirubinemia beyond infancy, it is necessary to know whether there has been exposure to contagion or a potential for sexual or vertical transmission of infections such as hepatitis or human immunodeficiency virus (HIV). Other risk factors for hepatitis (e.g., needle sticks, hemodialysis, transplant, transfusion of blood products or factor use) need to be excluded. The physician should pursue possible exposure to industrial toxins or foods previously implicated in hepatic injury (e.g., carbon tetrachloride, yellow phosphorus, tannic acid, alcohol, mushrooms of the Amanita species). The emergency physician must inquire about use of acetaminophen, salicylates, erythromycin estolate, ceftriaxone, rifampin, iron salts, nitrofurantoin, oxacillin, methimazole, tetracycline, trimethoprim–sulfamethoxazole, diphenylhydantoin, isoniazid, and para-aminosalicylic acid (PAS). The presence of prior episodes of jaundice, acholic stools, and/or abdominal pain may suggest an underlying disorder, predisposing the patient to obstruction of the biliary tree. Other historical points include the presence of fever, arthralgia, arthritis, conjunctivitis, rash, pruritus, vomiting, diarrhea, weight loss, color of the urine, abnormal bruising or spontaneous bleeding, and changes in mental status.

An examination that focuses on ongoing physical signs of liver disease may result in greater accuracy in clinical evaluation of the older jaundiced patient. These signs include skin changes (spider angiomata, excoriations, palmar erythema) and peripheral edema. The abdominal examination should include observations of the venous pattern, presence of ascites, mass, or peritoneal irritation. There should be an estimation of liver size, contour, and tenderness, as well as an estimate of spleen size. The clinician should exclude cardiovascular dysfunctions such as hypoxemia, systemic venous congestion, and low cardiac output. Observations should be made of mental status and neuromuscular changes.

Patients with cystic fibrosis, a_1-antitrypsin deficiency, Wilson's disease, or inflammatory bowel disease tend to have symptoms that remit and relax. However, slow progression is the rule. Patients with a_1-antitrypsin deficiency may have onset of respiratory or hepatic complaints at any age. Similarly, infants who have failure to thrive from cystic fibrosis may develop obstruction at any age in the extrahepatic or intrahepatic ducts and, transiently or persistently, may exhibit jaundice. Patients with ulcerative colitis and Crohn's disease may become symptomatic intermittently with episodes of cholestasis. The degree of hepatic derangement and expression of neurologic abnormality is variable with Wilson's disease. Patients typically exhibit dysarthria, tremors, rigidity, or psychic disturbances before the diagnosis is entertained. Rarely, patients at a younger age without prodromal events have acute jaundice and hepatomegaly and progress to hepatic failure.

Biliary calculi and acute inflammation of the gallbladder are uncommon causes of conjugated hyperbilirubinemia in the pediatric population. However, a subset of patients is predisposed to these complications. Cholelithiasis may complicate any of the hemolytic anemias, particularly in patients with sickle hemoglobinopathies. These patients have increased incidence of both liver and gallbladder disease. Liver or gallbladder dysfunction accounts for the jaundice when more than 10% of an elevated bilirubin in a patient with sickle cell disease is conjugated. Cholecystitis may accompany a variety of acute focal infections, such as pneumonia or peritonitis, and may occur in the course of bacterial sepsis. In this event, shock and hyperpyrexia may divert the clinician from the deranged biliary system. In less severe cases, fever, nausea, vomiting, abdominal distension, and right upper quadrant pain are prominent features of cholecystitis. Right upper quadrant abdominal mass, pain, and jaundice constitute the classic triad in the diagnosis of choledochal cyst. The clinical recognition may be delayed until there is a complication, such as cholangitis. An acute, painful right upper quadrant mass associated with jaundice also may occur in the course of acute hydrops of the gallbladder from Kawasaki disease or systemic streptococcal infection.

In the previously healthy child, the most common cause of conjugated hyperbilirubinemia is acute hepatitis. The illness may be abrupt in onset, with fever, urticaria, and arthralgia as primary manifestations. More often, the illness is insidious. Viral hepatitis is characterized by low-grade fever and gastrointestinal complaints such as anorexia, malaise, nausea,

vomiting, and abdominal pain before the jaundice. Liver enlargement with hepatitis (A, B, C, and non-A, non-B, non-C), varicella, herpesvirus, coxsackievirus, echovirus, Epstein-Barr virus, and adenovirus infection is inconstant. Hepatic tenderness is a more reliable finding. Rarely, ascites can accompany hepatitis virus infection. Splenomegaly is the rule with Epstein-Barr virus but is unusual with the other agents. On occasion, hepatitis may be associated with a distinctive erythematous papular eruption localized to the limbs (Gianotti disease).

Toxic hepatitis, unlike viral hepatitis, does not have a prolonged prodrome. Acute nausea, vomiting, and malaise are followed in 1 to 2 days by alterations in mental status and deterioration of liver function. Most patients with toxic hepatitis will have an identifiable exogenous precipitant. Children with fulminant hepatic failure typically experience anorexia, nausea, vomiting, malaise, and fatigue—all symptoms indistinguishable from those expected with viral hepatitis. The patient's jaundice becomes more profound, and vomiting becomes protracted. Hyperexcitability, mania, and subtle psychomotor abnormalities may be seen. Coagulopathy, ascites, and sudden decrease in liver size are often the prelude to the development of frank neuromuscular signs.

The objectives of the emergency physician are to render supportive care to those icteric patients with infectious and metabolic derangements and to identify those cases in which jaundice is caused by mechanical obstruction or hepatic failure. The impression based on a targeted history, physical examination, and clinical algorithms can be bolstered with the following laboratory examinations: complete blood count, platelet count, prothrombin time, total and direct bilirubin, transaminase, alkaline phosphatase, electrolytes, blood urea nitrogen, and creatinine. Urinalysis, culture, and toxicologic screen should be considered. Chest and abdominal radiographs are indicated when there are pulmonary parenchymal complaints or significant abdominal findings. Other laboratory tests that are often available immediately and that may provide useful information in specific circumstances are serum ammonia, albumin, total protein, cholesterol, pH, and carbon dioxide. If available, abdominal sonography or computed tomography may be helpful occasionally. In no circumstance will results of several important blood and urine tests be of immediate use. Such studies, which are appropriate, include serum for bile acids, ceruloplasmin, protein electrophoretic pattern, serologic evidence of recent infection (e.g., Ebstein-Barr virus, mycoplasmal or hepatitis profiles), polymerase chain reaction assays, and enzyme-linked immunosorbent assay and autoantibody test. Urinary analysis includes assessment of organic acids and copper. These investigations may be helpful, however, to the longitudinal caretaker who must maintain a vigilant watch over the jaundiced patient.

Children more than 8 weeks old with conjugated hyperbilirubinemia should be admitted to the hospital at the time of their presentation in all cases in which life-threatening conditions may exist (Table 42.3). Inpatient treatment also is suggested when intravenous fluids are necessary to treat symptomatic hypoglycemia or electrolyte imbalance and when operative intervention may prove necessary. Icteric patients who have been diagnosed previously with confidence and who have exacerbation of their symptoms may require admission to reappraise their status. The physician also may be influenced to admit the patient when poor parenting or geographic barriers inhibit consistent observations. Admission also is indicated for patients who require further diagnostic intervention, such as liver biopsy or scintigraphy, to arrive at a definitive diagnosis.

Suggested Readings

Fitzgerald JF. Cholestatic disorders of infancy. Pediatr Clin North Am 1988;35:357.

Franson TR, Hierbolzer WJ Jr, LaBrecque DR. Frequency and characteristics of hyperbilirubinemia associated with bacteremia. Rev Infect Dis 1985;7:1.

Flynn PM. Hepatitis C infections. Pediatr Ann 1996;25:496.

Gartner LM. Neonatal jaundice. Pediatr Rev 1994;15:422.

Grand RJ, Ulshen MH. Clinical and physiological abnormalities in hepatic function. Pediatr Clin North Am 1975;22:897.

Lai MW, Chang MH, Hsu HY. Non-A, non-B, non-C hepatitis: its significance in pediatric patients and the role of BG virus-C. Pediatrics 1997;131:536.

Maisels MJ, Kring E. Risk of sepsis in newborns with severe hyperbilirubinemia. Pediatrics 1992;90:741.

Mathis RK, Andres JM, Walker WA. Liver disease in infants. J Pediatr 1977;90:864.

Newman TB, Easterling J, Goldman ES, et al. Laboratory evaluation of jaundice in newborns. Am J Dis Child 1990;144:364.

Nowicki MJ, Balistreri WF. Hepatitis A to E: building up the alphabet. Contemp Pediatr 1992;9:118.

Nowicki MJ, Balistreri WF. The Cs, Ds, and Es of viral hepatitis. Contemp Pediatr 1992;9:23.

Persaud D, Bangaru B, Greco MA, et al. Cholestatic hepatitis in children infected with the human immunodeficiency virus. Pediatr Infect Dis J 1993;12:492.

Walker WA, Mathis RC. Hepatomegaly: an approach to differential diagnosis. Pediatr Clin North Am 1975;22:929.

CHAPTER 43

Limp

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Limping is a common complaint in the pediatric acute-care setting. A *limp* is defined as an alteration in the normal walking pattern for the child's age. The average child begins to walk between 12 and 18 months of age with a broad-based gait, gradually maturing into a normal (adult) gait pattern by the age of 3 years. Normal walking should appear smooth and effortless, although it involves a complex integration of the nervous and musculoskeletal systems. A normal gait cycle can be divided into two phases: stance and swing. The stance phase, the time from the heel striking the ground to the toe leaving the ground, encompasses about 60% of the gait cycle. The swing phase involves a sequence of hip then knee flexion, followed by foot dorsiflexion and knee extension as the heel strikes the ground to begin the next cycle.

The causes of limping are numerous, ranging from trivial to life-threatening, but most children who limp do so as a result of pain, weakness, or deformity. Pain results in an antalgic gait pattern with a shortened stance phase. The most common causes of a painful limp are trauma and infection. Neuromuscular disease may cause either spasticity (e.g., toe-walking) or weakness, which results in a steppage gait to compensate for weak ankle dorsiflexion. The Trendelenburg gait, characterized by a pelvic tilt away from the affected hip, is common in congenital or acquired hip disorders. A vaulting gait may be seen in children with limb-length discrepancy or abnormal knee mobility. A stooped, shuffling gait is common in patients with pelvic or lower abdominal pain.

The evaluation of a child with a limp demands a thorough history and physical examination, using an age-based approach. Toddlers generally provide the greatest diagnostic challenge because a history of trauma may be unclear, and the ability to describe or localize pain may be lacking. In all age groups, a detailed history of the circumstances surrounding the limp should be obtained, with focus on the issues of trauma, pain, and associated fever or systemic illness. The physical examination must be complete because limping may originate from abnormalities in any portion of the lower extremity, nervous system, abdomen, or genitourinary tract. The location of the pain may not represent the source of the pathology; for example, hip pain may be referred to the knee area. Laboratory and imaging studies should be tailored to the findings in the history and physical examination, keeping in mind an appropriate age-based differential diagnosis.

DIFFERENTIAL DIAGNOSIS

The extensive differential diagnosis of the child with a limp may be approached from several angles: by disease category, location of pathology, or age of the child. Table 43.1 presents the differential diagnosis by disease category; Table 43.2 organizes the differential diagnosis by age and location of pathology. The most common causes of limp are outlined in Table 43.3, and potentially life- or limb-threatening conditions are listed in Table 43.4. This section reviews the differential diagnosis within the framework of an algorithmic approach (Fig. 43.1).

FIGURE 43.1. Algorithmic approach to the child with a limp. *RSD,* reflex sympathetic dystrophy; *CBC,* complete blood count; *SCFE,* slipped capital femoral epiphysis; *AVN,* avascular necrosis; *DDH,* developmental dysplasia of the hip; *LCP,* Legg-Calvé-Perthes disease.

Trauma or Overuse	*Congenital*
Fracture	Vertical talus
Stress fracture	Tarsal coalition
Soft-tissue injury	Other congenital limb abnormalities
Spondylolisthesis	Spinal dysraphism
Herniated nucleus pulposis	Inguinal hernia
Infectious	*Neurologic*
Septic arthritis	Muscular dystrophy
Osteomyelitis	Peripheral neuropathy
Lyme arthritis	Reflex sympathetic dystrophy
Cholitis	*Neoplasia*
Pelvic inflammatory disease	Benign bone tumors
Inflammatory	Malignant bone tumors
Transient synovitis	Leukemia
Reactive arthritis	Intra-abdominal tumors
Rheumatic disease	Sacral tumors
Appendicitis	Spinal cord tumors
Developmental or Acquired	*Metabolic*
Developmental dysplasia of the hip	Rickets
Blount's disease	Hyperparathyroidism
Limb length discrepancy	*Hematologic*
Torsional deformities	Sickle cell disease
Avascular necrosis	Hemophilia
Slipped capital femoral epiphysis	
Tarsal coalition	

Table 43.1. Differential Diagnosis of Limp by Disease Category

	Long Bone	Skin/Soft Tissue	Any Joint	Hip	Knee	Ankle/Foot	Spine
Toddler	Fracture Toddler's Salter type I Periostitis Osteomyelitis Vaso-occlusive crisis Congenital anomaly	Contusion Strain Foreign body Immunization Infection	Septic arthritis Reactive arthritis Rheumatic disease Systemic JRA Hemarthrosis	Transient synovitis DDH	Occult trauma Blount's disease Referred hip pain	Poor shoe fit Occult trauma Vertical talus Kohler's disease	Dysraphism Infection Tumor
School age	Fracture Salter type I Discrepant limb length Osteomyelitis Tumor Vaso-occlusive crisis	Contusion Strain Myositis Growing pains Infection	Sprain Reactive arthritis Rheumatic disease SLE, HSP, RAF Septic arthritis Lyme arthritis	Transient synovitis JRA	Baker's cyst Referred hip pain	Poor shoe fit Salter type I fracture Tarsal coalition Kohler's disease	Dysraphism Infection Tumor
Adolescent	Fracture Osteomyelitis	Contusion Strain PFPS	Sprain Reactive arthritis IBD, SLE Septic arthritis Gonococcal Lyme arthritis	SCFE Legg-Calvé-Perthes disease Osteochondritis dissecans Chondromalacia Baker's cyst Referred hip pain	Osgood-Schlatter's disease Osteochondritis dissecans Chondromalacia Baker's cyst Referred hip pain	Poor shoe fit Hallux rigidus Bunion Freiberg's disease Sever's disease	Scoliosis Herniated disc Infection Tumor

JRA, juvenile rheumatoid arthritis; DDH, developmental dysplasia of the hip; EM, erythema multiforme; NSAID, nonsteroidal anti-inflammatory drug; AVN, avascular necrosis; RSD, reflex sympathetic dystrophy; IBD, inflammatory bowel disease; SLE, systemic lupus erythematosus; SCFE, slipped capital femoral epiphysis.

Table 43.2. Differential Diagnosis of Limp by Age and Location of Pathology

Trauma	*Rheumatic Disease*
Fracture	*Other Hip Disorders*
Soft-tissue injury	Developmental dysplasia
Overuse injuries	Legg-Calvé-Perthes disease
Transient Synovitis	Slipped capital femoral epiphysis
Infection	
Septic arthritis	
Osteomyelitis	

Table 43.3. Common Causes of Limp

Septic arthritis	Slipped capital femoral epiphysis
Osteomyelitis	Epidural abscess
Tumor	Appendicitis
Developmental dysplasia of the hip	

Table 43.4. Life- or Limb-Threatening Causes of Limp

The most common cause of limping in all ages is trauma, either acute or repetitive microtrauma (stress fractures). Older children who limp as a result of trauma can generally describe the mechanism of injury and localize pain well. The toddler and preschool age groups, with their limited verbal ability and cooperation skills, often provide a diagnostic challenge. A common type of injury in this population (often not witnessed) is the aptly named “toddler's fracture,” a nondisplaced spiral fracture of the tibial shaft that occurs as a result of torsion of the foot relative to the tibia. Occult fractures of the bones in the foot also occur in young children. Initial plain radiographic findings may be subtle, or at times nonexistent, but will become apparent in 1 to 2 weeks. Bone scans will identify these lesions sooner. Another fracture often lacking initial radiographic confirmation is a Salter-Harris type I fracture, which presents as tenderness over a physis after trauma to a joint area. Stress fractures may also lack overt radiographic findings. Common sites for overuse injury include the tibial tubercle (Osgood-Schlatter's disease), the anterior tibia (“shin splints”), and the calcaneous at the insertion of the

Achilles tendon (Sever's disease). More information on the subject of fractures is found in Chapter 115.

Trauma may also induce limping as a result of soft-tissue injury. Although children are more likely to sustain fractures than sprains and strains, the latter can occur. Joint swelling and pain out of proportion to the history of injury raises the possibility of a hemarthrosis as the initial presentation of a bleeding disorder (see Chapter 87). Severe soft-tissue pain and swelling in the setting of a contusion or crush injury suggests compartment syndrome. With compartment syndrome, pain is exacerbated by passive extension of the affected part; pallor and pulselessness are late findings. Severe pain of an entire limb out of proportion to the history of injury suggests reflex sympathetic dystrophy (RSD). RSD is most common in young adolescent girls. It may be accompanied by mottling and coolness of the extremity, presumably as a result of abnormalities in the peripheral sympathetic nervous system.

A limp that is accompanied by a history of fever or recent systemic illness is likely to be infectious or inflammatory in origin. However, the absence of fever does not preclude the possibility of a bacterial bone or joint infection, and many infections are preceded by a history of minor trauma. Septic arthritis is the most serious infectious cause of joint pain and limp. It is more common in younger children and typically presents with a warm, swollen joint. Exquisite pain with attempts to flex or extend the joint is characteristic of septic arthritis, and the degree of pain with motion serves as a helpful clinical sign in distinguishing bacterial joint infection from inflammatory conditions. A common diagnostic challenge is differentiating septic arthritis from transient (or toxic) synovitis in a young child with fever, limp, and pain localized to the hip. Transient synovitis, a postinfectious reactive arthritis, generally follows a milder course. It is usually preceded by a recent viral respiratory or gastrointestinal illness. Acute-phase reactants may be elevated in both conditions, although usually less so in synovitis. A joint effusion, which is better visualized with ultrasound than plain films, may be present in both. Orthopedic consultation for joint aspiration may be required for a definitive diagnosis because a septic hip is a surgical emergency requiring open drainage. Osteomyelitis is another potentially serious infectious cause of limp, although the presentation is typically more chronic than that of a septic joint. Osteomyelitis, which is also more common in younger children, presents with pain and occasionally warmth and swelling, usually over the metaphysis of a long bone. A reactive joint effusion may be present. Occasionally, osteomyelitis and septic arthritis will coexist. More detailed discussions of both septic joint and osteomyelitis are found in Chapter 84 and Chapter 123.

Rheumatic conditions that may result in limp are numerous; many are accompanied by systemic symptoms and characteristic skin rashes. Examples include Lyme disease, Henoch-Schönlein purpura, erythema multiforme, acute rheumatic fever, juvenile rheumatoid arthritis, and systemic lupus erythematosus. Occasionally, limping from arthralgia will precede the development of the arthritis and systemic involvement. An approach to the child with joint pain is found in Chapter 57, and a detailed discussion of arthritis is found in Chapter 101.

In the absence of obvious trauma, fever, or systemic symptoms, the next step in the approach to the differential diagnosis of a limp is to determine the focality of the findings and the degree of pain. Localized pain suggests repetitive microtrauma, bone tumor, or an acquired skeletal deformity. Repetitive microtrauma may be responsible for avascular necrosis of the foot bones in two locations: the tarsal navicular bone (Köhler's disease) in younger children and the metatarsal heads (Freiberg's disease) in adolescents. Both benign and malignant bone tumors may present with a painful limp. Benign lesions include bone cysts (unicameral or aneurysmal), fibrous dysplasia, and eosinophilic granulomas. Osteoid osteoma, caused by a painful nidus of vascular osteoid tissue, is another benign lesion unique to young people. The most common malignant pediatric bone tumors are osteogenic sarcoma and Ewing's sarcoma. Bone tumor pain may be acute or chronic, with acute pain usually related to a pathologic fracture. Examples of acquired skeletal abnormalities causing painful limp include tarsal coalition and osteochondritis dissecans. Tarsal coalition occurs as a result of gradual calcification of a congenital cartilaginous bar between tarsal bones; it presents most commonly as a painful flatfoot in school-aged children. Osteochondritis dissecans is related to separation of articular cartilage from underlying bone; it most commonly affects the knees of adolescent boys.

Localized findings without pain suggest congenital or slowly developing acquired limb abnormalities. Three disorders of the hip fit into this category, each of which is characteristic of a specific age group. Developmental dysplasia of the hip (DDH) includes a spectrum of abnormalities ranging from mild dysplasia to frank dislocation. Most affected children with access to primary care are diagnosed with abnormal hip abduction on routine examination in infancy. Occasionally, the diagnosis will be missed, and the child then presents at the onset of walking with a painless short-leg limp, or waddling gait if bilateral, with weakness of the abductor musculature. Legg-Calvé-Perthes (LCP) disease, an avascular necrosis of the capital femoral epiphysis, presents in young school-aged children as an insidious limp with mild, activity-related pain. Slipped capital femoral epiphysis (SCFE) presents in young, typically obese, adolescents with an externally rotated limp. The amount of pain experienced is related to the rate of displacement of the epiphysis, ranging from none to severe. LCP and SCFE are more common in boys. Other acquired skeletal deformities that may cause painless limp include limb length inequality, Blount's disease (with marked bowing of the proximal tibias), and torsional deformities. Baker's cyst of the popliteal tendon may cause limping with minimal local discomfort.

Limping in the absence of localized limb findings suggests a systemic (or nonlimb) source such as the spine or abdomen. A painful limp without localization or with migratory bone pain suggests a hematologic or oncologic cause such as sickle cell disease or leukemia. Limping with bilateral leg pain localized to the muscles, especially the calves, suggests myositis. Benign acute childhood myositis is common during influenza epidemics. Recurrent diffuse aches after periods of vigorous activity, usually worse at night, suggest benign hypermobility syndrome or “growing pains.” A painless, poorly localized limp may occur with metabolic bone disease (e.g., rickets). Spinal problems that can cause leg pain, weakness, or limp include dysraphism, vertebral infection, spondylolisthesis, and herniated disc. *Spinal dysraphism* refers to a spectrum of abnormalities in the development of the spinal cord and vertebrae ranging from obvious (myelomeningocele) to occult (tethered cord). Associated neurologic and musculoskeletal findings, including pain, atrophy, high arches, and tight heel cords, may develop in early childhood. Vertebral infection typically presents with fever and back pain. Spondylolisthesis and herniated disc are rare in young children but may be seen in adolescents who complain of back pain or radicular pain. Intra-abdominal pathology that can result in limp includes appendicitis, pelvic or psoas abscess, and renal disease. Solid tumors, most commonly neuroblastoma, can cause limp through retroperitoneal irritation or extension into the spinal canal. Likewise, a sacral teratoma may affect the nerves of the cauda equina or sacral plexus. Testicular pain may present with limping in a boy who is reluctant or embarrassed to admit the true source of his

discomfort.

EVALUATION AND DECISION

The conditions that lead to a presentation of limp range from mundane (poorly fitting shoes) to life-threatening (leukemia). The role of the pediatric acute-care physician is to rule out the possibility of life- and limb-threatening pathologic conditions. The serious conditions include bacterial infection of the bone or joint space, malignancy, and disorders that threaten the blood supply to the bone, such as avascular necrosis (AVN) and SCFE. Often, a definitive diagnosis will not be reached in the emergency department, and the patient will require follow-up with the primary care physician or specialist. Figure 43.1 provides an algorithmic approach to the child with a limp.

History

The history in a limping child should include information about the onset and duration of the limp, the family's perception of the origin of the problem, and associated symptoms such as pain, fever, and systemic illness. When pain is present, the physician should inquire about the location and severity. A history of trauma should be addressed, keeping in mind the inherent difficulty in obtaining an accurate trauma history in very young children. Conversely, obvious trauma in the absence of a consistent history raises the question of inflicted injury. In more chronic presentations, any cyclical or recurrent patterns should be noted. Stiffness and limp primarily in the morning suggest rheumatic disease, whereas evening symptoms suggest weakness or overuse injury. A history of joint or limb swelling should be investigated, with attention to the degree of swelling as well as any migratory or recurrent patterns.

The medical history should include birth and developmental history. Breech position is associated with DDH, and mild cerebral palsy may present in childhood with abnormal gait. History of viral infections, streptococcal pharyngitis, medication use, and immunizations may provide clues to the cause of limping. A family history of rheumatic or autoimmune disease, inflammatory bowel disease, hemoglobinopathy, or other bleeding disorders may help facilitate diagnosis. Finally, the review of systems should include questions about past trauma, infections, neoplasia, endocrine disease, metabolic disease, and congenital anomalies.

Physical Examination

The physical examination in a limping child should begin with observation of the child's gait. Ideally, the child should be observed walking in bare feet and wearing minimal clothing, preferably in a long hallway. The physician should attempt to observe the child unobtrusively to avoid gait changes caused by self-consciousness. The observer should note the symmetry of stride length, the proportion of the gait cycle spent in stance phase, hip abductor muscle strength (with abnormal strength manifested by Trendelenburg or waddling gait), in-toeing or out-toeing, and joint flexibility. Muscle strength may be tested by asking the child to run, hop, and walk on toes and heels.

After observing the child in action, the physician should perform a complete examination with attention to the musculoskeletal and neurologic systems. The musculoskeletal examination begins with inspection of the limbs and feet for swelling or deformity. The spine should be inspected for curvature, both standing and bending forward, and the soles of feet and toes should be checked for foreign bodies and callouses. The bones, muscles, and joints should be palpated for areas of tenderness; range of motion of all joints should be checked; and limb lengths (from anterior superior iliac spine to medial malleolus) and thigh and calf circumference should be measured for asymmetry. The neurologic examination should include inspection of the spine for lumbosacral hair or dimple (indicating possible spinal dysraphism), and testing of strength, sensation, and reflexes. The abdomen and external genitalia should be examined for tenderness or masses, and the skin for rashes. A rectal examination may be indicated if sacral pathology is suspected. Finally, wear patterns on the child's shoes may provide clues to the nature and duration of the limp.

Laboratory and Imaging

Plain radiographs remain a mainstay of the workup of a limping child. They provide an excellent means of screening for fracture, effusion, lytic lesions, periosteal reaction, and avascular necrosis. In a child with an obvious focus of pain, the radiographs may be obtained with views specific to that area, noting that children with knee pain may have hip pathology. The need for comparative views (of the normal extremity) depends on the experience of the physician interpreting the films. Some radiographic findings can be subtle, and comparison with the opposite side may be helpful. In a young child or a child lacking obvious focus for the limp, anteroposterior (AP) and lateral views of both lower extremities (including the feet) should be ordered as an initial screen. In toddlers lacking a focus of pain and in older children in whom hip pathology is suspected, AP and frog-leg lateral views of the pelvis are required. The frog-leg lateral view, obtained with the hips abducted and externally rotated, allows excellent visualization of the femoral heads. These radiographs should always include both hips to enable comparison of the femoral heads and width of the joint spaces. Radiographs of the spine are necessary if the child has neurologic signs or symptoms.

In children whose limp is associated with fever or systemic illness, laboratory studies, including a complete blood count (CBC) and an erythrocyte sedimentation rate (ESR), are indicated. In some institutions, a C-reactive protein (CRP) is obtained as a more sensitive acute-phase reactant. These studies serve as screens for infection, inflammation, malignancy, and hemoglobinopathy. Laboratory studies are also indicated in the absence of fever if the child has been limping for several days without evidence of trauma on plain films. Children with evidence of infection or inflammation with a joint effusion may require arthrocentesis for definitive diagnosis. In areas of endemic Lyme disease, a Lyme titer is a reasonable initial screening test in a patient with arthritis. A creatine phosphokinase (CPK) level may be helpful if muscle inflammation is suspected.

When the initial history, physical examination, imaging, and laboratory evaluation indicate the cause of the limp, specific treatment can be initiated. Abnormalities in the initial workup without a definitive diagnosis should prompt further imaging or laboratory studies. Bone scintigraphy is more sensitive than plain radiographs for occult fracture, infection, avascular

necrosis, and tumor; however, it is not specific for a given pathologic process. Computed tomography (CT) is an excellent imaging modality for cortical bone; it serves as a useful diagnostic adjunct in certain fractures, bony coalitions, and bone tumors. Ultrasound (US) is the preferred modality for diagnosing hip effusions; it is also useful for guiding needle aspirations of the hip joint. Magnetic resonance imaging (MRI) is useful in imaging the spinal cord, avascular necrosis, and bone marrow disease.

If the initial workup in a limping child is completely normal, including screening radiographs and laboratory studies, the child may be followed closely as an outpatient. These children should be examined every few days until improvement is noted or a cause is determined. If the limp persists beyond 1 to 2 weeks without a diagnosis, further workup or consultation with a specialist is indicated.

Suggested Readings

Clark MC. The limping child: meeting the challenges of an accurate assessment and diagnosis. Pediatr Emerg Med Reports 1997;2:123–134.

Dabney KW, Lipton G. Evaluation of limp in children. Curr Opin Pediatr 1995;7:88–94.

Del Beccaro MA, Champoux AN, et al. Septic arthritis versus transient synovitis of the hip: the value of screening laboratory tests. Ann Emerg Med 1992;21:1418–1422.

Myers MT, Thompson GH. Imaging the child with a limp. Pediatr Clin North Am 1997;44:637–658.

Renshaw TS. The child who has a limp. Pediatr Rev 1995;16:458–465.

Tenenbein M, Reed MH, Black GB. The toddler's fracture revisited. Am J Emerg Med 1990;8:208–211.

CHAPTER 44

Lymphadenopathy

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Lymphadenopathy is defined as swelling of the lymph nodes. Swollen lymph nodes are a common presenting sign in children, mainly because children have more pronounced lymphoid responses to inflammation than adults do, and they also have relatively more lymphoid tissue. Because the differential diagnosis of lymphadenopathy is extensive, it is helpful to distinguish localized from generalized lymphadenopathy. *Localized, or regional, adenopathy* generally occurs in response to a focal infectious process, although rarely other causes may need to be considered. Because a large number of organisms can cause localized adenopathy, it is often helpful to differentiate between acute and subacute/chronic regional adenopathy. *Generalized lymphadenopathy* is defined as enlargement of more than two noncontiguous lymph node regions. The most common causes of generalized adenopathy are systemic infections (bacterial or viral), autoimmune diseases, and neoplastic processes.

DIFFERENTIAL DIAGNOSIS

Acute Regional Adenopathy

The clinician caring for a child with acute regional adenopathy will benefit from knowledge of the anatomic distribution of nodes in the area and their drainage areas, as described in Table 44.1. The location of lymphadenopathy is often suggestive of a possible cause. For instance, in the head and neck region, swollen nodes are often a response to focal infectious processes occurring in areas that drain in the region of the nodes. Occipital nodes most commonly enlarge in response to bacterial or fungal scalp infections or to chronic inflammation such as occurs in seborrheic dermatitis. Because preauricular nodes drain the conjunctiva and lateral eyelids, these often enlarge in viral conjunctivitis. Epidemic keratoconjuctivitis caused by adenoviruses often presents with an enlarged preauricular node. The combination of conjunctivitis and ipsilateral preauricular adenopathy is called oculoglandular syndrome, or Parinaud's syndrome. Another infection that can present as Parinaud's syndrome is chlamydial conjunctivitis, also called neonatal-inclusion conjunctivitis. Chlamydial conjunctivitis, which generally presents within 5 to 7 days after birth, is diagnosed by the finding of intracytoplasmic inclusion bodies in conjunctival scrapings or, more commonly, by detection of the pathogen by immunofluorescent staining of ocular secretions. Parinaud's syndrome is also occasionally seen in cat-scratch disease, tularemia, and listeriosis. Similarly, the presence of submaxillary and submental nodes points to the possibility of an infectious process in the oral cavity. Therefore, the physician should perform a careful oral and dental examination in these cases. Dental abscesses or gingival infections may be responsible for lymphadenopathy in these regions.

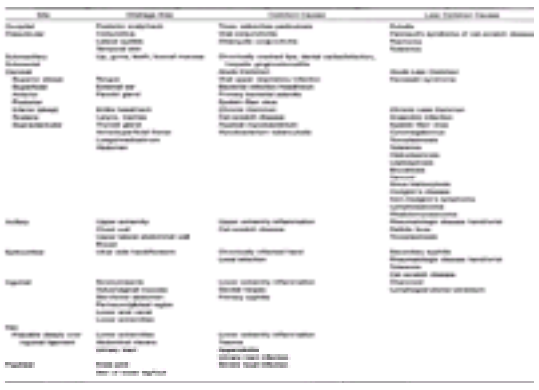

Table 44.1. Regional Adenopathy

The differential diagnosis of cervical adenopathy is more extensive, mainly because the anatomy of the region is more complex. As can be seen in Table 44.1, nodes in the cervical region can be divided into three areas: the superior deep nodes below the angle of the mandible, the superficial cervical nodes found anteriorly and posteriorly along the sternocleidomastoid muscle, and the inferior deep nodes at the base of the neck. Enlargement of superior deep or superficial nodes raises the possibility of a lingual, external ear, or parotid gland process. In contrast, the inferior deep

nodes have a much wider drainage area, including the head and neck, upper extremities, and the thoracic and abdominal regions. Swelling of these nodes, in particular scalene and supraclavicular nodes, can be the first sign of occult thoracic or abdominal pathology, such as malignancy. Therefore, nodes found in these regions must be investigated carefully, with thorough physical examinations and, if necessary, radiographic examinations.

By far, the most common cause of acute cervical adenopathy is a viral upper respiratory tract infection. In these cases, lymph nodes are generally symmetrically enlarged and are soft and minimally tender, if at all. The reactive adenopathy may persist for 2 to 3 weeks beyond the resolution of the viral illness, but there should be no progression in the size or extent of the adenopathy. Bacterial cervical adenitis is also a common cause of cervical lymphadenopathy in children, particularly in pre–school-age children. It is usually caused by group A b-hemolytic *Streptococcus* or *Staphylococcus aureus*, although anaerobes (usually penicillin-sensitive) may also be involved, particularly in oral infections. A history of a sore throat may be present in a minority of patients. Bacterial adenitis is most often unilateral and presents with firm, tender, and warm lymph nodes. In addition to the cervical area, other common sites of involvement include submaxillary, inguinal, and axillary nodes. If left untreated, these nodes may become erythematous and eventually fluctuant. Drainage of the nodes is sometimes required, even in appropriately treated cases (see Chapter 84).

Epstein-Barr virus (EBV), the agent of infectious mononucleosis, commonly causes posterior cervical lymphadenopathy in older children and adolescents. EBV infections do not always cause generalized adenopathy. The classic presentation of a child with EBV includes malaise, fever, an exudative tonsillopharyngitis, and hepatosplenomegaly. Facial edema may accompany significant EBV adenopathy, presumably reflecting obstructed lymph drainage. Younger children and infants with EBV infection may present less typically with fever alone or with symptoms suggestive of a mild upper respiratory infection. The diagnosis is made most easily with the detection of a positive heterophile agglutinating antibody (monospot), although it is important to remember that this test may be falsely negative in children under the age of 7 years.

A rarer, but important, cause of acute cervical lymphadenopathy is Kawasaki disease, a systemic febrile syndrome of as yet undefined cause (see Chapter 101. Kawasaki disease, also called mucocutaneous lymph node syndrome, occurs most often in children less than 4 years of age and is rare after 8 years of age. It is important to diagnose Kawasaki disease early because prompt treatment with intravenous gamma globulin can prevent coronary artery aneurysms, the most serious complications of this illness. The cervical lymphadenopathy in Kawasaki disease, seen in approximately 50 to 70% of patients, occurs during the early phase of the illness and may be unilateral or bilateral. The nodes are firm and mildly tender and should be at least 1.5 cm in diameter. The presence of a large node in the cervical area, in association with fever of greater than 5 days' duration, bilateral conjunctival injection with limbal sparing, mucous membrane involvement, peripheral edema or erythema, and a polymorphous truncal rash, should alert the physician to the possibility of this disorder.

Axillary adenopathy is commonly present with any infection or inflammation of the upper extremities. Most commonly, injuries to the hand, such as occur after falling or with puncture wounds or bites, may present with concomitant axillary adenopathy. Similarly, epitrochlear nodes, which are not normally palpable in children, may become inflamed after infections of the third, fourth, or fifth finger; medial portion of the hand; or ulnar portion of the forearm. Most commonly, these infections are caused by pyogenic bacteria (e.g., *S. pyogenes, S. aureus*), but depending on the inciting event, other pathogens may be responsible (e.g., *S. moniliformis, Spirillum minus* in rat-bite fever).

Inguinal adenopathy most often results from lower extremity infection, although sexually transmitted diseases may also be responsible. For example, acute genital infection with herpes simplex virus (HSV)-2 often presents with tender inguinal adenopathy, occasionally as the only sign. Similarly, chancroid, lymphogranuloma venereum, and syphilis may present with inguinal nodal swelling and tenderness. The presence of genital lesions, which may be either painful (as in HSV or chancroid) or painless (as in syphilis), offers clues to these diagnoses. Therefore, careful history taking and physical examination are necessary to exclude these possibilities. Enlarged iliac nodes are palpable deeply over the inguinal ligament and become inflamed with lower extremity infection, urinary tract infection, abdominal trauma, and appendicitis. Of note, iliac adenitis, which can present with fever, limp, and inability to fully extend the leg, may mimic the signs and symptoms of septic hip arthritis. Unlike in hip disease, however, hip motion is not limited on examination. Iliac adenitis may also be confused with appendicitis, but the pain initially occurs in the thigh and hip rather than in the periumbilical region or right lower quadrant.

Chronic Regional Adenopathy

Numerous agents can cause chronic regional lymphadenopathy. Organisms such as *Bartonella* (the etiologic agent of cat-scratch disease), mycobacteria, and atypical mycobacteria are most commonly responsible for chronic adenopathy. Cat-scratch disease, caused by *Bartonella henselae*, is a relatively common cause of chronic axillary or cervical adenopathy (see Chapter 84). Cat-scratch disease is characterized by a history of exposure to kittens (although other animals have also been implicated) and the development of a primary lesion at the site of a scratch. The primary lesion is 2 to 5 mm in size and is typically papular initially, and it may then progress to a pustule. About 2 weeks later, lymphadenopathy develops proximal to the site of the lesion. Typically, the nodes are enlarged but may or may not be inflamed. Lymphangitis does not occur in cat-scratch disease. Fever is present in only about 30% of patients. Other symptoms, such as seizures, may occur but are rare. The diagnosis of cat-scratch disease is confirmed by serology.

Tuberculous cervical lymphadenitis, otherwise known as scrofula, most commonly involves the posterior cervical nodes. Scrofula has become less common in the United States, although recent epidemiologic studies in this country have suggested that the incidence of tuberculosis is rising. A history of exposure to an individual with active tuberculosis is often elicited, and several family members may have positive skin tests. Pulmonary and other systemic symptoms, such as fever, fatigue, and weight loss, are often present. The affected nodes are typically bilateral, fixed, and matted. Fluctuance is a late and rare finding in tuberculous adenitis. The diagnosis of tuberculous cervical lymphadenitis is made by a combination of skin testing, chest radiographs, and if possible, culture data from the involved node.

In contrast, atypical mycobacterial adenitis usually involves young children, less than 5 years of age, and is generally unilateral. The node is rarely more than 3 cm in size. Overlying skin may turn a deep purple and gradually thins, developing a parchment-paper appearance. Fluctuance and ulceration occur commonly. Infected patients generally appear well, with a notable absence of any systemic symptoms. Chest radiographs are normal. A clear history of exposure to atypical mycobacteria (e.g., acquiring the infection via a fish tank) is the exception rather than the rule. Diagnosis is made by culture of the infected node. Treatment generally involves excision of the node, although recently reports of treatment with newer macrolides have suggested a possible role for antimicrobial therapy of these infections.

Other less common causes of chronic adenopathy deserve mention. A prolonged heterophile-negative adenopathy unresponsive to a trial of antibiotics should raise suspicion for one of these possibilities. Cytomegalovirus (CMV) infection, which is characterized by cervical adenopathy, pharyngitis, and atypical lymphocytosis, may cause prolonged adenopathy in younger children. Toxoplasmosis typically presents as a single, nontender posterior cervical node. Brucellosis, associated generally with axillary and cervical lymphadenopathy, and tularemia with cervical adenopathy, are rare infectious causes of chronic adenopathy in children.

Noninfectious etiologies may also cause chronic regional adenopathy. Various malignancies, such as Hodgkin's disease, lymphosarcoma, neuroblastoma, and rhabdomyosarcoma, may all present with chronic cervical lymphadenopathy (see Chapter 100). For example, Hodgkin's disease usually presents as a slowly growing, painless firm node in the upper third of the neck. Lymphosarcoma also presents as a firm painless node, but it occurs in younger children than those with Hodgkin's and more commonly involves extranodal sites such as tonsils. Rhabdomyosarcoma, the most common solid tumor of the head and neck in children, often involves the nasopharynx, middle ear, mastoid, or orbit, but it can also occur as a painless mass anywhere in the head and neck.

In African-American children, sarcoidosis must be entertained in a child with bilateral chronic cervical adenopathy. Scalene nodes are involved in more than 80% of cases. An abnormal chest film, with hilar adenopathy and peribronchial fibrosis, suggests sarcoidosis. Sinus histiocytosis, a benign form of histiocytosis, can present as a large painless cervical adenopathy. The clinical presentation often includes fever, anemia, leukocytosis, and elevated erythrocyte sedimentation rate. Although the initial clinical presentation may be confused with lymphoma, the disease usually has a benign course, with resolution over a prolonged period.

Generalized Lymphadenopathy

Various systemic illnesses are associated with generalized lymphadenopathy (Table 44.2). The most common causes of generalized lymphadenopathy include bacterial or viral illnesses that disseminate systemically. As an example, the high incidence of vomiting and abdominal pain in streptococcal pharyngitis has been attributed to abdominal node inflammation and swelling, suggesting a more systemic pattern of adenopathy in streptococcal disease. Rarer bacterial causes of generalized lymphadenopathy include bacterial illnesses such as brucellosis and leptospirosis, diagnoses that may be suggested by occupational or dietary history. Common viral causes of generalized adenopathy include EBV or CMV mononucleosis, rubella, and measles in parts of the world where the disease is endemic. Another cause of generalized adenopathy includes human immunodeficiency virus (HIV) infection. HIV infection in children can present with persistent generalized adenopathy, hepatosplenomegaly, and failure to thrive. Generalized lymphadenopathy may occasionally be the only presenting symptom in a child with vertical HIV infection.

Systemic Infection	*Autoimmune Disease*
Bacterial	Juvenile rheumatoid arthritis
Bacteremia	Systemic lupus erythematosus
Scarlet fever	Serum sickness
Subacute bacterial endocarditis	Autoimmune hemolytic anemia
Syphilis	*Primary Lymphoid Neoplasm*
Tuberculosis	Hodgkin's disease
Brucellosis	Non-Hodgkin's lymphoma
Viral	*Metastatic Neoplasm*
Varicella	Acute lymphocytic leukemia
Rubella	Acute myelogenous leukemia
Rubeola	Neuroblastoma
Epstein-Barr virus	*Histiocytosis*
Cytomegalovirus	Letterer-Siwe
Human immunodeficiency virus	Histiocytic medullary reticulosis
Fungal	*Storage Disease*
Histoplasmosis	Gaucher's disease
Coccidioidomycosis	Niemann-Pick's disease
Parasitic	*Drugs*
Toxoplasmosis	Phenytoin
Malaria	Isoniazid
	Miscellaneous
	Hyperthyroidism

Table 44.2. Generalized Adenopathy

Noninfectious systemic disease may also present with generalized adenopathy. Approximately 70% of patients with systemic lupus erythematosus (SLE) or juvenile rheumatoid arthritis (JRA) manifest generalized adenopathy during the acute phase of illness (see Chapter 101). The lymphadenopathy of serum sickness often occurs in the presence of the exanthem but may be seen without rash. The lymphadenopathy of autoimmune hemolytic anemia coincides with each episode of hemolysis.

Neoplastic disease that causes generalized adenopathy may be primary to the lymph node as in Hodgkin's and non-Hodgkin's lymphoma, or it may be metastatic to the node with invasion of the node by extrinsic malignant cells as in leukemia or neuroblastoma (see Chapter 100). Hodgkin's disease, as discussed previously under regional adenopathy, usually manifests as cervical adenopathy. In contrast, non-Hodgkin's lymphoma may present with rapidly enlarging, diffuse adenopathy, often accompanied by abdominal pain, vomiting, and diarrhea secondary to abdominal node involvement. Another neoplastic condition that can present with generalized adenopathy includes leukemia. Approximately 70% of patients with acute lymphocytic leukemia (ALL) and 30% of patients with acute myelogenous leukemia (AML) have generalized adenopathy (see Chapter 100). These children usually appear ill, having other

systemic signs—hepatosplenomegaly, anemia, and thrombocytopenia with petechiae, purpura, and hemorrhage.

Histiocytosis presents as a spectrum of disease, ranging from a benign, isolated eosinophilic granuloma found in a long bone of an older child to the malignant multiorgan histiocytic infiltration found in infants with Letterer-Siwe disease (see Chapter 100). Lymphadenopathy often occurs in histiocytosis and can be an isolated finding; however, it usually occurs in association with other manifestations of disease.

Rarer causes of systemic adenopathy include lipid storage diseases (Gaucher's and Niemann-Pick's disease), which can cause diffuse adenopathy and are almost always associated with hepatosplenomegaly. Bone marrow biopsy, showing lipid-laden histiocytes, is diagnostic.

Certain drugs, most notably phenytoin (Dilantin), are associated with generalized adenopathy, appearing 1 to 2 weeks after drug initiation and disappearing 3 to 4 weeks after the drug is discontinued. This generalized adenopathy may or may not be associated with other systemic signs (hepatosplenomegaly, pruritic rash, fever, anemia, and leukopenia).

Finally, hyperthyroidism can be associated with a nonspecific lymph node hyperplasia, but one should see other signs and symptoms of the illness, such as tachycardia, hypertension, diaphoresis, weight loss, goiter, lid lag, and hyperreflexia, on physical examination.

Life-Threatening Lymphadenopathy

Several disorders associated with lymphadenopathy, primarily but not exclusively oncologic, can be life-threatening (Table 44.3). The superior vena cava (SVC) syndrome is an example of life-threatening adenopathy. The SVC is a thin-walled vessel with low intravascular pressure that is approximated tightly to the right mainstem bronchus and completely encircled by the lymph nodes that drain the thoracic cavity. SVC syndrome is obstruction of the SVC, usually caused by massive adenopathy, and manifests as dilated chest wall and neck veins, facial edema, and plethora. Drowsiness or stupor, called “wet brain” syndrome, may also be seen. Superior mediastinal syndrome is a variant of SVC syndrome, with additional respiratory symptoms caused by trachea or bronchus compression. In contrast with patients with SVC syndrome, those with superior mediastinal syndrome present in respiratory distress with coughing and wheezing.

Superior vena cava syndrome	Acute myelogenous leukemia
Hodgkin's disease	Neuroblastoma
Non-Hodgkin's lymphoma	Letterer-Siwe disease
Neuroblastoma	Coronary artery aneurysm
Bone marrow failure/multiorgan infiltration	Kawasaki disease
Acute lymphocytic leukemia	

Table 44.3. Life-Threatening Conditions Associated with Lymphadenopathy

Almost all patients with SVC or superior mediastinal syndrome have a malignant etiology (see Chapter 100). In children, Hodgkin's and non-Hodgkin's lymphoma are the most common causes, followed by metastatic neuroblastoma. Emergency physicians who treat patients with SVC syndrome must be careful to administer all intravenous therapy in the lower extremities. Poor circulation in the upper extremities and torso because of SVC obstruction results in poor drug distribution and places the patient with SVC syndrome at increased risk of thrombus formation.

EVALUATION AND DECISION

The clinician who evaluates lymphadenopathy is faced with an extensive differential diagnosis. A meticulous history and physical examination can help focus the evaluation of the patient. Historical data that need to be obtained include the time of onset, the rate of growth, and the duration of symptoms. Lymphadenopathy of more than 3 weeks' duration is considered chronic. The presence and duration of fever, history of rash or pruritus, cough, weight loss, anorexia, and nausea are important systemic symptoms. Recent illnesses must be considered, particularly because lymphadenopathy may persist for 2 to 3 weeks after the resolution of common viral illnesses. Certain medications, most notably phenytoin (Dilantin) and isoniazid, can cause generalized lymphadenopathy. In addition, the presence of certain risk factors, such as young cats in the home (cat-scratch disease) or other animals (e.g., dogs, rabbits, rats), exposure to patients with active tuberculosis, consumption of unpasteurized milk, or exposure to fish tanks (atypical mycobacteria), among others, need to be ascertained. Finally, the clinician must ask whether any prior treatment, such as antibiotic therapy or attempted aspiration with cultures, has been initiated. For example, children with atypical mycobacterial adenitis may present to the emergency department after a prolonged course of antistaphylococcal antibiotic therapy failed to reduce the size of the node. Knowledge of the response to specific antimicrobial therapy can often guide the physician to exclude certain diagnoses.

The physical examination should include a careful determination of the size of the enlarged nodes and documentation of the number of nodes involved to provide an adequate baseline for follow-up. In general, lymph nodes larger than 1 cm are significant in any location. The presence of erythema, warmth, and tenderness often points to an acute pyogenic bacterial process. In most disease processes that cause lymphadenopathy in children, the nodes will be firm, rubbery,

and mobile. Lymph nodes fixed to underlying tissues or located in deeper fascial planes are rare in children, but when present, they should prompt the physician to consider early surgical evaluation. Finally, because several systemic diseases manifest a specific pattern of adenopathy, examination of all lymph node regions must be performed. Likewise, hepatosplenomegaly, rash, and other signs of systemic involvement must be sought.

The approach to the patient with lymphadenopathy focuses initially on the history and examination findings as noted, with emphasis on the distribution of enlarged nodes: regional or generalized (Fig. 44.1). Regional lymphadenopathy should be categorized as acute or subacute/chronic.

FIGURE 44.1. The diagnostic approach to the child with lymphadenopathy. *PPD,* purified protein derivative; *EBV,* Epstein-Barr virus; *CMV,* cytomegalovirus; *CBC,* complete blood count; *HIV,* human immunodeficiency virus.

The most common causes of acute regional lymphadenopathy include reactive hyperplasia, acute bacterial adenitis, and EBV infection (infectious mononucleosis). Findings of acute inflammation, such as erythema and tenderness, point to bacterial adenitis. The emergency physician must decide whether the patient would benefit from aspiration and drainage of the lymph node, particularly if the lesion is fluctuant and easily amenable to the procedure. Treatment of acute bacterial adenitis should include antistaphylococcal and antistreptococcal antibiotics as well as careful follow-up. It is important to note and to inform the patient's parents that these infections often are slow to resolve and may eventually require incision and drainage despite adequate antimicrobial therapy (see also Chapter 84).

The presence of systemic symptoms may suggest other causes of acute regional adenopathy. For example, the presence of pharyngitis, hepatosplenomegaly, and periorbital edema should suggest EBV infection. In the absence of any respiratory compromise, the treatment of EBV infection is supportive. Several days of high fever, rash, and swelling of the extremities in the presence of a large node should alert the physician to the possibility of Kawasaki disease. Early identification of these patients is essential in preventing serious sequelae of this disease. Therefore, a low index of suspicion for Kawasaki disease is prudent.

The evaluation of subacute or chronic regional adenopathy includes consideration of various infectious and noninfectious causes. Exposure to cats should alert the physician to the possibility of cat-scratch disease. The possibility of tuberculosis or atypical mycobacteria can be evaluated by placing a positive purified protein derivative (PPD) test on the patient or can be elicited by a history of exposure to a patient with active tuberculosis. Malignancies and chronic systemic disorders (sarcoid) are less common causes of subacute or chronic regional adenopathy. Either the location (e.g., supraclavicular) or persistence of the node points to a neoplastic disease or to another serious process.

The evaluation of generalized lymphadenopathy involves consideration of systemic diseases that may be associated with adenopathy. The presence of systemic signs of illness, such as weight loss and fever, may be seen in subacute bacterial endocarditis, HIV, tuberculosis, brucellosis, and syphilis. A recent, brief febrile illness, at times with a rash, is characteristic of EBV, tuberculosis, mononucleosis, or acute HIV infection. Signs of toxicity suggest less commonly encountered causes (tumors, collagen vascular disease, sarcoid). In the absence of toxicity, and particularly if the adenopathy begins to resolve within 4 weeks of presentation, the diagnosis of reactive hyperplasia is most likely.

The decision to perform a biopsy on an enlarged node remains a clinical one. In general, early node biopsy should be considered in all neonates with lymphadenopathy and in older children who are ill with systemic symptoms, persistent fever, or weight loss. Deep inferior cervical or supraclavicular adenopathy with or without an abnormal chest film showing hilar adenopathy should be aggressively pursued with biopsy. Beyond this, in the face of an otherwise negative diagnostic workup that included a complete blood count, tuberculosis skin test, EBV heterophile, and chest film, serial measurement over a period of weeks showing progressive or rapid enlargement of the affected node raises suspicion for malignant disease and biopsy should be strongly considered. Biopsy should also be considered if an enlarged node fails to regress in size after approximately 6 weeks of observation.

Suggested Readings

Bass JW, Freitas BC, Freitas AD, et al. Prospective randomized double blind placebo-controlled evaluation of azithromycin for treatment of cat-scratch disease. Pediatr Infect Dis J 1998;17:447–452.

Bodenstein L, Altman RP. Cervical lymphadenitis in infants and children. Semin Pediatr Surg 1994;3:134–141.

Hazra R, Robson CD, Perez-Atayde AR, Husson RN. Lymphadenitis due to nontubercolous mycobacteria in children: presentation and response to therapy. Clin Infect Dis 1999;28:123–129.

Jacobs RF. Tularemia. Adv Pediatr Infect Dis 1996;12:55–69.

Knight PJ, Mulne AF, Vassy LE. When is lymph node biopsy indicated in children with enlarged peripheral nodes? Pediatrics 1982;69:391.

Maurin M, Raoult D. *Bartonella (Rochalimaea) quintana* infections. Clin Microb Rev 1996;3:273–292.

Rowley AH, Gonzalez-Crussi F, Shulman ST. Kawasaki syndrome. Rev Infect Dis 1988;10:1.

Schaller RJ, Counselman FL. Infectious mononucleosis in young children. Am J Emerg Med 1995;13:438–440.

Starke JR. Nontuberculous mycobacterial infections in children. Adv Pediatr Infect Dis 1992;7:123–159.

Suskind DL, Handler SD, Tom LW, et al. Nontuberculous mycobacterial cervical adenitis. Clin Pediatr 1997;36:403–409.

Zitelli BJ. Neck masses in children. Pediatr Clin North Am 1981;28:813.

CHAPTER 45

Neck Mass

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Neck masses are common in children, and the diagnosis encompasses a multitude of disorders. By definition, neck masses include any visible swelling that disturbs the normal contour of the neck between the shoulder and the angle of the jaw. In the pediatric population, the four basic classifications of neck lesions are congenital, inflammatory, traumatic, and neoplastic. Congenital anatomic defects of the neck are often inapparent or minimally recognizable at birth but develop into significant cystic masses later. Included in this category are cystic hygromas, branchial cleft cysts, hemangiomas, thyroglossal duct cysts, and dermoids. It is important to have a working understanding of the embryology of the neck to assist in diagnosis and treatment. Inflammatory masses are the most common and usually represent structures normally present, such as lymph nodes that are undergoing changes from the infectious causes. By far, the most common causes of neck masses in children are reactive adenopathy and adenitis. Traumatic neck masses are usually caused by hematoma surrounding vital structures and may lead to significant distress. Malignant lesions of the head and neck must be ruled out, but fortunately, they are fairly uncommon and are often from the cancer of the lymphatic system.

True medical emergencies arise if neck masses compromise adjacent vital structures, including the airway, carotid blood vessels, and cervical spinal cord. In rare cases, the principal threat to life is from systemic toxicity. Infection that leads to septicemia or the effects of excess hormone secretion in thyroid storm can lead to uncompensated shock. Most large neck masses do not encroach on vital structures because their growth points outward. Embarrassment about personal appearance or a concern of malignancy may be factors, however, in the initiation of the emergency department (ED) visit.

This chapter first emphasizes recognition of masses that represent true emergencies (Table 45.1). Then, the approach to nonemergent, but commonly seen, lesions is described (Table 45.2). Table 45.3 lists causes of neck masses of children by origin.

Hematoma secondary to trauma
Cervical spine injury
Vascular compromise or acute bleeding
Late arteriovenous fistula
Subcutaneous emphysema with associated airway or pulmonary injury
Local hypersensitivity reaction (sting/bite) with airway edema
Airway compromise with epiglottitis, tonsillar abscess, or infection of floor of mouth or retropharyngeal space (with adenopathy)
Bacteremia/sepsis associated with local infection of a cyst (cystic hygroma, thyroglossal, or branchial cleft cyst)
Non-Hodgkin's lymphoma with mediastinal mass and airway compromise
Thyroid storm
Mucocutaneous lymph node syndrome with coronary vasculitis
Tumor—leukemia, lymphoma, rhabdomyosarcoma, histiocytosis X

Table 45.1. Life-Threatening Causes of Neck Mass

Lymphadenopathy secondary to viral or bacterial infection
Cervical adenitis (bacterial)
Hematoma
Benign tumors—lipoma, keloid
Congenital cyst (squamous epithelial cysts)

Table 45.2. Common Causes of Neck Mass

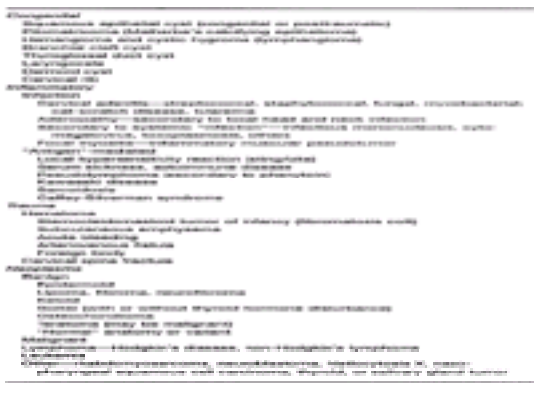

Table 45.3. Differential Diagnosis of Neck Mass by Etiology

EVALUATION AND DECISION

The initial history and physical examination should screen rapidly for airway or vascular compromise with consideration of integrity of the cervical spine. The presence of stridor, hoarseness, dysphagia, and drooling indicates respiratory compromise. The quality of breathing, level of consciousness, and integrity of the cervical spine should also be assessed. Appropriate resuscitative measures should be taken if respiratory or vascular compromise is evident. The cervical spine should be immobilized if there is history of trauma or the initial evaluation leads to suspicion. Table 45.1 lists disorders that constitute true emergencies because of local pressure on vital structures or because of systemic toxicity.

Child with Neck Mass and Respiratory Distress or Systemic Toxicity

Trauma from vehicular accidents, falls from heights, or sports injuries may cause bleeding or hematoma formation near vital structures such as the carotid artery or trachea. If the trauma involves the cervical spine, a hematoma may occur over fractured vertebrae. Even mild injuries may lead to severe hemorrhage and compression of vital structures of the neck in children who have clotting factor disorders (i.e., hemophilia) or platelet disorders (i.e., idiopathic thrombocytopenic purpura). Symptomatic arteriovenous fistulas may appear weeks after neck trauma. The emergency physician should be wary of severe trauma or ecchymoses with an insignificant “history” and should consider possible child abuse or an acquired bleeding problem. The progression of a pneumomediastinum to pneumothorax can be rapid and requires close observation of the tachypneic child with a “crepitant” neck mass. Acutely, this may be caused by trauma to the chest and rib cage or by severe airway obstruction caused by asthma or a foreign body. In children with obstructive lung diseases, such as asthma and cystic fibrosis, high transpulmonary pressure generated in these diseases forces air through small alveolar leaks into the mediastinum or pleural space. This may produce a pneumomediastinum that dissects into the neck. Anaphylactic reaction with neck swelling may precipitate an acute emergency if the swelling compromises the airway. Severe, local reactions to bee stings or to other sensitizing allergens may cause enough tissue edema to obstruct the trachea.

Infections associated with life-threatening processes include retropharyngeal, lateral pharyngeal, and peritonsillar abscesses. Rarely, epiglottitis may present with associated cervical adenitis or the appearance of submandibular mass from ballooning of the hypopharynx. These patients may have cervical adenitis and concomitant dysphagia, drooling, and stridor. Occasionally, branchial cleft cysts or cystic hygromas become infected and progress to mediastinitis, or laryngoceles may become acutely infected and obstruct air flow. Massive tonsillar hypertrophy with infectious mononucleosis or dental infection that spreads to the floor of the mouth (Ludwig's angina) and neck may cause neck masses and airway compression. More recently, children with human immunodeficiency virus (HIV) infection (see Chapter 85) are reported to have parotitis or generalized lymphadenopathy, particularly visible in the neck as a presenting complaint. Children may have hyperthyroid symptoms as part of the presentation of a neck mass. Similarly, patients with the mucocutaneous lymph node syndrome often have cervical lymphadenopathy and, on rare occasions, have active life-threatening vasculitis of the coronary vessels.

Neck tumors in children may become large enough to encroach on vital structures. Cystic hygromas and hemangiomas occasionally enlarge sufficiently enough to interfere with feeding or to obstruct the airway. Lymphoma, an uncommon but important cause of neck mass, is suggested especially by painless enlargement (often of supraclavicular nodes) that occurs over several weeks in the older school-age child. When mediastinal nodes are involved, the patient may rapidly develop a blockage of the intrathoracic trachea that is accentuated on lying down. These children may be fine when sitting, but when supine, the anterior mediastinal masses compress the trachea, causing the airway to collapse. Other tumors, such as rhabdomyosarcoma, leukemia, neuroblastoma, and histocytosis X, are life-threatening because of local invasion and metabolic and hematologic effects.

Child with Neck Mass and No Distress

Most children in the ED with a neck mass are not in distress; the leading diagnoses are reactive adenopathy and acute lymphadenitis from viral or bacterial infection. A common concern, however, is which neck mass bears the diagnosis of

malignancy and requires biopsy or further evaluation.

History

A careful history, establishing the duration of signs and symptoms as well as ascertaining the involvement of other organ systems (fatigue, weight loss, night sweats, adenopathy elsewhere), often suggests the diagnosis. Location of the mass, size and shape of the mass, duration of symptoms, and a history of injury are important. The patient's age at discovery of the lesion should be noted because those found early in infancy increase the risk of a cyst of congenital origin. Birth trauma, with bleeding into the sternomastoid muscle, may cause torticollis, which presents at several weeks of age with a neck mass. It is important to note that not all congenital lesions present at birth or in the first months of life. They are brought to medical attention with acute infection or inflammation, sometimes as a recurrent unilateral neck mass in young infants. Changes in the size of the mass with time or with a child's growth should be noted. Presence of a dimple or sinuses, history of drainage, and other symptoms of infection may assist in making the diagnosis. History of exposure to an infectious agent, including streptococcal pharyngitis, infectious mononucleosis, and cat-scratch disease, should be sought. Systemic symptoms that suggest serum sickness or pseudolymphoma ought to prompt an exposure history for medications (e.g., antibiotics, phenytoin, respectively). Figure 45.1 describes a pathway to facilitate some of the differential diagnoses by category.

FIGURE 45.1. Evaluation of the child with a neck mass.

Physical Examination

After assessment for critical emergency is done, the clinician should perform a thorough examination on a child with a neck mass. It is often valuable to examine the patient thoroughly and then come to examine the area of the head and neck last. Palpation of the mass, noting its location, size, shape, relationship, and attachment to normal structures in the neck, should be completed. Figure 45.2 diagrams the locations of many of the causes of neck mass. It is important to ascertain whether crepitation is present and the degree to which an inflammatory mass is fluctuant. The surrounding area should be palpated for additional lesions (10 to 20% of branchial lesions are bilateral) and to evaluate normal structures of the neck such as the thyroid gland, sternomastoid muscles, trachea, and cervical spine. Inspection of the oral cavity should be performed, noting oral mucosa, dentition, and the orifices such as Stenson's duct (parotid gland) and other glands. The presence of movement of the mass with swallowing or with protrusion of the tongue is important. The examination of the head should be meticulous, including the scalp, ears, sinuses, and nasopharynx.

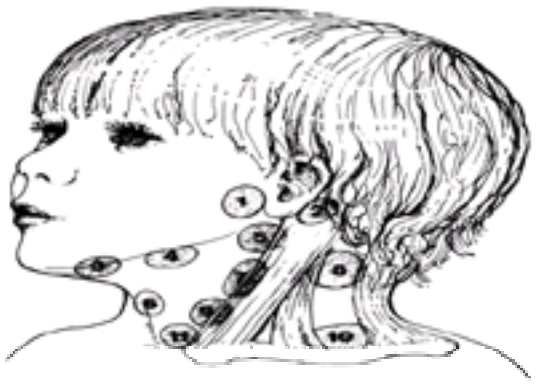

FIGURE 45.2. Differential diagnosis of neck mass by location.
Area 1. Parotid: Cystic hygroma, hemangioma, lymphadenitis, parotitis, Sjögren's and Caffey-Silverman syndrome, lymphoma.
Area 2. Postauricular: Lymphadenitis, branchial cleft cyst (1st), squamous epithelial cyst.
Area 3. Submental: Lymphadenitis, cystic hygroma, sialadenitis, tumor, cystic fibrosis.
Area 4. Submandibular: Lymphadenitis, cystic hygroma, sialadenitis, tumor, cystic fibrosis.
Area 5. Jugulodigastric: Lymphadenitis, squamous epithelial cyst, branchial cleft cyst (1st), parotid tumor, *normal*--transverse process C2, styloid process.
Area 6. Midline neck: Lymphadenitis, thyroglossal duct cyst, dermoid, laryngocele, *normal*--hyoid, thyroid.
Area 7. Sternomastoid (anterior): Lymphadenitis, branchial cleft cyst (2nd, 3rd), pilomatrixoma, rare tumors.
Area 8. Spinal accessory: Lymphadenitis, lymphoma, metastasis (from nasopharynx).
Area 9. Paratracheal: Thyroid, parathyroid, esophageal diverticulum.
Area 10. Supraclavicular: Cystic hygroma, lipoma, lymphoma, metastasis,*normal*--fat pad, pneumatocele of upper lobe.
Area 11. Suprasternal: Thyroid, lipoma, dermoid, thymus, mediastinal mass.

(Modified with permission from May M. Neck masses in children: diagnosis and treatment. Clin Pediatr 1976;5:17.)

Details of chronicity, size, and progression and evidence of inflammation help distinguish between infection and neoplasm. Characteristics that some authors have found associated with malignancy include masses that are firm and larger than 3 cm in diameter, nonpainful, progressively enlarging, ulcerating, deep to fascia or fixed to tissue, or discovered in a newborn. These criteria are sensitive but not specific for cancer. Even with these characteristics, most lesions are benign congenital cysts or inflammatory masses. The length of time the “node” is present is not discriminating in that, often, inflammatory nodes that are biopsied have been present for longer than 3 to 6 months.

During auscultation of the chest, special attention should be paid to inspiration because extrathoracic airway obstruction from the trachea or upper airway may produce only faint stridor. Respiratory distress or wheezing that worsens in the supine position may be an early sign of an anterior mediastinal mass. The physical examination should be completed, looking for signs of systemic illness as a cause for the neck mass. The general appearance and color of the child is important, as is the presence of hepatosplenomegaly or an abdominal mass, indicating a high suspicion for a malignancy. Signs of thyroid hormone excess or deficiency may be associated with a goiter. Rashes, generalized lymphadenopathy, and fever may indicate an inflammatory or oncologic process. Failure to thrive or weight loss may be found with a number of causes of infection or oncologic illness, including HIV disease, histiocytosis X, and others.

DIFFERENTIAL DIAGNOSIS

Congenital Masses

Thyroglossal duct cysts are the most common congenital cyst of the neck. They develop along the line of descent of the thyroid gland in the neck anywhere from the base of the tongue to the sternal notch. Thyroglossal duct cysts are usually midline, adjacent to the hyoid bone, and diagnosed in children 2 to 10 years of age. Thyroglossal duct cysts are soft, nontender, smooth, and may move when the child swallows or protrudes the tongue. If infected, they may be warm and erythematous and drain externally. Antibiotics (for mouth and skin flora), warm compresses, and incision and drainage (if indicated) should be initiated for signs of infection. Complete excision is the treatment of choice after complete resolution of infection.

Cystic hygromas are cystic lymphatic malformations occurring in the posterior triangle of the neck. Most are identified at birth, but some may be recognized after injury or upper respiratory infection when “herniation” has occurred after crying, coughing, or other forceful Valsalva maneuvers. Cystic hygromas appear discrete, soft, mobile, nontender, and vary greatly in size. Extension to the mediastinum is rare but can occur and can be seen on chest radiographs. Respiratory distress can occur in infants with large masses compressing the airway. Infection is uncommon, but signs of it would be as expected. Ultrasonography is useful in establishing if the mass is cystic. Computed tomography (CT) imaging can determine extent and involvement of surrounding structures. Spontaneous regression is rare; therefore, complete excision is the treatment of choice.

Branchial cleft anomalies are lesions most commonly occurring from defects in the development of the second branchial arch, giving rise to firm masses along the anterior border of the sternocleidomastoid muscle. Branchial cleft sinuses are painless and present with drainage at the junction of the middle and lower thirds of the sternocleidomastoid muscle. Cysts that are usually fluctuant, mobile, and nontender may occur if the sinus tract becomes blocked. Probing or injecting the tract may lead to infection. Ultrasonography may be useful in identifying cystic structures. Treatment with antibiotics with complete resolution of infection is necessary if the sinus or cyst is infected. Excision of the entire tract and cyst is important to prevent recurrence.

Hemangiomas are common head and neck lesions identified in infancy. They are three times more common in females than in males. Hemangiomas are soft, mobile, nontender, and bluish or reddish in color. They tend to get larger in the first year of life and then involute over the next several years. Rare complications include thrombocytopenia from platelet consumption, disseminated intravascular coagulation, hemorrhage, airway obstruction, congestive heart failure, ulceration, infection, and necrosis. Treatment for most hemangiomas is conservative and nonoperative because the issues are almost solely cosmetic and short term. Other treatments are reserved for rapidly growing lesions that are impairing vision or hearing or are life-threatening.

Neonatal torticollis results from sternocleidomastoid fibrosis and shortening of the muscle. Presenting symptoms of torticollis occur in the first 3 weeks of life, with the infant holding his or her face and chin away from the affected side and the head tilted toward the fibrous mass. The mass is firm and seems attached to the muscle. Physical therapy, including massage, range-of-motion exercises, stretching exercises, and positional changes, is the preferred treatment. Facial and cranial asymmetry can develop without intervention. Surgical intervention is rarely needed.

Inflammatory Masses

Cervical lymphadenopathy is the most common reason for neck masses in children. Up to 90% of children between the ages of 4 and 8 years can have cervical adenopathy without obvious associated infection or systemic illness. Lymphadenopathy in newborns and young infants is rare and warrants investigation. Supraclavicular lymphadenopathy is considered pathologic and should be biopsied. Etiology for cervical adenopathy includes bacterial or viral infection either from local, regional, or systemic illness. Anterior cervical nodes drain the oropharynx and become enlarged with upper respiratory, oral, and pharyngeal infections. Posterior cervical lymph nodes drain the scalp and nasopharynx and become enlarged with inflammation or infection in these areas. With treatment of the underlying infection, cervical lymphadenopathy should resolve.

Cervical lymphadenitis occurs when acute infection is present within the lymph node (see Chapter 84). Bacteria are the most common causes and include penicillin-resistant *Staphylococcus aureus* and group A b-hemolytic streptococci. Common presentation is usually one or more cervical lymph nodes that become acutely enlarged, tender, warm, and erythematous after an upper respiratory illness, pharyngitis, tonsillitis, or otitis media. Systemic symptoms of fever and malaise may be present. Treatment includes antibiotics and warm soaks. If the patient appears toxic, admission and treatment with intravenous antibiotics is appropriate. Without antibiotic treatment, enlargement with the development of fluctuation and regional cellulitis may progress. Most cases of acute cervical lymphadenitis resolve with antibiotics. If fluctuation is present, needle aspiration is indicated. The purulent fluid should be sent for Gram stain and culture with antibiotic selection based on test results. If resolution of the lymphadenitis does not occur after needle aspiration and antibiotics, incision and drainage should be performed.

Cat-scratch disease is another common cause of lymph node enlargement in children. Typically, regional lymph nodes enlarge 2 to 4 weeks after a cat scratch (usually a kitten). The lymphadenopathy can be cervical if the head or neck have been scratched. Fever and malaise may have been present initially and usually a single node is involved. The area around the lymph node is warm, tender, indurated, and erythematous. *Bartonella henselae* is the organism most likely responsible. Serologic titers for antibody and polymerase chain reaction assays are available in some laboratories. Warthin-Starry silver stain of the lymph node or inoculation site will identify the organism. Management is symptomatic with resolution in 2 to 4 months. Needle aspiration provides relief to those with tender, suppurative nodes and aids in the diagnosis. Surgical excision is unnecessary and can lead to formation of a draining sinus. Antibiotics should be considered for acutely ill patients with systemic symptoms. Rifampin, trimethoprim–sulfamethoxazole, and ciprofloxacin have been shown to be effective.

Mycobacterial infection of the cervical lymph nodes are most often caused by the atypical strains of *Mycobacterium avium-intracellulare and M. scrofulaceum*. The enlarged lymph nodes are generally submandibular in region and red, rubbery, and minimally tender to palpation. If systemic manifestations are present, an immune deficiency should be considered. On the other hand, clinical systemic signs of tuberculosis accompany cervical lymphadenopathy caused by *M. tuberculosis*. The supraclavicular lymph nodes are commonly involved. Children with suspected mycobacterium infection should have a purified protein derivative (PPD) tuberculin skin test and chest radiograph performed. An excisional biopsy may need to be performed to differentiate between tuberculous and nontuberculous mycobacteria as the offending organism. The PPD tuberculin test may be negative in atypical mycobacterium infections. Treatment for atypical mycobacterial cervical lymphadenitis is complete surgical excision. Incision and drainage results in a draining sinus. Treatment for *M. tuberculosis* lymphadenitis is the same as for pulmonary tuberculosis: 6 to 9 months of antituberculosis chemotherapy.

Cervical lymphadenitis can be the result of viral infections, most commonly mononucleosis. Classically, the patient has diffuse lymphadenopathy with prominent cervical lymphadenopathy and large, hypertrophied tonsils. *Epstein-Barr virus* is the most common cause of mononucleosis. Systemic symptoms of fever, malaise, and the presence of hepatosplenomegaly are common. Exudative pharyngitis may be present, and the throat should be cultured for group A b-hemolytic streptococci. If bacterial pharyngitis is present, the child should be treated with antibiotics. Generally, treatment for mononucleosis is supportive. Corticosteroids (prednisolone at 1 mg/kg per day) have been found useful in reducing tonsillar inflammation and preventing airway compromise.

Kawasaki disease (mucocutaneous lymph node syndrome) is associated with a single enlarged cervical lymph node, conjunctival injection without drainage, erythematous mouth, cracked lips, strawberry tongue, erythematous rash, induration of the palms of hands and soles of the feet, and fever of at least 3 days' duration. The cause is unknown but is thought to be infectious. Anti-immune therapy should be initiated if the diagnosis is strongly suspected. An echocardiogram should be performed to rule out coronary artery aneurysms.

Neoplasms

Fortunately, neoplasms of the head and neck in children are less commonly seen than infection or congenital lesions. Presentation is usually a painless, firm, fixed cervical mass. Systemic symptoms may not be present. Differentiating between a benign and malignant lesion can be difficult. Cervical lymphadenopathy that does not resolve with standard therapy should raise suspicion for a malignancy. Neoplastic etiologies for neck mass in children include Hodgkin's and non-Hodgkin's lymphoma, rhabdomyosarcoma, neuroblastoma, thyroid carcinoma, and nasopharyngeal carcinoma. As described in the section on evaluation, duration and characteristics of the neck mass will lead to increased risk of cancer. If a malignancy is suspected, a complete blood count, chest radiograph, and selective CT or magnetic resonance imaging (MRI) should be obtained. Treatment is individualized according to specific tumor and extent of disease (see Chapter 100 and Chapter 121).

LABORATORY TESTING

The clinical impression should be used to ascertain the need for laboratory studies or radiologic imaging. Many of the common conditions are inflammatory or acute infections, and no studies need to be done. Oxygenation may be determined by pulse oximetry. In processes for which the risk of critical airway obstruction is pending, the utility of the arterial blood gas adds little initially, and the stress may lead to worsening of the obstruction. A complete white blood count and differential are most helpful when an oncologic cause is suspected. When bleeding from trivial trauma is being considered as a cause of neck mass, the platelet count, prothrombin time, and partial thromboplastin time should be obtained. Consider also a bleeding time when a coagulopathy is in the differential. Serum thyroid hormone levels and thyroid-stimulating hormone (TSH) may be warranted for goiter. Throat culture for streptococcal disease (or a Rapid Strep screen) should be obtained when pharyngitis is found. A monospot or, preferably, Epstein-Barr virus titers should be done to confirm infectious mononucleosis.

Cervical spine radiographs need to be obtained for trauma patients when instability or fracture of the cervical spine is

suspected. Facial or mandibular films may be necessary to evaluate for some lower face trauma or for oral infections. Soft-tissue lateral neck films are helpful to evaluate for intraoral, retropharyngeal, or airway infectious problems. More specific diagnostic imaging uses CT with cuts to evaluate the sinuses and other cavities with better detail. In the child with respiratory distress, a chest radiograph is necessary to view the mediastinum, pleura, and lung for infection, tumor, pneumothorax, or pneumomediastinum. Ultrasound may be useful in defining the mass; a cystic mass with linear septations is characteristic of a cystic hygroma. Other masses (lymphadenopathy, thyroglossal duct) and fibromatosis colli (congenital torticollis) have fairly definitive patterns. Although not specific, the finding of calcification within a mass may suggest a teratoma or neuroblastoma. In some instances, the use of CT or MRI has improved the diagnostic accuracy.

In studies in which biopsies of neck masses were obtained, several authors have found preoperative diagnoses to be correct as infrequently as 60% of the time. Biopsy of the lesion is required when the suspicion is high for malignancy. As in adults, fine needle aspiration in children is becoming more popular at some centers because it offers high sensitivity and specificity for tumors. This may reduce the need for open biopsies as often as 75% of the time by identifying inflammatory or self-limiting processes.

THERAPY

In the ED, the clinical evaluation most often reveals adenopathy that requires no acute therapy or adenitis that necessitates a course of systemic oral antibiotics and local care. Important to the approach to adenitis is the follow-up in several days to monitor clinical response and need for aspiration and drainage. When the mass is suspicious for tumor or congenital cyst, surgical consultation for biopsy or excision is indicated. Hospitalization and institution of definitive therapy are indicated for the patients with neck masses who present with systemic toxicity, airway compromise, or severe local disease.

Suggested Readings

Athreya B, Silverman BK. The neck. In: Pediatric Physical Diagnosis. Norwalk, CT: Appleton-Century-Crofts, 1985:96–100.

Barton L, Feigin R. Childhood cervcal lymphadenitis: a reappraisal. J Pediatr 1974;84:846–852.

Bergman KS, Harris BH. Scalp and neck masses. Pediatr Clin North Am 1993;40:1151–1160.

Brown RL, Azizkhan RG. Pediatric head and neck lesions. Pediatr Clin North Am 1998;45:889–905.

Conn H, Conn R. Lymphadenopathy. In: Current Diagnosis. Philadelphia: WB Saunders, 1974:25–59.

Green M. The neck and lymphadenopathy. In: Pediatric Diagnosis. Philadelphia: WB Saunders, 1986:61–65, 398–403.

Herzog LW. Prevalence of lymphadenopathy of the head and neck in children. Clin Pediatr 1983;22:485–487.

Knight PJ, Mulne AF, Vassy LE. When is a lymph node biopsy indicated in children with enlarged peripheral nodes? Pediatrics 1982;69:391–396.

Knight PJ, Reiner CB. Superficial lumps in children: what, when, and why? Pediatrics 1983;72:147–153.

Kraus AU, Han BK, Babcock DS, Ostreich AE. Sonography of neck masses in children. Am J Roentgenol 1986;146:609–613.

Lusk RP. Neck masses. In: Pediatric Otolaryngology. Philadelphia: WB Saunders, 1990:1294–1302.

Nicklaus PJ, Kelley PE. Management of deep neck infection. Pediatr Clin North Am 1996;43:1277–1296.

Marcy SM. Infections of the lymph nodes of the head and neck. J Pediatr Inf Dis 1983;2:397–405.

May M. Neck masses in children: diagnosis and treatment. Pediatr Ann 1976;5:517–535.

Mobley DL, Wakely PE Jr, Frable MA. Fine needle aspiration: application to pediatric head and neck masses. Laryngoscope 1991;101:469–472.

Stewart MG, Starke JR, Coker NJ. Nontuberculous mycobacterial infections of the head and neck. Arch Otolaryngol Head Neck Surg 1994;120: 873–876.

Torsiglieri AJ Jr, Tom LW, Ross AJ III, et al. Pediatric neck masses: guidelines for evaluation. Int J Pediatr Otorhinolaryngol 1988;16:199–210.

CHAPTER 46

Neck Stiffness

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Neck stiffness is an important chief complaint in children evaluated in the emergency department. Commonly, neck stiffness is accompanied by neck pain. However, certain clinical conditions may lead a child to hold the neck in an abnormal posture without neck pain. The underlying causes of neck stiffness or malposition in children range from relatively benign (e.g., cervical adenitis) to life-threatening (e.g., meningitis, fracture or subluxation of the cervical spine).

Torticollis (meaning “twisted neck” from the Latin roots tortus and collum) is a variation of neck stiffness. The child holds the head tilted to one side and the chin rotated in the opposite direction, reflecting unilateral neck muscle contraction. This may result from various pathologic processes and may or may not be associated with neck pain. Torticollis is often congenital and muscular in origin; however, it can also be associated with acquired processes such as trauma, infectious or inflammatory illnesses, central nervous system (CNS) neoplasms, drug reactions, and a variety of different syndromes.

This chapter reviews the differential diagnosis of neck stiffness or malposition, including torticollis, both with and without neck pain, in children. The proposed algorithm at the end of the chapter will help distinguish potentially life-threatening from benign causes of neck stiffness, while providing a broad differential diagnosis for this important clinical finding.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of neck stiffness is best organized around a few important historical/clinical questions: 1) Is there a history of trauma? 2) Is there evidence of an infectious or inflammatory process (e.g., history or presence of fever)? and 3) Is there evidence of spinal cord involvement? Table 46.1 lists most causes of neck stiffness in children, Table 46.2 lists the common causes, and Table 46.3 lists the life-threatening causes. The following description categorizes the causes of neck stiffness in children by underlying mechanism and severity.

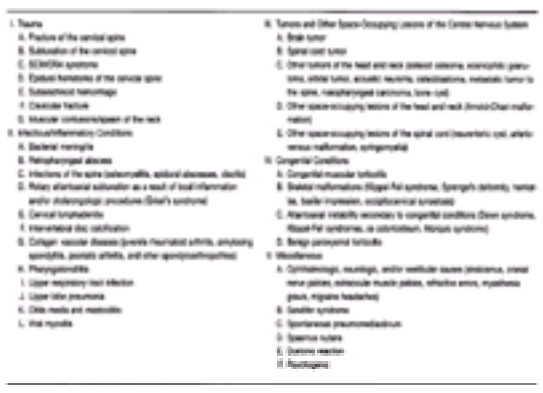

Table 46.1. Causes of Neck Stiffness or Malposition

I. Trauma	C. Pharyngotonsillitis and other upper respiratory tract infections
A. Minor trauma (cervical muscular contusions, strains, and spasm)	D. Viral myositis
B. Clavicular fracture	E. Muscle spasm
C. Rotary atlantoaxial subluxation	F. Rotary atlantoaxial subluxation
II. Infectious/Inflammatory Conditions	III. Congenital Conditions
A. Bacterial meningitis	A. Congenital muscular torticollis
B. Cervical lymphadenitis	IV. Miscellaneous
	A. Dystonic reaction

Table 46.2. Common Causes of Neck Stiffness or Malposition

I. Trauma	III. Tumors and Other Space-Occupying Lesions of the Central Nervous System
A. Injuries to the cervical spine (fractures, subluxation, SCIWORA, epidural hematoma)	A. Brain tumor
B. Subarachnoid hemorrhage	B. Spinal cord tumor
II. Infection	C. Other tumors and space-occupying lesions of the head, neck, and spinal cord
A. Bacterial meningitis	IV. Congenital Conditions
B. Retropharyngeal abscess	A. Atlantoaxial instability secondary to congenital conditions
C. Infections of the spine (osteomyelitis, epidural abscesses, discitis)	
D. Atlantoaxial subluxation with anterior displacement of the atlas as a result of local inflammation	

Table 46.3. Life-Threatening Causes of Neck Stiffness or Malposition

Neck Stiffness Associated with Trauma

Potentially Life-Threatening Causes

Trauma to the neck is a common cause of neck pain and stiffness in children (see Chapter 106). Fortunately, serious injuries to the cervical spine (fractures, subluxations) in children are uncommon. These injuries generally occur in the upper cervical spine, as opposed to neck injuries in adults, which more commonly involve the lower cervical spine. Neck injuries in children most commonly result from motor vehicle–related accidents, sports injuries, and falls.

Fractures of the Cervical Spine

Fractures of the cervical spine in children are uncommon, occurring in 1 to 2% of hospitalized pediatric trauma patients. Although some children with fractures of the cervical spine are unresponsive at the time of evaluation, most are alert and verbal, complain of neck pain, and have no demonstrable neurologic deficit. At the minimum, the cervical spine should be immobilized and multiple-view radiographs of the cervical spine should be obtained on any child with an altered level of consciousness, pain or stiffness of the neck, any neurologic deficits, or distracting painful injuries; they should also be obtained in those who are unable to perceive pain (as a result of alcohol or drugs) or describe their symptoms.

Subluxation of the Cervical Spine

Subluxations of the cervical spine are more common than fractures and may result from minor trauma (e.g., falls from low heights) as well as more severe trauma (see Chapter 106). The most commonly occurring of these is rotary atlantoaxial subluxation, which generally does not compromise the spinal canal because the transverse ligament of the atlas remains intact. Rotary subluxation typically causes neck pain and torticollis. Sternocleidomastoid (SCM) spasm and neck tenderness are localized to the same side as the head rotation in contrast to inflammatory muscular torticollis, in which the spastic, tender SCM muscle is opposite to the direction of head rotation. Neurologic deficits are rare in patients with rotary subluxation. With rotary atlantoaxial subluxation, an anteroposterior open-mouth radiograph typically shows the rotation of C1 on C2, with the odontoid in an eccentric position relative to C1. Computed tomography (CT) scans of the neck can confirm equivocal cases. Most patients with rotary subluxation can be treated with a soft collar and anti-inflammatory medications. Traction and immobilization are necessary for more severe rotary subluxation or if reduction is not achieved by conservative measures.

Atlantoaxial subluxation with compromise of the spinal canal results from ligamentous laxity or rupture and resultant anterior movement of the atlas on the axis. Children with Down syndrome are susceptible to atlantoaxial subluxation because of laxity of the transverse ligament of the atlas. Radiographic findings of atlantoaxial subluxation may include a widened predental space and prevertebral soft-tissue swelling. Treatment involves immobilization and cervical traction.

SCIWORA Syndrome

The ligamentous laxity and hypermobility of the pediatric cervical spine predispose children to spinal cord injuries without radiographic abnormality (SCIWORA syndrome). This may occur in more than 50% of children with cervical spinal cord injuries. Children younger than 8 years of age are most susceptible. Various mechanisms, including longitudinal distraction, hyperflexion, hyperextension, and ischemia to the spinal cord, may result in SCIWORA syndrome. Children with SCIWORA syndrome generally experience significant or progressive paralysis within 48 hours of a traumatic injury. Some children, however, may have transient neurologic symptoms that remit, then recur within the next day with worsening neurologic abnormalities.

Therefore, a careful neurologic history and examination are necessary for any child with traumatic injury to the neck, even in the absence of radiographic abnormalities. Cervical spine immobilization and hospitalization should be seriously considered even for children with transient neurological deficits.

Epidural Hematomas of the Cervical Spine

Epidural hematomas of the cervical spine are uncommon but may occur even after apparently minor trauma. These may compress the spinal cord, leading to progressive neurologic symptoms and signs. Magnetic resonance imaging (MRI) of the spinal cord clearly demonstrates this injury. Emergent neurosurgical consultation and surgical decompression are indicated.

Subarachnoid Hemorrhage

Subarachnoid hemorrhage after trauma may lead to neck stiffness but is accompanied by headache and/or other physical findings of head trauma.

Generally Non–Life-Threatening Causes

Clavicular Fracture

Fracture of the clavicle in children is common and may cause torticollis because of SCM muscle spasm. However, the diagnosis is usually clear because pain is localized to the clavicle and not to the neck.

Traumatic Muscular Contusions of the Neck

Blunt trauma to the neck may result in neck pain as a result of muscular contusion and/or spasm. This is a diagnosis of exclusion, however, and should not be entertained until a detailed physical (neurologic) examination and radiographs of the cervical spine exclude the possibility of a more serious injury. Treatment should include a soft cervical collar and analgesic medication.

Neck Stiffness Associated with Infectious/Inflammatory Conditions

Potentially Life-Threatening Causes

Bacterial Meningitis

Bacterial meningitis is the most important infectious cause of neck stiffness and is almost always accompanied by fever. Children with meningitis typically have findings of neck stiffness on physical examination, although this may not be apparent in young infants and in children with meningeal infection who lack an inflammatory response. Since the introduction of the *Haemophilus influenzae* type b protein conjugate vaccine, the two most common bacterial pathogens causing meningitis in children are *Neisseria meningitidis* and *Streptococcus pneumoniae.* Children with meningococcal meningitis with minimal cerebrospinal fluid (CSF) pleocytosis may not have significant neck findings. However, these children generally have a toxic clinical appearance because the lack of CSF pleocytosis often indicates overwhelming infection. Torticollis has also been reported in patients with bacterial meningitis, although far less commonly than meningismus.

Retropharyngeal Abscess

There are several other important infectious processes for which neck stiffness and usually fever are presenting signs. Retropharyngeal abscess is an infection that occupies the potential space between the posterior pharyngeal wall and the anterior border of the cervical vertebrae. Most commonly caused by group A streptococcus, oral anaerobic organisms and *Staphylococcus aureus,* these infections cause clinical toxicity, drooling, and stridor. Neck stiffness often is an associated clinical finding in children with these infections. Lateral radiographs of the neck reveal soft-tissue swelling anterior to the upper cervical vertebral bodies.

Infections of the Spine

Infectious processes involving the spine (osteomyelitis, epidural abscess, discitis) in children can involve the cervical region, although they occur most commonly in the thoracic and lumbar areas. Localized pain, fever, and elevation of erythrocyte sedimentation rate (ESR) generally accompany all of these infections. Vertebral osteomyelitis occurring in the cervical spine may lead to neck stiffness. Vertebral osteomyelitis is usually bacterial in origin (most commonly caused by *S. aureus*) but may be caused by mycobacteria (tuberculous or nontuberculous) as well. If the cervical spine is involved, radiographs of this area may reveal destruction of the vertebral body, local soft-tissue swelling, or narrowing of the disc space. Radionuclide scanning will reveal uptake at areas of increased metabolic activity of the spine before bony destruction is visible on radiography of the spine.

Although uncommon, spinal epidural abscesses are associated with significant morbidity and mortality. Epidural abscesses may occur in the cervical spine, although lower spine involvement is much more common. When these abscess occur in the cervical region, severe neurologic deficits may occur, and emergent neurosurgical referral is essential.

Infectious discitis is uncommon in children. This disease is often caused by infection with *S. aureus,* although bacterial cultures are commonly negative and the cause has been debated. Most children with infectious discitis are younger than 3 years of age. Disease is usually in the lumbar or thoracic vertebrae rather than in the cervical region, with lower back pain and limp being the most common presenting complaints (see Chapter 51). If conventional radiography is nondiagnostic, technetium bone scanning or MRI is helpful.

Generally Non–Life-Threatening Causes

Atlantoaxial Subluxation as a Result of Local Inflammation and/or Otolaryngologic Procedures

Atlantoaxial subluxation rarely may occur as a result of inflammatory processes in the head and neck region (e.g., rheumatoid arthritis, systemic lupus erythematosus, tonsillitis, pharyngitis) or after otolaryngologic procedures (e.g., tonsillectomy, adenoidectomy). This condition, also called Grisel's syndrome, is believed to occur as a result of ligamentous laxity after an infectious or inflammatory process. The subluxation is rotary, with or without displacement of the atlas, depending on the degree of involvement of the transverse ligament of the atlas. Most children with Grisel's syndrome have torticollis and neck pain, often localized to the SCM muscle. Fever and dysphagia are common as well. The child's head is tilted to one side and rotated to the side opposite of the facet dislocation. Routine radiographs of the neck may or may not reveal asymmetry between the facet joints and increased space between the dens of the axis and the anterior arch of the atlas. High-resolution CT scan with three-dimensional reconstruction is the best way to visualize the subluxation. Most commonly, the condition is mild and there is no anterior displacement of the axis. If mild, the condition usually responds to analgesic medication, physical therapy, and a soft collar. In the uncommon likelihood of severe disease (and certainly in the rare likelihood of spinal cord compression), neurosurgical consultation should be obtained because cervical traction and immobilization are needed. In addition to treating the subluxation, antibiotics to treat an underlying bacterial infection, if present, are needed.

Cervical Lymphadenitis

Cervical lymphadenitis, either acute or chronic, is a common cause of neck stiffness. The child with this condition typically has tender swelling over the lateral aspect of the neck, with or without fever. Most cases of cervical lymphadenitis are caused by *S. aureus* or group A streptococcus; however, other bacteria, mycobacteria, and noninfectious conditions may be involved (including *Rochalimaea,* the cause of cat-scratch disease, and Kawasaki disease). A purified protein derivative (PPD) skin test to screen for tuberculosis and empirical antibiotics to treat the most common bacterial pathogens are usually sufficient therapy.

Intervertebral Disc Calcification

Intervertebral disc calcification (IDC) in children is an uncommon, generally self-limited condition in which the nucleus pulposus of one or more intervertebral discs calcifies. Both the underlying cause of the condition and the cause of acute symptoms are unknown. It is generally thought that acute symptoms are secondary to some inciting event (e.g., mild trauma, viral infection) that results in an inflammatory response, possibly because of the release of calcium crystals. Children typically present with 24 to 48 hours of neck pain associated with neck stiffness or torticollis; fever is often present as well. A lumbar puncture may be necessary to exclude the possibility of meningitis or parameningeal infection. The ESR is usually elevated in IDC, and leukocytosis occurs in one-third of patients. Radiographs of the spine usually show the disc calcification, and CT scans help localize the calcification within the nucleus pulposus. The calcification resorbs spontaneously, and the disease is generally benign and self-limited, although disc protrusion and cord compression uncommonly may occur. Most importantly, one must distinguish acute infectious discitis (see previous discussion) from IDC. Distinguishing features include the lack of disc calcification and the single disc involvement of acute infectious discitis (IDC may involve one or more discs). Furthermore, infectious discitis is most common in the lumbar spine, whereas IDC in children more commonly involves the cervical spine. Finally, radiographic changes demonstrating erosion of vertebral bodies and collapse of disc spaces are seen with infectious discitis, but not with IDC.

Collagen Vascular Disease

Collagen vascular disease (see Chapter 101) in children may involve the cervical spine and lead to neck stiffness and/or pain. Children with juvenile rheumatoid arthritis may have either insidious or acute onset of symptoms, which commonly include neck stiffness. However, isolated cervical disease is unusual. Cervical involvement in ankylosing spondylitis is a late finding, as it is in other spondyloarthropathies. Girls with psoriatic arthritis, however, may have cervical involvement preceding sacroiliac and lumbar involvement.

Other Infectious/Inflammatory Conditions

Pharyngotonsillitis and upper respiratory tract infections may cause neck pain, although this is generally localized to tender cervical lymph nodes. Torticollis (i.e., Grisel's syndrome) may be seen as well. If neck pain is posterior in location and accompanied by fever, a lumbar puncture should be strongly considered to exclude the possibility of meningitis. Similarly, the diagnosis of viral myositis involving the neck can be made only after excluding the possibility of meningitis in a child with neck pain and fever. Otitis media and mastoiditis have also been reported as causes of torticollis. Upper lobe pneumonia may cause pain referred to the neck. Muscle spasm as a cause of torticollis is a diagnosis of exclusion after eliminating the possibility of more serious underlying causes.

Neck Stiffness Associated with Space-Occupying Lesions of the Central Nervous System

Potentially Life-Threatening Causes

Space-occupying lesions of the brain and spinal cord may lead to neck stiffness, malposition, pain, and/or torticollis. Even if the histology of these lesions is benign, they are potentially life-threatening because of the complications of intracranial pressure elevation and the potential for brain and spinal cord compression.

Brain Tumors

Children with tumors of the posterior fossa, the most common location for pediatric brain tumors, may present with head tilt or torticollis. Posterior fossa tumors may cause any of a number of other symptoms and signs (vomiting, headache, ataxia, disturbances in vision including diplopia, papilledema, cranial nerve deficits, corticospinal or corticobulbar signs). Head tilt may result from attempts to compensate for diplopia. However, neck stiffness is thought to result from irritation of the accessory nerve by the cerebellar tonsils trapped in the occipital foramen or by tonsillar herniation.

Spinal Cord Tumors

Tumors of the spinal cord are uncommon in children and account for a small fraction of all CNS tumors in childhood. The most common of the spinal cord tumors is astrocytoma. Typically, spinal cord tumors cause pain at the site of the tumor and neurologic defects (sensory and motor defects, impaired bowel and bladder function), but symptoms may be very slow to develop, often leading to delays in diagnosis. Spinal cord tumors may also cause torticollis. In one reported case, chiropractic manipulation of a child with persistent torticollis and a spinal cord tumor resulted in quadriplegia. Patients with these tumors may also hold their heads in a forward flexed position (“hanging head sign”). An MRI of the spine should be obtained on any child with symptoms and signs suggestive of spinal cord tumor and emergency neurosurgical consultation should be obtained.

Other Space-Occupying Lesions of the Head and Neck

Nasopharyngeal carcinoma is an uncommon tumor in children but may present with epistaxis, neck pain, and cervical adenopathy. Diagnosis requires a high index of suspicion. Other tumors of the head and neck, including orbital tumors, acoustic neuromas, osteoblastomas, and metastatic tumors to the spine, may cause torticollis. Arnold-Chiari malformations may also cause torticollis.

Other Space-Occupying Lesions of the Spinal Cord

Other uncommon space-occupying lesions of the cervical spine such as neurenteric cysts, arteriovenous malformations, and syringomyelia may also cause neck pain and stiffness, generally accompanied by neurologic findings. Early diagnosis by MRI is essential.

Generally Non–Life-Threatening Causes

Benign Tumors of the Head and Neck

Osteoid osteoma is a benign bone tumor that typically affects older children and adolescents. Pain is the typical presenting symptom, often worse at night. If the osteoma is in the cervical spine, neck pain results. Plain radiography is usually diagnostic (showing a well demarcated radiolucent lesion surrounded by sclerotic bone), and treatment is surgical. Eosinophilic granulomas and bone cysts are other benign (and rare) lesions of the spine that may cause neck pain.

Congenital Causes of Neck Stiffness

Neck stiffness and/or torticollis from congenital abnormalities are usually not life-threatening. These congenital causes are usually muscular or skeletal in origin.

Congenital Muscular Torticollis

Congenital muscular torticollis is the most common cause of torticollis in infancy. The etiology of this condition is unclear but is thought to be related to birth trauma causing an injury to the SCM muscle with hematoma formation, followed by fibrous contracture of the muscle. Other theories include those suggesting intrauterine malposition, infection, neurogenic causes, and intrauterine compartment syndrome of the SCM muscle. On examination, a palpable mass can often be detected in the inferior aspect of the SCM. The mass is generally not present at birth but appears in the neonatal period. The head is held in the characteristic position, with the patient's chin pointing away from the affected, contracted SCM muscle. Craniofacial asymmetry is commonly found to some degree in these patients, typically with contralateral flattening of the occiput and ipsilateral depression of the malar prominence. Radiographs of the cervical spine are necessary to exclude other causes of torticollis. Treatment is conservative with passive stretching of the involved muscle. If the deformity persists after 6 to 12 months, surgical release of the SCM is required (fewer than 5% of cases).

Skeletal Malformations

Klippel-Feil syndrome is characterized by congenital fusion of a variable number of cervical vertebrae, which may result in atlantoaxial instability. The cause of this syndrome is unknown. It is often associated with many other bony abnormalities, and significant scoliosis develops in more than 50% of affected children. Limitation in range of motion of the neck is the most common physical sign. In addition to limited neck motion, the classic triad also includes a low hairline and a short neck; the triad, however, is seen in fewer than half of patients.

Sprengel's deformity is characterized by congenital failure of the scapula to descend to its correct position. The scapula rests in a high position in relation to the neck and thorax. In its most severe form, the scapula may be connected by bone to the cervical spine and limit neck movement.

Hemiatlas is a malformation of the first cervical vertebra, which may cause severe, progressive torticollis. In time, the deformity becomes fixed; therefore, posterior fusion is recommended. Basilar impression is a condition resulting from anomalies at the base of the skull and vertebrae, which lead to a short neck, headache, neck pain, and cranial nerve

palsies due to compression of the cranial nerves. Many congenital conditions, including Klippel-Feil syndrome, achondroplasia, and neurofibromatosis, may cause basilar impression. Commonly associated with basilar impression is occipitocervical synostosis, a condition in which fibrous or bony connections between the base of the skull and the atlas cause neck pain, torticollis, high scapula, and several neurologic symptoms.

Atlantoaxial Instability

Several congenital conditions may be associated with atlantoaxial instability and predispose the patient to cervical subluxation. In addition to Down and Klippel-Feil syndromes, these include other skeletal dysplasias and os odontoideum (aplasia or hypoplasia of the odontoid). Children with these conditions should be screened for atlantoaxial instability. Morquio syndrome is a mucopolysaccharidosis resulting in flattening of the vertebrae and multiple skeletal dysplasias. The odontoid process of the axis is underdeveloped and may lead to atlantoaxial subluxation.

Other Congenital Causes

Benign paroxysmal torticollis of infancy presents as recurrent episodes of torticollis in association with pallor, agitation, and vomiting. Typical onset is between 2 and 8 months of age, and the condition tends to remit by 2 to 3 years.

Miscellaneous Causes of Neck Stiffness

Head tilt, neck stiffness, and/or torticollis has been reported in several other conditions, some of which are life-threatening and others generally benign.

Ophthalmologic, Neurologic, and/or Vestibular Causes

Head tilt or neck malposition may result from abnormalities of vision (strabismus, cranial nerve palsies, extraocular muscle palsies, refractive errors) or the vestibular apparatus. The child attempts to correct for the disturbance through changes in neck position. Careful ophthalmologic and neurologic examinations of the child with head tilt are necessary to exclude these possibilities. Torticollis has also been reported in patients with migraine headaches.

Myasthenia Gravis

Patients with myasthenia gravis may develop torticollis, but ptosis, impairment of extraocular muscular movement, and other cranial nerve palsies are generally earlier signs.

Sandifer Syndrome

Sandifer syndrome is the constellation of torticollis, gastroesophageal reflux, and hiatal hernia. Children with this syndrome may have recurrent vomiting and failure to thrive.

Spontaneous Pneumomediastinum

Spontaneous pneumomediastinum may present with neck pain and torticollis. A history of severe coughing and/or retching is usually elicited. Crepitus is generally palpated along the neck. Initial therapy is directed at the underlying disease process.

Spasmus Nutans

Spasmus nutans is an acquired condition of childhood, characterized by nystagmus, head nodding, and torticollis. Children with these findings typically become symptomatic in the first 2 years of life. The condition is generally benign and self-limited. However, some children with the symptoms of spasmus nutans have underlying brain tumors. Therefore, imaging of the brain is necessary to exclude this possibility.

Dystonic Reaction

Certain drugs with dopamine-2 receptor antagonism can cause acute dystonic reactions with torticollis. These include many neuroleptic and antiemetic agents, such as haloperidol, prochlorperazine, and metoclopramide. Treatment with diphenhydramine (1 to 2 mg/kg per dose) may be diagnostic and therapeutic.

Psychogenic

Hysterical patients may present with torticollis. This diagnosis can be made only after excluding other more serious causes.

EVALUATION AND DECISION

The approach to the child with a stiff or malpositioned neck should focus initially on whether there is spinal cord involvement, as detailed in Figure 46.1. For any child with neck stiffness or pain, a history of weakness or paresthesias of the extremities or of functional abnormalities of the bowel or bladder should be sought. In addition, a complete ophthalmologic and neurologic examination should be performed, with the latter focusing on spinal cord function. Included in this examination should be an assessment of muscle strength, sensation, deep tendon reflexes, the Babinski reflex, and anal tone. Extra vigilance must be used if the patient is too young or incapacitated to give an accurate history.

FIGURE 46.1. Approach to the child with stiff or malpositioned neck. *C-spine,* cervical spine radiograph; *MRI,* magnetic resonance imaging; *ENT,* ear, nose, throat; *AVM,* arteriovenous malformation; *CT,* computed tomography; *CBC,* complete blood count; *ESR,* erythrocyte sedimentation rate; *TB,* tuberculosis; *SCM,* sternocleidomastoid.

If spinal cord involvement is detected, neurosurgical consultation and imaging of the cervical spine (radiographs and MRI of the cervical spine) are necessary. Conditions causing cervical spinal cord compromise may rapidly lead to permanent disability or death if not immediately addressed. If secondary to trauma, one should suspect cervical spine fracture or subluxation, spinal epidural hematoma, or SCIWORA syndrome. In the setting of fever, a spinal epidural abscess should be considered. Atlantoaxial subluxation secondary to otolaryngologic diseases or procedures should be considered in children with spinal cord involvement and consistent histories. Finally, spinal cord tumors and other space-occupying lesions should be considered if the development of symptoms is gradual and not associated with trauma or fever.

The next issue that should be considered is whether the neck stiffness is the result of an acute traumatic event. If acute trauma is the cause of the neck stiffness, the cervical spine should be properly immobilized (see Chapter 106) and multiple radiographic views of the cervical spine obtained. Fractures and subluxations/dislocations will generally be identified on plain radiography of the cervical spine. Other modalities (e.g., flexion-extension views, CT, MRI) may be useful to detect ligamentous injury, rotary subluxation, or spinal epidural hematomas. Muscle strain and/or contusion is a diagnosis of exclusion in the setting of trauma and neck stiffness after the possibility of more serious conditions are excluded. If other symptoms in addition to the neck stiffness are present, appropriate studies should be obtained. For example, the patient with neck stiffness and headache may have a subarachnoid hemorrhage for which a head CT scan would be indicated. The patient with clavicle fracture may have spasm of the SCM muscle and torticollis. However, tenderness is localized over the injured clavicle, and radiographs will confirm the diagnosis.

Fever in the setting of neck stiffness suggests the presence of an infectious, inflammatory, or neoplastic process. The presence of meningitis must be excluded either clinically or with a lumbar puncture (see Chapter 84). On examination, the presence of meningismus must be determined. A lumbar puncture should be seriously considered in the presence of fever and neck stiffness of any type because meningitis may present with fever and atypical neck signs. Helpful supporting signs include Brudzinski's sign (flexing the neck, eliciting flexion of the knee and hip) and Kernig's sign (with the hip flexed, pain with extension of the leg). Other conditions (e.g., subarachnoid hemorrhage) may also present with fever and meningismus, and a lumbar puncture is helpful in evaluating these conditions as well.

After the presence of meningitis has been excluded in the febrile patient with neck stiffness, the examination should focus on the presence or absence of a cervical mass. If a cervical mass is identified, a history of contact with cats and constitutional symptoms suggestive of malignancy should be elicited. If the cervical mass is tender, a trial of antibiotics directed at the most common bacterial pathogens and the placement of a PPD skin test to screen for tuberculosis may be all that is necessary. If the cervical mass does not respond to an appropriate trial of antibiotics, cat-scratch disease, atypical mycobacterial infection, or malignancy may be the cause.

If no palpable cervical mass is present, a more in-depth evaluation may be necessary, based on the history and physical examination of the child. Radiographs of the cervical spine will diagnose retropharyngeal abscess in the febrile child with stridor, drooling, and neck stiffness and may detect atlantoaxial subluxation in the child with otolaryngologic disease or who has recently had an otolaryngologic procedure. Radiographs of the cervical spine may also be useful in detecting other diseases involving the cervical spine, including vertebral osteomyelitis, infectious discitis, intervertebral disk calcification, and neck stiffness from collagen vascular disease. White blood count or ESR will be elevated in most children with these conditions, as well as those with spinal epidural abscesses and infections of the head and neck (e.g., tonsillitis, mastoiditis). If plain radiography is not diagnostic, technetium scans will identify vertebral osteomyelitis or discitis. CT or MRI of the spine will identify spinal epidural abscesses and can be helpful if routine radiographs are equivocal in several of the previously described conditions. Finally, an upper lobe pneumonia identified on chest film may be the cause of neck stiffness in the febrile child.

In the afebrile child with neck stiffness, the presence of a cervical mass within the SCM suggests congenital muscular torticollis (in an infant) or a SCM hematoma or tear. Radiographs of the cervical spine should be obtained to exclude more serious conditions. If the cervical mass is not within the SCM, a malignancy or atypical infection may be the cause and a complete blood count and biopsy of the mass should be considered.

For the afebrile child with neck stiffness and/or malposition of the neck and no cervical mass, a careful ophthalmologic and neurologic examination should be performed to exclude the possibility of a brain tumor, other space-occupying lesions of the brain, visual disturbances, and vestibular disturbance causing the abnormal neck posture. Often, the patient does not have true neck pain but is attempting to correct for these disturbances through changes in head position. A head CT scan is necessary to exclude the possibility of a space-occupying lesion of the brain, including brain tumor.

The child with myasthenia gravis generally has ptosis and weakness of extraocular muscles and may develop torticollis. A trial of intravenous edrophonium chloride (Tensilon) is diagnostic because symptoms will improve immediately; however, edrophonium chloride should not be given to young infants, who are especially prone to this agent's ability to cause cardiac arrhythmias. Children with torticollis after taking neuroleptic or antiemetic medications will usually respond to intravenous diphenhydramine.

Finally, the child with neck stiffness without fever, cervical mass, or abnormal ophthalmologic or neurologic examination may have any of a number of conditions. Many of the disorders mentioned as typically being associated with fever are commonly seen without fever as well (atlantoaxial subluxation in the child with otolaryngologic diseases or after otolaryngologic procedures, intervertebral disc calcification, collagen vascular disease). Furthermore, infants with congenital muscular torticollis may not have SCM masses that are detectable on physical examination. Some children with neck stiffness may have dysmorphic features, suggesting specific skeletal malformation syndromes or cervical subluxation in a child with Down syndrome. Chronic symptoms may suggest a congenital syndrome, collagen vascular disease, or a neoplastic process, but children with these conditions may also present with acute onset of symptoms. Osteoid osteoma and other benign tumors of the head and neck may be detected by plain radiography of the cervical spine. A chest radiograph is indicated for the child with neck stiffness in association with a history of severe coughing and/or retching because an upper lobe pneumonia or spontaneous pneumomediastinum may be the cause. Finally, if no cause can be identified after a complete history, detailed examination, and careful radiographic and laboratory evaluation, muscle spasm or hysteria may be the cause of torticollis.

In conclusion, neck stiffness and/or malposition may indicate a wide array of medical and traumatic conditions, both life-threatening and relatively benign. A careful examination must be performed to exclude the presence of spinal cord involvement. Trauma and infection are the most important causes of neck stiffness in children, and a history of trauma or fever will help guide the evaluation and decision making. Cervical spine fracture, subluxation/dislocation, and meningitis remain the most important diagnoses to exclude.

Suggested Readings

Cheng JCY, Au AWY. Infantile torticollis: a review of 624 cases. J Pediatr Orthop 1994;14:802–808.

Cushing AH. Diskitis in children. Clin Infect Dis 1993;17:1–6.

Dias MS, Pang D. Juvenile intervertebral disc calcification: recognition, management and pathogenesis. Neurosurgery 1991;28:130–135.

Dietrich AM, Ginn-Pease ME, Bartkowski HM, King DR. Pediatric cervical spine fractures: predominantly subtle presentation. J Pediatr Surg 1991;26:995–1000.

Epps HR, Salter RB. Orthopedic conditions of the cervical spine and shoulder. Pediatr Clin North Am 1996;43:919–931.

Grogaard B, Dullerud R, Magnaes B. Acute torticollis in children due to atlanto-axial rotary fixation. Arch Orthop Trauma Surg 1993;112: 185–188.

Gupta AK, Roy DR, Conlan ES, Crawford AH. Torticollis secondary to posterior fossa tumors. J Pediatr Orthop 1996;16:505–507.

Rigamonti D, Liem L, Wolf AL, et al. Epidural abscess in the cervical spine. Mount Sinai J Med 1994;61:357–362.

Samuel D, Thomas DM, Tierney PA, Patel KS. Atlanto-axial subluxation (Grisel's syndrome) following otolaryngological diseases and procedures. J Laryngol Otol 1995;109:1005–1009.

Schwartz GR, Wright SW, Fein JA, et al. Pediatric cervical spine injury sustained in falls from low heights. Ann Emerg Med 1997;30: 249–252.

CHAPTER 47

Odor—Unusual

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The human nose is able to discriminate approximately 4000 odors! Occasionally, parents bring an infant or child to the emergency department (ED) complaining of an unusual smell. Adolescents are more likely to note a new or unusual odor themselves and present to the ED with specific complaints.

Unfortunately, olfaction is a sense that most medical professionals are not trained to use, quantify, or describe. Before the development of sophisticated laboratory tests, clinicians relied heavily on the sense of smell. Even today, early diagnostic clues that can lead the clinician to a more selective workup, rapid diagnosis, and prompt therapeutic intervention may be obtained. In some situations, a prompt diagnosis can be lifesaving or can improve the quality of life.

PATHOPHYSIOLOGY

The olfactory area extends from the roof of the nasal cavity approximately 10 mm down the septum and superior turbinates bilaterally. The exact mechanism of stimulation of the olfactory receptors is unknown. Smell is more acute in the darkness and is believed to be linked to blood cortisol levels.

The unique odor emitted by a person is produced by a combination of body secretions and excretions, particularly those from the oropharynx and nasopharynx and the respiratory tract, plus aromas from the skin and cutaneous lesions, urine, feces, and flatus. The most significant components of odor in healthy humans are the apocrine glands. These secretions are initially odorless, but bacterial breakdown that results in fatty acid production can cause an offensive odor. Body odor is altered by hygiene, metabolism, toxins, infections, and systemic diseases.

When a child is unable to detect odor, anosmia should be considered. When a child complains of strange odors, especially if no one else is able to identify them, temporal lobe epilepsy should be contemplated.

DIFFERENTIAL DIAGNOSIS

A number of conditions, including metabolic disorders, dermatologic conditions, intoxications, infections, foreign bodies, various abnormalities of the body orifices, and a variety of systemic diseases, may result in an abnormal body odor (Table 47.1).

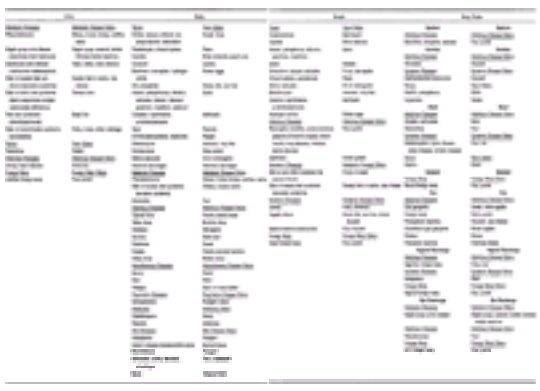

Table 47.1. Clinical Source and Cause of Unusual Odors

Metabolic Disorders Table 47.2)

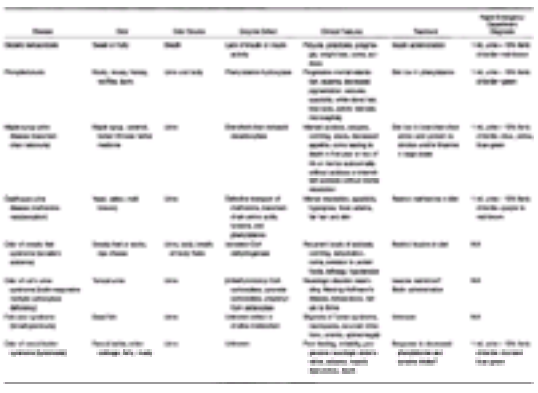

Table 47.2. Metabolic Disease Associated with Unusual Odors

The most common metabolic disorder that has a characteristic odor is diabetic ketoacidosis (DKA). The characteristic breath odor is caused by acetone and is described as sweet or fruity. It is important to note that any condition that results in a marked metabolic acidosis and ketosis will result in the characteristic sweet or fruity breath.

Inborn errors of metabolism that result in altered body, breath, or skin odors are unusual individually, but as a composite, they reflect a significant percentage of life-threatening illnesses of infancy (see Chapter 98). Although definitive diagnosis depends on specific identification of serum and urine amino and organic acid levels, many such conditions are associated with a positive ferric chloride test, which when performed in the ED can yield presumptive diagnosis.

Phenylketonuria is a disorder of amino acid metabolism associated with a deficiency of phenylalanine dehydroxylase and dihydropteridine reductase, which forces use of minor metabolic pathways of phenylalanine, resulting in the buildup of phenylacetic acid. It is the buildup of phenylacetic acid in the sweat and urine that causes a musty, mousy, horsey, wolflike, or barny odor. Clinical features of untreated phenylketonuria include white-blond hair, blue eyes, fair complexion, eczema, microcephaly, hypertonicity, increased risk for pyloric stenosis, seizures, and progressive mental deterioration. Although neonatal screening detects most of these cases, the observation of a characteristic odor in an infant should prompt appropriate laboratory studies, which may include a ferric chloride test in the ED. Prompt diagnosis and dietary restriction of phenylalanine promote a normal outcome.

Maple syrup urine disease is caused by a metabolic defect in the decarboxylation of the ketoacids of the branch chain amino acids (leucine, isoleucine, and valine), which results in their accumulation in the blood. It is apparently a metabolite of isoleucine in the urine that results in the characteristic odor of maple syrup, caramelized sugar, or boiled Chinese herbal medicine. Children with this disorder can have variable clinical manifestations, ranging from decreased appetite, vomiting, and ataxia to progressive acidosis, seizures, coma, and death. Prompt diagnosis and limitation of dietary branched chain amino acids promotes normal development.

Oasthouse urine disease, or methionine malabsorption syndrome, is caused by defective transport of methionine and, to a lesser extent, leucine, isoleucine, valine, tyrosine, and phenylalanine by the intestines and kidneys. The unabsorbed methionine in the gut is broken down by colonic bacteria to a-hydroxybutyric acid, which causes the characteristic odor described as yeast, celery, malt, or a brewery. Clinical presentation includes fair hair and skin, hyperpnea, extensor spasms, fever, edema, and mental retardation. Successful treatment consists of a methionine-restricted diet.

The odor of sweaty feet syndrome, or isovaleric acidemia, is caused by a defect in the catabolism of leucine. The characteristic odor described as sweaty feet or socks or ripe cheese comes from the buildup of isovaleric acid. Clinically, children experience vomiting, dehydration, acidosis, and slowly progressive mental deterioration. Treatment consists of restriction of leucine in the diet.

In the odor of cat's urine syndrome, the enzymatic defects are in the biotin-dependent enzymes b-methylcrotonyl-CoA carboxylase, pyruvate carboxylase, and propionyl-CoA carboxylase. The cause of the distinctive aroma of cat urine is unknown. Clinically, children have failure to thrive, ketoacidosis, and neurologic symptoms similar to Werdnig-Hoffmann's disease. Treatment consists of a low-leucine diet and the addition of biotin.

Fish odor syndrome, trimethylaminuria, results from an unidentified defect that possibly relates to choline metabolism. The dead fish odor in the urine results from buildup of trimethylamine. Clinical presentation includes stigmata of Turner syndrome, normal complement of chromosomes, neutropenia, recurrent pulmonary infections, and abnormal platelet function.

The odor of rancid butter syndrome, tyrosinosis, results from an unidentified defect in metabolism. It is hypothesized that a build-up of a-ketogammamethiolbutyric acid in the urine results in the characteristic smell of rancid butter. Clinical presentation includes poor feeding, irritability, seizures, coma, progressive neurologic deterioration, and early death secondary to infection and liver failure. In some cases, restriction of dietary phenylalanine and tyrosine has been helpful.

Dermatological Conditions

Many dermatologic diseases (Table 47.1) are associated with specific odors. Any cause of hyperhidrosis results in an

offensive body odor. Hidradenitis has a characteristic pungent odor, whereas Darier's disease is noted to have a pervasive aroma of burned tissue. An abscess or cellulitis is identified by the characteristic odors of the responsible microorganisms.

In burn patients, there is the typical odor of charred flesh, which when infected with *Pseudomonas,* takes on a characteristic sweet, grapelike odor.

Toxicologic Considerations

Recognition of a characteristic odor is vital for rapid, accurate diagnosis and treatment of some potentially lethal ingestions before laboratory identification (Table 47.1) (see also Chapter 88).

Penicillins give off an ammoniacal scent, whereas cephalosporins are noted to have a musty odor. Topical benzoyl peroxide, applied in large quantities, emits a pungent, pervasive aroma.

A strong garlic odor is typical of arsenic, arsine gas, phosphorus, tellurium, parathion, malathion, selenium, dimethyl sulfoxide, and thallium. The odor of bitter almonds or peach pits is indicative of cyanide poisoning, in which the degree of excretion of the odor parallels toxicity. Vacor (pyridylmethyl nitrophenylurea), an extremely potent rat poison that works as a potent pancreatic b-cell toxin, caused numerous cases of severe diabetes mellitus after overdose before it was withdrawn from the U.S. market in 1979. It often presented with an odor of peanuts.

Diagnostic odors are found in several sedative-hypnotic medications that primarily have central nervous system (CNS) manifestations. Ethchlorvynol (Placidyl) is a volatile agent that has an aromatic plastic or vinyl-like breath odor. Ingestion results in coma, hypothermia, respiratory depression, hypotension, and bradycardia. An overdose of chloral hydrate can result in CNS depression ranging from slurred speech, ataxia, and incoordination to deep coma, gastritis, and cardiac arrhythmias. It may be seen in children or as an intentional overdose in adults, and it imparts a fruity, pearlike scent. Disulfiram (Antabuse) gives the breath a rotten egg odor because of the sulfide metabolites. The pleasant smell of oil of wintergreen indicates methyl salicylate poisoning.

Infectious Diseases

Many microorganisms produce characteristic odors that suggest the diagnosis of their respective infectious diseases by olfaction alone (Table 47.1) (see also Chapter 84). Omphalitis in the newborn can be life-threatening. It presents with a foul or putrid odor associated with a draining, erythematous umbilical area. Less common infections that have been historically associated with characteristic odors include typhoid's aroma of freshly baked bread, yellow fever's butcher shop smell, smallpox's menagerie odor, scrofula's odor of stale beer, diphtheria's sweet smell, and rubella's scent of freshly plucked feathers.

Foreign Bodies

Foreign bodies are capable of producing a foul odor that results from secondary bacterial colonization or infection. Foreign body odors can be localized to a particular orifice, or they may pervade a patient's clothing, body, and surrounding environment. Foul-smelling, fetid, or feculent odors indicate anaerobic infections, whereas a sickly sweet odor is associated with *Escherichia coli,* and *Clostridia* is associated with a mousy odor.

Orifice Odors

Specific orifice odors can be diagnostic of infectious disease processes.

Oropharynx

A healthy mouth does not give off an offensive odor. Halitosis, or bad breath, is the result of a release of volatile sulfur compounds formed when the oral flora metabolizes amino acids from compounds in the saliva that adhere to the tongue, teeth, and gums. Halitosis is increased in states of diminished solid and liquid intake. Tonsillitis (see Chapter 71 and Chapter 84) has an offensive odor, and group A b-hemolytic streptococcus gives off a characteristic “strep breath” smell. Dental abscesses (see Chapter 124) and acute ulcerative gingivitis (Vincent's stomatitis or trench mouth) are associated with a penetrating, offensive odor. The oropharynx is also the portal of exit for deeper infections. Lung abscesses, empyema, bronchitis, and bronchiectasis result in foul breath and sputum. Nasal foreign bodies in toddlers are usually associated with an odor identified by parents as bad breath.

Nose

Nasal drainage can be clear and odorless or mucopurulent and odiferous. Nasal drainage and bleeding can reflect local infections, foreign bodies, irritations of the nasal passage, and sinus drainage.

Ear

Sterile inner ear fluid is odorless but gives off a rank smell when infected. Acute otitis externa usually is associated with a mucoid drainage, whereas chronic otitis externa produces a purulent, discolored drainage with a foul odor, usually secondary to*Pseudomonas aeruginosa* or *Staphylococcus aureus.*

Genitalia

Vaginal secretions are combinations of vulvar secretions from sebaceous, sweat, Bartholin's and Skene's glands,

transudate through the vaginal wall, exfoliated cells, cervical mucus, endometrial and oviductal fluids, plus vaginal microorganisms and menstrual blood. These secretions are hormonally mediated and vary with the menstrual cycle. Odors are exacerbated by the presence of retained foreign bodies, including tampons and diaphragms.

Bacterial vaginosis (nonspecific vaginitis, *Gardnerella* vaginitis, *Corynebacterium* vaginitis, *Haemophilus* vaginitis, nonspecific vaginosis, and anaerobic vaginosis) is caused by an increase in anaerobic bacteria and a decrease in lactobacilli (see Chapter 94). The anaerobic bacteria act synergistically with *Gardnerella vaginosa* to produce enzymes and aminopeptidases that degrade protein and decarboxylases that convert amino acids and other compounds to amines. The amines produce the characteristic “fishy” odor, which is best detected by alkalinization using 10% potassium hydroxide placed directly on a vaginal swab and smelling immediately. This odor also can be indicative of sexual abuse in children. Vaginal infection with *Trichomonas* often is associated with a fishy odor, whereas *Candida* vaginitis is notably free of odor (see Chapter 94).

A male counterpart, balanoposthitis, is associated with a urethral discharge that produces a fishy odor when alkalinized because of the same process and organisms as occur in bacterial vaginosis.

Urethral Meatus

A urinary tract infection caused by urea-splitting bacteria will emit an ammoniacal odor.

Rectum

Stool odors vary with diet, medications, and microbiologic flora. Various malabsorptive syndromes, such as sprue, cystic fibrosis (see Chapter 96), and Whipple's disease, are associated with foul-smelling stool. The presence of blood in the stool has a distinctive, pungent odor, as does pus. *Shigella* and *Salmonella* (see Chapter 84) have distinctive rank odors.

Systemic Diseases

Several nutritional syndromes (Table 47.1), such as pellagra's stench of sour or musty butter and the putrid or fetid odors of scurvy and gout, have unique odors. Schizophrenia has a characteristic body odor described as heavy, unpleasant, and pungent. The odor-producing substance is *trans*-3-methyl-2-hexanoic acid, which is produced in the sweat. Uremic breath is produced by secondary and tertiary amines, dimethylamines, and trimethylamines that produce a fishy odor. Malignancy—especially when associated with an expanding external mass, bleeding, and necrosis—gives off a trenchant odor because of tissue and cellular breakdown plus gas formation. Hepatic failure gives an odor of “fetor hepaticus” (described as musty, rotten eggs, or garlic) and is noted in the breath or urine. In Crohn's disease (see Chapter 93), the development of gastric fistulae often are heralded by a feculent odor.

A physiologic odor that often heralds the onset of puberty is underarm body odor. This is usually the earliest sign of puberty and precedes all other physical changes. Age of onset is around 6 to 8 years and reflects the onset of adrenarche. Dehydroepiandrosterone sulfate (DHEAS) is the androgen believed to be responsible for the pungent aroma of underarm body odor and can be measured for confirmation. Although the adrenal and hypothalamic–pituitary–gonadal axis are separate systems involved with the onset of puberty, they often become active nearly simultaneously.

EVALUATION AND DECISION

The evaluation of a child who presents to the ED should incorporate all of the senses, including smell (Fig. 47.1). Both presence and absence of odors can be diagnostic. Each person has a unique odor, ranging from pleasant to offensive. Using the sense of smell should be done in stages; an initial evaluation of the prevailing odor of the examination room, followed by attention to overall body odor and identification of odors from individual orifices and body fluids. Body fluids such as ocular, ear, nasal, sinus tract, or umbilical drainage; vomitus; sputum; genital discharge; stool; ulcers; and superinfection of the dermis have unique identifiable odors.

FIGURE 47.1. Evaluation and decision for unusual odors. *DKA,* diabetic ketoacidosis.

Good or poor hygiene is readily detected in a closed examination room. When an unusual odor is detected, the history should include information about medications (topical, oral, or rectal), onset and duration of odor, methods used to alter odor, unusual drainage from body orifices, suspicion of foreign body, fever, and other pertinent symptoms.

In the evaluation of the significance of odors, attention must be paid to the child's age and developmental level. At birth,

infant odors are a conglomeration of their own and their mother's physical environment. After birth, a well-cared-for, healthy infant should have a very pleasing aroma and odorless breath. Offensive body odor in a newborn suggests an inborn error of metabolism, a localized infection such as omphalitis, or neglect. During infancy, inborn errors of metabolism are relatively common (Table 47.3) and potentially life-threatening causes of unusual odor (Table 47.4). Infection localized to the umbilicus, omphalitis, produces a foul odor and is easily diagnosed on the basis of erythema, induration, and discharge. Other sources found in older children (foreign bodies, ingestions, and pharyngitis) are unusual in the infant.

Infants	Localized infections
Inborn errors of metabolism	Stomatitis
Omphalitis	*Adolescents*
Neglect	Localized infections
Toddlers	Pharyngitis
Foreign bodies	Tonsillitis
Otic	Vaginitis
Nasal	Toxins
Vaginal	Alcoholic beverages
Urethral	Puberty
Rectal	

Table 47.3. Common Causes of Unusual Odors

Metabolic Disease	Isopropyl alcohol
Inborn errors of metabolism	Methyl salicyate
DKA	Vacor
Infectious Diseases	*Systemic Disease*
Omphalitis	Uremia
Diphtheria	Liver failure
Lung abscess	Gastrointestinal obstruction
Toxins	Peritonitis
Arsenic	
Cyanide	

Table 47.4. Life-Threatening Causes of Unusual Odor

In an older child, the physician should determine a child's history and whether clinical signs of chronic systemic diseases such as diabetes, liver failure, or uremia are present. The child with ketoacidosis usually appears dehydrated and manifests deep, rapid (Kussmaul) respirations; the breath may smell of ketones. Uremia develops in patients with renal failure who may have short stature, edema, hypertension, and a characteristic fishy odor of their breath. With liver failure, the patient may have jaundice, ascites, lethargy, or mental status changes, as well as breath and urine that takes on the odor of rotten eggs or garlic.

Body odors change with puberty through hormonally induced metabolic changes. The most significant change is the development of axillary odor related to apocrine secretions that are retained or spread by axillary hair. Normal quantities of sweat have a barely perceptible odor, whereas increasing quantities of sweat production cause increasingly noticeable and offensive odor.

Early on, the physician should determine the potential risk of ingestion of a toxic substance. In the adolescent, the risk of a significant ingestion increases and can be life-threatening (Table 47.4). Next, the physician should ascertain whether the odor emanates from a particular body orifice such as the ear, nose, pharynx, vagina, urethra, or rectum. Nasal foreign bodies are particularly common in children between the ages of 1 and 5 years. They may remain hidden for weeks, long beyond the child's memory of placing the object, and eventually, they produce a secondary infection that leads to a foul discharge. In some cases of foreign body, the odor may be so strong that it appears to be generalized. Thus, a careful examination of the various orifices, with particular attention to the nares, always is advisable. The presence or absence of fever is another crucial variable in the evaluation of odor. Fever suggests an infectious cause, either systemic or localized. Pharyngitis and tonsillitis characteristically cause foul odor to the breath, much like a lung abscess. Occasionally, parents state that their child smells as if he or she had a “strep throat.” Although foreign bodies occasionally produce obstruction and a secondary infection of severity sufficient enough to evoke a febrile response, in most cases, the inflammation is localized and the child is afebrile.

In the absence of a known history or obvious findings of chronic systemic disease, a visible foreign body, known ingestion, or fever, the clinician should perform a careful physical examination and a urinalysis. DKA always causes glucosuria and ketonuria, but hepatic or renal failure may be less obvious. In addition, inborn errors of metabolism may first manifest well beyond the newborn period. Foreign bodies may defy routine attempts at visualization and, based on a high index of suspicion, require endoscopy or imaging procedures. Therefore, persistence or an unusual odor or concern for chronic toxicity merits further laboratory evaluation. In cases in which an explanation of the odor is not uncovered, a follow-up evaluation in 2 to 3 days is prudent.

In patients who have died in the ED, a faint fecal odor usually is noted, possibly related to the release of intestinal

contents to the atmosphere. Unique identifiable odors detected at the time of death or autopsy can direct laboratory evaluation toward possible causes of death.

Suggested Readings

Chiang WK. Otolaryngologic principles. In: Goldfrank LR, Flomenbaum NE, Lewin NA, et al, eds. Toxicologic Emergencies. 6th ed. Stanford, CT: Appleton & Lange, 1998:457–471.

Rezvani I, Rosenblatt DS. An approach to inborn errors of metabolism. In: Behrman RE, Kliegman RM, Arvin AM, eds. Textbook of Pediatrics. 15th ed. Philadelphia: WB Saunders, 1996:328–329.

CHAPTER 48

Oligomenorrhea

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In this chapter, possible causes of oligomenorrhea and secondary amenorrhea are reviewed. *Oligomenorrhea* means infrequent menstruation and can be defined for the pediatric emergency physician as an interval of more than 6 weeks between two menstrual periods. If menstrual cycles do not resume within 3 to 6 months, the term *secondary amenorrhea* is applied. Some patients with anovulatory menstrual cycles have oligomenorrhea punctuated by episodes of excessive bleeding. An approach to the evaluation of abnormal vaginal bleeding is presented in Chapter 76.

Oligomenorrhea should be distinguished from hypomenorrhea, a nonpathologic pattern of light but regular menstrual periods. This chapter does not include a separate consideration of primary amenorrhea—that is, failure to menstruate by a specified age, often 16 years. However, some disorders discussed here can produce primary rather than secondary amenorrhea as part of an overall delay in pubertal development.

The differential diagnosis of oligomenorrhea is given in Table 48.1.

I. Hypothalamic-Pituitary Axis Disorders	3. Pelvic irradiation
A. Disorders of weight and/or energy expenditure	4. Autoimmune diseases
1. Anorexia nervosa	B. Hormone-secreting tumors
2. Strenuous exercise	III. Uterine Disorders
3. Marked thinness or weight loss	A. Endometrial destruction
4. Chronic illness	1. Surgical
B. Delayed maturation	2. Tuberculous
C. Psychological stress	IV. Hyperprolactinemia
D. Central nervous system tumors	A. Lactation
E. Pseudocyesis	B. Drugs (see Table 48.2)
II. Ovarian Disorders	C. Pituitary adenoma
A. Ovarian failure	D. Hypothyroidism
1. Gonadal dysgenesis	V. Hyperandrogenism
2. Cancer chemotherapeutic agents	A. Polycystic ovary syndrome
	B. Adrenal disease
	VI. Miscellaneous conditions
	A. Pregnancy
	B. Hormonal contraception
	C. Hypothyroidism or hyperthyroidism

Table 48.1. Differential Diagnosis of Oligomenorrhea Organized by Pathophysiology of Disorder or Condition

EVALUATION AND DECISION

Diagnosis of Pregnancy

“Is she pregnant?” is always the first question to answer in evaluating an adolescent with one or several missed menstrual periods (Fig. 48.1). If the patient is not pregnant, her evaluation can proceed at a more deliberate pace. If she is pregnant, however, prompt diagnosis and referral are important for the teenager who intends to seek a therapeutic abortion, as well as for the one who plans to continue her pregnancy. Early and regular prenatal care are associated with reduced morbidity and mortality among pregnant teenagers and their offspring. Early diagnosis also affords the pregnant adolescent more time to decide on and arrange for a therapeutic abortion, if that is her choice. Because an adolescent under the age of 18 years must obtain parental or judicial consent for a therapeutic abortion, a delayed diagnosis of pregnancy may push the procedure into the second trimester, resulting in higher morbidity and higher cost.

FIGURE 48.1. Differential diagnosis of oligomenorrhea. *T,* testosterone; *LH,* luteinizing hormone; *FSH,* follicle-stimulating hormone; *PCOS,* polycystic ovary syndrome.

Early pregnancy is not always easy to recognize. Symptoms of fatigue, nausea, vomiting (not necessarily in the morning), urinary frequency, and breast growth or tenderness are common, but by no means are they universal or specific. On pelvic examination, the first indications of pregnancy are softening of the lower uterine segment (Hegar's sign) and of the cervix (Goodell's sign) at 4 to 6 weeks after the last menstrual period. By 6 weeks' gestation, the uterus changes from pear-shaped to globular, and by about 8 weeks, the vagina and cervix acquire a bluish hue (Chadwick's sign). These changes occur in ectopic as well as in intrauterine pregnancies. A serviceable rule is that the pregnant uterus grows to about the size of a tennis ball at 8 weeks after the last menstrual period, becomes baseball-sized at 10 weeks, and softball-sized at 12 weeks. When the uterus is retroflexed, its size is more difficult to assess and rectovaginal palpation should be done. After 12 weeks, the uterine fundus is palpable above the symphysis pubis on abdominal examination. Fetal movement can be discerned after about 16 weeks. The fundus reaches the level of the umbilicus at 20 weeks' gestation.

Some patients may report the result of a home pregnancy test. Although manufacturers claim an accuracy rate of 99% for home pregnancy tests, accuracy of patient-administered tests has been as low as 77%, with false-positive test results being somewhat more common than false-negative ones. The emergency physician is well-advised to order a standard laboratory test to confirm the results of a home pregnancy test.

The emergency physician should know which urine or serum pregnancy test is used by his or her laboratory and that test's level of sensitivity. Radioimmunoassays and immunoenzymometric assays detect the core fragment of human chorionic gonadotropin (hCG), regardless of whether the fragment is free or a component of the intact hCG molecule. These tests, commonly known as b-hCG tests, are specific and sensitive, identifying total levels of hCG as low as 5 mIU/mL in serum and 25 mIU/mL in urine. Because the serum concentration of hCG in pregnant patients reaches 100 mIU/mL by about the time of the next anticipated menstrual period, the clinician can detect pregnancy in nearly all patients within about 9 days after conception (before the menstrual period has been missed) and in essentially all patients who have missed a period, including those with ectopic pregnancies that produce abnormally low levels of hCG.

If a patient with one or several missed menstrual periods also complains of abdominal pain or abnormal vaginal bleeding, the diagnosis of *ectopic pregnancy* must be entertained. (The diagnosis of ectopic pregnancy is discussed at greater length in Chapter 76.) Three-fourths of women with ectopic pregnancies experience 1 to 12 weeks of amenorrhea before abnormal bleeding or pain eventually prompts them to seek medical attention. Pelvic inflammatory disease is an important risk factor for ectopic pregnancy (see Chapter 94). The rate of ectopic pregnancy per 1,000 reported pregnancies in the United States increased dramatically, from 4.5 in 1970 to 14.3 in 1986, and (based on outpatient as well as inpatient hospital data) to an estimated 19.7 in 1992. Although ectopic pregnancies occur rarely in adolescents, the increase in sexual activity and pelvic inflammatory disease rates among adolescents during the last quarter century has heightened their susceptibility to ectopic pregnancy. Between 1970 and 1986, the rate of ectopic pregnancy among young women between the ages of 15 and 24 years was 6.1 per 100,000 pregnancies.

Pseudocyesis is a rare cause of amenorrhea in women who believe they are pregnant and who exhibit many presumptive symptoms and signs of pregnancy, including nausea, vomiting, hyperpigmented areolae, galactorrhea, and abdominal distension. The diagnosis is made when a patient who insists that she is pregnant nevertheless has no true uterine enlargement, no demonstrable fetal parts or heart sounds, and a negative pregnancy test. Psychiatric consultation should be obtained for such patients.

Evaluation of Nonpregnant Patients

If the physician can answer “no” to our original question—“Is she pregnant?”—the evaluation of an adolescent with oligomenorrhea can proceed at a more leisurely pace (Fig. 48.1). During the first 2 years after menarche, irregular menstrual cycles are common. The average girl requires about 15 months to complete her first 10 cycles, and some girls take much longer. As a rule, if an adolescent who complains of oligomenorrhea is fewer than 2 years past menarche, is not sexually active, and has no signs suggestive of any specific cause of oligomenorrhea (hirsutism, obesity, galactorrhea, extreme thinness), further investigation is not warranted. She can be considered likely to have maturation that is delayed but within the range of normal. Of course, she should be reassured and followed.

Adolescents with a pattern of oligomenorrhea that has continued for longer than 2 years after menarche or that began after a regular menstrual pattern had already been established need further evaluation. In the interview, historical details about the patient's menstrual pattern, growth, endocrine and central nervous systems, psychological status, and medications should be sought specifically. On physical examination, the patient's height and weight, skin, breasts, and pelvis should be checked carefully. Because galactorrhea is not always spontaneous, the examiner should try to express

fluid manually from the patient's breasts. The completed examination will separate the majority of patients who have no notable abnormalities from a minority with the important findings of hirsutism, obesity, and galactorrhea.

HIRSUTE OR OBESE PATIENTS

Classically, hirsutism, obesity, ovarian enlargement, and amenorrhea or infertility constitute the clinical features of *polycystic ovary syndrome* (PCOS, previously the Stein-Leventhal syndrome). However, patients with PCOS are a heterogeneous group with varying combinations of these features. In one sample of women with polycystic ovaries diagnosed ultrasonographically, 61% were hirsute, 35% were obese, 71% had oligoamenorrhea, and 4% had menometrorrhagia. In another sample of women with polycystic ovaries on ultrasonography, 80% had menstrual irregularities, but the remaining 20% had both regular menstrual cycles and no biochemical evidence of hyperandrogenism. In some cases, clinically evident hyperandrogenism is accompanied by peripheral insulin resistance, hyperinsulinemia, and acanthosis nigricans (HAIR-AN syndrome). Most adolescents with biochemical evidence of PCOS do not have enlarged ovaries.

Pathophysiologically, PCOS involves chronic anovulation in association with hyperandrogenism and, in some patients, hyperinsulinemia. The anovulation results from a mildly increased production rate and circulating level of testosterone, decreased sex hormone–binding globulin (SHBG), and tonically rather than cyclically elevated estrogens. Perhaps as a result of a disturbance in the hypothalamic–pituitary– ovarian feedback system that may occur during puberty or even earlier, patients with PCOS have increases in both the amplitude and frequency of luteinizing hormone (LH) secretion and normal to low follicle-stimulating hormone (FSH) concentrations. Increased amounts of androstenedione and testosterone are secreted by the ovarian theca cells in response to stimulation by LH, and then are not converted to estradiol by aromatase because this enzyme's activity depends on local concentrations of FSH. In patients with hyperinsulinemia, increased insulin binding of insulinlike growth factor-I receptors also appears to augment the production of androgens by ovarian theca cells. The local excess of androgens inhibits the normal development of ovarian follicles. Peripherally, androstenedione is increased and is converted to estrone in adipose tissue. The excess peripheral androgens also cause a decrease in the concentration of SHBG. The resulting higher proportion of testosterone unbound to SHBG can stimulate excessive hair growth, even in patients whose total serum testosterone level (bound plus free) is normal or only minimally elevated. The reduction in SHBG also results in higher circulating levels of free estradiol. The persistently, rather than cyclically, elevated peripheral estrogen concentration stimulates continuing secretion of LH and maintains the state of chronic anovulation.

The goals of treatment for PCOS are to restore monthly menstrual cycles, to minimize hirsutism, to prevent the development of endometrial hyperplasia, and, it is hoped, to reduce the risk of endometrial adenocarcinoma, which occurs with increased frequency among patients with PCOS. Medroxyprogesterone or estrogen–progestin birth control pills are used to suppress ovarian or adrenal androgen production and to stimulate monthly menstrual bleeding.

Partial or late onset *congenital adrenal hyperplasia* is a rare cause of oligomenorrhea associated with hyperandrogenism that is usually indistinguishable clinically from PCOS. Other rare causes, including *Cushing's disease* and *ovarian and adrenal tumors*, should be suspected in patients with hirsutism accompanied by signs of glucocorticoid excess or virilization (marked acne, deepening of the voice, or clitoromegaly), and in those with testosterone levels above 200 ng/dL.

GALACTORRHEA

Hyperprolactinemia occurs in approximately 25% of adult women with secondary amenorrhea but is a much less common cause of oligomenorrhea in adolescents. Nevertheless, the possibility of hyperprolactinemia must be considered in all adolescents with oligomenorrhea because only 40 to 50% of hyperprolactinemic patients have spontaneous or expressible galactorrhea. The constellation of oligomenorrhea, galactorrhea, and hyperprolactinemia can be produced by drugs (Table 48.2) that block pituitary dopamine receptors or interfere in other ways with dopaminergic or serotoninergic central nervous system pathways, by the discontinuation of birth control pills, by cutaneous or neurogenic stimulation of the breasts, and by excessive secretion of prolactin itself (e.g., primary hypothyroidism, pituitary adenoma). Rarely, in hypothyroid patients, hypothalamic thyroid-releasing hormone acts as a prolactin-releasing factor, resulting in galactorrhea. Breast-feeding is an obvious physiologic cause of prolactin secretion and oligomenorrhea. The occasional patient with galactorrhea but a normal prolactin level should be reevaluated periodically in an effort to identify a treatable cause of the problem.

Antipsychotic and Antidepressant Agents	*Drugs Used to Treat Gastrointestinal Disorders*
Amoxapine (Asendin),	Cimetidine (Tagamet)
domipramine (Anafranil)	Metoclopramide (Reglan)
chlorpromazine (Thorazine),	*Antihypertensive Agents*
thioridazine (Mellaril),	Methyldopa (Aldomet)
prochlorperazine	Reserpine (Hydromox, Serpasil,
(Compazine), fluphenazine	others)
(Prolixin), other phenothiazines	Verapamil (Calan, Isoptin)
Haloperidol (Haldol)	*Opiates*
Pimozide (Orap)	Codeine
Risperidone (Risperdal)	Morphine
Thiothixene (Navane)	

Table 48.2. Partial List of Drugs That Can Cause Hyperprolactinemia and/or Galactorrhea

NORMAL AND THIN PATIENTS

Among adolescents who do not have hirsutism, obesity, or galactorrhea, suppression of the hypothalamic–pituitary axis is the most common cause of oligomenorrhea that occurs or persists for at least 2 years after menarche. *Abnormalities of body weight* are probably the most common sources of this central disturbance. A quarter of a century ago, Frisch and McArthur observed that menarche tends to occur only after adolescents have achieved a critical fat-to-body weight ratio of 17% and that, similarly, fat must constitute 22% of body weight for menstrual cycles to reappear in women with secondary amenorrhea. This is in accordance with the clinical observation that many patients with serious chronic illnesses, malnutrition, or rapid weight loss develop amenorrhea. The observations that amenorrhea often precedes substantial weight loss in patients with anorexia nervosa and that amenorrheic ballet dancers experience menarche and resume menses during intervals of rest unaccompanied by weight gain give credence to the hypothesis that psychological stress and energy expenditure deficits may have additional, independent effects on the hypothalamic–pituitary axis.

Accordingly, in assessing the nonpregnant adolescent with oligomenorrhea, one should inquire routinely about potential sources of significant emotional upset (e.g., family disruption, peer difficulties, depression), recent weight loss, chronic illness or other causes of poor weight gain, behavior characteristic of anorexia nervosa (Table 48.3), and *strenuous exercise* (especially long distance running, dancing, or gymnastics).

A. Refusal to maintain body weight over 85% of the expected weight for age or height, with the low weight a result of either weight loss or failure to gain.
B. Intense fear of gaining weight or becoming obese, even though underweight.
C. Disturbed perception of body weight or shape, undue influence of weight or shape on self-evaluation, or denial of the seriousness of the low weight.
D. In postmenarchal females, failure to menstruate during three or more consecutive anticipated cycles. (Menstruation induced by hormonal treatment is excluded.)

Table 48.3. DSM-IV Criteria for the Diagnosis of Anorexia Nervosa

Other diagnostic possibilities for oligomenorrheic patients with no abnormal physical findings include a wide variety of conditions. About half of women using contraceptive *medroxyprogesterone* injections for 12 months have amenorrhea; after 2 years of use, the proportion with amenorrhea is 68%. Amenorrhea also occurs in about 2% of menstrual cycles among patients taking *birth control pills* that contain 50 µg or less of estrogen. However, amenorrhea persisting 12 months after the last injection of medroxyprogesterone or 6 months after birth control pills have been stopped should be evaluated in the standard fashion. The diagnoses uncovered among patients with “postpill amenorrhea” are nearly as heterogeneous as those seen in amenorrheic patients who have never taken birth control pills.

Hypothyroidism and, less commonly, *hyperthyroidism* can produce menstrual irregularities. Many patients with hyperprolactinemia and oligomenorrhea do not have concomitant galactorrhea that would otherwise prompt a medical investigation. Similarly, although hirsutism and obesity are classic features of PCOS, many adolescent patients with oligomenorrhea and the endocrinologic abnormalities of PCOS lack one or both of these signs. A history of hot flashes, antineoplastic chemotherapy, pelvic irradiation, or autoimmune disease suggests the diagnosis of *premature ovarian failure*. *Endometrial destruction* that results from overly vigorous curettage or pelvic tuberculosis is a rare cause of oligomenorrhea.

DIAGNOSTIC PROCEDURES

Patients with oligomenorrhea but few other symptoms or signs of disease require laboratory evaluation to differentiate among the many potential causes of oligomenorrhea after pregnancy has been excluded (Fig. 48.1). Determinations of serum levels of FSH, testosterone (T) or free testosterone (free T), and prolactin are needed in order to corroborate the suspected diagnosis or to categorize the patient whose history and physical examination have provided few diagnostic clues. Other laboratory tests should be performed when warranted by the clinical situation. Prepubertal levels of LH and FSH indicate hypothalamic–pituitary suppression in a postmenarchal patient; this pattern is commonly found. An elevated LH:FSH ratio of greater than 2.5 can be found in about one-third of patients with PCOS. The finding of a mildly elevated total or free T level constitutes strong evidence for a diagnosis of PCOS. FSH values over 40 mIU/mL suggest ovarian failure as the cause for oligomenorrhea. An elevated prolactin level is likely to indicate a pituitary microadenoma in patients who are not using any of the drugs known to cause hyperprolactinemia and galactorrhea (Table 48.2).

The administration of exogenous progestin often is advocated as an in vivo test of ovarian and endometrial function. If the hypothalamic–pituitary axis is producing some gonadotropin, the ovaries are responding with some estradiol production, and the uterine endometrium is growing appropriately, then the addition of exogenous progestin (medroxyprogesterone acetate, 10 mg/day for 7 days) will be followed by at least scanty menstrual bleeding within 7 days after the treatment is completed. This menstrual flow, if it appears, provides the patient and her physician with tangible evidence of the basic integrity of these organs and indicates that anovulation is the source of the amenorrhea. For diagnosis in adolescents, however, laboratory investigation is much preferable to progestin administration because the latter procedure does not pinpoint the cause of the anovulation and because the selection of an appropriate treatment for

oligomenorrhea depends necessarily on the physician's identifying a specific explanation for the disorder.

Suggested Readings

Alper MM, Garner PR. Premature ovarian failure: its relationship to autoimmune disease. Obstet Gynecol 1985;66:27–30.

Bastian LA, Piscitelli JT. Is this patient pregnant? Can you reliably rule in or rule out early pregnancy by clinical examination? JAMA 1997;278:586–591.

Blythe MJ. Common menstrual problems of adolescence. Adolesc Med: State of the Art Rev 1997;8:87–109.

Chard T. Pregnancy tests: a review. Hum Reprod 1992;7:701–710.

Franks S. Polycystic ovary syndrome. N Engl J Med 1995;333:853–861.

Goldman S, Johnson FL. Effects of chemotherapy and irradiation on the gonads. Endocrinol Metab Clin North Am 1993;22:617–629.

Goudas VT, Dumesic DA. Polycystic ovary syndrome. Endocrinol Metab Clin North Am 1997;26:893–912.

Neinstein LS, Himebaugh KS. Galactorrhea. In: Neinstein LS, ed. Adolescent Health Care. 3rd ed. Baltimore: Williams & Wilkins, 1996:826–831.

Robinson S, Robin DA, Deacon A, Wheeler MJ, Clayton RN. Which hormone tests for the diagnosis of polycystic ovary syndrome? Br J Obstet Gynaecol 1992;99:232–238.

CHAPTER 49

Oral Lesions

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Oral lesions commonly occur in infancy and childhood and may represent a wide range of illnesses—from benign lesions that completely resolve without intervention to those associated with life-threatening diseases. The differential diagnosis includes a large number of localized congenital and acquired causes; however, lesions associated with systemic disease must also be considered (Fig. 49.1 and Table 49.1). Most often, patients with isolated complaints (e.g., a mouth sore or mass, drooling, pain, fever) represent common, self- limited conditions (Table 49.2). Systemic, and potentially life-threatening, diseases (Table 49.3) may present initially with isolated mouth findings, necessitating a complete history and physical examination in all patients with oral lesions.

FIGURE 49.1. Oral lesions.

Congenital Oral Lesions	**Syphilis**
Epstein's pearls	Acquired (secondary)
Epithelial pearls	Congenital
Bohn's nodules	Hairy tongue
Dental lamina cysts	***Tumorous Oral Lesions***
Natal teeth	Eruption cyst
Sputs (gum boil)	Oral papilloma
Lymphangioma	Fibroma
Hemangioma	Mucocele
Infectious Oral Lesions	Ranula
Candidiasis	Pyogenic granuloma
Herpes simplex virus (HSV)	Rhabdomyosarcoma
Gingivostomatitis—primary	***Oral Lesions Associated with Systemic Disease***
Labialis—recurrent (cold sore)	Systemic Disease
Hand–foot–mouth disease	Stevens–Johnson syndrome
Herpangina	Toxic shock syndrome
Scarlet fever	Mucositis
Streptococcal pharyngitis	Kawasaki disease
Measles	Crohn's disease
Varicella	Behçet's syndrome
Human immunodeficiency virus (HIV)	Epidermolysis bullosa
	Miscellaneous Oral Lesions
Dental–alveolar abscess	Aphthous stomatitis
Periadontitis	Geographic tongue
Acute necrotizing ulcerative gingivitis (trench mouth)	Gingival hyperplasia
	Leukoplakia

Table 49.1. Differential Diagnosis of Oral Lesions

Candidiasis	Hand–foot–mouth disease
Aphthous stomatitis	Herpangina
Herpes simplex virus	
Gingivostomatitis—primary	
Labialis—recurrent	

Table 49.2. Common Causes of Oral Lesions

Stevens-Johnson syndrome	Toxic shock syndrome
Kawasaki disease	Human immunodeficiency virus

Table 49.3. Life-Threatening Causes of Oral Lesions

PATHOPHYSIOLOGY

Oral lesions may result from localized or systemic pathophysiologic processes. Localized causes include congenital masses and cysts, infectious diseases, and oral tumors. Systemic illnesses with prominent oral involvement include a number of infectious and other inflammatory or toxin-mediated conditions. Given the broad spectrum of illnesses presenting with oral lesions, it is convenient to discuss individual causes under specific headings within the differential diagnosis. Several conditions with typical oral lesions exist that do not comfortably fit under any of these headings and are discussed in the section on miscellaneous oral lesions.

DIFFERENTIAL DIAGNOSIS

Congenital Oral Lesions

Most oral lesions present at birth or early infancy represent benign findings. Patients are largely asymptomatic, and the lesions resolve spontaneously.

Epstein's pearls occur in more than 60% of newborns as small, white milia in the midline of the hard palate. These epithelial inclusion cysts are often found in clusters and resolve over the first few months of life. Epithelial pearls are similar to Epstein's pearls and appear as shiny, small white, self-limited lesions that occur on the gums.

Bohn's nodules are also self-limited cysts that appear on the mandibular or maxillary dental ridges. Dental lamina cysts occur on the alveolar ridge of newborns and represent trapped remnants of the dental lamina.

Natal teeth are the premature eruption of primary teeth and are found at birth (natal) or within the first month of life (neonatal). These teeth are either supernumerary or true deciduous teeth and are usually found in the lower incisor region. Natal teeth may lead to ulcerations of the underside of the tongue, called Riga-Fede disease.

Epulis is a congenital fibrous, sarcomatous tumor that arises from the periosteum of the mandible or maxilla. The mass is firm and pedunculated and may regress spontaneously. Excision is required if the epulis interferes with feeding or breathing or for cosmetic reasons.

Lymphangioma is a benign congenital tumor of lymphatic vessels appearing on the tongue, lips or buccal mucosa at birth or in early infancy. Hemangiomas are benign vascular malformations present at birth that may become more apparent as the patient grows. Oral hemangiomas are typically accompanied by vascular lesions elsewhere in the body, especially on the skin.

Infectious Oral Lesions

Infectious oral lesions are typically manifestations of viral infections but may be caused by bacterial or fungal infections as well (see also Chapter 84).

Candidiasis, or thrush, is white plaques on the buccal mucosa, gingivae, and palate that will not “rub off” with a tongue

blade. Caused by *Candida albicans,* thrush is common in neonates and infants. When thrush occurs after infancy, the immune status of the patient must be considered. Oral candidiasis is the most common infection of patients with human immunodeficiency virus (HIV).

The typical lesions of herpes simplex virus (HSV) are groups of vesicles on an erythematous base that may become unroofed and appear as erosions in and around the mouth. Infections may be primary or recurrent. *Herpes gingivostomatitis*, most commonly caused by HSV type 1 (HSV-1), represents primary infection that typically occurs in young children and infants. These patients have pain, fever, and drooling. *Herpes labialis* manifests as recurrent painful lesions that occur on the lips, most often the lower lip. Herpes labialis, or “cold sores,” may be accompanied by an acute febrile illness, extensive sun exposure, or stress.

Hand–foot–mouth disease is characterized by discreet shallow erosions in the mouth, especially on the soft palate, accompanied by erythematous papulovesicular lesions on the hands and feet. High fever may be associated with this enteroviral, typically coxsackievirus, mediated disease that is self-limited in nature. Supportive treatment, specifically antipyretics and adequate oral hydration, is usually sufficient therapy.

Herpangina is also a group A coxsackievirus infection that causes vesicles or ulcers on the pharynx of patients with fever, muscle aches, and malaise.

The characteristic “strawberry tongue” seen in streptococcal scarlet fever is the result of hypertrophic red papillae on a thick white coat. Palatal petechiae are often present as is the typical “sandpaper” papular rash on an erythematous base that blanches on palpation, involving the trunk and back. Streptococcal pharyngitis without exanthem often presents with strawberry tongue and palatal petechiae.

Koplik's spots, pinpoint white macules on markedly erythematous mucous membranes, occur during the prodrome of measles, which includes cough, coryza, conjunctivitis, and fever. By the time the characteristic rash occurs, Koplik's spots typically have resolved.

Varicella lesions occurring in the mouth result in painful vesicles, which may become unroofed, on an erythematous base. Patients may be reluctant to swallow because of pain. Unless bacterial secondary infection occurs, these lesions are self-limited.

Oral lesions commonly associated with HIV-infected patients include candidiasis, hairy leukoplakia, herpes simplex, aphthous ulcers, and necrotizing ulcerative gingivitis. Blue, purple, or red macules, papules or nodules on the palate suggest oral Kaposi's sarcoma, whereas diffuse swelling, discrete nodules, or ulcers of any oral mucosal surface may indicate non-Hodgkin's lymphoma.

The pain, erythema, and swelling of the gingiva seen with dental-alveolar abscesses may be associated with fever and loosening or extrusion of the associated tooth. Significant lymphadenopathy and facial cellulitis may develop. Causative organisms are streptococci and anaerobes, although antibiotic therapy with penicillin is secondary in importance to drainage of the abscess.

Pericoronitis is local infection of the gingiva surrounding an erupting tooth. Although penicillin therapy may be required, good oral hygiene is essential. Lymphadenopathy and facial swelling may accompany pericoronitis.

Acute necrotizing ulcerative gingivitis, also called Trench mouth or Vincent's angina, is a spirochetal infection of the gingiva that occurs in adolescents. Patients report tender, bleeding gums and breath that has a fetid odor. Gums are hyperemic and appear “punched-out” secondary to tissue loss between the teeth. Treatment involves attention to oral hygiene, mouth rinses with a dilute hydrogen peroxide solution, oral penicillin, and debridement of necrotic tissue.

Although infection is present at birth, the oral lesions of congenital syphilis may not become obvious until several months of age. Erythematous papules are seen in the mouth and other mucocutaneous sites. *Hutchinson's teeth*, peg-shaped, superior, central incisors, are not present until later in life. The secondary stage of acquired syphilis is characterized by patches of ulcers or raised lesions in the mouth and is seen in association with generalized rash, fever, malaise, and adenopathy.

Patients receiving long-term antibiotic therapy may develop elongation of filiform papillae of the dorsum of the tongue and a “hairy” appearance from fungal overgrowth called hairy tongue. Hairy leukoplakia of the lateral aspects of the tongue is found in HIV-infected patients in association with intraepithelial proliferation of Epstein-Barr virus infection.

Tumorous Oral Lesions

Eruption cysts are associated with the eruption of teeth and appear on the alveolar ridge and may contain blood.

Oral papilloma are typically benign, although a small percentage of papillomas may become malignant. They are fingerlike extensions from the epithelium of the tongue, gums, lips, or buccal mucosa.

Fibroma is found on the tongue, lips, buccal mucosa, or palate and is a benign, smooth mass with a sessile base.

Ranula is a retention cyst or mucocele of the submaxillary or sublingual ducts. Ranulas are typically seen on the underside of the tongue or on either side of the frenulum on the floor of the mouth.

Mucoceles arise secondary to obstruction of salivary glands. Mucoceles are soft, well-demarcated masses. Patients are

typically asymptomatic. Excision or marsupialization is required.

Pyogenic granuloma represents granulation tissue that develops in response to an irritant such as trauma or foreign body. Most commonly found on the gingiva, pyogenic granulomas are also found on the tongue, lips, and buccal mucosa. Treatment is incision and drainage. Recurrence is common, especially when a foreign body is present.

Although 35 to 40% of cases will present in the head and neck region, rhabdomyosarcoma is a rare malignant tumor of the oral cavity. These lesions are characterized by rapid growth. They are ulcerative in nature and may present with bleeding. Associated signs and symptoms are usually attributed to the mass lesion or obstructive sequelae.

Oral Lesions Associated With Systemic Disease

Stevens-Johnson syndrome, a severe form of erythema multiforme, consists of an inflammatory process that typically involves the skin and mucous membranes. Oral lesions are erythematous plaques on the mucosa of the oral cavity and lips that develop into vesicles or bullae and may become hemorrhagic. This potentially life-threatening disorder is believed to be secondary to a drug reaction or to follow infections.

Toxic shock syndrome may manifest erythema of the oropharynx and a strawberry tongue in patients with a diffuse erythematous macular exanthem, hyperemic mucous membranes, fever, and signs of shock. This toxin-mediated disease is caused by *Staphylococcus aureus* and may be associated with tampon use or nasal carriage of this organism. Toxin-mediated disease associated with streptococci also presents with diffuse erythema of the skin and oropharynx and may progress to septic shock.

Mucositis presents as ulcers, exudate, and pseudomembranes on the gingivae and buccal mucosa of patients with neutropenia oftentimes secondary to chemotherapy. Lesions are extremely painful, and the breath becomes fetid.

Kawasaki disease is a potentially life-threatening disorder that typically presents with an array of findings, including prolonged fever, rash, lymphadenopathy, nonpurulent conjunctivitis, and edema of the hands and feet. Oral changes of Kawasaki disease include red, dry, cracked lips; erythematous oropharynx; and strawberry tongue. Therapy is directed toward prevention of coronary aneurysm development.

The inflammatory lesions of Crohn's disease may occur in any portion of the gastrointestinal tract. Oral lesions, seen most often in adolescents and young adults, consist of ulcers, polypoid papulous hyperplastic mucosa, and edema found on the lips, gingiva, vestibular sulci, and buccal mucosa. Immunosuppressive therapy with steroids and azathioprine has yielded mixed results.

Chronic, recurrent ulcers surrounded by erythema and gray exudate are found anywhere in the oral cavity in patients with Behçet's syndrome. Similar lesions occur on the skin, and the genitourinary tract may also be involved. Behçet's syndrome is rare, affecting older children and adolescents, usually boys.

More than 15 types of hereditary epidermolysis bullosa have been described. This rare, vesiculobullous condition effects mucous membranes and teeth as well as the skin. Scarring may lead to restriction of mouth opening.

Miscellaneous Oral Lesions

Aphthous stomatitis is ulceration of the oral epidermis of unknown cause. These recurrent lesions typically present as 5- to 10-mm ulcerations with a rim of erythema on the buccal mucosa, lips, and lateral aspect of the tongue. The lesions are painful, but patients do not experience fever. The lesions resolve spontaneously after 7 to 10 days.

Geographic tongue represents a benign inflammatory disorder that results in migratory smooth annular patches on the tongue. Although typically asymptomatic, patients may complain of pain. Geographic tongue is typically seen in children less than 4 years of age. No treatment is required.

Gingival hyperplasia is seen in patients receiving long-term anticonvulsant therapy with phenytoin. There appears to be an etiologic interaction with poor dental hygiene. The gingivae undergo fibrous enlargement but are not inflamed or painful. Gingival fibromatosis is an inherited form of gingival hyperplasia.

Leukoplakia of the oral mucosa develops secondary to chronic smokeless tobacco use. These painless, leathery, white patches or plaques occur in areas of greatest tobacco exposure, typically on the mucosa of the buccal sulcus. Significant exposure to tobacco may result in dysplasia or carcinoma.

EVALUATION AND DECISION

When evaluating patients with complaints of oral lesions, it is important to consider a myriad of associated signs and symptoms. The patient's age, general health and appearance, presence of an exanthem or fever, and whether the lesions are painful must be considered before the mouth is examined. Once the lesions are identified, they should be further characterized by color, type, and location.

Neonates with oral lesions can be divided into two groups based on the morphology of the lesions. Discrete masses usually represent congenital disorders, most of which are self-limited. Candidiasis, which involves the oral cavity more diffusely, is also common.

Among older children, toxic-appearing patients require immediate evaluation for potentially life-threatening disease. Patients with conditions listed in Table 49.3 have associated findings such as diffuse cutaneous rash, hyperemia of other

mucous membranes, or poor perfusion indicative of shock. However, Stevens-Johnson syndrome may cause isolated oral lesions initially and then rapidly progress to systemic involvement.

Once life-threatening causes have been considered, additional history and physical examination may lead to diagnosis of other systemic diseases. Weight loss, abdominal pain, and diarrhea with or without blood suggest Crohn's disease, whereas genital ulceration in an adolescent boy points to Behçet's syndrome or secondary syphilis.

The presence of rash and fever makes disorders of infectious etiology more likely. Measles, varicella, scarlet fever, and hand–foot–mouth disease are generally diagnosed by history and physical examination alone. Laboratory evaluation might include a throat culture for streptococci, and serologic testing for measles or HIV when these infections are suspected.

Infectious causes of oral lesions without exanthem may display obvious findings such as cachexia and alopecia in the neutropenic patient with mucositis, or they may be relatively localized to the oropharynx as in herpangina, herpes gingivostomatitis or labialis, and dental infections, which may or may not have fever and lymphadenopathy.

Oral lesions without overt signs of systemic disease are mostly congenital or tumorous in nature. Lesions found in the newborn and during infancy are largely self-limited, and most will resolve spontaneously. A few, including lymphangioma, hemangioma, and congenital epulis, may require intervention.

Children and adolescents experience an array of oral lesions not associated with obvious signs of systemic disease that are typically further delineated by considering the type of lesion (i.e., mass, vesicle, ulcer) and whether they are painful. Most of these processes require little or no therapy. Rhabdomyosarcoma is an obvious exception to this observation.

Suggested Readings

Dilley DC, Siegel MA, Budnick S. Diagnosing and treating common oral pathologies. Pediatr Clin North Am 1991;38:1227–1264.

Kline MW. Oral manifestations of pediatric human immunodeficiency virus infection: a review of the literature. Pediatrics 1996;97:380–388.

Peter JR, Haney HM. Infections of the oral cavity. Pediatr Ann 1996; 25:572–576.

Plauth M, Jenss H, Meyle J. Oral manifestations of Crohn's disease: an analysis of 79 cases. J Clin Gastroenterol 1991;13:29–37.

CHAPTER 50

Pain—Abdomen

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Abdominal pain is a common complaint of children who come to the emergency department (ED). Children with abdominal 50pain have a sensation of “discomfort,” varying from mild to agonizing, often hard to describe, and localized to the abdomen. Although most children with acute abdominal pain have self-limiting conditions, the pain may herald a serious medical or surgical emergency. Abdominal pain tests the clinician's talents for uncovering the cause and for calming the family. The diverse causes include acute surgical diseases (e.g., appendicitis, strangulated hernia, trauma to a viscus); intra-abdominal medical ailments (e.g., gastroenteritis, food poisoning, ulcer disease, urinary infections); extra-abdominal conditions (e.g., pneumonia, tonsillitis, contusions of muscle and bone); systemic illnesses (e.g., “viral syndrome,” leukemia, diabetes mellitus, sickle cell anemia); and functional abdominal pain. Clearly, the most difficult challenge continues to be making a timely diagnosis of appendicitis and other causes of an acute condition in the abdomen early enough to reduce the rate of complications.

PATHOPHYSIOLOGY

Abdominal pain can be stimulated by at least three neural pathways: visceral, somatic, and referred. Visceral pain generally is a dull, aching sensation primarily in the midabdominal, epigastric, or lower abdominal regions. Distension of a viscus stimulates nerves locally, initiating an impulse that travels through autonomic afferent fibers to the spinal tract and central nervous system (CNS). The nerve fibers from different abdominal organs overlap and are bilateral, accounting for the lack of specificity to the discomfort. Children perceive the sensation of visceral pain generally in one of three regions: the epigastric, periumbilical, or suprapubic midline area. Somatic pain usually is well localized and intense (often sharp) in character. It is carried by somatic nerves in the parietal peritoneum, muscle, or skin unilaterally to the spinal cord level from T6 to L1. An intra-abdominal process will manifest somatic pain if the affected viscus introduces an inflammatory process that touches the innervated organ. Referred pain is felt at a location distant from the diseased organ, either as a sharp, localized sensation or as a vague ache. Afferent nerves from different sites, such as the parietal pleura of the lung and the abdominal wall, share pathways centrally. All three types of pain may be modified by the child's level of tolerance. Psychogenic and environmental factors augment or inhibit the “sensation” to varying degrees in different persons. It is amazing that at times pain may be minimal with an appendiceal abscess and, conversely, severe with functional etiology.

Certain illnesses that cannot be explained neurophysiologically are associated with abdominal pain. These include conditions such as tonsillitis with high fever, viral syndromes, and streptococcal pharyngitis (although there may be painful intra-abdominal lymphadenopathy as in mesenteric lymphadenitis syndrome). Other systemic or local conditions may present with abdominal pain as a primary manifestation. Despite abdominal pain appearing localized, the examination needs to be complete. The principal causes of abdominal pain in children and adolescents are summarized in Table 50.1. Table 50.2 highlights those disorders that are life-threatening.

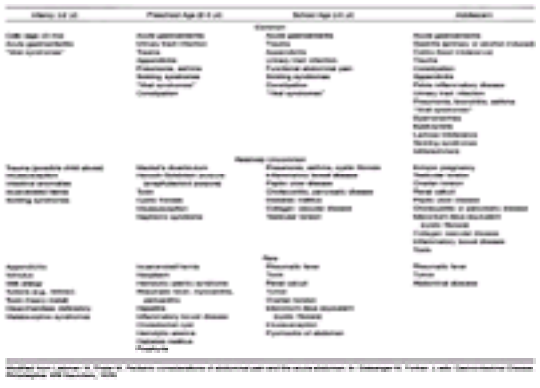

Table 50.1. Causes of Acute Abdominal Pain

Table 50.2. Life-Threatening Causes of Acute Abdominal Pain

EVALUATION AND DECISION

The evaluation of the child with abdominal pain is an important and challenging task. The assessment must focus on any history of trauma, the patient's age, the onset and chronicity of the pain, the related symptoms and pertinent history, and the physical findings (Fig. 50.1).

FIGURE 50.1. Evaluation of the child with abdominal pain.

Abdominal Pain With Trauma

Abdominal pain associated with trauma is covered in detail in the trauma section (see Chapter 103, Chapter 104, and Chapter 108). The first priority is the primary survey, with assessment of cardiovascular status (vital signs and clinical peripheral perfusion), altered breathing, and the extent of neurologic and visible injuries, while simultaneously establishing intravenous access. Exposure of the patient occurs during this initial assessment. The physician should perform a rapid, gentle physical examination to separate superficial injury (e.g., muscle contusion) from significant intra-abdominal trauma (e.g., splenic rupture or hepatic hematoma). In children who are unstable at presentation and have obvious serious or multiple injuries or a severe mechanism of injury (penetrating injury, severe blunt trauma, fall from above 20 feet, ejection from a vehicle, impact velocity more than 35 miles per hour), a rapid, aggressive workup is indicated. It is best to pass a large-bore nasogastric tube to avoid gastric distension. Radiographs (e.g., chest and abdomen with pelvis) and laboratory tests (e.g., complete blood count [CBC], urinalysis, alanine aminotransferase [ALT] and aspartate aminotransferase [AST], and amylase) are indicated in most such cases and with any objective findings. Children with localized and acute pain after blunt trauma may appear surprisingly well but have significant solid organ or hollow viscus trauma. When an intra-abdominal injury is suspected in a stable patient, an urgent computed tomographic (CT) scan should be obtained to assist in pinning down a diagnosis.

Abdominal Pain without Trauma

In assessing the child who develops abdominal pain without a history of trauma, the first priority is stabilization if the child is seriously ill. Attention to airway, breathing, and circulation (ABCs) is critical because cardiorespiratory disease and shock may present with abdominal pain as the major complaint; abdominal emergencies left untreated can lead to cardiorespiratory failure. The next priority is to identify the child who requires immediate or potential surgical intervention, whether for appendicitis, intussusception, or other congenital or acquired lesions. Third, an effort is directed to diagnose any of the medical illnesses from among a large group of acute and chronic abdominal and extra-abdominal inflammatory disorders that require emergency nonsurgical management. Table 50.2 lists life-threatening causes of abdominal pain by age groups. Finally, the physician is left to deal with a host of self-limiting or nonspecific causes of abdominal pain, including nonorganic etiologies. The algorithm presented in this chapter for the approach to abdominal pain without trauma has been designed on the basis of three branch points: age; chronicity; and the presence of obstruction, peritonitis, or a mass.

Infant Less Than 2 Years Old

The infant less than 2 years old with abdominal pain is the most difficult to evaluate because the child cannot describe or localize the complaint. To the parent, the pain may consist of “crying out,” of constantly drawing the legs up with sudden movements or jerks, of being inconsolable, of moaning with lethargy, or of irritability accentuated with fondling or rocking.

Acute Pain

In evaluating the uncomfortable infant, as described in the algorithm, the clinician looks first at the onset of “pain,”

separating acute from recurrent. Then, an evaluation is made of additional symptoms as they occurred chronologically. The bowel pattern (last stool, consistency, diarrhea), presence of fever, and amount of vomiting along with timing are noted. Obstruction may present with isolated vomiting, and a low-grade fever suggests an inflammatory process, including peritonitis. Diarrhea as an early feature often heralds gastroenteritis. Cough (sometimes with posttussive emesis) may suggest pneumonia, bronchiolitis, or asthma. The story of episodic colicky pain with interposed quiet intervals, even in the absence of a “currant jelly” stool, makes one suspicious of intussusception or, occasionally, midgut volvulus.

The physical examination must be used in conjunction with the history to determine which diagnosis should be pursued aggressively. An ileus, manifesting clinically with distension and absent bowel sounds, often accompanies surgical conditions, sepsis, and infectious enterocolitis. Ileus may be seen with pneumonia or a urinary tract infection (UTI). If an abdominal mass is palpable, intussusception, abscess, or neoplasm (commonly of renal origin) is likely. An incarcerated hernia and intussusception are the most common causes of obstruction in this age range. Inguinal hernias may incidentally incarcerate during febrile illnesses in young, crying infants or may be a cause of abdominal obstruction. Signs of partial or complete obstruction with peritonitis indicate a perforated viscus from intussusception, volvulus, or occasionally, appendicitis or Hirschsprung's disease. Rectal examination helps define pelvic masses, localize tenderness in retrocecal appendicitis, and identify a currant jelly stool in some cases of intussusception.

On auscultation of the chest, locally decreased or tubular breath sounds or adventitious sounds (i.e., crackles) suggest pneumonia. A “silent” pneumonia, particularly in a lower lobe, may present with abdominal pain and normal chest and abdominal examination. Atelectasis from the splinting secondary to abdominal pain may manifest with decreased breath sounds. Abdominal pain and pallor can occur in neoplasia, as with bleeding into an abdominal Wilms' tumor, hepatoma, or neuroblastoma. The presence of pallor and pain also raises the possibility of sickling hemoglobinopathies with the development of either a vaso-occlusive crisis or a splenic sequestration. Abdominal pain may be associated with jaundice when rapid hemolysis is related to acute splenic enlargement or with every dysfunction in acute hepatitis. If bruising is noted, hemophilia or leukemia may be the cause of abdominal pain in the ill child. At times, an intra-abdominal vasculitis that causes pain may precede the rash of Henoch-Schönlein purpura.

Chronic Pain

When apparent abdominal pain is recurrent or chronic in infants less than 3 months of age and is not accompanied by other findings or symptoms, the physician often makes a diagnosis of “colic” (see Chapter 17). However, several serious uncommon causes of recurrent abdominal pain in infancy must be considered. These include recurrent intussusception; malrotation with intermittent volvulus; milk allergy syndrome; and various malabsorptive diseases such as cystic fibrosis, celiac disease, and lactase deficiency.

Laboratory Testing

In most instances, the history and physical examination lead the emergency physician to the diagnosis. When the cause is gastroenteritis, most infants and toddlers have a history and physical examination clearly suggestive of this diagnosis and need no further evaluation. When clinical evidence of obstruction, peritonitis, or a mass is present, a CBC and urinalysis are always indicated. It may be wise to obtain these tests when appendicitis is under consideration but the history and physical examination are nondiagnostic. Serum electrolytes, glucose, and blood urea nitrogen (BUN) are not always helpful but should be obtained with peritonitis, obstruction, mass, dehydration, and renal diseases. Abdominal radiographs may be useful in confirming obstruction or the presence of a mass; a barium or air-contrast enema is indicated urgently in the case of suspected, uncomplicated intussusception. Ultrasound may yield preliminary findings that, in the right setting, suggest the need for a therapeutic study for intussusception. An upper gastrointestinal (GI) series may help delineate malrotation, and an intravenous pyelogram, renal scan, or abdominal or renal ultrasound is indicated if abnormalities of the kidney are likely.

Child 2 to 5 Years Old

Similar to the infant, the child who is 2 to 5 years of age usually has an organic cause of abdominal pain. The most common causes of abdominal pain are inflammatory processes, such as gastroenteritis and UTI. As with the younger child, the emergency physician must first ascertain whether the abdominal pain is acute or chronic in onset. With acute pain, the clinician should look for surgical conditions. The ill-appearing child with subacute symptoms may still harbor surgical diseases with complications (e.g., ruptured appendix). In every case, the physician must search for signs of obstruction, peritoneal inflammation, or peritonitis before attributing the cause to a nonsurgical disease.

Acute Pain

The preschool child may be able to describe pain and other symptoms verbally. Although such history is not consistently reliable, it almost always is to be taken seriously. More difficult to detect is the true chronologic order of symptoms. Classic presentations for surgical diseases are rarely volunteered, but occasionally, they may be elicited. Symptoms such as anorexia and vomiting suggest distension of an intra-abdominal viscus; rectal bleeding points to infectious enterocolitis, intussusception, Meckel's diverticulum, or more rarely, inflammatory bowel disease (IBD). Extra-abdominal complaints, such as cough, sore throat, and headache, are commonly present; they often indicate a viral syndrome, pharyngitis, or pneumonia. Urinary symptoms may occur with pyelonephritis, and polydipsia with polyuria may herald the onset of diabetes mellitus with abdominal pain from ketoacidosis. Children with known past intra-abdominal pathology or surgery may develop complications of their prior illnesses.

The physical examination helps separate acute abdominal conditions with peritoneal inflammation or obstruction from less emergent and more common conditions. The most important surgical causes of abdominal pain are acute appendicitis, often with an atypical presentation, and occasionally, intussusception or malrotation. The presence of guarding or persistent abdominal tenderness with gentle palpation warns the emergency physician of a serious

abdominal emergency. Persistent right lower quadrant pain or tenderness on palpation alone suggests a need for surgical evaluation in the ED. This is the most consistent finding in patients discharged to home with early appendicitis. Usually, during a quiet, relaxed examination, the pain from gastroenteritis abates; other “referred” abdominal pains (e.g., from pneumonia or tonsillitis) often seem to disappear when the child is reassessed in a calm fashion. Ill children with abdominal pain occasionally have life-threatening diseases (Table 50.2) or the more uncommon diseases (Table 50.1). The physical examination of such patients may show jaundice (hepatitis, hemolytic anemia), rash or arthritis (Henoch-Schönlein purpura), cardiac murmurs (rheumatic fever), friction rubs (pericarditis), or “acetone” on the breath (diabetes mellitus).

Chronic Pain

A history of recurrent abdominal pain suggests conditions such as sickle cell anemia, inflammatory bowel disease (IBD), cystic fibrosis, or asthma. Chronic constipation can occur in children between 2 and 5 years of age, but true psychogenic or other nonorganic abdominal pain is fairly uncommon in this preschool age group.

Laboratory Testing

The use of the laboratory parallels its role in young infants. A CBC and urinalysis may be useful in this age range, either to point toward or away from an inflammatory process such as appendicitis. If the appendix has not ruptured, the white blood cell (WBC) count is usually normal or minimally elevated. WBC counts that are higher than 16,000 to 18,000/mm ^{3} suggest an acute bacterial infection or complicated intra-abdominal process, such as an abscess. A grossly abnormal urinary sediment points to UTI or, occasionally, glomerulonephritis but does not exclude an inflamed appendix that is lying anteriorly near the bladder. Occasionally, a hemoglobin electrophoresis is needed to confirm the diagnosis of sickling syndromes.

Radiographic findings are usually normal or nonspecific in children with GI infections or appendicitis;therefore, a normal film should not reduce the suspicion of potential surgical disease. Radiographic signs of appendicitis include localized bowel obstruction (a sentinel loop), an appendicolith, or obliteration of the psoas shadow. Radiographs are of greater benefit in younger children in whom atypical presentations are more common. An abnormally thickened intestinal mucosa from IBD or ascites from nephrotic syndrome favors the diagnosis of these nonsurgical emergencies. Chest radiographs may detect lower lobe pneumonia or asthma with atelectasis in this age group. Once again, the recent literature suggests that abdominal ultrasound may help establish a diagnosis in children with atypical abdominal pain, particularly of genitourinary origin.

Child 5 to 12 Years Old

The preadolescent child adds another dimension to the spectrum of abdominal pain—that of nonorganic or psychogenic illness. The leading organic causes of abdominal pain still are inflammatory and include gastroenteritis, appendicitis, and UTIs. Occasionally, the child is the victim of chronic disease (Table 50.1). Colicky abdominal pain is more rarely associated with intussusception than in younger children. Usually a “lead” point for an intussusceptum is seen in older children (e.g., mesenteric adenitis, lymphoma, polyp, cystic fibrosis, anaphylactoid purpura). Abdominal pain with other symptoms may herald the presentation of inflammatory bowel, collagen vascular, ulcer, gallbladder, pancreatic, or liver disease. Constipation is a common cause of acute abdominal pain in older children, especially in the those with delayed development.

Acute Pain

The history of abdominal pain generally is reliable in the older child. The presence of fever, cough, vomiting, and/or sore throat suggests an infectious cause. Associated diarrhea may be from infectious colitis, IBD, or an appendiceal abscess irritating the bowel. A genitourinary history of discharge or suspicion of sexual abuse may be difficult to elicit in this age range but must be sought. With UTIs, urinary frequency and dysuria usually occur. Finally, pain that begins periumbilically and migrates to the right lower quadrant after several to 24 hours suggests appendicitis, a more common diagnosis in the older child.

The findings on physical examination, including the rectal and genitourinary examination in girls, are often revealing in this age group. Localized tenderness in the right lower quadrant or diffuse tenderness with involuntary guarding raise the suspicion of appendicitis or other diseases that cause peritonitis. As in younger children, it is critical to assess for an atypical presentation of appendicitis. Careful extra-abdominal examination is paramount to discover many of the infectious causes of abdominal pain. Rectal examination should be performed along with visualization of the genitalia in the young girl while in the knee–chest or frog leg position (see Chapter 94).

Chronic Pain

Chronic abdominal pain may occur as a result of many of the conditions listed in Table 50.1. Important considerations are chronic infection with an enteric pathogen and IBD. When the history and physical examination suggest a mild, self-limiting disease or a nonorganic basis for the abdominal pain, however, the emergency physician should refrain from overusing the laboratory or radiography department to allay the parents' fear of organicity. It often is difficult to provide reassurance and counseling in the busy ED to enmeshed parents and their child with recurrent abdominal pain; however, time spent in this endeavor can be worth the effort. Most important is the recognition that the child feels real pain, despite reinforcement of the idea that an organic diagnosis is not likely to be forthcoming. Counseling needs to be tailored to the level of sophistication of the family. Sometimes, revisits to the ED or acute-care office setting may be necessary for reappraisal of an acute episode if referral to a primary physician cannot be arranged.

The syndrome of functional abdominal pain precipitates more than 80% of outpatient physician visits by children for abdominal pain. The presentation of such pain is generally episodic, emanating from the umbilicus and, by definition, has

no organic cause. The pain rarely occurs during sleep and has no particular associations with eating, exercise, or other activities. There may be a positive family history of GI symptoms or migraine. The child's growth and development are normal, and the abdominal examination is unremarkable; occasionally, mild midabdominal tenderness, without involuntary guarding, is elicited. If performed, screening tests such as a CBC, sedimentation rate, and urinalysis all are normal.

The emergency physician's task is to allay any fears of serious organic disease during the acute episode, which is not always easy. At the same time, the physician must support the child, who is truly feeling pain. Because the long-term solution to a functional complaint is generally not in the realm of the ED, the physician should explain all of the organic illnesses that the pain is *not* felt to be and suggest a nonorganic cause of the pain. The emergency physician should provide an avenue for continued supportive follow-up through referral to the primary physician.

Laboratory Testing

In most children over the age or 5 years with abdominal pain, laboratory tests are needed only to confirm a diagnosis that is suspected clinically. If appendicitis is being considered, a CBC and urinalysis should be obtained unless the history and examination appear diagnostic, wherein clinical pathway studies have demonstrated minimal need for laboratory testing. Before appendiceal rupture, the WBC count usually is minimally elevated and the urinalysis is normal. With perforation, the WBC count usually rises above 16,000/mm ^{3}, and pyuria occurs occasionally. Again, radiographic findings with appendicitis may include a normal film, a sentinel loop, an appendicolith, or obliteration of the psoas shadow. In patients with significant fever, a chest radiograph may assist in the evaluation. When IBD is suspected in the child with abdominal pain, a sedimentation rate may prove useful because it is almost invariably elevated with this group of disorders. In addition, a CBC and serum protein to evaluate for anemia or hypoalbuminemia can be useful. Often, other laboratory tests are indicated to corroborate clinical findings (e.g., chest radiograph in lower lobe pneumonia; urinalysis and culture for pyelonephritis or cystitis). Only the rare, complicated patient with abdominal pain requires a battery of tests.

Adolescent More Than 12 Years Old

The adolescent patient, particularly the female, taxes the emergency physician's skills, generating an extensive differential diagnosis of both life-threatening and less serious but common causes for abdominal pain (Table 50.1 and Table 50.2). Of the gynecologic causes, potentially catastrophic ectopic pregnancies and the more common pelvic inflammatory disease (PID) head the list in importance.

Acute Pain

The history of acute abdominal pain needs to be explored carefully to avoid missing a pertinent diagnosis. The location of the pain and its character may assist in differentiating the acute surgical abdomen from gastroenteritis or psychogenic pain. A menstrual history and ascertainment of sexual activity are essential. Obtaining this information is often difficult, however, and one must assume that pregnancy and sexually transmitted diseases are possible despite rigorous denials of sexual contact. Sensitivity to the family regarding this possibility is important. Obviously, a complete physical assessment of the teenage girl routinely includes a pelvic examination.

Acute pain with peritonitis in the adolescent boy usually results from appendicitis. Assessment also must include that for testicular torsion or epididymitis. The adolescent girl with peritoneal findings may have appendicitis, PID, or less commonly, cholecystitis or ectopic pregnancy.

Boys without peritoneal signs often have gastroenteritis or a viral syndrome. Girls are more prone to UTIs or pyelonephritis; ectopic pregnancy, although uncommon, must be considered because this diagnosis may not produce peritoneal irritation before rupture. Gravid girls may be suffering from a complication of pregnancy or any of the usual disorders that cause abdominal pain as listed in Table 50.1 and Table 50.2.

Chronic Pain

Chronic abdominal pain in the adolescent is similar to that in the younger child, with the exception of an increased prevalence of IBD and the need in girls to consider long-standing gynecologic ailments such as dysmenorrhea, endometriosis, and chronic PID. It is particularly difficult to establish the cause of chronic pain when dealing with adolescents on an episodic basis, making appropriate referral essential.

Laboratory Testing

The use of the laboratory in diagnosing abdominal pain is similar to that for the child, with the exceptions of the need for abdominal or pelvic ultrasound, pregnancy testing, or microbiologic evaluations for sexually transmitted diseases. In the ED, the postmenarchal girl with abdominal pain should have a pregnancy test (see Chapter 48) when pregnancy is suspected or no definite diagnosis has been made. Ultrasound often is helpful to establish the location of a gestational sac or to evaluate the adnexa for a possible mass. In equivocal cases of gynecologic disorders versus an acute appendiceal inflammation, ultrasound and CT have been helpful in eliciting the cause of pain. Appropriate cultures and microscopic examinations for sexually transmitted diseases often are indicated. Recently, laparoscopic evaluation has been used increasingly as both a diagnostic and therapeutic procedure for older children and adolescents.

SUMMARY

Abdominal pain is one of the most common complaints of children who seek treatment in the ED. The history and physical examination should distinguish most cases that require surgical intervention or admission to the hospital;

laboratory tests are indicated only to confirm diagnoses suspected clinically.

Inflammation, usually in the form of gastroenteritis, is the most common organic cause at all ages for abdominal pain. Acute surgical conditions that are not always easy to diagnose, however, must be excluded. Appendicitis, which is often atypical, is by far the most important diagnosis to exclude, especially in the older child and adolescent. Reexamination during the same visit or in the next 6 to 12 hours and surgical consultation often are necessary to decide on the need for hospitalization or surgery.

Nonabdominal causes of abdominal pain should receive consideration in children of all ages. A particularly common cause of abdominal discomfort in the older child is recurrent, functional pain.

Suggested Readings

Apley J. The Child with Abdominal Pains. Philadelphia: FA Davis, 1959.

Hofter FA. Interventional radiology in the acute pediatric abdomen. Radiol Clin North Am 1997;35:977–987.

Liebman W, Thaler M. Pediatric considerations of abdominal pain and the acute abdomen. In: Sleisenger B, Fortran J, eds. Gastrointestinal Disease. Philadelphia: WB Saunders, 1978;1:411.

Reynolds SL. Missed appendicitis in a pediatric emergency department. Pediatr Emerg Care 1993;9:1–13.

Scharff L. Recurrent abdominal pain in children: a review of psychological factors and treatment. Clin Psych Rev 1997;17:145–166.

Schwartz MZ, Bulas D. Acute abdomen. Laboratory evaluation and imaging. Semin Pediatric Surg 1997;6:65–73.

Siegel MN, Carel C, Surrot S. Ultrasonography of acute abdominal pain in children. JAMA 1991;266:1987–1989.

Wolf SG. Abdominal Diagnosis. Philadelphia: Lea & Febiger, 1979.

CHAPTER 51

Pain—Back

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As a rule, back pain in children is meaningful until proved otherwise. If back pain is less common in children than in adults, it is also far more likely to signify pathology. Furthermore, children are subject to conditions causing back pain that are seldom seen in adults, and the common causes in adults occur uncommonly in children. The diagnosis may be difficult because children have less ability to describe their pain and, in younger children, even to localize it.

DIFFERENTIAL DIAGNOSIS

In the famous joke, Dr. Loeb asks the medical student how he arrived at the correct diagnosis of splenic sarcoma and the student replies, “What else causes back pain?” A complete list of causes might run for pages, but for our purposes, we can divide the likely childhood causes into a few categories (Table 51.1 and Table 51.2).

Table 51.1. Cause of Back Pain

I. Traumatic	C. Pneumonia
A. Compression fracture	D. Urinary tract infection
B. Spondylolysis/spondylolisthesis	III. Neoplastic
II. Infectious	A. Benign tumors
A. Discitis	B. Malignant tumors
B. Vertebral osteomyelitis	

Table 51.2. Common Causes of Back Pain in Children

Trauma is a major problem in children. Spinal trauma is covered in Chapter 125, and cervical spine injuries are covered in Chapter 106. Acute injury, especially with axial loading, may cause compression fractures, which are more common in children. Compression fractures present with localized pain over the affected vertebra or vertebrae. Radicular pain may develop and may occasionally be so severe as to cause a sympathetic ileus and vomiting. The signs of back strain and lumbar disc herniation are similar to those in the adult, but in adolescence, these diagnoses should be made with caution, and in childhood, they are distinctly rare.

Injury may be less obvious when it is chronic or recurrent. Especially common among adolescents is spondylolysis. This represents a weakening or discontinuity of the pars interarticularis, which connects the vertebrae at the facet joints. The condition is believed to be associated with overuse and microfractures. If the vertebra slips forward on the one beneath, the resulting condition is called spondylolisthesis. Either condition can cause back pain, most often in the lower lumbar area or at the L5–S1 level. These lesions often present during the adolescent growth spurt and are associated with

repeated lifting and back extension, especially in sports. Another condition linked to overuse, Scheuermann's disease, is seen especially in adolescents as anterior wedging of several vertebrae, especially in the thoracic spine. This condition is painful in about half of cases.

Rare but dangerous causes of back pain deserve special vigilance in the child (Table 51.3). A spinal epidural hematoma may follow a fall or blow, sometimes presenting days later, or it may arise spontaneously, especially in patients with bleeding disorders or those who are receiving anticoagulant therapy. Back pain may only briefly precede symptoms of spinal cord compression. Spinal epidural abscess may present with back pain, low-grade fever, and signs of spinal cord compression. Percussion usually elicits spinal tenderness. Appreciation of the signs of spinal cord compromise should lead to emergent neurosurgical referral.

I. Traumatic	D. Meningitis
A. Spondylolisthesis	E. Transverse myelitis
B. Spinal epidural hematoma	F. Spinal epidural abscess
II. Infectious	G. Aortic dissection
A. Discitis	III. Neoplastic
B. Vertebral osteomyelitis	A. Malignant tumors
C. Paraspinal, retroperitoneal abscess, or pyomyositis	B. Benign tumors
	C. Leukemia/lymphoma

Table 51.3. Serious Causes of Back Pain in Children

Infectious causes of back pain are many. Infection in the back itself may prove difficult to diagnose. Vertebral osteomyelitis and discitis present in a similar fashion. A limp or failure to bear weight may be more obvious than back pain in the young child, who is most susceptible to these infections.

Paraspinal or psoas abscess and pyomyositis in paraspinal or pelvic muscles may present as back pain. Iliac osteomyelitis and sacroiliac joint infection also may present as back pain. Osteomyelitis of the ribs occurs rarely. The more rare infections include spinal tuberculosis (Pott's disease) and brucellosis, in which small vertebral abscesses may accompany lymphadenopathy and hepatosplenomegaly.

Infections outside the musculoskeletal axis that cause back pain include urinary tract infection (UTI), which may cause flank pain with or without upper tract infection; pneumonia; and meningitis. These infections in children may lack the obvious symptoms seen in older patients. The young child with a UTI often shows no urinary symptoms as such, although fever or gastrointestinal symptoms may occur. Pneumonia can be surprisingly silent. Cough may be minimal. Auscultation and percussion are of limited sensitivity in the small chest. Meningitis occasionally may be identified by parents primarily as back pain. This is especially true in infants who are noted to fuss when moved; a parent may note that it hurts the child to move the back. Myalgias and generalized backache may be seen in influenza, mononucleosis, streptococcal pharyngitis, and other generalized infections. Postinfectious conditions include transverse myelitis, which may follow an upper respiratory infection; back pain may precede weakness by as much as 1 to 2 days.

Abdominal conditions that may present as back pain include pancreatitis, in which the steady, penetrating pain radiates prominently to the back. Gallbladder pain may radiate to the back as well, and appendicitis, especially in a retrocecal location, can cause radiation to the back or pain and tenderness in the flank.

Collagen vascular diseases that cause back pain include ankylosing spondylitis and juvenile rheumatoid arthritis (with sacroiliitis), especially pauciarticular type II disease. Both of these conditions chiefly affect boys over 8 years of age. The back is notably stiff to flexion, especially in the lumbar region. Spondylitis also may be seen in association with Reiter syndrome, regional enteritis, ulcerative colitis, and psoriasis.

A painful, rapidly progressive scoliosis or atypical curvature suggests serious spinal pathology, often with neuromuscular involvement. Idiopathic scoliosis demonstrates back pain only in the most severe cases.

A host of neoplastic causes, both benign and malignant, may present with back pain (Table 51.1). These are not so rare that they can be ignored in any evaluation of back pain. Ewing's sarcoma may mimic infection, with fever, leukocytosis, and rarefaction of bone on radiographs. Leukemia, and especially lymphoma, may present as back pain.

Sickle cell disease and other hemoglobinopathies may cause back pain. Dissecting aortic aneurysm has been reported rarely in children, usually with hypertension or with Marfan's syndrome and other connective tissue disorders. Table 51.1 lists other miscellaneous causes of back pain.

Psychogenic back pain should be diagnosed with caution in children. The young child lacks the psychological machinery for conversion and the motivation to malinger. Even in adolescents, an overly prompt diagnosis of functional pain may lead to neglect of organic illness. Psychosomatic pain may be suggested, however, by a cheerful affect when describing symptoms, by disproportional school absence, or by unusual family dynamics, especially in the adolescent patient. Even if suspicion of a psychological component exists, a thorough investigation should be undertaken to exclude organic causes.

EVALUATION AND DECISION

The first task in a child with back pain is to rule out any sign of neurologic involvement. (Fig. 51.1). Conditions that affect the spinal cord are seen often enough in children that their gravity should be borne in mind throughout the evaluation. The history should be reviewed for limb weakness or disuse or for a change in function of the bowel or bladder. Although seen with some purely musculoskeletal problems, limp in the presence of back pain should suggest possible spinal cord involvement (see Chapter 43). A thorough examination of spinal cord function is indicated in any child with back pain of unknown cause. This should include evaluation of muscle bulk, tone, and strength; sensitivity to pinprick in addition to light touch; the deep tendon reflexes and Babinski reflex; and anal tone, the anal contracture reflex, and in boys, the cremasteric reflex. When any neurologic compromise is suspected, additional sensory examination of proprioception, heat, and cold is warranted. Any suspicion of spinal cord involvement warrants prompt neurosurgical consultation.

FIGURE 51.1. Approach to the diagnosis of back pain. *WBC,* white blood cell; *ESR,* erythrocyte sedimentation rate.

Systemic symptoms should be carefully reviewed. Fever, fatigue, poor appetite, weight loss, or a decrease in walking or weight bearing usually signify a worrisome illness. Night pain that awakens the patient from sleep is associated with spinal tumor and, if relieved by aspirin or nonsteroidal anti-inflammatory drugs, suggests osteoid osteoma or osteoblastoma. Sciatica may signify disc herniation, especially in the adolescent athlete or laborer.

A history of trauma is important in evaluating back pain but may be misleading in the young child whose frequent minor injuries may be seen by the parents as the trigger for a problem that actually is nontraumatic. Adolescents with stress injuries may not identify trauma in connection with back pain. Certain injury histories suggest specific diagnoses; vertical loading of the spine, as when a child lands in a seated position after a fall, often is associated with compression fractures of the vertebrae.

In adolescents with back pain, a history should be sought of sports-related, lifting-related, or work-related exposures to back stress. Weight lifters, gymnasts, football players, and participants in many similar sports have well-known tendencies to hyperextend the back. Unfortunately, not all teenagers who develop stress injuries are engaged in such easily identified activities. As excessive competition in sports is introduced to younger and younger children, such injuries may be seen earlier in childhood.

Fever is likely, of course, to signify infection, and infection both in the back and in adjoining areas is a common cause of back pain. However, fever is sometimes seen with neoplastic and collagen vascular causes of back pain. Even more importantly, children with musculoskeletal infections are afebrile at presentation in a significant proportion of cases.

Age may be suggestive of cause. The preschool child is unlikely to have overuse injuries, spondylolysis, or ankylosing spondylitis; however, infectious causes are more likely at this age. Family history may be positive in ankylosing spondylitis and related conditions. History may reveal an underlying disease, chronic inactivity (in the bedridden child), or drug therapy that may cause osteoporosis, which increases the risk of bony injury, especially compression fracture.

Chronicity of back pain may suggest collagen vascular conditions, Scheuermann's disease, or perhaps, a developmental defect in the spine, but tumors may progress slowly with chronic back pain being the only noted symptom. Likewise, the acute onset of pain or neurologic symptoms often may be the first signs of a chronic (but expanding) mass lesion.

In addition to the neurologic examination previously mentioned, the physical examination should cover the appearance of the spine; percussion of the spine, ribs, and flank; palpation of the sacroiliac and paraspinal areas; rotation of each hip as well as straight-leg raising; and rectal and genital examinations. (Hydrometrocolpos has been reported as a cause of back pain, even in premenarchal girls.) Flexibility of the spine should be checked, especially to flexion and extension, and unusual kyphosis, lordosis, or scoliosis should be noted. Careful chest and abdominal examinations and the absence of meningeal signs require documentation.

In many cases, the cause of back pain remains obscure even after a thorough history and physical examination. Here, the physician faces an important decision. In the younger child, it is appropriate to seek consultation or to proceed to imaging studies. Plain films may reveal a narrowed disc space in discitis; bone scan may reveal discitis or vertebral osteomyelitis, even in the absence of fever and bony changes; magnetic resonance imaging (MRI) may reveal a spinal or paraspinal tumor. In the preteen or teenage patient, a history of likely back strain (or the appearance of clues to psychosomatic pain previously mentioned) may warrant a trial of empiric therapy; even here a complete physical examination and screening radiographs and laboratory studies should be documented, and close follow-up should be

ensured. (Further treatment of the varying causes of back pain is discussed in specific sections of this book.)

Plain radiography of the spine is much more likely to be revealing in children with back pain than in adults. Plain films should be ordered in cases with trauma, with exacerbation by activity (as may be the case in spondylolysis and spondylolisthesis), when chronicity or progression are noted, or when the diagnosis remains uncertain. Although two-view studies may show compression fractures, spondylolisthesis, discitis, and lesions of the vertebral body, oblique views are recommended with any history of trauma or possible stress-induced injury to assess integrity of the pars interarticularis. Plain chest radiography is indicated when pneumonia is considered.

Bone scintigraphy is most useful for the definition of osteomyelitis, discitis, or other early osseous pathology when radiography proves unrevealing. Newer single-photon emission computed tomographic (SPECT) scans may reveal stress injury, such as spondylolysis, before it is seen on planar bone scans or plain radiographs. Radiolabeled white blood cell scans may detect paraspinal infection or abscess.

Computed tomography (CT) remains a useful method for emergent evaluation of trauma or when lesions outside the spine are suspected. However, this technique is limited in its ability to delineate densities within the bony spinal canal and to study the alignment of spinal structures in the vertical axis without special methods or reconstructions. MRI overcomes these limitations and provides highly detailed longitudinal images of the spine, the canal, the cord, and other tissues. MRI is preferred for evaluations of possible intraspinal pathology. It has largely supplanted myelography.

Laboratory investigations are directed by the presentation. A complete blood count and sedimentation rate are often useful. Urinalysis, serum amylase, and biliary studies may rule out abdominal pathology. Sickle cell preparations or hemoglobin electrophoresis may be indicated. Other evaluations may be used selectively.

In summary, back pain in children warrants more concern and closer investigation than back pain in adults. Because back pain is less common in children, we can afford to pay closer attention, and because it more often implies significant pathology, we must do so.

Suggested Readings

Afshani E, Kuhn JP. Common causes of low back pain in children. Radiographics 1991;11:269–291.

Bellah RD, Summerville DA, Treves ST, et al. Low-back pain in adolescent athletes: detection of stress injury to the pars interarticularis with SPECT. Radiology 1991;180:509–512.

Gerbino PG, Micheli LJ. Back injuries in the young athlete. Clin Sports Med 1995;14:571–590.

Hollingworth P. Back pain in children. Br J Rheumatol 1996;35:1022–1028.

Payne WK, Ogilvie JW. Back pain in children and adolescents. Pediatr Clin North Am 1996;43:899–917.

Sherry DD, McGuire T, Mellins E, et al. Psychosomatic musculoskeletal pain in childhood: clinical and psychological analyses of 100 children. Pediatrics 1991;88:1093–1099.

Tunnessen WW. Back pain. In: Signs and Symptoms in Pediatrics. 2nd ed. Philadelphia: Lippincott, 1988:467–470.

Wolfe MW, Bennett JT. Pyomyositis with toxic shock syndrome presenting as back pain and fever: a case report and literature review. Am J Orthop 1997;26:135–137.

CHAPTER 52

Pain–Chest

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The complaint of chest pain uncommonly represents a life-threatening emergency in children, in contrast to the same complaint in adults. Recently, the epidemiology has changed with the increase of cocaine abuse in which chest pain, myocardiopathy, and even myocardial infarction (MI) may occur. Although heart disease is an uncommon source of chest pain in children, the fear of a cardiac origin for the pain may evoke anxiety in the child or in the parents. Thus, a careful approach to the patient is imperative even in the pediatric setting. This chapter first briefly reviews the pathophysiology of chest pain, then outlines the differential diagnosis in children, and finally presents the evaluation, as appropriate in the emergency department (ED).

PATHOPHYSIOLOGY

To understand the possible origins of chest pain or discomfort, it is important to review how this sensation is transmitted. Musculoskeletal pain is produced by irritation of these tissues and is transmitted through the sensory nerves. The stimulus is carried through the nerve in the dermatomal or intercostal distribution to the dorsal root ganglia, up the spinal afferents, and into the central nervous system (CNS). This local, peripheral, sharp pain also can be produced by primary dorsal root irritation in the spine. Because of overlap of nerve distribution, pain may be sensed in locations distal to the irritation. For example, the third and fourth cervical nerves evoke pain as far caudally as the nipple line of the chest.

Tracheobronchial pain is transmitted by vagal afferents in the large bronchi and trachea to fibers in the cervical spinal column. Dull, aching, or sharp pain is felt in the anterior chest or neck. The irritation or sensation of cough is transmitted in a similar fashion. Pleural pain arises in the pain-sensitive parietal pleura and then travels through the intercostal nerves in the chest wall, giving rise to sharp, well-localized pain. The visceral pleura is insensitive to pain. The intercostal or phrenic nerves transmit diaphragmatic pain. Peripheral diaphragmatic irritation may cause local chest wall pain because of the intercostal innervation. Central diaphragmatic stimulation travels by the phrenic nerve, with the distribution of pain referred to the shoulder of the affected side.

The esophagus appears to be more pain sensitive in its proximal portion. Pain is transmitted by afferents to corresponding spinal segments, with resultant anterior chest or neck pain. The pericardium is innervated by portions of the phrenic, vagal, and recurrent laryngeal nerves, as well as by the esophageal plexus. This appears to give rise to various sensations, including chest or abdominal pain, dull pressure, and even referred anginalike pain.

Other mediastinal structures, such as the aorta, have pain fibers in the adventitia of the vessel wall. They transmit pain through the thoracic sympathetic chain to the spinal dorsal roots, giving rise to sharp, variably localized chest pain. Cardiac pain probably is transmitted by a number of routes, including the thoracic sympathetic chain and the cardiac nerves through the cervical and stellate ganglia. It has been proposed recently that pain arises from abnormal ventricular wall movement and stimulation of the pericardial pain fibers. These routes account for the sensation of cardiac chest pain as pressure or crushing pain substernally or as sharp pain in the shoulder, neck, or arm.

DIFFERENTIAL DIAGNOSIS

A differential diagnosis of chest pain in children is included in Table 52.1. Most chest pain in children is caused by acute respiratory disease, musculoskeletal injury, anxiety, or inflammation (Table 52.2). Often, the physician does not make a causative diagnosis of the chest pain and calls it idiopathic in origin. This idiopathic chest pain actually may be unrecognized organic disease, such as gastroesophageal reflux. Although much less common, chest pain in association with cardiorespiratory distress demands immediate attention. Table 52.3 lists the life-threatening causes of chest pain by disease and mechanisms for decompensation.

Table 52.1. Causes of Chest Pain

Functional (anxiety/psychosomatic)
Musculoskeletal contusion/strain
Costochondritis/myositis
Cough or respiratory infections (bronchitis, pneumonia, pleurisy, upper respiratory infections)
Asthma
Gastroesophageal reflux
Idiopathic

Table 52.2. Common Causes of Chest Pain

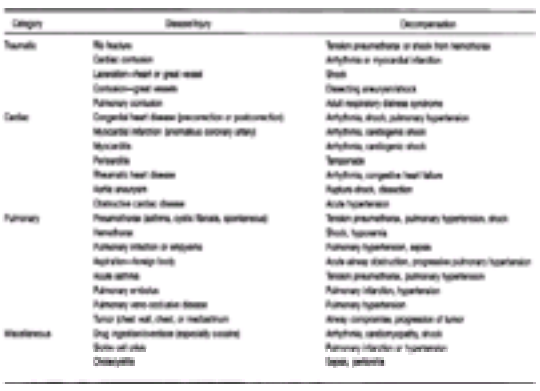

Table 52.3. Life-Threatening Causes of Chest Pain

In the case of trauma, cardiac or pulmonary compromise may arise from direct injury to the heart, great vessels, or lung (see Chapter 107). Chest pain in the nontraumatized, yet dyspneic or cyanotic patient, most often stems from a respiratory problem, such as acute pneumonitis, empyema, pleurisy, asthma, or pneumothorax (either spontaneous or associated with cystic fibrosis [CF] or asthma). Rarely does severe chest pain in an acutely ill child result from an MI (see Chapter 8) that comes from aberrant coronary vessels, other underlying cardiac diseases (aortic stenosis), an acute arrhythmia, pericardial disease, or pulmonary embolus. Pediatricians do not often think of pulmonary embolus early on. Usually, this may have associated risk factors, such as recent trauma, particularly spinal injury, or known cardiorespiratory problems. More recently, acute chest pain associated with ischemia, arrhythmia, or myocardiopathy may be secondary to acute or chronic cocaine exposure. Other toxins have cardiac effects also. Nonorganic chest pain may appear to cause respiratory distress in the hyperventilating teenager (see Chapter 131), but close examination should distinguish this syndrome from serious problems.

Chest pain in children usually occurs without associated cardiorespiratory signs or symptoms, often as a chronic problem. By the time of the ED visit, the pain has often resolved. Mild to moderate strain or injury from exercise or trauma may produce a contusion or rib fracture. Inflammation of nerves, muscles, bones, costochondral junctions, the esophagus, or the lower respiratory tract often causes organic chest pain. Both respiratory infection (pneumonia or bronchitis) and allergic respiratory disease (asthma) are important causes to consider. Spontaneous pneumomediastinum and pneumothorax may occur in patients with reactive airway disease, CF, or as a result of barotrauma (e.g., Valsalva maneuver, forceful vomiting or coughing). Aspiration of a foreign body into the trachea or esophagus may occur without such history in a toddler or even in an older child. Unrecognized disease rarely causes isolated chest pain in a child who otherwise appears well, but the physician should consider drug exposure (e.g., cocaine, methamphetamine, nicotine, b-agonist abuse). In addition, attention should be paid to diagnosing the rare patient with progressive obstructive heart disease, angina, mitral valve prolapse, or early pericardial or myocardial inflammation (see Chapter 82). A large group of children (up to 50%) will be left whose pain best fits into an anxiety-induced or idiopathic category. Every individual evaluation must be started, however, with a broad differential diagnosis in mind to ensure proper diagnosis and management of the child with chest pain.

EVALUATION AND DECISION

Child with Thoracic Trauma

The first step in evaluating the child with chest pain is to perform a thorough history and physical examination. If any evidence of trauma to the chest exists (see Chapter 107), the patient requires rapid evaluation and may need immediate resuscitation as well (Fig. 52.1A). Correction of cardiac or respiratory insufficiency may diagnose, as well as treat, the cause of chest pain. Alveolar ventilation should be assessed for adequacy and bilateral symmetry to distinguish acute respiratory failure from hemothorax or pneumothorax. In children with chest trauma, tachycardia with hypotension generally is caused by hypovolemia secondary to a hemothorax, hemopneumothorax, or vascular injury. Reduced cardiac output and perfusion, however, also may be secondary to a rhythm disturbance (from a myocardial contusion, tension pneumothorax) or cardiac tamponade (which causes muffling of the heart sounds and pulsus paradoxus). A discrepancy of the pulse or blood pressure between the extremities points to aortic diseases, such as traumatic avulsion or aneurysm, as the cause of chest pain. Ruptured esophagus and tracheobronchial disruption may result from rapid deceleration injuries and may present with chest pain, respiratory distress, and hypotension.

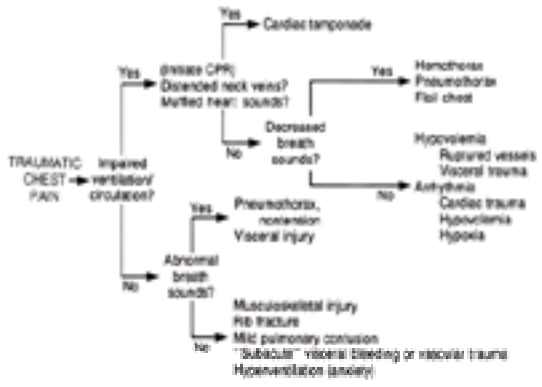

FIGURE 52.1A. Diagnostic approach to traumatic chest pain. *CPR*, cardiopulmonary resuscitation.

Many children with thoracic injuries but no respiratory distress also complain of chest pain. Although a careful examination is mandatory in an effort to exclude significant intrathoracic trauma, the cause of the pain usually resides in the chest wall: contusions of the soft tissues or rib fractures. A history of significant trauma even in the absence of cardiovascular abnormality dictates that radiographs and an electrocardiogram (ECG) be obtained. Rib fractures in young infants suggest child abuse. In older children, a predisposing cause for fracture (e.g., bone cyst, tumor) should be sought.

Child with No Thoracic Trauma

Initially, the physician needs to assess for cardiorespiratory instability. Next, the physician should inquire about a history suggestive of prior cardiorespiratory disease (Fig. 52.1B). Children with respiratory illnesses such as asthma or CF are at risk for pneumothorax, acute respiratory failure from mucous plugging or pneumonitis, and acute pulmonary hypertension. Severe hypoxemia may accompany their chest pain. Auscultatory findings, such as rales or wheezing, may be minimal when obstructive pulmonary disease is moderately severe. In the child with a history of cardiac arrhythmias (see Chapter 82), congenital heart disease, cardiac surgery, or pericardial effusions, chest pain may signal an exacerbation of the underlying problem. Although uncommon in children, acute pulmonary embolus should be considered when chest pain with the sense of impending doom and/or risk factors (e.g., obesity, birth control pills, pregnancy, collagen vascular disease, nephrotic syndrome, cigarette smoking, recent surgery, a positive family history) is present. Pulmonary embolus may occur as a complication of an underlying disease, medical therapy, or surgical repair.

FIGURE 52.1B. Diagnostic approach to atraumatic chest pain. *CPR*, cardiopulmonary resuscitation; *CF*, cystic fibrosis.

In the absence of prior cardiopulmonary disease or trauma, the approach must be directed toward unmasking evidence for any of the serious cardiorespiratory illnesses listed in Table 52.3. In particular, chest pain associated with exertion, syncope, or palpitations is concerning. A history of untreated Kawasaki disease or hyperlipidemia has been associated with MI at an early age. However, most children with chest pain will be found to have less severe acute inflammatory

processes of the respiratory tract or musculoskeletal system or a psychosomatic disturbance.

Infectious diseases of the respiratory tract are associated with fever, malaise, cough, and coryza and may involve several family members simultaneously. A first asthmatic attack should be suspected when an associated night cough, history of wheezing, or family history of atopy is present. The physical examination in asthma shows a prolonged expiratory phase of respiration, variable degrees of chest hyperinflation, and wheezing accentuated by a forced expiratory effort. In musculoskeletal inflammation, one should be able to elicit tenderness of the chest wall and a “trigger point,” where palpation reproduces the pain. Reproduction of the pain by a “hooking maneuver” performed over the lower anterior ribs implicates the “slipping rib syndrome.” Pain following a dermatome unilaterally suggests intercostal neuritis; children with zoster (shingles) may have pain preceding the development of rash.

When focal, peripheral pain is found without a trigger point, the physician should consider pain referred from areas of sensory nerve overlap. A relationship of the pain to eating or swallowing suggests esophageal disease, and often, the physical examination may appear normal. Some of these patients will have a thin body habitus and/or cardiac findings of mitral valve prolapse. A foreign body (e.g., a coin) in the proximal esophagus commonly manifests with chest discomfort and drooling in the young child. Similarly, an aspirated foreign body may cause dull, aching chest pain associated with cough. Auscultation and plain radiographs of the chest do not always reveal the object or signs of an obstructed upper or lower airway. “Texidor's twinge,” or pericardial “catch,” is a relatively common cause of short duration and sharp pain in healthy teenagers and young adults, often related to exercise and located in the left substernal region. It may be produced by stretching of the supporting ligaments of the heart and is easily distinguishable from angina by its sudden, stabbing onset, a duration of less than 60 seconds, and the absence of referral to other areas. Cigarette smoking has been associated with chest pain in teenagers and adults.

A thorough examination usually uncovers evidence of the cardiac and respiratory causes of chest pain listed in the tables. In addition to the usual cardiac and pulmonary examination, one should search for trigger points on the chest wall and changes in pain associated with positional changes. Chest pain relieved by leaning forward is consistent with pericarditis; whereas that which is worsened by reclining may represent gastroesophageal reflux or hiatal hernia. Extrathoracic abnormalities, such as a rash or arthritis, may provide clues to collagen disorders (see Chapter 101) or other systemic illness. Marfan's syndrome should be suspected in the tall, thin patient whose upper extremity span exceeds his or her height. During examination of the heart and lungs, it is useful to relate normal findings to the child and family because this reassurance often serves as the major “treatment” of self-limiting or functional problems.

Laboratory studies sometimes are indicated to help confirm a diagnosis or to relieve the anxiety of the child or family. Pulse oximetry is a quick and inexpensive screen that is helpful in determining the severity of any suspected pulmonary disease. Chest radiographs may reveal findings consistent with asthma, pneumonitis, pleurisy, or spontaneous pneumothorax. Foreign bodies ingested and lodged in the esophagus can be visualized if radiopaque (e.g., coins). In the cervical esophagus, they will lie flat in the posteroanterior view of the chest. However, foreign body aspiration most often manifests by hyperinflation or atelectasis on radiographs because most tracheobronchial foreign bodies are not radiopaque. Inspiratory and expiratory films or decubitus chest radiographs may help demonstrate focal hyperinflation (i.e., the lobe with an obstructed bronchus remains inflated in expiration or when placed on the down side on a decubitus film). The wide mediastinum from an aortic aneurysm, abnormal cardiac silhouette related to a pericardial effusion or cardiomegaly, rib fractures or bone changes of metabolic bone derangements, and cysts all produce characteristic radiographic changes. A calcified ring may be visualized in approximately one-third of patients with a history of Kawasaki disease. The presence of atelectasis may suggest mucous plugging or may be subtle evidence of pulmonary infarction from an embolus or a vaso-occlusive crisis of sickle cell anemia.

An ECG should be performed if cardiac disease is suspected. The ECG will be normal in most cases in which the physical examination is unremarkable. It may show signs of cardiac strain or ischemia with valvular heart disease, diseases of outflow obstruction, or angina. Acute cocaine exposure may present with classic signs of myocardial ischemia or myocardiopathy. A decreased QRS wave voltage and electrical alternans suggest the presence of a pericardial effusion in the child with muffled heart sounds. Arrhythmias, such as atrial fibrillation and supraventricular tachycardia, may be identified by careful evaluation of a rhythm strip.

Studies other than chest radiographs and ECGs are rarely necessary. An elevated leukocyte count with a shift to the left may point toward infection as the cause of pain. Examination of a peripheral smear and a hemoglobin electrophoresis are indicated in the child suspected of having sickle cell disease as the cause of chest pain. If an intra-abdominal source for chest pain from diaphragmatic irritation is under consideration, a serum amylase may be obtained when considering pancreatitis. If a right-sided subdiaphragmatic abscess is possible, alanine aminotransferase (ALT) or aspartate transferaminase (AST) elevation may reveal the inflammatory nature of the process further delineated by ultrasound or computed tomography (CT) scan. Esophageal causes of chest pain often may be diagnosed clinically in the ED with a trial of antacid therapy. To confirm the findings of a hiatal hernia, esophagitis, or a nonopaque foreign body, a barium study or endoscopy may be required. Pulmonary embolus, presenting occasionally in the older teenager or debilitated patient, may be suspected clinically. Low Pa_{O_2} and/or ECG abnormalities are suggestive. This suspected diagnosis requires the performance of nuclear ventilation-perfusion scans or angiography for confirmation. Toxicologic screens are useful if the patient is considered at risk of drug abuse, particularly cocaine, or the diagnosis remains unclear.

A large group of children with chest pain will have no evidence of organic disease and no history of underlying cardiorespiratory disease or trauma. They may have a family history of chest pain. Often, there will be a stressful situation that has precipitated the episode. Complaints of chest pain and other somatic aches often are chronic, with no abnormalities noted on the physical examination. Such children have psychogenic chest pain. To elicit the predisposing factors, the physician should interview the child and family away from the chaos of the ED, if possible. Often, a chest radiograph and an ECG are helpful in allaying parental fears of cardiac disease. Definitive ongoing management, however, requires referral to a primary care physician.

SUMMARY

Chest pain in children is a relatively uncommon sign of serious disease but often has great importance to the patient or family. Most cases can be diagnosed by the emergency physician from the history and physical examination alone, although at times, a chest radiograph or an ECG is helpful. The physician should always consider drug-induced chest pain (especially associated with cocaine) and other life-threatening conditions. Psychogenic chest pain is a common occurrence and may be chronic or related to an acute stressful event. The possibility of cardiac disease needs to be addressed directly by the examining physician to alleviate fully the patient's (or family's) anxiety. The most common causes of organic chest pain are musculoskeletal (traumatic or inflammatory) and infectious disorders, usually self-limiting or easily treated diseases. Occasionally, serious abdominal, pulmonary, or cardiac problems require immediate attention.

Suggested Readings

Asnes R, Santulli R, Bemborad J. Psychogenic chest pain in children. Clin Pediatr 1981;20:788.

Burns JC, Shike H, Gordon JB, et al. Sequelae of Kawasaki disease in adolescents and young adults. J Am Coll Cardiol 1996;28:253–257.

Dekel B, Paret G, Szeinberg A, et al. Spontaneous pneumomediastinum in children: clinical and natural history. Eur J Pediatr 1996;155:695–697.

Driscoll D, Glicklich L, Gallen N. Chest pain in children: a prospective study. Pediatrics 1976;57:648.

Editorial. Texidor's twinge. Lancet 1979;2:133.

Gitter MJ, Goldsmith SR, Dunbar DN, et al. Cocaine and chest pain: clinical features and outcome of patients hospitalized to rule out myocardial infarction. Ann Intern Med 1991;115:277–282.

Green M. Chest pain. In: Pediatric Diagnosis. 4th ed. Philadelphia: WB Saunders, 1986:390–391.

MacBryde CM, Blacklow RS. Thoracic pain. In: Signs and Symptoms. Philadelphia: JB Lippincott, 1970.

Pittman JAL, Pounsford JC. Spontaneous pneumomediastinum and Ectasy abuse. J Accid Emerg Med 1997;14:335–336.

Selbst SM, Ruddy RM, Clark BJ, et al. Pediatric chest pain: a prospective study. Pediatrics 1988;82:319–323.

Selbst SM, Ruddy RM, Clark BJ, et al. Follow up of patients previously reported. Clin Pediatr 1990;29:374–377.

Taubman B, Vetter VL. Slipping rib syndrome as a cause of chest pain in children. Clin Pediatr 1996;35:403–405.

Woolf PK, Gewitz M, Berezin S, et al. Noncardiac chest pain in adolescents and children with mitral valve prolapse. J Adolesc Health 1991;12:247–250.

CHAPTER 53

Pain—Dysphagia

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The primary function of swallowing is the ingestion, preparation, and transport of nutrients to the digestive tract. Secondary functions of swallowing are the control of secretions, clearance of respiratory contaminants, protection of the upper airway, and equalization of pressure across the tympanic membrane through the eustachian tube. Dysphagia is defined as any difficulty or abnormality of swallowing. Dysphagia is not a specific disease entity but is a symptom of other, often clinically occult, conditions and may be life-threatening if respiration or nutrition are compromised. Odynophagia (pain on swallowing) or sialorrhea (drooling) may also be present in the dysphagic pediatric patient. This chapter briefly presents the normal anatomy and physiology of swallowing, the differential diagnosis of disturbances of this process, and the evaluation and treatment of the pediatric patient with dysphagia.

PATHOPHYSIOLOGY

Swallowing begins in utero as early as the 16th week of gestation, playing an important role in gastrointestinal development and regulation of amniotic fluid volume. By the 34th week of gestation, this complex process, involving 26 muscles, 6 cranial nerves (V, VII, IX, X, XI, XII), and cervical nerves C1–C3, is functional, although incompletely coordinated with breathing. In the first few days after birth, each infant develops an individual pattern of sucking, swallowing, and breathing, usually with a 1:1 or 1:2 ratio of breaths per suckle, to prevent aspiration of material into the larynx. This stage of suckling, or suckle feeding, is primarily under medullary control, with minimal input from the cerebral cortex. A transitional period begins at 6 months of age, as the cortex gradually exerts more control over the preesophageal phase of swallowing, allowing for the introduction of solid foods. The preesophageal region depends on normal sensorimotor function of cranial nerves V, VII, IX, and XII, which enervate voluntary skeletal muscles of the face, tongue, and neck, as well as the involuntary muscles of the posterior pharynx. Swallowing in the esophageal region remains an autonomic process, with vagal sensorimotor control coordinating peristalsis of the upper striated and lower smooth muscle of the esophagus. By 3 years of age, the swallowing pattern is mature, although the pediatric patient, unlike the adult, may regress to a less mature stage if normal swallowing is disrupted.

To facilitate suckle feeding and breathing, the infant oropharynx is anatomically different from the adult, with a relatively larger tongue, smaller oral cavity, and more anterior and superior epiglottis and larynx. As the face and mandible grow, the oropharynx enlarges, creating more room for the eventual voluntary use of the tongue and dentition, and the larynx descends, eventually allowing for mouth breathing. Although breathing continues to cease during swallows, the older child depends less on close coordination between eating and breathing.

A normal swallow, using the suckling infant as an example, begins with rhythmic movement of the lips, tongue, and mandible. These parts function as a unit, creating negative intraoral pressure, while also compressing the nipple. The milk expressed from each suckle is stored in the posterior oral cavity until a larger fluid bolus is formed. As the tongue delivers the bolus to the pharynx, the nasopharynx is closed off by the posterior tongue and by elevation of the soft palate. The larynx elevates to a position under the tongue, closing the airway, as the epiglottis inclines to direct the bolus posterior. A pharyngeal wave of contraction sweeps the bolus toward the upper esophagus, where the cricopharyngeal sphincter relaxes, allowing passage into the esophagus. As the esophagus begins peristaltic contractions and the bolus moves past a relaxed lower esophageal sphincter into the stomach, the airway reopens, the cricopharyngeal sphincter constricts to close the upper esophagus, and respirations resume. Dysphagia can result from disruption of normal mechanisms at any stage of the swallowing process.

DIFFERENTIAL DIAGNOSIS

The differential diagnostic list for dysphagia is extensive and is commonly divided into preesophageal or esophageal disorders (Table 53.1). Preesophageal causes of dysphagia are further subdivided into anatomic categories, including nasopharyngeal, oropharyngeal, laryngeal, and generalized problems. Infectious and inflammatory disorders of either anatomic region may disrupt swallowing, whereas neuromuscular problems tend to be predominantly preesophageal, given the autonomic function of the esophagus. However, the esophagus can be affected by motility disorders intrinsic to smooth muscle. Finally, the differential diagnosis includes several systemic conditions that may affect the normal swallowing process.

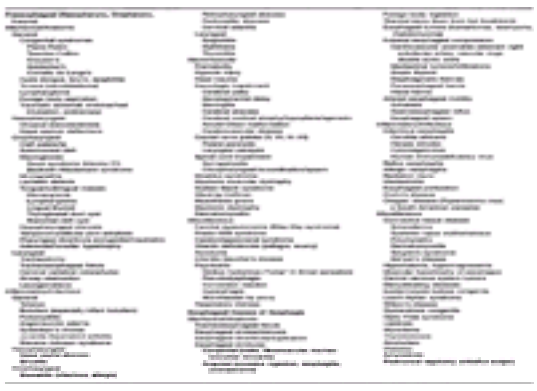

Table 53.1. Differential Diagnosis of Dysphagia

In the adult patient, dysphagia most commonly results from a variety of neuromuscular disorders, whereas the pediatric patient more often has swallowing difficulty from congenital, infectious, inflammatory, or obstructive causes (Table 53.2). In the newborn or infant, swallowing may be disturbed as a result of prematurity, often associated with respiratory and neurologic disabilities. Gastroesophageal reflux is common in infants, although in a small percentage of patients, it may persist into childhood with reflux esophagitis. Ingestion or aspiration of a foreign body must always be considered in the toddler who has either the acute or chronic onset of dysphagia.

Newborn/Infant	*Child*
Prematurity	Foreign body aspiration/ingestion
Tracheoesophageal fistula	Caustic ingestion
Choanal stenosis/atresia	Infectious
Birth trauma	Neurologic impairment (cerebral palsy, mental retardation, head trauma)
Congenital abnormalities	Inflammatory
Gastroesophageal reflux	
Respiratory illness	
Neurologic/neuromuscular disease	
Infectious (botulism, candidiasis, herpetic esophagitis)	
Inflammatory	

Table 53.2. Common Causes of Dysphagia

Life-threatening causes of dysphagia may involve airway compromise, serious local or systemic infection, and inflammatory disease (Table 53.3). The newborn may have a congenital anatomic abnormality such as tracheoesophageal fistula, with aspiration of swallowed fluid into the lungs, or may have traumatic injury to the upper airway and esophagus from iatrogenic instrumentation in the delivery room. The older child may have a foreign body in the airway or esophagus, with the possibility of complete airway obstruction (see Chapter 29). Numerous infectious processes may present with dysphagia and can threaten airway integrity. These include epiglottitis, retropharyngeal abscess, Stevens-Johnson syndrome, and central nervous system (CNS) infections.

Foreign body aspiration/ingestion	Polio
Tracheoesophageal fistula	Diphtheria
Upper airway obstruction	Central nervous system infection/ abscess
Traumatic esophageal perforation	Stevens-Johnson syndrome
Epiglottitis	Corrosive ingestion
Retropharyngeal abscess	Laryngeal paralysis
Botulism	
Tetanus	

Table 53.3. Life-Threatening Causes of Dysphagia

EVALUATION AND DECISION

The evaluation of dysphagia in the pediatric patient begins with a detailed history, including pregnancy and delivery, family history, feeding history, growth and development, and a history of other illness (Table 53.4). An accurate and complete history should suggest the diagnosis in approximately 80% of patients. Prenatal polyhydramnios, maternal infection, maternal drug or medication use, bleeding disorders, thyroid dysfunction, toxemia, or irradiation may lead to swallowing problems in the newborn or infant. Fetal neurologic development may be altered by prenatal difficulties and may result in dysphagia after birth. Maternal myasthenia gravis may also cause temporary feeding problems in the newborn.

Table 53.4. Important Historical Features for Dysphagia

A history of traumatic delivery may result in neurologic injury or laryngeal paralysis. Newborn intubation may be associated with trauma to the trachea, larynx, or esophagus, as well as hypoxic brain injury. A history of prematurity, developmental delay, failure to thrive, hypotonia, or associated congenital abnormalities may indicate a neuromuscular cause for dysphagia. The feeding history should include acute or chronic onset of symptoms, age at onset, weight loss, failure to thrive, and type and amount of food the child eats. Presence of fever, pain, respiratory symptoms, facial color, stridor, liquid or solid food intolerance, vomiting, regurgitation, drooling, voice change, position during feeding, and the timing of symptoms in relation to feedings should also be documented. For example, the infant with an upper airway obstruction may become fatigued or begin coughing and choking shortly after beginning to eat. Anatomic abnormalities of the trachea, larynx, or esophagus commonly present in infancy as respiratory problems during feeding, although vascular lesions that result in extrinsic compression of the esophagus may remain silent until the introduction of solid foods, and occasionally into adulthood. Gastroesophageal reflux in infants may manifest as vomiting shortly after feeding or with a history of nighttime cough or emesis. Intrinsic lesions, from inflammation, tumor, or foreign body, may create problems with solid food but cause no difficulty with liquids. Infants with previously unrecognized neuromuscular disorders commonly present initially with dysphagia, particularly for liquids; drooling; prolonged feeding time; weak suckle; or nasal reflux of swallowed material. A history of fever may indicate an aspiration pneumonia or other infectious or inflammatory causes of dysphagia.

The child with dysphagia should undergo a thorough general physical examination, initially focusing on the patient's cardiopulmonary status. Evidence of respiratory distress or cardiovascular compromise should be treated promptly in the appropriate manner, as outlined elsewhere in this text (see Chapter 1, Chapter 2, and Chapter 5). Assurance of a secure and stable airway should precede attempts to examine the oropharynx or to remove a foreign body (see Chapter 29).

In the stable dysphagic patient, evaluation of head size and shape, facial structure, mandibular development, tongue disproportion, and ear configuration may provide evidence of an underlying congenital abnormality, such as Pierre-Robin, Treacher-Collins, Crouzon's, and Goldenhar's syndrome. Evaluation of nasal airway patency in the infant can be determined by gently passing an 8-Fr catheter through the nares into the stomach. If the catheter fails to pass easily, choanal stenosis, atresia, or esophageal obstruction must be considered. Inspection of the oral cavity, pharynx, and neck may reveal a cyst, mass, localized infection, or inflammatory cause for dysphagia. Cervical auscultation over the thyroid cartilage during feeding may note evidence of aspiration if upper airway breath sounds are abnormal or if the timing of breathing and swallowing is uncoordinated. The pulmonary examination may also detect signs of aspiration or respiratory compromise, including elevated respiratory rate, increased respiratory effort, stridor, stertor, rales, rhonchi, wheezing, or change in voice quality. Neurologic examination may reveal an altered level of arousal from an underlying brain injury or depressed sensorium from drugs or infection that may limit effective swallowing. Examination of the cranial nerves, particularly V, VII, IX, X, and XII, may reveal abnormalities from traumatic or surgical injury, tumor, or congenital disorder. Evaluation of muscle tone, strength, and reflexes in consideration of other neuromuscular causes of dysphagia completes the general physical examination.

Provided oral intake is not contraindicated by an expected procedure or intervention, observation of a typical feeding, given by a parent or primary caretaker, may help elucidate the cause of dysphagia. The manner of presentation of food to the patient, the consistency and amount given, patient position, duration of feeding, regurgitation (oral or nasal), agitation or behavior change, or the development of respiratory symptoms may further guide the diagnostic evaluation. Patients with upper airway obstruction may have an exacerbation of symptoms when attempting to drink. Patients with lesions such as tracheoesophageal fistula, vascular rings, or esophageal obstruction may begin coughing and choking soon after drinking without any initial difficulty. However, esophageal disorders such as extrinsic compression, strictures, tumors, or altered motility commonly are clinically silent and typically require use of radiographic or direct visual techniques for diagnosis.

Evaluation of the stable dysphagic patient may proceed on the basis of age and acute versus chronic onset of symptom development (Fig. 53.1). The neonate and young infant will require evaluation techniques and consideration of the age-related differential diagnoses outlined in Table 53.2, whereas the older child with an acute onset of dysphagia generally requires a more urgent approach. Witnessed or suspected foreign bodies, either ingested or aspirated, should be investigated with plain radiographs (or contrast studies if a radiolucent object is considered) and, if identified, emergently removed (see Chapter 29). A history of neck trauma or caustic ingestion should lead to the suspicion of aerodigestive tract abnormalities. These patients may present dramatically with neck pain, drooling, and evidence of facial or other trauma, but they may also have a subacute presentation (see Chapter 88, Chapter 106, and Chapter 112). Presence of fever or signs of systemic illness may result from potentially life-threatening infectious or inflammatory conditions (Table 53.3). Less severe problems (gingivostomatitis or thrush) may present with mouth lesions and can be managed on an outpatient basis after careful assessment of hydration status. Severe problems, including Stevens-Johnson syndrome, herpetic esophagitis, and diphtheria, may be disc overed on a detailed examination and

may require inpatient management.

FIGURE 53.1. Evaluation scheme for the child with dysphagia or odynophagia. *Radiographic options:* neck, chest, abdomen, inspiratory/expiratory films, lateral decubitus films, fluoroscopy, contrast studies, ultrasonography, echocardiography, angiography, computed tomography, magnetic resonance imaging. *Laboratory options:* complete blood count, blood gas, cultures, toxin identifications, nutritional and electrolyte profile. *Consultant's options:* pediatrics, general surgery, otolaryngology, gastroenterology, neurology, infectious disease, cardiology, pulmonology, rheumatology, oncology, nutrition, speech therapy, occupational therapy.

Patients with a nonacute history of swallowing difficulty can be evaluated and treated as shown in Figure 53.1. The initial emphasis with these patients lies more in determination of nutritional status and development issues than in acute emergency department (ED) intervention, although prolonged feeding difficulty can develop into a life-threatening problem. The child with obvious anatomic abnormalities, neurologic impairment, specific syndromes, or a tracheostomy may need referral to appropriate subspecialists after initial evaluation. The child without obvious anatomic or neurologic abnormality who has weight loss or failure to thrive may be evaluated as an outpatient.

Radiographic evaluation of the stable dysphagic patient usually begins with an examination of the airway and soft tissues of the neck, looking for evidence of a foreign body, mass, airway impingement, or other abnormality. A chest radiograph may suggest aspiration pneumonia, congenital heart disease, or mediastinal abnormality or, as in the patient with achalasia, demonstrate fluid levels within an enlarged esophagus. Computed tomography (CT) scan, echocardiography, or angiography may further identify problems suspected from initial studies.

A videofluoroscopic swallowing study (VFSS or modified barium swallow) is currently the gold standard for evaluating preesophageal disorders. The patient is fed a typical solid or liquid diet (mixed with contrast material) by his or her parent or caretaker, while the radiologist records the preesophageal and esophageal swallowing phases on videotape. With a swallowing specialist present, such as a speech–language pathologist, the feeding presentation and position, consistency, amount, and type of foods can be varied, both to diagnose problems resulting in dysphagia or aspiration and to evaluate possible therapeutic interventions. This dynamic study may reveal evidence of aspiration, nasopharyngeal reflux, motility disorders, obstructions, masses, cricopharyngeal dysfunction, fistulas, inflammatory processes, or other causes of dysphagia. VFSS differs from the standard barium swallow (BS) or upper gastrointestinal (UGI) series in that it does not use pure contrast but instead uses food mixed with contrast in an attempt to simulate the normal feeding pattern as closely as possible. VFSS is less effective than a UGI series or BS at diagnosing gastroesophageal reflux or lower esophageal, gastric outlet, and small bowel abnormalities (Fig. 53.2), but it is superior in identifying preesophageal causes of dysphagia.

FIGURE 53.2. Upper gastrointestinal series (UGI) of a 14 year old girl presenting with dysphagia. The UGI demonstrates a significantly dilated upper esophagus, with a functional spasmodic obstruction of the lower esophagus, characteristic of achalasia.

Fiberoptic endoscopy under local or general anesthesia may be indicated for suspected mass lesion, stricture, caustic ingestion, inflammatory lesion, or foreign body. Endoscopy may also be used to obtain biopsy specimens to aid in diagnosis. In addition, nasopharyngoscopy allows direct visualization of the swallowing process and can document aspiration and functional esophageal disorders.

Other tests, such as a complete blood count, appropriate cultures in the febrile patient, or arterial blood gas for the

patient with respiratory distress, also may be indicated. Cervical ultrasonography has been used to identify abnormalities with the tissues and function of the palate, tongue, and floor of the mouth but is less useful than contrast studies for assessing airway problems and aspiration. Manometry may be useful in the dysphagic patient with an esophageal motility disorder, but it is better tolerated and more typically used in adults. Esophageal pH testing or radionuclide scintigraphy (milk scans) may document previously unsuspected gastroesophageal reflux. Additional neurologic testing may include studies of brainstem-evoked responses, peripheral nerve conduction, or electromyography.

Treatment of dysphagia is dictated by the diagnosis. Disorders with the potential to become life-threatening should be treated in the hospital under the care of appropriate specialists. If nutrition has been severely compromised from chronic dysphagia, one should consider nasogastric, nasojejunal, or gastrostomy tube feedings. Many pediatric facilities have developed multidisciplinary feeding/swallowing teams to provide subspecialty expertise, while maintaining continuity and coordination of patient care. Such pediatric teams may include developmental or general pediatricians, speech–language pathologists, pulmonologists, otolaryngologists, gastroenterologists, neurologists, nutritionists, psychologists, occupational or physical therapists, and social service workers. If such a specialty service is not available, involvement of appropriate individual specialists for the management of the patient with dysphagia is imperative as mentioned in Figure 53.1. However, therapy for many disorders can be initiated on an outpatient basis. Gastroesophageal reflux and resultant esophagitis can often be successfully managed with small volume thickened feeds, positioning, and elevation of the head of the bed. Medical therapy consists of liquid antacids, cisapride (a gastric motility agent), and H_2-blockers. Mycostatin will be helpful in candidal esophagitis, whereas herpetic esophagitis often is self-limiting.

Pediatric dysphagia is an uncommon complaint in the pediatric ED but may be the presenting symptom for a wide variety of underlying clinical problems. The history and physical examination first must focus on potentially life-threatening causes and will often lead to a specific diagnosis. Causes of dysphagia not identified from the initial evaluation may require radiographic or subspecialty referral for further diagnostic and therapeutic management.

Suggested Readings

Arvedson JC. Dysphagia in pediatric patients with neurologic damage. Semin Neurol 1996;16(4):371–386.

Brodsky L. Dysphagia with respiratory/pulmonary presentation: assessment and management. Semin Speech Lang 1997;18(1):13–22.

Derkay CS, Schechter GL. Dysphagia. In: Bluestone CD, Stool SE, Kenna MA, eds. Pediatric Otolaryngology. Vol. 3. Philadelphia: WB Saunders, 1996.

Gilardeau C, Kazandjian MS, Bach JR, et al. Evaluation and management of dysphagia. Semin Neurol 1995;15(1):46–51.

Jolley SG, McClelland KK, Mosesso-Rousseau M. Pharyngeal and swallowing disorders in infants. Semin Pediatr Surg 1995;4(3):157–165.

Jones B, Donner MW. Examination of the patient with dysphagia. Radiology 1988;167:319–326.

Kramer SS. Radiologic examination of the swallowing impaired child. Dysphagia 1989;3:117–125.

Lefton-Greif MA, Arvedson JC. Pediatric feeding/swallowing teams. Semin Speech Lang 1997;18(1):5–11.

Lefton-Greif MA, Loughlin GM. Specialized studies in pediatric dysphagia. Semin Speech Lang 1996;17(4):311–329.

Loughlin GM, Lefton-Greif MA. Dysfunctional swallowing and respiratory disease in children. Adv Pediatr 1994;41(2):135–162.

Myers MA. Esophageal disorders associated with emesis in infants and children. Semin Pediatr Surg 1995;4(3):166–175.

Reilly S, Skuse D, Poblete X. Prevalence of feeding problems and oral motor dysfunction in children with cerebral palsy: a community study. J Pediatr 1996;129:877–882.

Shapiro J, Healy GB. Dysphagia in infants. Otolaryngologic Clin North Am 1988;21(4):737–741.

Tuchman DN. Dysfunctional swallowing in the pediatric patient: clinical considerations. Dysphagia 1988;2:203–208.

Tuchman DN. Cough, choke, sputter: the evaluation of the child with dysfunctional swallowing. Dysphagia 1989;3:111–116.

Weiss MH. Dysphagia in infants and children. Otolaryngologic Clin North Am 1988;21(4):727–735.

Werlin SL. Dysphagia. In: Hoekelman RA, ed. Primary Pediatric Care. 3rd ed. St Louis: Mosby, 1997:918–922.

CHAPTER 54

Dysuria

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Dysuria, or the sensation of pain when voiding, stems from irritation of the bladder, the urethra, or both. The sensation is produced by the contraction of the bladder and the peristaltic activity of the urethra, both of which stimulate the pain fibers of the edematous and erythematous mucosa. In addition, 1) young children may complain of painful urination when they are instead experiencing related symptoms, such as pruritus, and 2) parents may interpret various nonspecific statements or behaviors by their children as indicative of painful urination.

Dysuria is a commonly reported symptom that has a number of different causes (Table 54.1), but it usually stems from one of several common disorders of childhood and adolescence (Table 54.2). Most children with dysuria as a chief complaint have disorders of the genitourinary tract. Although patients with urethritis secondary to systemic illnesses may have dysuria as one of their many symptoms, it is only occasionally the principal reason for a visit to an emergency department (ED).

I. Systemic Conditions
  A. Stevens-Johnson syndrome
  B. Reiter's syndrome
  C. Behçet's syndrome
II. Localized Conditions
  A. Infection
    1. Pyelonephritis
    2. Cystitis
      a. Viral
      b. Bacterial (*Escherichia coli* and other organisms)
    3. Urethritis/balanitis
      a. *Neisseria gonorrhoeae*
      b. *Chlamydia* species
      c. Herpes simplex
      d. Other
  B. Chemical Irritation
    1. Detergents
    2. Fabric softeners
    3. Perfumed soaps
    4. Bubble baths (?)
    5. Medication
  C. Trauma
    1. Local injury
    2. Masturbation
  D. Miscellaneous
    1. Hypercalciuria/urinary stones
    2. Labial adhesions
    3. Psychogenic dysuria
III. Complaints Misinterpreted as Dysuria
  A. Pinworms
  B. Sexual abuse

Table 54.1. Causes of Dysuria

Disorder	Cause	Age	Fever	Tenderness
Pyelonephritis	*E. coli* other bacteria	All	Common, >38.8°C	Flank
Cystitis	Viruses*E. coli* other bacteria	All	Occasional, <38.8°C	Suprapubic
Infectious arthritis	*N. gonorrhoeae* *C. trachomatis*	Adolescents	None	Prominent joint (occasional)
Chemical/traumatic arthritis	Physical insult	Children	None	None

Table 54.2. Differential Diagnosis of Common Causes for Dysuria

Most diseases causing dysuria are self-limited or easily treated; however, the rarely seen systemic causes of urethritis or the spread of some bacterial pathogens beyond the genitourinary tract may be life-threatening (Table 54.3)

Stevens-Johnson syndrome
Gonococcal urethritis/vaginitis (when complicated by pelvic inflammatory disease or systemic spread)

Table 54.3. Life-Threatening Causes of Dysuria

DIFFERENTIAL DIAGNOSIS

Systemic Conditions (Table 54.1)

Stevens-Johnson syndrome is a severe manifestation of erythema multiforme, which may affect the mucous membranes throughout the body, producing conjunctivitis, oral ulceration, and urethritis. The rash that occurs in most patients often has the appearance of target lesions. Although usually self-limited, in some cases, pulmonary involvement leads to death.

Reiter's syndrome is characterized by conjunctivitis, arthritis, and urethritis. Rarely diagnosed, it is more common in males.

Also rare, Behçet's syndrome is another multisystem disease that may cause urethral ulceration and dysuria.

Localized Conditions (Table 54.1)

Infection of the genitourinary tract is the predominant cause of dysuria. Pyelonephritis is the most serious of these disorders, usually manifesting with fever, often above 39°C (102.2°F), and flank pain or tenderness (older children and adolescents). Patients with cystitis may or may not have fever, which is usually low grade, and suprapubic pain or tenderness. Urethritis is a more localized infection, which often produces a discharge. In adolescents, *Neisseria gonorrhoeae* and *Chlamydia trachomatis* are the most common pathogens. When herpes simplex causes urethritis, vesicles are usually apparent on examination. Younger children may develop a nonspecific bacterial urethritis with involvement of the glans penis (balanitis) or both the glans and the prepuce (balanoposthitis). In the setting of an urban ED, infectious urethritis and/or cervicitis may be the source of isolated dysuria in up to 30% of adolescent girls and often goes undiagnosed when only a urine specimen, and not a cervical swab, is sent for culture.

In young children, certain drugs taken systemically and topical exposures to a variety of chemicals have been reported to irritate the urethral mucosa; however, these findings have not been well documented. Potential local irritants include detergents, fabric softeners, perfumed soaps, and possibly bubble baths. These patients have either no physical findings or only mild erythema but no discharge.

Minor injury is another relatively common cause of urethral irritation. In older children and adolescents, normal self-exploratory sexual play, masturbation, voluntary sexual activity, or sexual abuse may be the source of the trauma. As for patients with chemical urethritis, the examination is generally unremarkable.

The passage of a stone is an uncommon cause of dysuria in children. In these cases, the dysuria is usually proceeded by flank pain and is often accompanied by hematuria. Generally, urinary stones in children develop in the setting of anatomic abnormalities and/or recurrent infection. Exceptions to this rule are children who are otherwise healthy but have idiopathic hypercalciuria; these children may develop calculi. In addition, these patients have been reported to complain of dysuria even in the absence of an observable stone.

Labial adhesions occur relatively often in young girls. Although they are most often asymptomatic, they may cause dysuria on occasion.

Throughout childhood and into adolescence, a complaint of dysuria may be psychogenic in origin, occurring in the absence of inflammation in the genitourinary tract.

Complaints Misinterpreted as Dysuria (Table 54.1)

Enterobius vermicularis (pinworms) infests the perianal area, but occasionally spreads to the vagina in young girls. The pruritus that accompanies this infestation may be expressed as dysuria.

Young children who have experienced sexual abuse may complain of pain in their genital area or may exhibit behaviors that are interpreted by adult observers as indicative of genital pain. These patients may appear in the ED with a chief complaint of dysuria.

EVALUATION AND DECISION

When evaluating a child with dysuria, the physician should ask about trauma and exposure to chemicals such as detergents, fabric softeners, perfumed soaps, bubble baths, and medications that have been reported to irritate the mucosal lining of the urethra or bladder. A negative history for injury may not be accurate because most trauma is not recalled by young patients or, in the case of masturbation or abuse, may be denied. The detection of sexually transmitted diseases, a common cause of dysuria in adolescents, may be facilitated by obtaining a history about the nature and extent of sexual activity.

Some children may have a suggestive constellation of symptoms and signs in addition to dysuria. As examples, the adolescent girl with abdominal and pelvic discomfort probably has pelvic inflammatory disease and the 6-year-old child with fever and flank pain most likely has pyelonephritis. Various systemic disorders (Table 54.1) may cause conjunctivitis, oral ulceration, arthritis, and cutaneous lesions.

In addition to a routine physical examination, special attention should be directed to the genitourinary tract. A swollen or edematous urethral meatus or a discharge from the urethra or vagina will localize the source of the symptoms in an otherwise well child to the genitalia, obviating the need to look extensively for other diseases. In particular, vesicles occur with infections caused by herpes simplex. Labial adhesions are easily recognized on inspection in young girls.

The most important tasks for the emergency physician are to recognize the rare but serious systemic syndromes and to diagnose or exclude infections (Fig. 54.1). If the history and general examination do not suggest a systemic condition, the next step is to examine the genitalia. Lesions, such as vesicles or ulcers, in this area are seen with Stevens-Johnson and Behçet's syndrome, but they more likely represent an infection with herpes simplex, even in the preadolescent child.

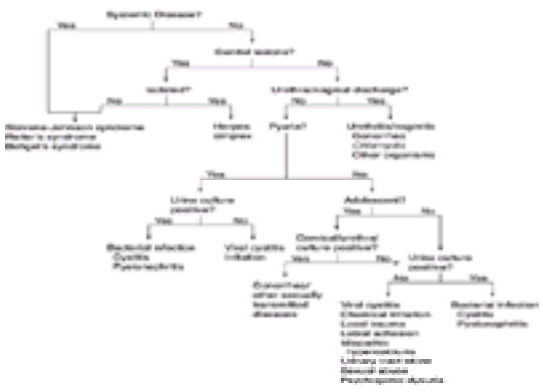

FIGURE 54.1. Approach to the diagnosis of dysuria.

A urethral or vaginal discharge suggests an infection of the genitalia: urethritis in the boy, and urethritis or vaginitis in the girl. *N. gonorrhoeae* is an organism that commonly causes disease in this area. A Gram stain often provides a clue to the diagnosis. In prepubertal girls or boys of any age, the finding of Gram-negative intracellular diplococci points to the diagnosis of gonorrhea. In contrast, because nonpathogenic organisms that colonize the vagina after puberty have the same appearance as *N. gonorrhoeae* on Gram stain, the smear is an unreliable tool in teenage girls. A urethral or vaginal culture should be obtained for confirmation and susceptibility testing. Postpubertal patients may be treated in the ED, but treatment in young children should await the results of cultures because of medical-legal implications.

If no discharge is seen, the physician should obtain a urinalysis and urine culture. A positive result on testing by dipstick (leukocyte esterase and/or nitrites) or the finding of pyuria (more than 5 to 10 white blood cells per high-power field) increases the likelihood of bacterial infection (urethritis, cystitis, or pyelonephritis) but does not prove the diagnosis. Inflammatory conditions, such as chemical urethritis, and nonbacterial infections may also evoke a leukocyte response. Thus, the physician may choose to allow the results of cultures to guide the management of children with pyuria, rather than immediately beginning antibiotic therapy, unless fever and flank pain point to pyelonephritis.

In the young child with dysuria in the absence of pyuria, local trauma and chemical irritation are the most likely causes for the pain. Because a few children with urinary tract infections (UTIs) do not have either a positive result on testing by dipstick or pyuria, most physicians obtain a urine sample for culture, particularly from febrile patients. On the other hand, some experts would argue that the likelihood of infection is low enough in the absence of positive indicators on urine analysis that no further testing is needed. Adolescents require cultures of the genital tract to diagnose mild gonococcal or chlamydial infections.

When no other cause for dysuria is found in a prepubertal girl who has adhesions of her labia minora, these adhesions may be responsible for the painful urination. Most girls with labial adhesions, however, are asymptomatic. Thus, infection or another cause for dysuria should be excluded in girls with this finding.

A few patients with a normal examination and negative cultures may complain persistently of dysuria. In this setting, idiopathic hypercalciuria provides a potential diagnosis. If suspected, the diagnosis can be confirmed by measurement of calcium excretion in the urine. Another possible explanation is that the patient is experiencing vaginal pruritus secondary to pinworms. Confirmation of this diagnosis requires either identification of the larvae or ova or a response to a trial of mebendazole.

Last, the physician should give consideration to both sexual abuse and psychogenic dysuria. In most of these cases, further evaluation outside of the ED will be needed.

INITIAL MANAGEMENT

When a specific diagnosis has not been established and the physician is awaiting the results of cultures, therapy directed at the symptom of dysuria can provide some relief. Generally, a dilute urine causes less irritation than a concentrated one, so a generous fluid intake should be recommended. For the child over 6 years of age, phenazopyridine (Pyridium) at a dosage of 100 mg given three times per day may be helpful as a urinary tract anesthetic.

Suggested Readings

Alon A, Warady BA, Hellerstein S. Hypercalciuria in the frequency dysuria syndrome of childhood. J Pediatr 1990;116:103.

Bachur R, Caputo GL. Bacteremia and meningitis among infants with urinary tract infections. Pediatr Emerg Care 1995;11:280.

Baker RB. Dysuria: presenting complaint in labial fusion. Am J Dis Child 1986;140:1100.

Brunham RC, Paavonem J, Stevens CE, et al. Mucopurulent cervicitis: the ignored counterpart in women of urethritis in men. N Engl J Med 1984;311:1.

Demetriou E, Emans SJ, Masland RP. Dysuria in adolescent girls: urinary tract infection or vaginitis? Pediatrics 1982;70:299.

Hoberman A, Wald ER. Urinary tract infections in young febrile children. Pediatr Infect Dis J 1997;16:11.

Hoberman A, Wald ER, Reynolds EA, et al. Is urine culture necessary to rule out urinary tract infections in young febrile children? Pediatr Infect Dis J 1996;15:304.

Kaplan KE, Fleisher GR, Paradise JE, Friedman HM. Social significance of genital herpes simplex in children. Am J Dis Child 1984;138:872.

Komaroff AL. Urinalysis and urine culture in women with dysuria. Ann Intern Med 1986;104:212.

Kunin CM. The quantitative significance of bacteria visualized in the unstained urinary sediment. N Engl J Med 1961;265:589.

Lohr JA, Portilla MG, Geuder TG, et al. Making a presumptive diagnosis of urinary tract infection by using a urinalysis performed in an on-site laboratory. J Pediatr 1993;122:22.

Paradise JE, Campos JM, Friedman HM, et al. Vulvovaginitis in premenarcheal girls: clinical features and diagnostic evaluation. Pediatrics 1982;70:193.

Pryles CV, Eliot CR. Pyuria and bacteriuria in infants and children. Am J Dis Child 1965;110:628.

Shaw KN, McGowan KL, Schwartz JS. Screening for urinary tract infection in infants in the emergency department: which test is best? Pediatrics 1998;101:E6.

Stamm WE, Counts GW, Running KR, et al. Diagnosis of coliform infection in acutely dysuric women. N Engl J Med 1982;307:463.

Stunsfeld JM. The measurement and meaning of pyuria. Arch Dis Child 1962;37:257.

CHAPTER 55

Pain—Earache

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Ear pain is a common symptom of many different conditions in or around the ear (Table 55.1, Table 55.2 and Table 55.3). Preverbal children may present with fussiness, crying, or waking intermittently at night. Children may have difficulty differentiating tinnitus from ear pain. Ear pain may be otogenic or nonotogenic in origin. A complete history and physical examination is necessary, focusing not only on the ear but on adjacent areas and those regions innervated by nerves which also innervate the ear. Areas that might be a source of referred pain include the oral cavity, larynx, pharynx, and cervical spine; rarely, referred pain may be from a site below the clavicles. Patients with accompanying central nervous system symptoms, vertigo, or cranial nerve deficits need more extensive evaluation.

Table 55.1. Causes of Earache

Otogenic	Nonotogenic
Acute otitis media	Dental caries or abscess
Otitis externa	Pharyngitis
Foreign body	Sinusitis
	Cervical adenopathy

Table 55.2. Common Causes of Earache

Hemotympanum secondary
to basal skull fracture
Local spread of acute
otitis media to:
mastoid cavity—subperiosteal abscess with acute mastoid osteitis
vascular structures—lateral sinus thrombosis
intracranial region—meningitis, extradural abscess, subdural empyema,
focal encephalitis, brain abscess
temporal bone—facial nerve paralysis
inner ear—labyrinthitis

Table 55.3. Life-Threatening Causes of Earache

DIFFERENTIAL DIAGNOSIS

Otogenic Causes

Trauma is usually evident by physical examination. Hematomas of the pinna resulting from blunt trauma may occur over the cartilaginous portion or upper half of the ear lobe between the perichondrium and the underlying cartilage. The hematoma may appear as a boggy, purple swelling. Sterile aspiration and a molded pressure dressing are necessary to avoid pressure necrosis of the underlying cartilage and a resulting “boxer's” or “cauliflower” ear.

The most common complication of ear piercing is superimposed infection—usually with *Staphylococcus aureus.* Treatment involves removal of the foreign body, the earring, and administration of an oral antistaphylococcal antibiotic such as dicloxacillin or a topical antibacterial ointment. Nickel earrings may cause earlobe dermatitis. These earrings should be replaced with gold or stainless steel earrings, and topical corticosteroids should be applied.

Preauricular pits are asymptomatic until they become infected. Physical examination reveals a warm, erythematous, tender area anterior to the tragus. Treatment includes oral antibiotics to cover skin flora, occasional incision and drainage, and referral for eventual resection.

Frostbite of the auricle is painful. The ear usually appears pallid secondary to vasoconstriction. With thawing, it becomes hyperemic and edematous, and vesicles may appear. It is treated by rapid rewarming with application of moist gauze or cloths soaked in water 37.8° to 40°C (100° to 104°F). Vesicles should be left intact. Analgesics are given for pain control. No debridement of damaged tissue is done until full demarcation of tissue loss is determined.

Furunculosis is a Gram-positive, usually staphylococcal infection of a hair follicle at the external auditory meatus that causes marked pain and occasional otorrhea. This generally occurs in the cartilaginous part of the canal and may cause cervical adenopathy. Treatment consists of heat application to encourage spontaneous drainage. Topical antistaphylococcal treatment or oral antistaphylococcal antibiotics such as dicloxacillin are rarely indicated. If spontaneous drainage does not occur, incision and drainage may be necessary. Coalescence of furuncles produces a carbuncle or abscess, which requires drainage.

Herpes zoster oticus, or Ramsay Hunt's syndrome, is a painful viral infection characterized by vesicles on the auricle, the external auditory meatus, and occasionally, the tympanic membrane (TM). It is caused by a residual viral infection of the seventh and eighth cranial nerves. Complications include facial paralysis, hearing loss, and vertigo. Other cranial nerves may be affected. Although this normally presents in adults, it may occur at any age. Patients in the early stages may be treated with acyclovir; otherwise, treatment is mainly symptomatic.

Otitis externa (OE) usually presents in warm, humid weather. Because swimming can predispose the canal to this condition, it is often called swimmer's ear. The initial presenting symptom may be pruritus. Scratching the canal may damage the skin and predispose the area to secondary bacterial infection. Ear pain, itching, and discharge are the primary complaints. Examination reveals a tender auricle, a mildly erythematous canal with edema, and foul-smelling, gray–green discharge. *Pseudomonas aeruginosa, S. aureus, Proteus*, and *Streptococcus* are causative organisms. Treatment includes cleaning and acidifying the ear canal, and possible antibiotic coverage for Gram-negative and Gram-positive organisms. An otic suspension combining antibiotics and an acidic medium with hydrocortisone for anti-inflammatory effect is the usual treatment. A wick may be used if the canal is too edematous to allow the drops to enter. If there is any accompanying cellulitis, oral antibiotics should be added. Ear plugs for swimmers and acidifying drops after swimming can be suggested to prevent recurrence. A serious complication of OE is malignant or necrotizing external otitis, which is a fulminant bacterial otitis externa with *P. aeruginosa*. This may extend beyond the limits of the external auditory canal, producing cellulitis, chondritis, osteitis, osteomyelitis, and facial nerve paralysis. These patients require further evaluation with computed tomography (CT), bone scan or gallium scan, otorhinolaryngology (ORL) consultation, and probable admission for combination intravenous antibiotics.

Impacted cerumen in the canal is a common cause of ear pain or discomfort. Ear wax removal is often necessary for evaluating ears. Careful immobilization is essential. Removal under direct visualization through an otoscope is ideal. An ear curette or small cotton swab may be used. Hard cerumen may cause bleeding with removal when adherent to the canal. The wax may be softened with a ceruminolytic agent, then irrigated with warm water. A plastic syringe attached to the plastic tubing end cut from a butterfly needle may be used. A water jet device may also be used, but it must be set on low pressure to prevent damage to the TM. A perforated TM is a contraindication to irrigation. An antibiotic/hydrocortisone otic suspension as prophylaxis to prevent OE after canal irritation or irrigation is sometimes recommended.

Foreign bodies can elicit a painful inflammatory reaction. Physical examination visually confirms a foreign body. Treatment is accomplished by removal with an ear curette, alligator forceps, or irrigation with warm water. Inflamed canals may be treated with topical antibiotic/hydrocortisone otic drops.

Otomycosis is a fungal infection of the external canal. It may be acute or chronic, primary or secondary to bacterial infection and prolonged use of antibiotic drops. *Aspergillus* and *Candida* are the most common pathogens. The primary complaint may be pruritus. On physical examination, the canal contains gray, white, or blackish debris that resembles dirty cotton and has an erythematous and edematous canal wall. Treatment consists of cleaning and acidifying the ear

canal and possibly applying topical antifungal drops such as nystatin.

Dermatoses of the ear can cause ear discomfort as well. Seborrheic dermatitis and psoriasis can affect the external auditory canal and produce scaling or drainage. The primary complaint may be itching. Usually, other areas are affected as well, including the retroauricular region and scalp. Removal of crusts can be helpful. Topical treatment consists of hydrocortisone.

Acute otitis media (AOM) is the most common illness resulting in office visits to physicians who care for children (see also Chapter 84). In 1990, approximately 25 million office visits in the United States were related to AOM. Reasons attributed to children having more infections include eustachian tube dysfunction secondary to its more horizontal position and shorter length in the child compared with the adult, immature or impaired immunology, and day-care center exposure. History often includes recent viral upper respiratory infection followed by deep-seated otalgia, fever, and decreased hearing. The three major bacterial pathogens are *Streptococcus pneumoniae* (pneumococcus), nontypeable *Haemophilus influenzae*, and *Moraxella catarrhalis. Streptococcus progenes, S. aureus,* and Gram-negative enteric bacilli are rare causes. The four major criteria when examining the TM for AOM are position, color, translucency, and mobility. AOM may show a hyperemic, opaque, bulging TM with poor mobility. The drum may be bulging with serous fluid or yellow pus in the middle ear space. The auricle is usually normal. A conductive hearing loss may result from accumulation of fluid in the middle ear and impairment of the drum motion. Treatment is administration of an oral antibiotic for 10 days. Amoxicillin remains the drug of choice because of its safety, low cost, and good taste. If symptoms do not abate in 48 hours, the patient should be examined, with consideration for changing therapy to cover resistant strains of *Haemophilus, Moraxella,* or pneumococcus. Second-line antibiotic choices include second- and third-generation cephalosporins, loracarbef, ampicillin/clavulanate, azithromycin, or intramuscular ceftriaxone. Oral decongestants, antihistamines, and intranasal decongestants have not proven to be effective. Approximately one-third of children have sterile effusions, which are presumed to be viral or unresolved effusions from previously treated infections. Pain may be treated with analgesics, such as acetaminophen or ibuprofen orally. Topical anesthetic drops may be used to provide temporary relief if there is no perforation. For those with tympanic perforation and purulent drainage, a topical antibiotic drop, such as an antibiotic/hydrocortisone otic suspension, is sometimes given to treat the inflammatory reaction in the ear canal. Perforated TMs usually heal spontaneously, but they may lead to more chronic unhealed perforations, cholesteatoma, or tympanosclerosis. Children with significant recurrent infections may require further evaluation for immunologic problems and/or an ORL consultation for myringotomy tube placement.

Although rare, dangerous complications of AOM include local spread to the mastoid cavity, soft-tissue and vascular structures of the neck, intracranial region and temporal bone, or inner ear. On examination for subperiosteal abscess with acute mastoid osteitis, the pinna may be displaced inferiorly and anteriorly with obliteration of the postauricular crease as a result of swelling. The ear drum may appear gray, not bulging or perforated. This most commonly occurs in infants 4 to 12 months old. Treatment includes antimicrobials and, in some cases, drainage. Facial or abducens nerve paralysis is rarely associated with AOM and is treated with antibiotics and consideration of surgical nerve decompression. Because of the location of the middle ear and mastoid air cells adjacent to the posterior and middle cranial fossa and sigmoid venous sinus of the brain, infection may spread to these adjacent structures, leading to intracranial complications such as meningitis, extradural abscess, subdural empyema, focal encephalitis, brain abscess, lateral sinus thrombosis, and otitic hydrocephalus. All patients with facial or abducens nerve palsy, vertigo, or central nervous system signs require further radiographic evaluation, such as CT scan.

Otitis media with effusion (OME) or serous otitis media is an inflammation of the middle ear in which a collection of liquid is present in the middle ear space, which is usually mucoid or serous without signs or symptoms of infection. These represent 25 to 35% of all otitis cases. In 1994, the U.S. Department of Health and Human Services published a Clinical Practice Guideline for treating otitis media with effusion in children 1 to 3 years of age, developed with the cooperation of several expert advisory groups. Although most effusions resolve spontaneously, the length of time for resolution is variable and there is significant concern regarding conductive hearing loss during this critical age of speech and language development. Physical examination should include pneumatic otoscopy to test TM mobility. Tympanometry may be used to confirm an effusion. For healthy children with no craniofacial or neurologic abnormalities or sensory deficits, the effusion may be managed with observation or antibiotic therapy. For OME, bacterial pathogens are identified in about one-third of children. Many studies have shown that antibiotics provide only temporary resolution of fluid. Steroids, antihistamines, and decongestants are not recommended. Chronic effusions may require tympanostomy tube placement for drainage by ORL.

TM perforation may be caused by pressure from fluid behind the membrane or by external trauma. Trauma often occurs with compressive injuries such as a slap with an open hand or injuries from instruments such as cotton-tipped applicators being placed into the ear canal. Pain may be severe immediately after the injury, but it becomes duller with time. Most perforations heal spontaneously. The use of prophylactic antibiotic ear drops is controversial—if chosen, most suggest suspension over solution. Oral antibiotics are reserved for injuries from contaminated objects or those in a location believed to be infectious. If the perforation involves more than 20% of the drum, a referral for possible repair is needed. Acute hearing loss, facial paralysis, and severe vertigo associated with the perforation call for ORL evaluation because of possible ossicular damage or direct injury to the facial nerve or labyrinth.

Bullous myringitis is a painful, usually viral but possibly bacterial or mycoplasma infection of the TM characterized by serous or hemorrhagic vesicles or blebs. Treatment consists of analgesics and possible oral antibiotics.

Hemotympanum secondary to basal skull fracture may be a serious finding on examination of the ear. The TM may appear dark red or purple secondary to the blood behind it. These patients may have other findings consistent with basal skull fractures such as Battle's sign—ecchymoses behind the ear, raccoon eyes—periorbital ecchymoses, or cerebrospinal fluid (CSF) drainage from the nose or ears. A CT scan to determine the extent of the injuries is indicated. Often, no therapy is needed and antibiotic prophylaxis is controversial.

Aerotitis, or acute barotitis, is a special type of AOM caused by middle ear barotrauma. A sudden change in altitude in an airplane or the pressure exerted during deep sea diving can cause eustachian tube closure and produce a severe and

painful pressure change in the middle ear with extravasation of blood into the middle ear space. The drum may appear blue because of bleeding behind it. The patient has severe pain and hearing loss. Physical examination reveals a hemorrhagic TM. The process is self-limited, lasting 2 to 3 days and resolving spontaneously. Treatment consists of analgesics for pain and decongestants to encourage opening of the eustachian tube. Myringotomy is performed only for severe pain or persistent fluid.

Cholesteatomas may be visualized in the middle ear. These cystlike structures, which consist of epithelial cells and cholesterol, may be congenital or acquired, often secondary to previous perforation with residual TM epithelial cells in the middle ear. Enzymes formed within the sac may cause erosion of adjacent bones. Although rare, abnormal growths in the canal or middle ear must be evaluated for possible neoplasm and should be referred for evaluation by ORL.

Nonotogenic Causes

Inflammation, infection, neoplasm, or trauma along the course of any nerves innervating the auricle or the external auditory canal, including cranial nerves V, VII, IX, and X and cervical nerves C2 and C3, can produce pain that the patient may interpret as originating in the region of the ear. Therefore, a full head and neck examination, as well as radiographic examinations may be necessary to disclose the cause of ear pain.

The trigeminal nerve (V) supplies some of the most common areas of referred ear pain, including those of dental origin such as erupting teeth or abscesses and oral mucosal ulcerations from aphthous ulcers or viral stomatitis. Sinusitis, sialadenitis, or lymphadenitis in these regions may also cause pain. Early mumps may present as ear pain before obvious parotid swelling.

Facial nerve (VII) pain may be a precursor of Bell's palsy or herpes zoster oticus.

The glossopharyngeal nerve (IX) supplies the oropharynx, nasopharynx, and posterior third of the tongue. Inflammation of these areas, such as with pharyngitis or tonsillitis, is another common cause of referred earache. Peritonsillar abscess or cellulitis may produce unilateral pain. Earache may also occur after adenotonsillectomy. Nasopharyngeal or oropharyngeal tumors, such as lymphoma or rhabdomyosarcoma, although rare in children, may be associated with ear pain.

The vagus nerve (X) supplies the base of the tongue, larynx, and trachea. Inflammatory or mass lesions in these areas may refer pain to the ear.

Cervical nerves C2 and C3 supply the mastoid and posterior pinna; therefore, ear pain may result from cervical spine injuries, arthritis, or disc disease as well as any generalized neck disorder.

When otologic examination is normal and no pathology is found in the distribution of the cranial or cervical nerves the pain may be psychogenic, especially in a person with anxiety or depression. Also, children may not be able to describe tinnitus and refer to it as pain. Certain drug ingestions such as quinine, quinidine, salicylates, nicotine, ethacrynic acid, and aminoglycosides are possible causes.

EVALUATION AND DECISION

A history is always important in determining the cause of pain or discomfort and should include questions about other ear symptoms, including tinnitus, hearing loss, vertigo, drainage, and itching, as well as systemic symptoms, such as fussiness, crying when lying down, upper respiratory infection, fever, and alteration in oral intake. Children may have difficulty describing pain. Parents may attribute pulling on ears or fussiness to ear pain. Preverbal children being evaluated for suspected ear pain need close evaluation for other causes of their fussiness.

Physical examination should always be complete, especially in children who are not old enough to verbalize that their fussiness is ear pain. Most children do not like to be immobilized for evaluation of the ears and throat, so otoscopy and visualization of the pharynx should be the last part of the examination. Initial examination of the ear includes the auricle and external auditory canal, followed by the middle ear by pneumatic otoscopy. Important aids to the examination include positioning of the child and removal of cerumen. The child may be positioned on the parent's lap or shoulder or placed supine on the examination table. Cerumen removal may be accomplished with the use of a small, cotton-tipped applicator; wire loop or plastic cerumen curette; irrigation with warm water; or application of cerumenolytics before irrigation.

Particularly important components of the examination include the external ear, the auditory canal, the TM, the surrounding structures of the head and neck, and the neurologic evaluation (Fig. 55.1).

FIGURE 55.1. Approach to the diagnosis of earache.

EXTERNAL EAR EXAMINATION

External problems are usually obvious on initial examination. Trauma, erythema, or vesicles on the auricle may be easily seen. Palpation of the area around the ear may reveal swelling or tenderness either from nodes, mastoiditis, parotitis, or preauricular pit infection.

Abnormal Otoscopy

Otogenic causes are readily diagnosed by visualization with otoscopy. Inflammation in the canal or middle ear, foreign bodies, abnormal lesions, perforations, or cholesteatomas may be seen. Evaluation of the TM for signs of infection includes describing position, color, degree of translucency, and mobility. Erythema alone is not sensitive as a predictor of infection because it may occur from increased blood flow with crying. Mobility is evaluated by applying pressure to the rubber bulb attached to the otoscope with a good seal in the canal and looking for inward movement with positive pressure and outward movement with negative pressure. Purulent discharge in the canal may be wicked out with cotton to better visualize the TM for a perforation or foreign body. Any patient with accompanying vertigo, facial nerve palsy, hemotympanum, or central nervous system symptoms requires further evaluation with CT scan.

Normal Otoscopy

A normal otoscopic evaluation should prompt a search for referred pain. Evaluation of the cervical spine, oropharynx, and neck should reveal possible sources of inflammation from shared sensory nerves. Radiographs to evaluate dental sources are usually not required emergently. A history of possible drug ingestions should be obtained. If there is a clinical suspicion of disease in the nasopharynx or larynx, an examination with a nasopharyngoscope may be needed. A patient with a completely normal examination and no other accompanying complaints should be referred back to his or her physician for follow-up before a psychogenic cause is given.

Suggested Readings

Becker W, Naumann HH, Pfalz CR. Ear, Nose, and Throat Diseases: A Pocket Reference. 2nd revised ed. New York: Thieme, 1994.

Bluestone CD, Klein JO. Otitis Media in Infants and Children. 2nd ed. Philadelphia: WB Saunders, 1995.

Clinical Practice Guideline. Otitis Media with Effusion in Young Children, U.S. Department of Health and Human Services, Agency for Health Care Policy and Research Publication No. 94-0622, July 1994.

Jazbi B. Pediatric Otorhinolaryngology: A Review of Ear, Nose, and Throat Problems in Children. New York: Appleton-Century-Crofts, 1980.

Karma PH, Sipila MM, Kataja MJ, Penttila AM. Pneumatic otoscopy and otitis media: the value of different tympanic membrane findings and their combinations. In: Kim DJ, Bluestone CD, Klein JO, et al, eds. Recent Advances in Otitis Media. Proceedings of the Fifth International Symposium, Burlington, Ontario: Decker, 1993:41–45.

Lucente FE, Sobol SM, eds. Essentials of Otolaryngology. 3rd ed. New York: Raven Press, 1993.

Quick Reference Guide for Clinicians: Managing Otitis Media with Effusion in Young Children, U.S. Department of Health and Human Services, Agency for Health Care Policy and Research Publication No. 94-0623, July 1994.

CHAPTER 56

Headache

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Headache is a common complaint of pediatric patients in the emergency department (ED). It is estimated that by the age of 15 years, up to 75% of children have experienced headaches, although most are cared for at home. Parents may seek medical care if the child has a new-onset headache that is particularly painful and does not respond to nonprescription medications, if the child complains of progressively more severe headaches, or if the child has headaches that are recurrent over days, weeks, or months. Children with known migraine headaches are normally seen by a physician only when their standard medication regimen is not effective. Headache as an isolated complaint is a relatively unusual presentation in pediatric patients; it is more often one of a number of symptoms, such as fever, lethargy, sore throat, neck pain, and vomiting.

Like other challenging presentations, headache is seen with regularity and is almost always benign, but in a small subset of patients, it can portend a potentially life-threatening illness. Just as it can be difficult to identify the one child with appendicitis from a succession of patients with viral gastroenteritis, so too the clinician's skill can be tested in distinguishing which child among the many with headache has a serious underlying process. Therefore, the primary responsibility of the emergency physician is to make this important discrimination between “bad” headaches and benign headaches.

Fortunately, this can almost always be done successfully after a thorough history and physical examination. Laboratory and radiographic tests are rarely needed in the acute evaluation of headache in pediatric patients. One notable exception to this rule, however, is brain tumor. Although most serious illnesses (e.g., meningitis, encephalitis, ruptured vascular anomaly) will be apparent after an initial evaluation, the presence of a brain tumor may not be. The history can be subtle, and the examination is commonly unrevealing, often leading to a delay in diagnosis. Therefore, characteristics of headaches caused by a brain tumor are described in detail in this chapter. Above all, the key to propermanagement of these patients is ensuring appropriate follow-up care.

PATHOPHYSIOLOGY

For a headache to occur, there must obviously be some noxious stimulus that affects one or more pain-sensitive structures. Injury to an area that is insensitive to pain, as occurs with certain types of stroke syndrome, may cause significant morbidity but will not manifest as headache. It is therefore useful to consider the sensory innervation of the head and neck. All extracranial structures are sensitive to pain. Thus, processes that affect the sinuses, oropharynx, scalp, neck musculature, and so on, often cause patients to complain of headache. By contrast, some intracranial structures are sensitive to pain and some are not. For example, the brain, ependymal lining, choroid plexus, and much of the dura and pia-arachnoid over the hemispheres are insensitive to pain. Pathologic processes affecting these areas can cause headache, but only by impinging on adjacent pain-sensitive structures. The most pain-sensitive intracranial structures are the proximal portions of the large cerebral arteries at the base of the brain, the venous sinuses, and the large cerebral veins.

Various physiologic mechanisms come into play in causing headache. Painful stimuli can be broadly categorized as resulting from vascular effects, muscle contraction, inflammation, and traction/compression (Table 56.1). Examples of each of these types of headache etiology are described in the following discussion of differential diagnosis. It should be noted that visual problems are an unlikely cause of significant headaches in children. A child with persistent headaches that have previously been attributed to “eye strain” may therefore deserve a more careful evaluation.

I. Vascular	IV. Traction/Compression
A. Febrile illness	A. Increased intracranial pressure
B. Migraine	1. Cerebral edema
C. Systemic hypertension	2. Hydrocephalus
D. Hypoxia	3. Intracranial hemorrhage or hematoma
II. Muscle Contraction	4. Brain abscess
A. Tension	B. Tumor
B. Fatigue	C. Lumbar puncture
III. Inflammation	V. Others
A. Intracranial infections	A. Posttraumatic
1. Meningitis	B. Psychogenic
2. Encephalitis	C. Ocular
B. Dental infections	
C. Sinus infections	

Table 56.1. Pathophysiologic Classification of Headaches

Attempting to predict the neuroanatomic location of a pathologic process using only the site of headache pain described by a child is often difficult. In part, this is attributable to the unpredictable displacement of structures caused by a mass lesion. In addition, the extremely complex relationships of the various nerves involved in pain sensation of the head and neck lead to unexpected patterns of referred pain. Thus, a posterior fossa lesion can cause frontal or orbital pain, and supratentorial lesions may result in pain localized to the occiput or the back of the neck.

DIFFERENTIAL DIAGNOSIS

A comprehensive discussion of the various causes of headache in pediatric patients is beyond the scope of this chapter. The conditions described here are those most likely to be seen in acute- and emergency-care settings (Table 56.2) and those with the greatest potential for imminent morbidity or mortality (Table 56.3).

Vascular	Muscle Contraction
Febrile illness	Tension
Migraine	Fatigue
Inflammatory	Others
Sinus infections	Psychogenic
Dental infections	Ocular
	Posttraumatic

Table 56.2. Common Causes of Headache

Vascular	Traction/Compression
Hypertension	Increased intracranial pressure
Hypoxia	Tumor
Inflammatory	Hemorrhage/hematoma
Meningitis	
Encephalitis	

Table 56.3. Life-Threatening Causes of Headache

Vascular

Headaches associated with vascular changes are believed to be caused primarily by vasodilation, although the exact mechanism has yet to be fully described. One common example of this type of headache is migraine. Migraine headaches are typically chronic and remitting, with a characteristic pattern that is easily described by the patient or parents. Often, a strong family history of migraines is present. For the emergency physician, the main issue with patients with migraines is generally pain control because the diagnosis is already known. However, a significant change in the quality, severity, or timing of headaches in these patients may represent a separate and potentially more serious problem. In such cases, the clinician should not be dissuaded by the existing diagnosis from pursuing an appropriate workup as indicated.

Headaches accompanying fever are also thought to be mediated by vascular effects. Because fever is such a common symptom, this is probably the most common cause of headaches in pediatric patients seen in the ED. Hypertension is

another possible cause of vascular headaches in children. Hypertension causes not only global changes in cerebral vasculature, but also possibly a component of increased intracranial pressure that leads to headache.

Finally, hypoxia is a potent stimulus for cerebral vasodilation and can produce headaches on that basis. Therefore, children with an acute hypoxic insult (e.g., carbon monoxide poisoning) or those with disease states that predispose to hypoxia (e.g., cystic fibrosis, cyanotic heart disease) may present with headaches resulting from an acute process or an exacerbation of an underlying illness.

Muscle Contraction

Headaches can be caused by contraction of the scalp or neck muscles. This is the classic “tension” headache that so often plagues adults. These headaches usually occur when a patient has experienced prolonged periods of mental or emotional stress. This leads to recurrent episodes of muscle tension and/or spasm, which cause muscle soreness. The patient can often localize a specific site where the pain is felt, and the involved muscles may be tender to palpation. Although muscle contraction is an unlikely cause of headache in younger children, the stress of life during adolescence will often produce this type of headache. Onset is typically at the end of the day. A headache that is present on arising in the morning or that awakens a patient from sleep would be an unusual manifestation of muscle contraction.

Inflammation

A wide variety of inflammatory conditions can result in headache, ranging from benign to potentially life-threatening entities. Children with bacterial meningitis or encephalitis may present with headache, although this is usually only one of a constellation of symptoms, such as fever, lethargy, neck pain, confusion, or coma. Headache is unlikely to be the sole complaint in these patients. However, an older child or adolescent who has viral meningitis can present with a severe headache, minimal or mild neck discomfort, and no other signs of significant illness. Fortunately, viral meningitis is a benign process in most cases. Rare causes of inflammatory headache include retroorbital cellulitis or abscess and cerebral abscess. Focal findings on neurologic and/or ocular examination will normally provide clues to these unusual diagnoses.

Headaches can also be caused by inflammatory processes affecting other structures of the head and neck. For example, pediatric patients with pharyngitis caused by group A streptococcus will often complain of headaches. Indeed, the classic presentation for streptococcal pharyngitis in children is sore throat, fever, headache, and abdominal pain. In a child who has difficulty localizing pain, otitis media and otitis externa can also present as headache. Pediatric patients with sinusitis will sometimes complain of facial or periorbital pain, although younger children may simply have a persistent nasal discharge. Dental abscess can be overlooked as a cause of headache-type pain because it is a relatively uncommon finding in children. Therefore, a careful examination of the teeth and gingiva should be performed for all pediatric patients with unexplained headaches. Finally, inflammation of the temporomandibular joint (TMJ syndrome) is a rare cause of unilateral headaches in children. These patients typically report increased pain while chewing and have point tenderness over the mandibular condyle.

Traction/Compression

Headaches can be caused by mass effect from a pathologic lesion that produces traction and/or compression involving pain-sensitive intracranial structures. For the emergency physician, the most important conditions in this category are intracranial hemorrhage and brain tumor. An intracranial hemorrhage produces displacement of surrounding tissues and, in cases of more significant bleeding, increased intracranial pressure. In the pediatric population, this is most often the result of a severe head injury (see Chapter 105). However, in rare cases, a child can have a nontraumatic intracranial hemorrhage from a ruptured vascular anomaly (e.g., an arteriovenous malformation), which leads to bleeding into the brain parenchyma and ventricles. As with other vascular events, this type of hemorrhage is characterized by the abrupt onset of severe pain. By contrast, headaches resulting from a brain tumor typically have a more insidious onset. The child will often complain of progressively worsening headaches for several weeks or even months. Additional symptoms, such as persistent vomiting or gait abnormalities, may also be present. Unfortunately, the physical examination can be normal during the early phase of the illness, and as mentioned previously, this commonly leads to a delay in diagnosis. Other processes that cause headache as a result of traction and compression include pseudotumor cerebri, hydrocephalus, and persistent spinal fluid leak after lumbar puncture.

Psychogenic

Although less common than in adults, headaches of psychogenic origin are seen in children as well. Possible causes include school avoidance behavior, malingering with secondary gain issues, and a true conversion disorder. These patients often have a history of chronic headaches that have been unresponsive to various treatment methods, and they may have undergone a battery of tests without receiving a diagnosis. Parents of these children are usually worried and frustrated. Their reasoning in coming to the ED after an extensive prior workup is often simply “to get another opinion.” For the emergency physician, establishing definitively that a child's persistent headaches are the result of a psychogenic cause is generally impossible. Obviously, this should be considered a diagnosis of exclusion. However, if the history and physical examination do not suggest a more serious cause of headaches, the best management approach is to communicate genuine concern about the patient, attempt to allay some of the parental fears, and ensure appropriate outpatient follow-up.

EVALUATION AND DECISION

As stated previously, the diagnosis for pediatric patients presenting with headache will be evident in all but a small minority of cases after a thorough history and physical examination. Laboratory tests and imaging modalities are rarely needed. Even if a definitive diagnosis cannot be established immediately, the identification of a potentially life-threatening cause of headaches will almost always be possible before the child leaves the ED. Concern about the

possibility of a more serious cause warrants aggressive use of whatever diagnostic or therapeutic interventions are indicated, such as a computed tomography (CT) scan of the head, lumbar puncture, or intravenous antibiotics. Occasionally, a child with a suspected brain tumor will be appropriately discharged from the ED without undergoing any diagnostic tests. Such a disposition assumes that proper follow-up for such patients can be arranged and that magnetic resonance imaging (MRI) of the head will be performed within 24 to 48 hours. An approach to the diagnostic evaluation of a child with headaches is outlined in Figure 56.1.

FIGURE 56.1. Approach to the diagnosis of headache.

Clinical Assessment

History

Before proceeding to specific questions about headache symptoms, the clinician should inquire about the general health of the patient, particularly during the hours leading up to the current presentation. For example, the presence of a high fever, decreased activity, and poor oral intake is suggestive of an serious inflammatory cause such as meningitis. A patient with these same symptoms who also has an abrupt change in mental status may have encephalitis. If a child has been relatively well but has complained of headache associated with persistent nasal discharge (especially if it is purulent), this may be caused by a sinusitis. A child with tooth pain, ear pain, or sore throat may also have a readily apparent reason for headaches.

After general health issues are covered, the clinician should then obtain a complete history regarding the headache itself. As with many illnesses, the cause of headaches can usually be diagnosed with a high degree of accuracy solely based on history; the physical examination is often merely confirmatory. One of the more important points to investigate is the mode of onset. A headache that starts abruptly and causes extreme pain may represent a vascular event such as a ruptured arteriovenous malformation, whereas a headache with a more gradual onset would be inconsistent with this diagnosis. It is sometimes useful to question the patient about the severity of the pain, although in younger children, this history may not always be reliable. A youngster who is smiling and playing with toys may nod “yes” in response to the question “Is the pain very, very bad?” In such cases, the description of the severity of pain must obviously be correlated with the child's clinical appearance. Questions about the quality of pain (e.g., boring, throbbing) are often less useful in children for similar reasons.

The frequency and duration of headaches can also provide valuable clues about the origin of headaches. A child who complains of constant pain for several days without respite (i.e., goes to sleep with it, wakes up with it) will commonly have a tension headache or, perhaps more likely, a psychogenic headache. On the other hand, a patient who presents with headaches that have progressively become more frequent or prolonged may have a more serious underlying condition. Similarly, a child with headaches that have steadily worsened in severity over time warrants careful evaluation, again given the limitations of a child's description of pain. Parents can often help clarify such situations. For example, they may report that the child previously complained of headaches while continuing to play, but now the headaches cause the child to stop what he or she is doing, lie down, and hold his or her head crying. Such a progression of symptoms over several days or weeks will in most cases be a sufficient indication for performing a CT scan or MRI of the head before the child leaves the ED.

The time and circumstances of occurrence are also important historical points to ascertain. For example, headaches that are present when a child arises each morning or that awaken a child at night should raise suspicion about a possible brain tumor. In contrast, headaches that occur only later in the day are typically related to stress and result from muscle contraction. In addition, any precipitating events that consistently cause or exacerbate a headache should be identified. If an older child has a headache that is significantly worse when leaning down (e.g., to pick up something off the floor), this is most likely to be caused by sinusitis, although in rare cases, this history may be present in a child with a brain tumor.

Any relevant details about the patient's medical history and family history should routinely be obtained. As mentioned previously, children with cystic fibrosis or congenital heart disease may have headaches caused by worsening hypoxia. Likewise, a child with renal disease may develop headaches in response to an elevated blood pressure. In general, the most important question regarding family history is whether anyone has had migraine headaches. It should be remembered, however, that many people use the term *migraine* rather broadly to refer to any type of severe headache. Therefore, the clinician may find it useful to describe typical migraine symptoms before questioning parents about this aspect of the history. Abrupt onset of headache in several family members may suggest carbon monoxide poisoning.

Before leaving this subject, it is worth reemphasizing the importance of a thorough history in developing an appropriate clinical suspicion of a possible brain tumor. A variable period of time exists when a child with a brain tumor will

experience headaches before any abnormal physical findings are apparent. Making a presumptive diagnosis of brain tumor as a likely cause of headaches during this early stage of the illness will therefore depend entirely on the history. In their classic article, Honig and Charney described several historical points that are characteristic of children with brain tumor headaches (Table 56.4). Although no single pathognomonic response on history unerringly establishes the diagnosis, eliciting one or more of these findings should certainly raise the level of concern that a child's headaches may be caused by a brain tumor.

Nocturnal headache or pain on arising in the morning
Worsening over time (severity, frequency, and/or duration)
Associated with vomiting (although may also occur with migraine), especially if vomiting gets progressively worse
Behavioral changes
Polydipsia/polyuria (craniopharyngioma)
History of probable neurologic deficits (e.g., ataxia/incoordination/"clumsiness," blurred vision, or diplopia)

Reprinted with permission from Honig PJ, Charney EB. Children with brain tumor headaches: distinguishing features. Am J Dis Child 1982;136:121–141.

Table 56.4. Characteristic Historical Findings of Brain Tumor Headaches in Children

Physical Examination

Finding an abnormality on the physical examination of a child with headaches will be a relatively rare event. Nevertheless, a thorough head-to-toe examination should be performed in every case because identification of even a subtle finding (e.g., early papilledema) can significantly alter the course of evaluation and treatment. As with all children seen in the ED, the first step of the examination is to assess the patient's appearance. Does the child look sick or well? Does the child appear to be in severe pain, mild pain, or no pain at all? This initial evaluation represents the first important branch point in the decision algorithm for patients with headache (Fig. 56.1). Thus, a child who appears ill may have a more serious underlying condition, such as meningitis or an intracranial hemorrhage, requiring a rapid examination and prompt initiation of treatment.

The vital signs should also be assessed, particularly the temperature and blood pressure. Although omitting the blood pressure reading for younger children is a tendency, this is never acceptable for a patient who has headaches. Significant hypertension, usually resulting from undiagnosed renal disease, is a rare but potentially dangerous cause of headaches that can affect children of any age. Consequently, if a blood pressure is not taken initially by triage personnel, this must be performed as part of the child's evaluation in the ED. Measuring basic growth parameters for a pediatric patient with headaches can also provide valuable information. Macrocephaly may be the result of hydrocephalus, and short stature can be associated with a craniopharyngioma that causes impaired pituitary function.

The head and neck examination will sometimes reveal an obvious source of headache in a child. The scalp should be examined for evidence of head injury. Even when no history of trauma exists, the child may have had an unwitnessed event, or the history may be intentionally misleading with a victim of child abuse. Tenderness of the scalp or neck muscles is often present with headaches resulting from stress and muscle contraction. The eyes should be examined to detect any abnormalities in pupillary responses or extraocular movements. A sluggish pupil may be caused by an expanding mass lesion that causes compression of the third cranial nerve, and pain with extraocular movements may be elicited with a retroorbital cellulitis or abscess. The eyegrounds should also be carefully examined for signs of papilledema. The clinician may find an otitis media or otitis externa when the ears are examined. Streptococcal pharyngitis as a cause of headaches may be evident as swelling, erythema, and exudates of the tonsillar pillars. Facial tenderness and erythema are sometimes seen in children with maxillary or frontal sinusitis. The teeth and gingiva should be examined for evidence of inflammation or abscess. Nuchal rigidity can be a sign of meningitis, intracranial hemorrhage, or in rare cases, brain tumor. If a child has a ventricular shunt, assessment of shunt function should be performed when appropriate (see Procedures).

Examining the skin is also important for the child with headaches. Because the skin and central nervous system have a common embryologic origin, cutaneous lesions are sometimes seen with neurologic disorders. For example, a child with numerous hyperpigmented spots scattered over the body (café-au-lait spots) most likely has neurofibromatosis. Similarly, children with tuberous sclerosis will almost always have several small, hypopigmented spots (ash leaf spots) that are more apparent when viewed under a Wood's ultraviolet lamp.

Every child with a complaint of headaches obviously needs a complete neurologic examination. Any new focal finding suggests the presence of a focal lesion, such as a tumor or hemorrhage. Some children with migraine headaches develop focal neurologic abnormalities as part of their migraine syndrome (e.g., ophthalmoplegia), but parents can normally confirm that this is not a new problem. As mentioned previously, the mental status of a child with headaches must always be carefully assessed. A diminished level of consciousness may be the result of encephalitis, a large intracranial hemorrhage, or significantly elevated intracranial pressure. To the extent that the child can cooperate, cranial nerve function should also be evaluated. Cranial nerve abnormalities may result from an elevated intracranial pressure or direct compression by a mass lesion. Sensory and motor function should be examined, although here again the ability of a younger patient to cooperate may be limited. A reasonable evaluation can be accomplished by observing the child's gait while walking and/or running and by assessing the child's dexterity in performing age-appropriate activities, such as transferring a toy from hand to hand and tying shoe laces. Any evidence of gait abnormalities or deficits in fine motor

coordination warrants further investigation.

Laboratory and Radiographic Testing

By far, most children presenting in an acute-care setting with headache as the chief complaint will not require any laboratory tests. Most will have minor problems, such as otitis media, viral illness with low-grade fevers, or tension headaches. Laboratory testing rarely provides any useful information in such cases. Certainly, the child with a possible serious infectious process causing headaches can require a variety of tests, including a complete blood count, blood cultures, and lumbar puncture. Yet these patients are more likely to have other symptoms such as high fever and lethargy, rather than headache, as the primary complaint. Should a lumbar puncture be necessary for a child, it is important to remember that an emergent head CT scan should be obtained first when an intracranial space-occupying lesion is in the differential diagnosis. Failure to do this could theoretically lead to a herniation syndrome resulting from differential in cerebrospinal fluid (CSF) pressure. Serum electrolytes, blood urea nitrogen, creatinine, and a urinalysis should be obtained for any child with headaches who is found to have an elevated blood pressure. The patient with a ventricular shunt who has fever and headaches will likely require a shunt tap by a neurosurgical consultant. Finally, a child with a suspected subarachnoid hemorrhage should undergo a lumbar puncture if the head CT scan is negative. This is necessary because a small hemorrhage may not be detected by CT, and in such cases, blood in the CSF is the only diagnostic finding. However, this is an uncommon situation in the pediatric population.

As with laboratory testing, few children with headaches who come to the ED will require an emergent imaging study. In general, plain radiographs of the skull are of little or no value for these patients. A child with a ventricular shunt may require a shunt series, but this includes radiographs of the entire course of the shunt and not simply skull radiographs. Likewise, sinus radiographs are rarely indicated in pediatric patients because the diagnosis almost always is made on clinical grounds. Occasionally, a child with multiple episodes of an apparent sinus infection will require a CT scan of the sinuses, but this is normally done as an outpatient.

The two modalities currently in use that provide the best radiographic information about intracranial abnormalities are CT and MRI. Each has certain advantages and disadvantages. At present, CT is available on a more emergent basis, making it the test of choice to evaluate patients at risk for problems such as intracranial hemorrhage, cerebral edema, and herniation syndrome. CT is especially useful for patients with head trauma. However, CT does not offer the quality of image resolution provided by MRI. Smaller lesions, particularly those of the posterior fossa and brainstem, are more reliably detected by MRI. This is true even when the CT scan is performed using contrast material. Consequently, MRI is superior for children suspected of having a brain tumor who have a normal neurologic examination and no signs of elevated intracranial pressure. If these patients undergo a head CT scan in the ED, they will likely also require an outpatient MRI. Such duplication of testing is costly and usually unwarranted. Lack of availability is the primary drawback of MRI. In most institutions, an MRI is difficult if not impossible to obtain on an emergent basis, particularly after hours. As discussed in the following, the emergency physician must take these as well as other factors into account in determining which, if any, imaging modality is indicated for a child with headaches.

Treatment and Disposition

Patients with headaches caused by a potentially life-threatening process (e.g., meningitis, encephalitis, ruptured vascular anomaly) require specific treatment approaches discussed elsewhere in this textbook. Children with headaches that are presumptively diagnosed as benign can almost always be successfully treated with acetaminophen or ibuprofen. The various options available for treating pediatric migraine patients are described in Chapter 83.

Although most children complaining of headache can be safely discharged from the ED with an appropriate follow-up plan, some will require admission to the hospital for further evaluation and treatment. For example, a child with headaches who is found to be hypertensive must be admitted both for management of the blood pressure and investigation of the underlying cause. Patients with migraine who have intractable headache pain may also warrant admission to receive a more effective analgesic regimen. The child with a ventricular shunt who has severe headaches will usually require a shunt series, a CT scan of the head, and neurosurgical evaluation. If neurosurgical consultation is not immediately available, the patient should be transported to an appropriate receiving facility.

A potentially confusing issue that the emergency physician will inevitably face is how to properly manage a child who is suspected of having a brain tumor. Should all of these patients have a brain imaging study in the ED? As discussed previously, the resolution of even a contrast-enhanced head CT scan is inferior to MRI for detecting certain types of tumors. Also, a small but finite risk is associated with the administration of contrast material. On the other hand, MRI is generally far more difficult to obtain on an emergent basis than CT. What then is the appropriate diagnostic approach?

Without question, a child with headaches who has papilledema or a focal neurologic finding must have an immediate head CT scan, usually enhanced with contrast material as deemed necessary by the radiologist. Similarly, a patient who has headaches associated with visual changes, frequent vomiting, or other signs of elevated intracranial pressure requires an emergent head CT scan. Because mass lesions that cause such changes are usually larger and more easily detectable, the potential reduction in image resolution is more than offset by the greater speed with which a head CT scan can be performed. Time, not image quality, is the critical factor for these patients.

But what about the child with a suspicious history (e.g., increasing frequency or duration of pain, headaches that awaken the child from sleep or occur every morning) who has a normal neurologic examination and no signs of elevated intracranial pressure? In most cases, such patients can be safely discharged from the ED with an outpatient MRI scheduled within 24 to 48 hours. A delay in diagnosis of 1 or 2 days is usually acceptable if this allows the appropriate diagnostic study to be performed. Obviously, parents must be clearly instructed that any sign of deterioration, such as mental status changes or vomiting, requires that the child be immediately returned to the ED for a reevaluation.

Suggested Readings

Burton LJ, Quinn B, Pratt-Cheney JL, et al. Headache etiology in a pediatric emergency department. Pediatr Emerg Care 1997;13:1–4.

Chu ML, Shinnar S. Headaches in children younger than seven years of age. Arch Neurol 1992;49:79–82.

Ferrari P, Incorpora G, Cocuzza M, et al. Multicenter study of childhood headache. Childs Nerv Syst 1994;10:455–457.

Honig PJ, Charney EB. Children with brain tumor headaches: distinguishing features. Am J Dis Child 1982;136:121–141.

Rothner AD. Headaches in children and adolescents. Va Med Q 1996;123:90–93.

Shinnar MD, D'Souza BJ. The diagnosis and management of headaches in childhood. Pediatr Clin North Am 1981;29:79–94.

CHAPTER 57

Joint Pain

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Arthritis and arthralgia are common reasons for children to seek care in the emergency department. Arthritis is joint inflammation marked by swelling, warmth, and limitation of motion, whereas arthralgia is simply pain without inflammation of the joint space. The diagnostic approach is challenging because the differential diagnosis is lengthy (Table 57.1), clinical and laboratory findings are rarely specific for a particular disease, and the disease pattern for many of the conditions listed is often greatly variable. Among the most common causes of joint pain in children are bacterial infections, trauma, and postinfectious conditions (Table 57.2). Although rare, several life-threatening causes (Table 57.3), including acute rheumatic fever and leukemia, must be considered. This chapter serves as a guide to the approach to the child with arthritis or arthralgia, with an emphasis on historical points and physical examination findings that can serve to narrow the diagnostic possibilities.

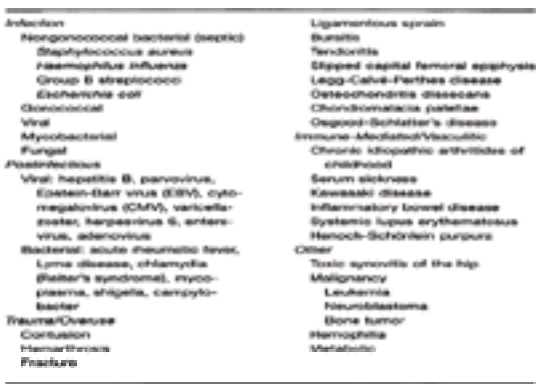

Table 57.1. Joint Pain—Differential Diagnosis

Nongonococcal bacterial (septic)	Postinfectious (reactive)
Gonococcal	Traumatic hemarthrosis
Lyme disease	Serum sickness
Toxic synovitis of the hip	Henoch-Schönlein purpura

Table 57.2. Common Causes of Joint Pain

Acute rheumatic fever
Kawasaki disease
Malignancy
Leukemia
Neuroblastoma
Bone tumor

Table 57.3. Life-Threatening Causes of Joint Pain

DIFFERENTIAL DIAGNOSIS

The possible causes of joint complaints in children are extensive (Table 57.1), but infectious and traumatic causes are the most common (Table 57.2). Children between 6 to 24 months of age have the highest incidence of nongonococcal bacterial (septic) arthritis, and boys are affected twice as often as girls. Septic arthritis results primarily from the hematogenous dissemination of an organism into the joint or the bony metaphysis. The diagnosis of septic arthritis of the hip should not be delayed because pressure in the joint space will compromise the vascular supply to the femoral head, leading to necrosis.

Among adolescents, disseminated gonococcal infection with polyarthritis occurs three to five times more often in girls, often during menstruation. Of note, few patients report lower abdominal pain or vaginal discharge concurrently and cultures of blood and synovial fluid are typically negative. The highest yield for establishing the diagnosis is by Gram stain of the skin lesions showing Gram-negative intracellular diplococci or by culturing the cervix, rectum, or throat.

In the absence of a clear history of a tick bite, Lyme disease is a challenging clinical diagnosis because only about 40 to 70% of children have the characteristic erythema migrans rash, constitutional symptoms may be mild, and serologic tests have a high incidence of false positivity. The diagnosis relies on the specific Western Blot confirmation of a positive serology.

Toxic synovitis is a poorly understood inflammation of the hip joint afflicting children 3 to 6 years of age. The diagnosis is typically made on clinical grounds, and this self-limited disease does not result in joint destruction.

Reactive, or postinfectious, arthritis is probably more common than septic arthritis. Arthritis secondary to various enteric infections is not rare in children and parvovirus B19 is also a common cause of acute arthritis in adolescents and young adults. *Chlamydia trachomatis* infection of the genitourinary tract should be considered in any sexually active adolescent with new onset arthritis. With postinfectious arthritis, no clear evidence supports that antimicrobial treatment modifies the disease course.

Traumatic injuries to a joint may cause periarticular swelling or an effusion indicative of a hemarthrosis. A host of other conditions may come to light in the setting of minor trauma but may be only indirectly related to that trauma.

Less common is chronic arthritis with an incidence of 5 to 10 per 100,000 children less than age 16 years. These diseases are characterized by arthritis persistent for at least 6 weeks in the absence of a defined diagnosis. For purposes of this discussion, we will group these disorders under the heading chronic idiopathic arthritides of childhood (CIAC). CIAC is much less benign than previously believed, with about 40% of children from all disease subtypes continuing to have active joint inflammation, and 20% of children having severe functional limitations after 10 years or more of follow-up. There may be a relatively limited window of opportunity in the first 2 years of disease to limit joint disease. Thus, it is imperative that a primary care physician suspect the diagnosis and make the appropriate referral.

EVALUATION AND DECISION

Figure 57.1 depicts an algorithm for the diagnostic approach to the child with joint pain. In evaluating such children, one should inquire about a history of trauma, fever, the specific joint(s) involved, duration, rash, tick bites, sexual risk factors, intravenous drug use, and recent illnesses. Radiographs of the affected joint are particularly useful in the setting of trauma or acute monarthritis without an obvious cause. A complete blood count and erythrocyte sedimentation rate are generally indicated for children with joint pain in the absence of trauma. For the febrile child with monarthritis and a joint effusion, an arthrocentesis will guide the evaluation. Finally, ultrasound is superior to plain radiographs in the detection of a hip effusion.

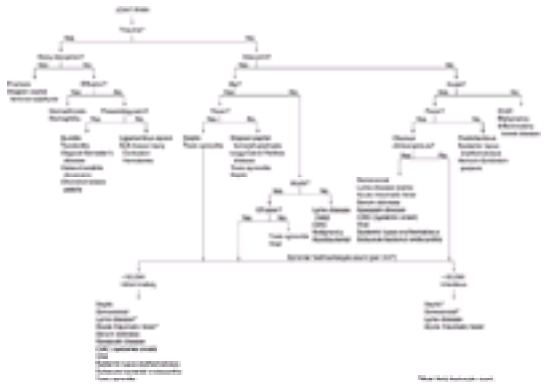

FIGURE 57.1. A diagnostic approach to joint pain. *CIAC,* chronic idiopathic arthritides of childhood.

A key initial point in the history is whether trauma preceded the pain. Because it is easy to be led astray by parents trying to recall what traumatic event could have led to the child's symptoms, one should remember that if the mechanism was not severe enough to immediately prevent the child from continuing an activity, it is unlikely to be the cause of a significantly swollen and painful joint.

On the other hand, if there was a definite traumatic event preceding the onset of symptoms, particularly in the absence of fever, one can proceed with that aspect of the evaluation.

A radiograph will detect fractures or a slipped capital femoral epiphysis. Radiographs aid in determining whether associated swelling is caused by a joint effusion or is simply soft-tissue swelling outside of the joint, a distinction that is often difficult to make on physical examination alone. In the setting of acute trauma and in the absence of fever, an effusion is indicative of a hemarthrosis and is rarely a diagnostic or therapeutic indication for performing an arthrocentesis because the patient will typically experience only temporary relief followed by a rapid reaccumulation of blood.

In the absence of an effusion, inquiries about the duration of symptoms should be made. Children with conditions such as bursitis, tendonitis, and Osgood-Schlatter's disease typically have chronic, low-grade pain and may inadvertently come to medical attention after minor trauma. New-onset periarticular swelling and pain immediately after acute trauma suggests ligamentous or other soft-tissue injury.

In the absence of trauma, monarthritis of the hip is uniquely problematic. In particular, bacterial infections of this and other joints are the most serious because of the rapidity with which destruction of cartilage can occur. The most important determinant of the outcome of a septic arthritis is the length of delay between the onset of infection and the institution of therapy. Unlike most other causes of fever and arthritis, in more than 90% of affected children, septic arthritis involves only a single joint, usually of the lower extremity; neonates, however, may have polyarthritis. Because just 60 to 70% of children with septic arthritis are febrile at presentation, the absence of fever does not preclude the diagnosis. However, if the child allows full range of motion, the diagnosis is unlikely.

A child with acute onset of monarthritis of the hip or any other large joint, defined by the presence of an effusion and marked by severely restricted range of motion, with or without fever, needs an arthrocentesis. The synovial fluid should be analyzed for glucose and protein, cell count and differential, Gram stain, and culture. Most patients with septic joints will have a decreased glucose level and greater than 50,000 white blood cells per cubic millimeter with a neutrophil predominance, but in one study, one-third had counts of less than 25,000/m ^{3}. In addition, Gram stain is negative in about 25% of patients, synovial fluid cultures are negative in about 25%, blood cultures are negative in about 50%, and 20 to 30% with septic arthritis have a peripheral white blood cell count of less than 10,000. The erythrocyte sedimentation rate is almost always elevated, with a median value of 50 mm in one case series. Thus, laboratory evaluation is used simply to confirm the clinical suspicion but should not be relied on to make the diagnosis.

In contrast to septic arthritis, children with toxic synovitis of the hip typically appear well; often present with a limp; may have only mildly elevated temperatures; allow almost complete range of motion of the affected joint; and the complete blood counts, erythrocyte sedimentation rates, and radiographs are typically normal. Usually, the physician can make a clinical diagnosis of toxic synovitis, but in a small subset, septic arthritis cannot be ruled out before the completion of an arthrocentesis. The disease is self-limited, usually lasting less than 1 week.

A slipped capital femoral epiphysis must not be missed. About half of patients will provide a history of trauma. This condition is most often seen in obese adolescent boys. Most children have chronic pain, although some have an acute limp with hip or knee pain and restricted range of motion. Plain radiographs (frog-leg view of the hip) showing a widened epiphysis and caudal displacement of the femoral head establish the diagnosis.

Legg-Calvé-Perthes disease, a condition of uncertain cause, occurs overwhelmingly in boys, with an onset between 4 and 8 years of age. The pain, which may be localized to the hip or referred to the thigh or knee, is insidious in onset. The aseptic necrosis of the femoral head will be manifest on plain radiographs as a small, osteopenic femoral head with a widened joint space.

Historical and physical examination findings help narrow the choices among the many causes of fever and polyarthritis (Table 57.4). The ill-appearing adolescent with migratory arthritis, tenosynovitis involving the extensor tendons of the wrist or ankle, and scattered crops of vesiculopustules on an erythematous base should be strongly suspected for gonococcal arthritis. Joint involvement with Lyme disease has two distinct patterns. In early disseminated disease, the child may develop episodic migratory polyarthritis, affecting mainly large joints. Each episode lasts a few days, helping distinguish it from idiopathic chronic arthritis. Two weeks to 2 years (mean 6 months) after the tick bite, 6% develop an intermittent monarthritis, typically of the knee, ankle, or wrist. The joint is significantly swollen but only mildly painful, and patients are usually afebrile at this stage. Painful migratory joint pain involving five or more joints in a school-age child with recent evidence of a group A streptococcal infection should raise the concern for acute rheumatic fever. Evidence of carditis, erythema marginatum, subcutaneous nodules, and positive serology for antistreptococcal antibodies support the diagnosis. The presence of diffuse urticaria and angioedema accompanying arthralgia or arthritis, especially 3 to 10 days after initiation of an antibiotic, helps distinguish serum sickness from other causes of polyarthritis and fever. Kawasaki disease is characterized by high and persistent fever, conjunctival injection without exudate, mouth and lip swelling and cracking, swelling and erythema of the hands and feet, a nonspecific rash, and lymphadenopathy. Ten days or so after the disease onset, there may be desquamation of the fingers and toes, myocardial dysfunction, and thrombocytosis; about 30% of patients will also develop arthritis or arthralgia. Daily temperature spikes exceeding 40°C (104°F), especially if accompanied by a transient pink rash, suggests one of the CIAC (formerly called Still's disease). A common viral-related arthritis is that caused by hepatitis B infection. The arthritis precedes the symptoms of hepatitis and resolves when the jaundice appears. During the 1- to 3-week prodromal phase, polyarthritis may be accompanied by moderate fever and sometimes by an urticarial or maculopapular rash. Parvovirus B19 causes a similar clinical syndrome in young women with a sudden onset of symmetric, self-limited polyarthritis, particularly in the hands. With subacute bacterial endocarditis, musculoskeletal symptoms are variable, ranging from asymptomatic joint effusions to frank arthritis of up to three joints. Preexisting congenital heart disease, a prolonged fever, a new murmur, and splinter hemorrhages may all be clues to the diagnosis of this rare entity in children.

Usually Febrile at Presentation	*May or May Not Be Febrile at Presentation*
Nongonococcal bacterial (septic)	Leukemia
Gonococcal	Mycobacterial
Acute rheumatic fever	Postinfectious (reactive)
Chronic idiopathic arthritides of childhood (systemic onset)	Lyme disease
Subacute bacterial endocarditis	Systemic lupus erythematosus
Serum sickness	Inflammatory bowel disease
Kawasaki disease	

Table 57.4. Fever and Joint Pain

A joint aspiration is rarely necessary to establish a diagnosis for a child with polyarthritis and fever or to rule out a septic process in this setting. If an arthrocentesis is obtained, typically the synovial fluid will be sterile and the leukocyte count will characterize the process as inflammatory (Fig. 57.1).

Postinfectious arthritis is one of the more common causes of acute polyarthritis without fever. One to two weeks after a bout of enteritis or urogenital infection (Reiter's syndrome), a child may develop an asymmetric postinfectious (reactive) polyarthritis, predominantly involving large joints of the lower extremities. The severity of the antecedent illness has little correlation with the arthritis, and the intensity of synovitis and fever is mild or absent at this stage.

As with many of the diseases discussed to this point, systemic lupus erythematosus (SLE) has a variable clinical presentation with regard to musculoskeletal involvement. In fact, no two patients have an identical pattern of immune complex formation or clinical disease expression. A symmetric polyarthritis involving peripheral joints of the hands or feet may be seen. However, small effusions of the knee are also common with active disease and the arthritis may be intermittent or migratory as well. Patients with this type of arthritis are usually afebrile, yet high fever may be a prominent finding. Although arthritis is 1 of the 11 diagnostic criteria for SLE established by the American College of Rheumatology, it is uncommon for SLE to present with isolated arthritis. Arthritis of the small joints, a positive test for antinuclear antibodies (ANA), and abnormalities of the skin, kidneys, or central nervous system should raise the clinician's suspicion for SLE.

Henoch-Schönlein purpura (HSP) is rarely a diagnostic challenge thanks to the presence of petechiae and purpura in the characteristic below-the-waist distribution. Classically, children will also have polyarthritis, colicky abdominal pain, and nephritis. As with the rash, periarticular swelling usually involves joints below the waist.

The CIAC are marked by their duration, typically 6 months or longer. That they appear at several points in the diagnostic algorithm reflects their diversity. Some children will have systemic distribution of symptoms, whereas others will have monoarticular or oligoarticular disease, and fever or rash may or may not be present. Tests for rheumatoid factor and ANA in children are unhelpful. When performed as a screen, rheumatoid factor tests are rarely positive and, when positive, are as likely to be in children with other diseases as in children with chronic arthritis.

In the absence of fever, chronic pain of one or more joints may also indicate malignancy. Specifically, leukemia or neuroblastoma can both present with true joint swelling, as can bony tumors. Pallor, weight loss, and other constitutional complaints, as well as anemia or cytopenias, would support this diagnosis.

A large joint oligoarthritis occurs as an extraintestinal complication of inflammatory bowel disease in about one-third of children, usually during times of active disease. Clues to the diagnosis include abdominal pain, hematochezia, anemia, and weight loss.

In summary, this review of joint pain in children should serve as a guide to the diagnostic evaluation. The clinician must choose from many different causes, each with variable and nonspecific characteristics. In addition, laboratory studies are rarely specific for a particular disease. However, by asking the appropriate questions, performing a careful physical examination, and selectively obtaining adjunct studies, the clinician can follow the correct diagnostic path.

Suggested Readings

Athreya BH. Vasculitis in children. Pediatr Clin North Am 1995;42: 1239–1261.

Baker DG, Schumacher HR. Acute monarthritis. N Engl J Med 1993;329:1013–1020.

da Silva NA, de Faria Pereira BA. Acute rheumatic fever: still a challenge. Rheum Dis Clin North Am 1997;23:545–568.

Fink CW, Fernandez-Vina M, Stastny P. Clinical and genetic evidence that juvenile arthritis is not a single disease. Pediatr Clin North Am 1995;42:1155–1169.

Lehman TJA. A practical guide to systemic lupus erythematosus. Pediatr Clin North Am 1995;42:1223–1238.

Malleson PN. Management of childhood arthritis. Part I: acute arthritis. Rheumatology 1997;76:460–462.

Malleson PN. Management of childhood arthritis. Part II: chronic arthritis. Rheumatology 1997;76:541–544.

Pinals RS. Polyarthritis and fever. N Engl J Med 1994;330:769–774.

Primm PA. Initial approach to the child who presents with infections of the bones and joints. Pediatr Infect Dis 1996;7:27–34.

Schumacher HR. Arthritis of recent onset. Arthritis 1995;97:52–63.

Shulman ST, DeInocencio J, Hirsch R. Kawasaki disease. Pediatr Clin North Am 1995;42:1205–1222.

Watts RA, Scott DGI. Rashes and vasculitis. Br Med J 1995;310:1128–1132.

CHAPTER 58

Pain—Scrotal

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Acute scrotal swelling or pain in a child should be considered a potential surgical emergency. Although some causes of acute scrotal swelling may be benign and require no more than observation and reassurance to the patient and parent, other causes may lead to the rapid loss of a testis if diagnosis and treatment are delayed. The patient with such a complaint should be evaluated promptly. Many diagnoses in cases of scrotal pain are most reliably made clinically, differentiating by age, historical features relating to the evolution of pain and associated symptoms, and physical examination findings.

PATHOPHYSIOLOGY

The anatomic structures contained in the scrotum include the testes; the epididymis; appendages of the testis; and the nerve, vascular, and lymphatic structures that constitute the spermatic cord and traverse the inguinal canal into the scrotum (Fig. 58.1). The anatomy of the testicle, its related structures, and the layers of tissue that surround each testicle in the scrotum may each relate to the pathology seen in this area. The descent of the testis through the inguinal canal from the abdomen until its eventual position in the scrotum also contributes to the risk of pathology in the scrotum and associated groin area. Pathophysiologic causes of acute conditions of the scrotum include ischemia, inflammation, trauma, and tumor. Because these processes often alter blood flow to structures within the scrotum, appropriate imaging modalities when correlated with clinical history and examination are often useful in coming to the correct diagnosis.

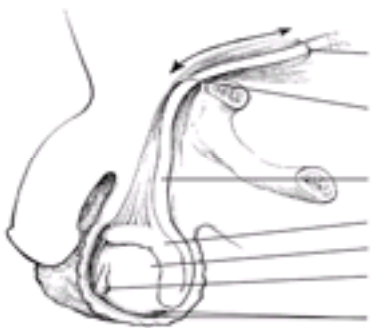

FIGURE 58.1. Anatomy of the scrotal contents.

DIFFERENTIAL DIAGNOSIS

Table 58.1 lists the principal causes of acute scrotal swelling and Table 58.2 provides the most common diagnoses by age.

Painful Scrotal Swelling	*Painless Scrotal Swelling*
Torsion of testis	Hydrocele
Torsion of appendage of testis	Hernia
Trauma—hematocele, hematoma, epididymitis, testicular rupture	Varicocele
	Spermatocele
Epididymitis	Idiopathic scrotal edema
Orchitis	Henoch-Schönlein purpura
Hernia—incarcerated	Kawasaki disease
Tumor—acute hemorrhage	Testis tumor
	Antenatal torsion of the testis

Table 58.1. Causes of Acute Scrotal Swelling

Infancy	*Adolescence*
Hydrocele	Epididymitis
Hernia	Torsion of the appendix testes
Childhood	Torsion of the testes
Hernia	Trauma
Torsion of the appendix testes	
Torsion of the testes	
Trauma	

Table 58.2. Common Causes of Acute Scrotal Swelling

Causes of Painful Scrotal Swelling

Torsion of the Testis

Testicular torsion is the most significant condition causing acute scrotal pain and represents a true surgical emergency. It is the most common cause of acute painful scrotal swelling in children, accounting for approximately 30% of cases of acute scrotal pain. Testicular torsion is more common in the newborn period and during the early stages of puberty. Approximately two-thirds of cases of intravaginal torsion occur in children between the ages of 12 and 18 years, overlapping the peak incidence of appendage torsion.

Torsion results from an inadequate fixation of the testis to the intrascrotal subcutaneous tissue (Fig. 58.2), resulting in the so-called “bell-clapper” deformity. The testis, which hangs more freely within the tunica vaginalis in this deformity, may rotate, producing torsion of the spermatic cord, venous engorgement of the testis, and subsequent arterial infarction (Fig. 58.3).

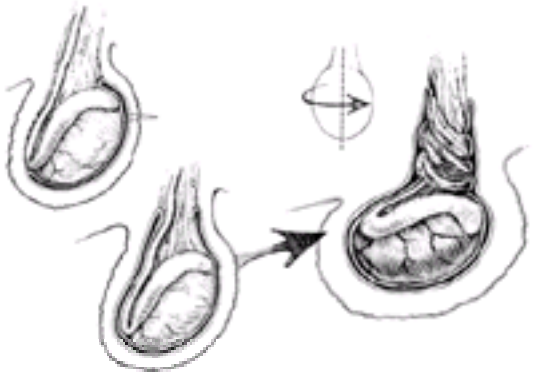

FIGURE 58.2. Torsion testis abnormality of testicular fixation—bell-clapper deformity—permits torsion of spermatic vessels with subsequent infarction of the gonad. *Epid.*, epididymis.

FIGURE 58.3. Torsion of testis. **A.** Swollen, diffusely tender testis high in the scrotum (from twisting of cord structures). **B.** Surgically exposed testis showing torsion of cord structures. Testis was infarcted and was removed.

The sudden onset of severe scrotal pain and tenderness, often with radiation to the abdomen, and associated nausea and vomiting is typical. Often, these episodes have their onset in the early morning. At other times, they may be associated with sports activity or mild testicular trauma that may be perceived by the patient as cause of the pain. A history of trauma often is misleading in patients with testicular torsion. The patient may recall prior episodes of similar pain that resolved spontaneously, suggesting intermittent torsion and spontaneous detorsion.

With torsion of the testis, typically the testis is acutely swollen and diffusely tender and usually lies higher (“horizontal or transverse lie”) in the scrotum than its contralateral mate. Because the pain may be referred to the abdomen, it is essential that the genitalia are examined carefully in every child who complains of abdominal pain. There may be overlying erythema of the skin of the scrotum. The cremasteric reflex is usually absent with testicular torsion but may be present in early or incomplete torsion. Urinalysis is usually negative.

Time is important in establishing the diagnosis of torsion of the testis. If a testis has been twisted sufficiently to fully obstruct its blood supply for more than 6 to 12 hours, surgical detorsion is unlikely to salvage the gonad. It is impossible to determine clinically, however, whether the torsion has been partial or total. Therefore, it is an oversimplification to assume that if symptoms have been present for more than 6 to 12 hours, an irreversible situation has developed that would preclude any attempt at testicular salvage. The duration of symptoms does not always determine functional recoverability.

Although the diagnosis continues to be established most reliably by a skilled examiner familiar with acute scrotal lesions in children, diagnostic imaging studies may be valuable. Nuclear testicular scanning or Doppler sonography reveals decreased or absent arterial blood flow within the affected testicle when compared with its mate. It must be stressed that if the history and physical examination strongly suggest testicular torsion or if any appreciable time would be lost in arranging for these studies, the preferred course is to proceed with surgical exploration or an attempt at manual detorsion (see Fig. 58.5) if surgical intervention is not readily available.

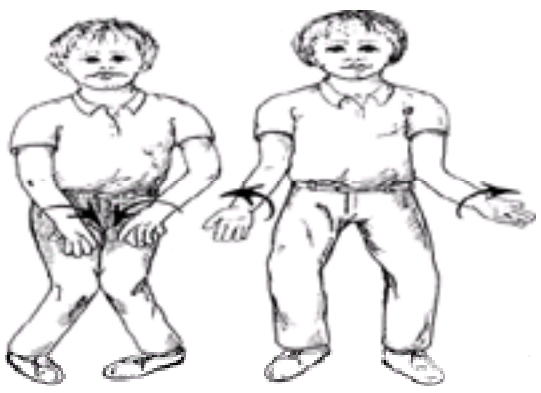

FIGURE 58.5. Torsion of testis. Because torsion typically occurs in a medial direction, manual detorsion should be attempted initially by rotating the testis outward toward the thigh.

The testicular nuclear perfusion scan with technetium-99 pertechnetate is helpful (Fig. 58.4) but has limitations. Technetium is injected and the scrotum scanned. Impeded blood flow to the torsed testicle results in a cold spot. The presence of a hydrocele, abscess, hematoma, or scrotal hernia may result in decreased counts on that side of the scrotum and may be confused with a torsion of the testis. False-negative scans may occur from spontaneous detorsion or in cases of late torsion in which a severe degree of overlying scrotal edema may be associated with sufficient, increased vascularity to obscure the underlying ischemic testis. Doppler flow ultrasound can assess anatomy and blood flow. Limitations associated with Doppler sonography must also be recognized, particularly related to small, lower flow prepubertal testes and the operator-dependent nature of this test. False-negative ultrasounds may occur for reasons similar to those discussed for nuclear scan. Another pitfall encountered with use of both sonography or nuclear scan is incomplete or intermittent torsion in which the study may indicate normal, increased, or decreased flow, depending on timing.

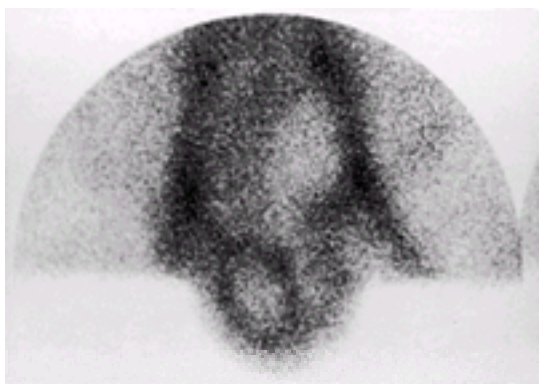

FIGURE 58.4. Torsion of testis (several days old). Testicular scan (technetium-99m) shows central photopenic area surrounded by photon dense area reflecting inflammatory reaction around necrotic testis.

The therapy for testicular torsion is surgical exploration, detorsion, and fixation of both the torsive and contralateral testicles. If a child is seen within a few hours of the onset of his torsion, before severe scrotal swelling has ensued, it may be possible to accomplish detorsion of the spermatic cord manually and thus restore blood supply to the testis. The Doppler ultrasound stethoscope provides a noninvasive evaluation of testicular blood flow and is a useful adjunct in manual detorsion of the testis. Initial examination reveals decreased arterial flow to the affected testis, compared with the contralateral one. Intravenous fentanyl (1 to 3 µg/kg) or morphine (0.1 mg/kg) is administered just before attempting detorsion. Because torsion typically occurs in a medial direction, the detorsion should be carried out by rotating the testis outward toward the thigh (Fig. 58.5). Relief of pain and reposition of the testis in a lower position in the scrotum suggests a successful outcome. This can be confirmed with the Doppler stethoscope by noting a return of normal arterial pulsations to the testis. Although successful completion of manual detorsion may avoid the necessity of an emergency anesthetic for surgical reduction, it does not remove the necessity for a fixation of the testis to prevent the recurrence of this condition. An orchiopexy of the affected testis, as well as of the contralateral one, which is malfixed in more than 50% of cases, is recommended during the same hospitalization.

Torsion of Testicular Appendage

Several vestigial embryologic remnants are commonly attached to the testis or epididymis that may twist around their base, producing venous engorgement, enlargement, and subsequent infarction. Appendage torsion is most common in boys aged 7 to 12 years but can occur at any age. Scrotal pain is the usual presenting feature, although the pain is typically less severe and more indolent in onset than the pain associated with testicular torsion. Although there may be associated nausea, vomiting, and diaphoresis, these symptoms are less common than with torsion of the testis.

If the child is seen early after the onset of pain, scrotal tenderness and swelling may be localized to the area of the twisted appendage, typically on the superior lateral aspect of the testis. It may be possible to hold the testis gently and have the patient point to the specific point of pain. If this site is indicated to be the upper pole of the testis with the remainder of the testis being nontender, the diagnosis of a torsion of a testicular appendage is likely. Although the classic “blue dot” sign of an infarcted appendage may be visible, often it cannot be seen because of overlying edema. In some cases evaluated later in the clinical course, the degree of scrotal tenderness and edema increases to the point at which differentiation from torsion of the testis becomes difficult. Nuclear scanning or Doppler sonography demonstrates normal or increased flow to the affected testicle and epididymis compared with the opposite side, representing the inflammation that occurs with torsion of the appendage. Occasionally, surgical exploration may be required to be certain that a torsion of the testis is not present.

If the examiner is confident in the diagnosis of a torsion of an appendix testis, surgical exploration is not needed. The child should be sent home with analgesics/anti-inflammatories, support to the scrotum, and instructions to rest quietly. The pain usually resolves in 2 to 12 days, but in most cases, pain should improve somewhat within a few days. The patient should return in 48 hours, having had nothing to eat or drink on the morning of the return visit. In most cases, the child's pain will have lessened, and nothing further is indicated. Occasionally, however, a child will seem to have a disproportionate degree of discomfort from the torsion of these tiny appendages. For these children, removal of the appendage may shorten their morbidity. In the older, cooperative child, this may be carried out under a spermatic cord block, but in most cases, a general anesthetic is required. Contralateral scrotal exploration for this condition is not indicated.

Trauma/Hematocele

In children, most trauma to the scrotum results from a direct blow to the perineum or a straddle injury that forcefully compresses the testicle against the pubic bone. Penetrating injuries are less common, and the small size and greater mobility of the prepubertal testis make testicular injuries rare in this group.

Scrotal trauma includes a spectrum of injuries that ranges from minimal scrotal swelling to rupture of the testis with a tense, blood-filled scrotum (Fig. 58.6A). Unless the testis clearly can be felt to be normal and without significant tenderness, urgent surgical evaluation should be undertaken. Often scrotal ultrasound examination is useful (Fig. 58.6B). When any question of testicular rupture remains, surgical exploration is indicated. This approach is based on two facts: 1) a ruptured testis has the best salvage rate when surgically repaired, and 2) testicular torsion may present with a spurious history of trauma.

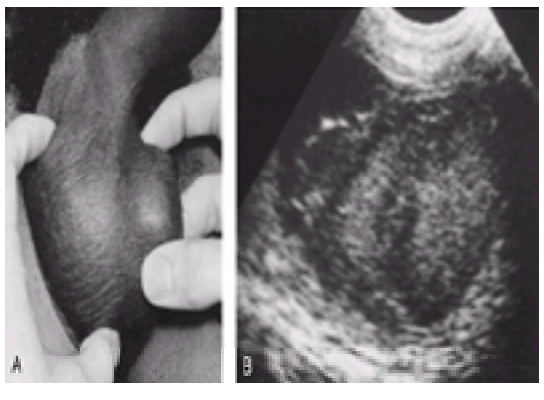

FIGURE 58.6. Rupture of testis. **A.** Testicular swelling and tenderness following kick to scrotum. **B.** Ultrasound examination of testis: central linear sonolucent area reflects site of testicular rupture. Surgical repair resulted in a well-preserved gonad.

A hematocele, or blood within the tunica vaginalis, may represent severe testicular injury. An obvious ecchymosis of the scrotal wall in the setting of trauma suggests a hematocele. Sonography can identify the fluid collection within the tunica because blood is more echogenic than hydrocele fluid. Scrotal exploration is indicated if testicular rupture is present, or in cases of large hematoceles, which heal more readily after surgical drainage.

Scrotal trauma can also result in an intratesticular hematoma or laceration of the tunica albuginea. Ultrasound can assist in determining the location of blood. Any question or indication of testicular laceration requires surgical exploration and drainage of the hematoma with repair of the laceration. If the tunica albuginea can be determined to be intact, no surgical intervention is necessary.

Traumatic epididymitis is local inflammation, resulting from blunt trauma to the scrotum, which usually occurs within a few days. Typically, short-lived acute pain associated with trauma is followed by a pain-free period after which pain returns. On examination, scrotal erythema, edema, and tenderness of the epididymis may be found. In this noninfectious variety of epididymitis, the urinalysis is negative. Sonography is helpful to rule out any more severe injury and will demonstrate hyperemia associated with the inflammation. Treatment is supportive.

If a scrotal laceration is present, it is essential that the testis and spermatic cord be evaluated for possible injury. This may require an examination under general anesthesia or an inguinal cord block with more severe injuries. For simple scrotal lacerations, careful hemostasis and closure of the laceration with chromic catgut is sufficient.

Epididymitis/Orchitis

Epididymitis is an infection or inflammation of the epididymis, which is most commonly seen in adolescents and adults. It is rarely seen in prepubertal boys in whom it is often associated with a urinary tract infection caused by structural abnormalities of the urinary tract. At any age when epididymitis is associated with a urinary tract infection and in all prepubertal boys with epididymitis, urinary tract imaging with sonogram and voiding cystourethrogram are necessary to rule out a structural problem.

The onset of swelling and tenderness is typically more gradual than with torsion of the testis or a testicular appendage. Associated symptoms of urinary frequency and dysuria may be present. Early on, the epididymis may be selectively enlarged and tender, readily distinguished from the testis. With time, inflammation spreads to the testis and surrounding scrotal wall, making localization impossible. Although elevation of the scrotum relieves pain in epididymo-orchitis (Prehn's sign) but causes increased pain in torsion, this finding has not been found to be reliable in children. The cremasteric reflex should be preserved.

Although white cells in the urinary sediment are seen more often in epididymitis than in torsion, they are not consistently present. A culture of the urine should always be obtained, as well as a culture of any penile discharge or the urethra in any sexually active male. Color Doppler sonography typically demonstrates an increase in size and blood flow to the affected testis and epididymis. Nuclear scan shows increased activity in the affected testis (Fig. 58.7).

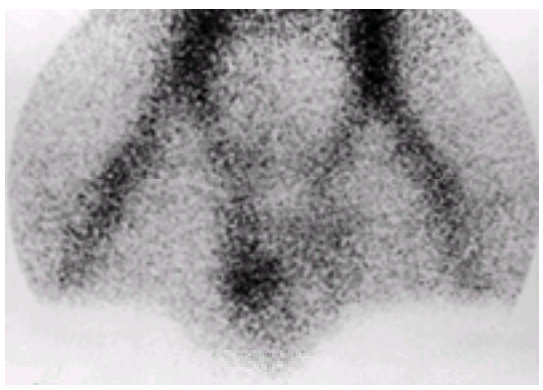

FIGURE 58.7. Epididymitis. Testicular scan (technetium-99m). Diffuse photon dense area in scrotum reflects uptake of radionuclide into inflamed epididymis.

Treatment consists of analgesics, sitz baths, elevation of the scrotum, and antibiotics. Trimethoprim–sulfamethoxazole (40 mg/kg per day sulfamethoxazole) and tetracycline (20 mg/kg per day) achieve best levels in epididymal tissue. In the older boy, as in the adult, *Chlamydia trachomatis* may account for many cases, and the patient should be treated for chlamydia and gonorrhea with doxycycline and ceftriaxone or suitable alternatives. Other organisms, including those bacteria responsible for urinary tract infections, should be considered in the younger boy. The patient should be warned that this process is frustratingly slow to resolve and that he may have several weeks of gradually subsiding discomfort and scrotal swelling.

Orchitis

Orchitis is an inflammation or infection of the testis resulting from the extension of epididymitis, rarely as hematogenous spread of a systemic bacterial infection, or following certain viral infections, including mumps. Other viruses implicated include adenovirus, Epstein-Barr virus, coxsackievirus, and echoviruses. Although rare before puberty, orchitis occurs in about 18% of postpubertal boys with mumps parotitis. In 70% of cases, it is unilateral. It results in testicular atrophy, but not necessarily sterility, in 50% of affected testes. Fortunately, it is much less common since the advent of vaccine against mumps. The onset of mumps orchitis occurs from 4 to 6 days after parotitis manifests. Although rare, orchitis has been reported in the absence of parotitis. Adrenocorticotropic hormone (ACTH) and corticosteroids in adults may produce some degree of local relief of symptoms, but the course of mumps orchitis is not altered.

Causes of Painless Scrotal Swelling

Hydrocele

An accumulation of fluid within the tunica vaginalis that surrounds the testis—a hydrocele—may be seen with torsion of the testis or an appendage, epididymitis, trauma, or tumor. In these cases, examination of the underlying testis is abnormal. If the testis can be felt to be normal and the hydrocele is not associated with any abnormality of the overlying scrotal soft tissues, it is much more likely to be a simple hydrocele. In the infant, this is the result of fluid being left in place after the processus vaginalis has closed. When the size of the hydrocele has no history of waxing or waning, it may simply be observed. Usually, the fluid will be reabsorbed in the first 12 to 18 months of life.

If the hydrocele has a clear-cut history of changing in size (often with crying or exertion), particularly if it is associated with thickening of the cord structures as they are felt against the pubic tubercle (the silk-glove sign), then the processus vaginalis is patent and the diagnosis is that of a communicating hydrocele (Fig. 58.8). Here the patent processus vaginalis does not generally close spontaneously and may enlarge to permit the development of hernia. Surgical exploration and high ligation of the processus vaginalis with a wide opening of the tunica vaginalis to complete the decompression of the hydrocele is appropriate treatment. Because a scrotal hernia may be confused with a hydrocele, aspiration should never be carried out in children.

FIGURE 58.8. Hydrocele. Waxing and waning of size indicates a communicating hydrocele with a patent processus vaginalis, requiring surgical correction.

Occasionally, a hydrocele of the cord presents as a scrotal swelling just above the testis. Differentiation from an incarcerated hernia may be difficult and occasionally may require surgical exploration. Surgical treatment like that for a hydrocele of the testis is appropriate.

Hernia

Although most inguinal hernias present in children with a mass in the groin, occasionally the hernia may extend and present as a scrotal swelling. An incarcerated hernia may produce pain in some patients. The diagnosis and treatment of inguinal hernias is discussed in Chapter 118.

Varicocele

A usually painless scrotal swelling caused by a collection of abnormally enlarged spermatic cord veins, called a varicocele, is most commonly found on routine examination of asymptomatic boys aged 10 to 15 years. Most varicoceles occur on the left, representing spermatic vein incompetence caused by the left spermatic vein draining into the renal vein at a sharp angle, whereas the right spermatic vein drains into the inferior vena cava.

On occasion, a varicocele can present with mild pain or discomfort. The hemiscrotum appears full but does not have overlying skin changes. The testis and epididymis should be palpated to be normal. A mass of varicose veins described as “a bag of worms” can be appreciated above the testicle. Standing and supine examination often reveals the varicocele, which is more prominent when standing. Doppler ultrasound is diagnostic, demonstrating both normal flow to the testis and the collection of tortuous veins. Some large varicoceles may require internal spermatic vein ligation and may have some impact on testicular size and fertility. Most varicoceles are asymptomatic and benign.

Spermatocele

Located above and posterior to the testicle in postpubertal boys, spermatoceles are sperm-containing cysts of the rete testes, ductuli efferentes, or epididymis. Multiple or bilateral spermatoceles may occur. On examination a small, nontender mass that transilluminates may be appreciated distinct from and posterior to the testicle. These masses must be differentiated from a hydrocele or tumor. Sonography may confirm the location distinct from the testis and help distinguish a spermatocele from tumor. Referral to a urologist is indicated for the excision of large uncomfortable spermatoceles or for aspiration to differentiate a hydrocele from a spermatocele. Otherwise, no specific treatment is needed.

Idiopathic Scrotal Edema

Idiopathic scrotal edema is a rare entity that represents only 2 to 5% of acute scrotal swellings in otherwise normal children. Typically, a prepubertal child presents with the rapid onset of painless but notable edema of the scrotal wall that may be bilateral and may extend up onto the abdominal wall. The skin of the scrotum may be erythematous. The child is usually afebrile and urinalysis is negative. Through the edematous scrotum, the testes can be felt to be normal in size and nontender. This edema of the scrotal wall is of unknown origin, although it is believed to represent a form of angioneurotic edema. Insect bites, allergic reactions, cellulitis, and contact dermatitis can also be contributors to localized scrotal swelling. No specific therapy has been demonstrated to be effective. Bed rest and scrotal elevation may help. Children spontaneously begin to improve within 48 hours, regardless of treatment. Occasionally, scrotal edema is seen secondary to diseases that cause generalized edema and/or ascites, such as nephrosis and cirrhosis.

Henoch-Schönlein Purpura

Occasionally, a child may be seen with a petechial rash on the scrotum as the initial presentation of this systemic vasculitic syndrome characterized by nonthrombocytopenic purpura, arthralgia, renal disease, abdominal pain, and gastrointestinal bleeding. More typically, the rash begins on the lower extremities or buttocks and later may involve the scrotum. If the associated swelling is not great, the cord structures and testes can be felt to be uninvolved and normal. In other cases with severe swelling, surgical exploration may be necessary to rule out testicular torsion, which rarely has been noted to coexist. When skin lesions are present the diagnosis of Henoch-Schönlein purpura (HSP) must be suspected. Occasionally, the acute scrotum is the dominant presenting symptom. Ultrasound may help rule out testicular torsion in these instances.

Kawasaki Disease

Another vasculitis that can produce scrotal swelling and mild pain is Kawasaki disease, which has characteristic features including fever, adenopathy, rash, conjunctivitis, and irritability. Although discussed in detail elsewhere (see Chapter 101), it is important to note the association of scrotal swelling with this systemic disease to avoid unnecessary surgical explorations or delay in diagnosis of the underlying vasculitis.

Testis Tumor

Testicular or paratesticular tumors are rare in children. They usually present as painless, unilateral, firm to hard scrotal swellings. Leukemic infiltration of the testis may present bilaterally. There may be an associated hydrocele. In children less than 2 years of age, the tumor usually is a yolk sac carcinoma or teratoma. After puberty, germinal cell tumors, as found in the adult population, are seen. Evaluation of a solid testicular mass involves an initial testicular ultrasound examination usually followed by surgical exploration through a groin incision to permit control of the spermatic vessels and a possible radical inguinal orchiectomy.

Antenatal Torsion Testis (Newborn)

A newborn boy may present with a painless, smooth, testicular enlargement that usually is dark in color. There should be no or minimal edema of the overlying scrotum. This is extravaginal torsion of the testis and probably occurs during the late period of embryonic development, as the testis descends into the scrotum. At this time, the testicular tunics are not yet attached to the scrotal tissue, and torsion of the entire testis with its tunics can occur. Even though the conventional course of action has been surgical exploration, salvage of the testis has been rare. It has been argued that the contralateral testis may be malfixed and at risk for subsequent torsion and therefore should undergo surgical fixation. Torsion has been reported rarely, however, and current practice is simply to observe these children. After 4 to 6 months, the torsed testis usually has been resorbed.

EVALUATION AND DECISION

Although this chapter is entitled scrotal pain, the most efficient approach to the differential diagnosis is through consideration of the important entities causing painful versus painless scrotal swelling. This approach is outlined in Figure 58.9. Testicular torsion, torsion of an appendage, orchitis, epididymitis, and trauma related injuries to the scrotum or testicles are further discussed as the common etiologies for painful scrotal swelling. Hemorrhage into a tumor, incarcerated hernias, HSP, and Kawasaki disease may cause either painful or painless scrotal swelling. No one aspect of the history or physical examination may be diagnostic, but collectively the clinical findings often suggest a diagnosis. More recently available adjunctive radiologic studies may be helpful when the clinician is fully aware of their capabilities and limitations.

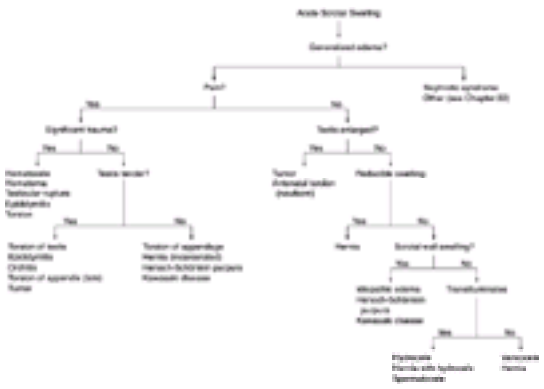

FIGURE 58.9. Diagnostic approach to acute scrotal swelling.

Initial Approach

As a first step in the evaluation of the child with a complaint of scrotal swelling or pain, the physician should determine whether the child is suffering from a generalized edematous state, such as the nephrotic syndrome. When the problem in localized to the scrotum, patients can be divided into those who have a painless swelling and those who are experiencing pain.

In the immediate neonatal period, antenatal torsion may cause painless scrotal enlargement. Beyond infancy, the most common causes (Table 58.2) are hernias and hydroceles; a hernia is often reducible. The physician must also consider idiopathic scrotal edema, spermatocele, and varicocele. Both Kawasaki disease and HSP may involve the scrotal sac and cause swelling that is either painless or mildly painful. When the swelling is within the scrotum, rather than involving the scrotal sac, possible diagnoses include hydrocele, hernia (reducible if incarcerated and nonreducible), spermatocele, and hydrocele.

Painful swelling may follow a well-documented injury, in which case the likely diagnoses are hematocele, hematoma, testicular rupture, and traumatic epididymitis. The physician should bear in mind that boys with testicular trauma often give a history of having had an incidental minor injury. Nontraumatic scrotal pain raises the suspicion of a testicular torsion, particularly if the testis is tender. Unless the diagnosis of a systemic disorder (HSP, Kawasaki disease) is obvious, the patient is an adolescent with the classic signs of epididymitis, or an incarcerated hernia is reduced, imaging via Doppler ultrasound or nuclear scan is usually indicated. All patients in whom torsion is suspected, after an initial evaluation by the emergency physician, require a surgical consultation.

History

The age of the child should be considered in evaluating scrotal pain and/or swelling, but overlaps in characteristic age at presentation exist. Testicular torsion occurs in the newborn or early pubertal age range. Torsion of an appendage of the testis, HSP, idiopathic scrotal edema, and Kawasaki disease commonly occur in the prepubertal age group. Epididymitis is more common in adolescents but may occur in prepubertal boys.

Onset of pain is more abrupt in testicular torsion, whereas, the pain of appendage torsion and epididymitis is more gradual. Children may have difficulty pinpointing the onset of pain and even the exact location of their pain because they may not initially localize sensation to the scrotum, but rather often complain of lower abdominal pain. Questions about recent activity and behavior may indicate a more insidious course. Nausea and vomiting often accompany testicular torsion, and fever and symptoms of urinary tract infection may suggest epididymitis or other inflammatory diseases (vasculitides).

A history of trauma should always be addressed recognizing the difference between significant trauma associated with severe acute pain and minor trauma to which the pain of torsion may mistakenly be attributed by the patient. Prior scrotal pain may also indicate intermittent torsion.

A history of prior genitourinary surgeries should be elicited because predisposition to urinary tract infections and epididymitis may be related to genitourinary abnormalities or prior instrumentation. Prior surgery for hernias, hydroceles, and undescended testis, unless associated with other genitourinary or anal rectal abnormalities, does not suggest a predisposition to infection. In addition, torsion can occur despite prior scrotal surgeries thought to secure the testis.

Physical Examination

Examination of the child with scrotal pain and/or swelling should be both careful and organized. Initial observation of the patient's gait, resting position, and facial expression are helpful. Writhing or an especially quiet supine posture versus active movement may best indicate the degree of pain. Observation of associated skin changes, presence and location of swelling, and the natural position of the testicle in the scrotum while standing should then be appreciated. The cremasteric reflex elicited by stroking the upper inner thigh should cause the testicle to elevate when intact. Next the lower abdomen, inguinal canal, and cord should be palpated. Finally the scrotum and its contents should be sequentially palpated. Asking the patient to localize his pain with one finger at this time may be especially helpful. The unaffected hemiscrotum should always be palpated first. Knowledge of the location and specific attempt at palpation of the appendix testis and epididymis is beneficial before palpation of the testis itself (Fig. 58.2). Appreciation of swelling, tenderness, and consistency should be noted for all intrascrotal structures. Transillumination may be helpful in some cases (Fig. 58.9).

Imaging Modalities

Technetium-99 pertechnetate radioisotope scanning has been used to assess testicular blood flow. In a child with torsion, little or no isotope appears in the testis, whereas normal or increased activity in the affected testis is associated with epididymitis and appendage torsion. This study provides no information about anatomy and, in addition to a procedure time of approximately 30 minutes, often requires some time in gathering staff, achieving intravenous access, and preparing the isotope. The interpreter of this study must also be confident in differentiating blood flow to the testis versus the scrotal wall.

Color Doppler imaging with pulsed Doppler allows both visualization of scrotal anatomy and intratesticular arterial blood vessel flow determination and comparison. This study can differentiate scrotal wall from testicular blood flow. With appropriate experience, Doppler imaging has comparable accuracy to that of nuclear scanning. This study is noninvasive and requires less preparation, making it more readily available. The operator-dependent nature of the study and the need for clinical correlation must be heeded. Torsion is diagnosed when blood flow is diminished or absent, as compared with the contralateral testis. Any doubt in interpretation or concern about the adequacy of the signal should result in consideration of surgical exploration.

Suggested Readings

Caldamone AA, Valvo JR, Altebarmakian VK, et al. Acute scrotal swelling in children. J Pediatr Surg 1984;19:581.

Dewire DM, Begun FP, Lawson RK, et al. Color Doppler ultrasonography in the evaluation of the acute scrotum. J Urol 1992;147:89.

Dresner ML. Torsed appendage: diagnosis and management; blue dot sign. Urology 1973;1:63.

Evans JP, Snyder HM. Idiopathic scrotal edema. Urology 1977;9:549.

Kaplan GW, King LR. Acute scrotal swelling in children. J Urol 1970;104:219.

Kass EJ, Lundak B. The acute scrotum. Pediatr Clin North Am 1977;44:1251.

Klein BL, Ochsenschlager DW. Scrotal masses in children and adolescents: a review for the emergency physician. Pediatr Emerg Care 1993;9:351.

Knight PJ, Vassy LE. Diagnosis and treatment of acute scrotum in children and adolescents. Ann Surg 1984;200:664.

Kugan SJ. Acute and chronic scrotal swellings. In: Gillenwater JY, Grayhack JT, Howards SS, et al, eds. Adult and Pediatric Urology. 2nd ed. St Louis: Mosby, 1991:2189.

Lindsey D, Stanisic TH. Diagnosis and management of testicular torsion: pitfalls and perils. Am J Emerg Med 1988;6:42.

Siegel A, Snyder HM, Duckett JW. Epididymitis in infants and boys: underlying urogenital anomalies and efficacy of imaging modalities. J Urol 1987;138:1100.

CHAPTER 59

Pallor

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Rosy cheeks in children always have been valued highly by grandmothers and other trendsetters in American life. Unfortunately, the loss or absence of this pleasing pink color is a relatively common problem in childhood. The development of pallor can be acute and associated with a life-threatening illness, or it can be chronic and subtle, occasionally first noted by someone who sees the child less often than the parents. The onset of pallor can provoke anxiety for parents who are familiar with descriptions of the presentation of leukemia in childhood. In some instances, only reassurance may be needed, as in the case of a light-complexioned or fair-skinned, nonanemic child. Even if there is a hematologic cause for the pallor, it often is a temporary condition readily amenable to therapy. Pallor can portend a severe disease, however, and when acute is onset, it can herald a true pediatric emergency for which rapid diagnosis and treatment are needed.

The degree of pallor depends on the concentration of hemoglobin in the blood and the distribution of blood in the blood vessels of the skin. Any condition that decreases the concentration of hemoglobin or alters the distribution of blood away from the body's surface may present as pallor. Clinically, pallor caused by anemia usually can be appreciated when the hemoglobin concentration is below 8 to 9 g/dL, although the complexion of the child and the rapidity of onset may influence this value. The hematologic causes for pallor in children are discussed later, and further details regarding their management may be found in Chapter 87. Nonhematologic causes of pallor are outlined briefly in Table 59.1.

Physiologic ("fair-skinned")	Respiratory distress
Shock: septic, hypovolemic, neurogenic, cardiogenic, anaphylactoid	Skin edema
	Pheochromocytoma
Hypoglycemia and other metabolic derangements	

Table 59.1. Pallor Without Anemia

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of the major hematologic causes of pallor in children is outlined in Table 59.2. The concentration of hemoglobin in the blood can be lowered by three basic mechanisms: decreased erythrocyte or hemoglobin production, increased erythrocyte destruction, and blood loss. The most common causes of pallor and anemia seen in the emergency department (ED) are iron deficiency, infections, and blood loss (Table 59.3), but several less common diseases remain important considerations.

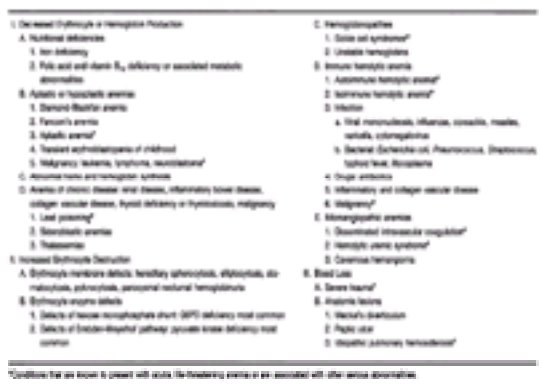

Table 59.2. Pallor With Anemia

Decreased Erythrocyte or Hemoglobin Production	Increased Erythrocyte Destruction
Iron deficiency	Sickle cell syndromes
Aplasia	Autoimmune hemolytic anemia
	Viral, bacterial, or parasitic infections
	Blood Loss

Table 59.3. Relatively Common Causes of Pallor or Anemia

Decreased Production of Hemoglobin and Red Cells

Nutritional Anemias

In children, the most common cause of decreased hemoglobin production is nutritional iron deficiency. This condition usually is seen in the first 2 years of life, at which time the dietary iron content often is insufficient to meet the demands of the rapidly increasing red cell mass. Premature infants are particularly likely to develop iron deficiency anemia because iron stores at birth are less than those found in term infants, whereas the growth (and therefore expansion of the red cell mass) of the premature infant often is faster than that of term infants. The early exhaustion of iron stores in premature babies may result in pallor by 6 months of age; in normal infants, signs of iron deficiency anemia are uncommon before 10 to 12 months.

The infant with severe iron deficiency usually is irritable and very pale. The lack of iron in the diet may be readily apparent or may be recognized only after careful questioning, particularly regarding the daily consumption of cow's milk. The hemoglobin concentration may be as low as 2 g/dL at the time of diagnosis. Severe anemia may result in a compensatory increase in cardiac output. Conditions that complicate anemia, such as fever, may make further demands on the heart and thereby provoke the development of congestive heart failure (see Chapter 82).

The red cells are markedly microcytic and hypochromic in severe iron deficiency anemia. Variation in red cell size and shape usually is present, and elongated, pencil-like cells are particularly common. The percentage of reticulocytes may be elevated moderately, but the absolute reticulocyte count is low. Treatment usually is instituted before confirmatory laboratory studies are available. This rarely poses a problem, however, because the diagnosis often can be made on the basis of the history alone. The rapidity of the assay for free erythrocyte protoporphyrin, which is increased in iron deficiency anemia, makes this test particularly useful in the evaluation of the severely anemic child. Serum iron and ferritin levels have too long of a turnaround time to be of much value in the emergency management of anemia, but they are valuable confirmatory tests.

Other nutritional anemias, such as vitamin B_{12} or folic acid deficiency, are uncommon in children in the United States and rarely develop in the absence of a grossly altered diet, extended hyperalimentation, intestinal resection, or chronic diarrhea. Unusual alterations of B_{12} and folic acid absorption and metabolism may cause symptoms similar to those of the nutritional megaloblastic anemias. Megaloblastic anemia is rarely severe enough to be life-threatening. The condition is characterized by normochromic, macrocytic red blood cells, hypersegmented neutrophils and an elevated serum level of lactic dehydrogenase (LDH). The diagnosis of nutritional disorders is confirmed by the finding of low serum levels of folic acid or vitamin B_{12} and the response to folic acid or vitamin B_{12} replacement therapy.

Hypoplastic and Aplastic Anemia

Pallor usually is the first sign of aplastic or hypoplastic anemia. Diamond-Blackfan syndrome is a congenital hypoplastic anemia commonly detected in the first few months of life. The anemia can be severe at the time of diagnosis. The red cells are normocytic or macrocytic. The reticulocyte count is low. The white cell count is low in approximately 10% of affected patients, but thrombocytopenia occurs only rarely. The diagnosis is made by examination of a bone marrow aspirate that shows markedly reduced or absent erythrocyte precursors. The second major congenital hypoplastic

anemia is Fanconi's anemia, a syndrome characterized by pancytopenia and associated abnormalities, including hyperpigmentation and hypopigmentation, microcephaly, strabismus, small stature, mental retardation, and abnormalities of the thumbs and radii. Unlike Diamond-Blackfan syndrome, all three cell lines of the bone marrow are affected, and the hematologic abnormalities rarely develop before 3 to 4 years of age. The anemia is normochromic and macrocytic.

Acquired aplastic anemia also can present with severe pallor in children. The anemia usually is associated with granulocytopenia and thrombocytopenia. The condition often is idiopathic but has been associated with exposure to certain drugs and chemicals (chloramphenicol, benzene, pesticides), radiation, and viral infections (especially hepatitis). The diagnosis is made by an examination of the bone marrow.

Transient erythroblastopenia of childhood (TEC) is a condition often associated with a recent viral illness and is characterized by moderate to severe anemia caused by diminished red cell production. The mean corpuscular volume (MCV) usually is normal at the time of diagnosis. The white cell count is normal or moderately decreased; the platelet count is normal. The reticulocyte count is decreased, and the Coombs test is negative. Bone marrow examination shows reduction or absence of erythrocytic precursors initially, followed by erythroid hyperplasia during recovery. Transient erythroblastopenia that occurs in the first 6 months of life may be difficult to distinguish from Diamond-Blackfan anemia. Spontaneous recovery ultimately confirms the diagnosis of TEC.

Hypoplastic anemia can be the presenting symptom of childhood malignancies. The pallor can be severe, and although all three cell lines of the bone marrow usually are affected, anemia may be the only notable hematologic abnormality. The diagnosis can be suspected from the presence of other symptoms or findings such as lymphadenopathy, bruising, limb pain, gum bleeding, or an abdominal mass.

Patients with hemolytic anemias such as spherocytosis or sickle cell disease may develop red cell aplasia, usually in association with parvovirus B19 infection. Decreased red cell production in the face of ongoing hemolysis causes an exacerbation of the anemia. The usually elevated reticulocyte count falls to inappropriately low levels, often less than 1%. Although the platelets and white cells are generally unaffected, they may be mildly deceased. Red cell transfusions are appropriate if the anemia is associated with cardiovascular signs or symptoms or if continuing reticulocytopenia indicates that the anemia is likely to become severe before the usual spontaneous recovery after 3 to 7 days. Hematologically normal children with underlying (but sometimes unrecognized) immunologic disorders may also develop parvovirus-induced anemia as a result of prolonged viremia.

Disorders of Heme and Globin Production

Pallor may be the presenting sign of nonnutritional disorders of hemoglobin synthesis, including the sideroblastic anemias and thalassemia syndromes. These disorders are characterized by a microcytic, hypochromic anemia. Sideroblastic anemia may be inherited (sex-linked) or acquired. Iron use within the developing red cell is abnormal, accounting for the presence of diagnostic ringed sideroblasts in the bone marrow. The serum iron and ferritin levels usually are markedly elevated.

In the thalassemias, production of the globin portion of the hemoglobin molecule is defective. Cooley's anemia (b-thalassemia major) presents with severe pallor usually between 6 and 12 months of age, as the fetal hemoglobin (HbF) level declines but the normal rise in adult hemoglobin (HbA) production fails to occur because of reduced or absent b-globin production. Although b-thalassemia is often associated with Mediterranean ancestry, this disease and related disorders (e.g., E-b thalassemia, HbH disease) also are seen commonly in children of Southeast Asian, Indian, Pakistani, and Chinese background. The presence of hepatosplenomegaly and characteristic red cell morphology, including marked variation in red cell shape, makes this diagnosis readily apparent.

Lead poisoning affects heme synthesis, but significant anemia is unusual unless blood lead levels are markedly elevated. Iron deficiency is common in children with increased lead levels and usually accounts for the microcytic anemia found in these patients. If a concomitant hematologic disorder cannot be found in the anemic patient with plumbism, particular care should be given to the possibility of severe lead intoxication.

Systemic Disease

Numerous disorders that are not primarily hematologic may be associated with pallor and anemia. Occasionally, pallor is the only presenting finding of a serious systemic disorder. Chronic inflammatory diseases, such as juvenile rheumatoid arthritis (JRA) and ulcerative colitis, often are accompanied by a normocytic or microcytic anemia related to impaired iron use. The serum iron is reduced. The low iron-binding capacity distinguishes the anemia of chronic inflammation from the anemia of iron deficiency. Similar clinical and laboratory findings may be associated with chronic infections such as acquired immunodeficiency syndrome (AIDS) and subacute bacterial endocarditis. Other diseases in which anemia may be a prominent component include chronic renal disease, hyperthyroidism, and hypothyroidism. The anemia in these disorders is not severe enough to be considered a hematologic emergency unless complicated by other hematologic abnormalities. However, the anemia may be the first clue to an underlying disease in which early treatment may improve the outcome substantially.

Increased Red Cell Destruction

The numerous conditions associated with shortened red cell survival can be congenital, as in the case of the hemoglobinopathies and membrane and enzyme defects, or acquired, as in the case of autoimmune hemolytic anemia, drug-associated hemolytic anemias, disseminated intravascular coagulation (DIC), and hemolytic uremic syndrome (HUS). The hemoglobin levels in these disorders can be normal, slightly depressed, or so low as to be life-threatening. This level is determined by the severity of the defect and the patient's ability to respond to the presence of a shortened red cell survival. Compensation is achieved by an increase in erythrocyte production as is evident from the elevated

reticulocyte count that usually is found in these conditions.

An alteration in the patient's ability to compensate for increased red cell destruction may result in a severe, life-threatening exacerbation of the underlying anemia (see the previous section). This aplastic crisis, the result of a transient decrease in erythrocyte production in the presence of shortened red cell survival, should be suspected in a patient with a known hemolytic anemia who develops increasing pallor and anemia associated with a reticulocyte count much lower than usual. Unfortunately, when the hemolytic anemia has not been diagnosed previously, the recognition of an aplastic crisis can be difficult because the findings are similar to those of transient erythroblastopenia of childhood or congenital erythrocyte hypoplasia (i.e., anemia with low or absent reticulocytes). Examination of the bone marrow may be helpful in distinguishing these disorders because production of early red cell precursors often has resumed shortly after detection of an aplastic crisis. Often, the diagnosis of an aplastic crisis is made only after the aplasia has ended and the underlying hemolytic anemia is apparent. After resolution of a suspected aplastic crisis, every effort should be made to uncover any underlying condition that might cause shortened red cell survival. The differential diagnosis of such conditions is presented next.

Membrane Disorders

The degree of pallor associated with anemia caused by erythrocyte membrane abnormalities depends on the hemoglobin level. In rare instances, patients with hereditary spherocytosis, the most common of the membrane disorders, may develop significant anemia and pallor in the newborn period. Moderate or severe anemia is less common in the other membrane disorders, such as hereditary elliptocytosis and hereditary stomatocytosis. The anemia of the erythrocyte membrane disorders is accompanied by reticulocytosis. The red cell morphology often permits the diagnosis to be made from the peripheral smear. Because these disorders often are inherited in an autosomal dominant fashion, a family history of anemia, splenomegaly, splenectomy, or cholecystectomy may be helpful. However, a particularly severe form of spherocytosis occurs as an autosomal recessive disorder, and some children with more typical disease lack an informative family history. Consequently, the diagnosis should not be dismissed in the absence of other affected family members.

Infantile pyknocytosis is a hemolytic anemia seen during the first few months of life and is characterized by distorted and contracted erythrocytes and burr cells. The disorder may be associated with pallor and hyperbilirubinemia. Spontaneous recovery usually occurs by 6 months of age.

Enzyme Disorders

Erythrocyte enzymatic defects, such as pyruvate kinase deficiency and certain variants of glucose-6-phosphate dehydrogenase (G6PD) deficiency, may be associated with pallor from increased red blood cell destruction. In the latter disorder, pallor may be accentuated by acute hemolytic crises after exposure to oxidant stress (e.g., naphthalene-containing mothballs, drugs, acidosis). Although alterations in red cell morphology sometimes are found in these enzyme disorders, assays of specific enzymes or substrates are required for definitive diagnosis.

Hemoglobinopathies

Pallor may result from the low hemoglobin level found in patients with sickle cell anemia and related hemoglobinopathies. Acute accentuation of pallor can result from an aplastic crisis, a complication of hemolytic disorders that is particularly common in sickle cell anemia. During an aplastic crisis, the normally elevated reticulocyte count may fall to zero, and the hemoglobin level may fall as low as 1 to 2 g/dL, resulting in severe pallor and signs of high-output cardiac failure.

The sequestration crisis of sickle cell anemia and related hemoglobin disorders (SC disease, S-b° thalassemia, S-b $^{+}$ thalassemia) results from acute pooling of red cells and plasma in the spleen. The sudden and severe anemia and the hypovolemia associated with this complication constitute a true hematologic emergency and, if untreated, may rapidly lead to death. The presence of increased pallor and acute enlargement of the spleen in a patient with a sickling disorder should prompt immediate investigation of a possible sequestration crisis. Although this complication rarely occurs in children with homozygous sickle cell disease or S-b° thalassemia after the age of 5 years, sequestration crises may occur much later in children with sickling disorders such as SC disease or S-b $^{+}$ thalassemia, in which early splenic infarction is less common.

Immune Hemolytic Anemia

Pallor caused by autoimmune hemolytic anemia usually is acute in onset and may be associated with severe anemia. The presence of only moderate anemia (6 to 8 g/dL) at diagnosis should not detract from consideration of this disease as a hematologic emergency because brisk hemolysis may result in a sudden, additional fall in hemoglobin level. Autoimmune hemolytic anemia usually, but not always, is characterized by a positive Coombs test and an increased reticulocyte count. Spherocytes are commonly seen in the peripheral smear. Other causes of immune hemolytic anemia include infections, drugs, inflammatory diseases, and malignancies.

Microangiopathic Anemia

Alterations in the normal flow of blood through the vascular system may cause increased red cell destruction. In DIC, abnormal fibrin deposition within small blood vessels results in mechanical injury to the erythrocytes. Thrombocytopenia and clotting abnormalities, which often herald the onset of DIC, also may contribute to the anemia by causing diffuse bleeding. The main diagnostic findings are red cell fragments in the peripheral blood smear, platelet and clotting abnormalities typical of a consumptive coagulopathy (see Chapter 87), and the clinical features of a disease such as septic shock, which is associated with DIC.

The increased red cell destruction in HUS and thrombotic thrombocytopenic purpura (TTP) also is caused by

intravascular fibrin deposition. Thrombocytopenia and uremia may lower the hemoglobin concentration even further by causing bleeding, impaired red cell production, shortened red cell survival, and increased plasma volume. In some instances, the anemia may be moderately severe when the uremia is only mild and thrombocytopenia is absent, leaving doubt about the correct diagnosis. In more typical cases, however, the diagnosis is readily apparent from the findings of oliguria, central nervous system abnormalities, increased blood urea nitrogen (BUN), thrombocytopenia, and abnormalities of red cell morphology, including fragments and helmet cells.

The proliferation of blood vessels within a cavernous hemangioma may trap red cells or may initiate a localized consumptive coagulopathy, causing erythrocyte destruction. Anemia is rarely severe unless the thrombocytopenia that is more typical of the disorder causes chronic blood loss.

Blood Loss

Although sudden, massive hemorrhage usually is accompanied by signs of hypovolemic shock, the repeated loss of smaller amounts of blood may be associated with few findings other than pallor. The finding of iron deficiency anemia despite normal dietary iron intake or iron supplementation may be a clue to the presence of chronic blood loss from the gastrointestinal (GI) tract or within the lungs.

EVALUATION AND DECISION

The initial assessment of the child with pallor should include an immediate determination of the degree of illness. Rapid treatment may be imperative for the severely ill child. In the presence of hypovolemic shock, immediate support of vascular volume is required. When high-output cardiac failure from severe anemia occurs, transfusion with small aliquots of packed red cells is necessary. Only after these initial therapeutic efforts have been completed can a thorough evaluation of the anemia proceed.

The presence of an underlying disease that requires immediate therapy may alter the usual approach to the investigation of pallor. For example, iron deficiency may be the most likely explanation for anemia in a pale 1-year-old African-American child with newly diagnosed meningitis, but the early recognition of this hematologic abnormality does not directly affect the management of the patient. If the anemia is caused by sickle cell disease, however, the management of a patient with meningitis may require modification. Therefore, specific studies for sickling disorders should be performed in the initial stages of the evaluation of the anemia in such a patient.

If the child with pallor is not acutely ill, a deliberate search for the cause of pallor should be undertaken (Fig. 59.1). A thorough yet relevant history should be obtained with particular attention to the type of onset of pallor. The slow development of pallor, often noticed only by a family member or friend who sees the child only occasionally, suggests diminished red cell production, as is found in bone marrow aplasia or iron deficiency. However, the acute onset of pallor is consistent with the brisk hemolysis found in autoimmune hemolytic anemia and often is accompanied by jaundice, dark urine, and cardiovascular changes.

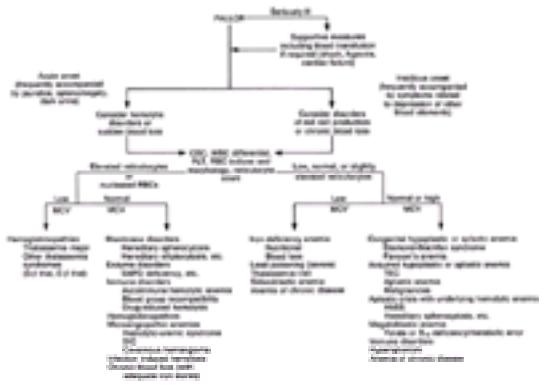

FIGURE 59.1. The diagnostic approach to pallor. *CBC,* complete blood count; *WBC,* white blood cells; *PLT,* platelets; *RBC,* red blood cells; *MCV,* mean corpuscular volume; *G6PD,* glucose-6-phosphate deficiency; *TEC,* transient erythroblastopenia of childhood; *DIC,* disseminated intravascular coagulation.

After establishing the type of onset of the anemia, the history can be directed toward more narrow categories of anemia or specific diseases. A detailed dietary history, with particular attention to milk intake, is important in young children with suspected iron deficiency; excessive consumption of cow's milk often results in iron deficiency. Vitamin B_{12} deficiency may accompany strict vegetarian diets from which meat and egg products are excluded and may occur in breast-fed infants of vegetarian mothers or mothers with pernicious anemia. Nutritional folic acid deficiency is rare and usually can be readily deduced from the presence of severe dietary alterations and evidence of other vitamin deficiencies.

Sources of internal or external blood loss should be carefully sought. Chronic GI bleeding may escape detection until iron deficiency anemia develops. Similarly, small pulmonary hemorrhages associated with idiopathic pulmonary hemosiderosis are often mistaken for other pulmonic processes until several recurrences of iron deficiency anemia suggest a hidden site of blood loss.

If increased bruising or bleeding accompanies pallor, multiple blood elements are probably affected. The circulation time of platelets is short in comparison with that of red cells. Therefore, clinical findings of thrombocytopenia often are present

by the time pallor develops in patients with acquired aplastic anemia, Fanconi's anemia, and acute leukemia.

The family history helps in the diagnosis of hemoglobinopathies and inherited disorders of red cell membranes and enzymes. Because results of previous hemoglobin testing may have been explained inadequately or recalled inaccurately, a negative family history or newborn screening for hemoglobinopathies should not preclude evaluation of the patient's hemoglobin phenotype if a sickling disorder is suspected. The presence of a microcytic anemia unresponsive to iron in the parents suggests a thalassemic disorder. A history of splenomegaly, splenectomy, or cholecystectomy in family members may help identify a hemolytic disorder such as hereditary spherocytosis or pyruvate kinase deficiency. Finally, a well-directed review of systems is essential in looking for systemic disorders such as chronic renal disease, hypothyroidism, or JRA. Pallor may be the presenting complaint in these and other disorders.

In the examination of the severely anemic patient, pallor of the skin and mucous membranes usually is readily apparent. When anemia is less severe or when the skin color is dark, pallor may be appreciated only in the nailbeds and palpebral conjunctivae. Blood pressure (BP) and pulse should be measured to be sure that hypovolemic shock and high-output cardiac failure are neither present nor imminent. If anemia or volume loss is mild, tachycardia may be present, but normal BP is preserved. A systolic flow murmur often is heard if the hemoglobin level is below 8 g/dL. Lymphadenopathy and splenomegaly may suggest a malignancy or an infectious disease such as mononucleosis. When splenomegaly occurs without lymphadenopathy, however, attention is drawn to hemolytic disorders such as hereditary spherocytosis and autoimmune hemolytic anemia or hemoglobinopathies (sickling disorders or thalassemia major). Scleral icterus also may be present in these disorders of shortened red cell survival. The finding of an unusually large and firm spleen in the absence of increasing scleral icterus suggests that red cells are being sequestered (e.g., splenic sequestration crisis of sickle cell disease, hypersplenism).

The skin should be examined for the presence of hemangiomas that might cause microangiopathic anemia. Careful auscultation of the abdomen and head may detect hemangiomas of the viscera. Bony abnormalities associated with red cell disorders include frontal bossing from compensatory expansion of the bone marrow in hemolytic diseases and radial and thumb anomalies found in some patients with Fanconi's anemia.

Numerous classifications of anemia have been used to assist the physician in the laboratory investigation of pallor. Historically, the reticulocyte count and the MCV have been helpful measurements in categorizing causes of anemia. The reticulocyte count can be performed rapidly and, as shown in Figure 59.1, distinguishes anemias caused by impaired red cell production (e.g., iron deficiency, hypoplastic anemia) from those caused by shortened red cell survival (e.g., hemoglobinopathies, membrane disorders). The MCV provides a quick, accurate, and readily available method of distinguishing the microcytic anemias (iron deficiency, thalassemia syndromes) from the normocytic (membrane disorders, enzyme deficiencies, autoimmune hemolytic anemia, most hemoglobinopathies) or macrocytic (bone marrow/stem cell failure, disorders of B_{12} and folic acid absorption or metabolism) anemias.

The reticulocyte count and MCV should be interpreted with caution. As shown in Figure 59.1, disorders of shortened red cell survival are not always characterized by an increased reticulocyte count. For example, reticulocytopenia may occur in autoimmune hemolytic anemia despite active hemolysis and increased erythropoiesis in the bone marrow. Chronic hemolytic disorders, such as sickle cell anemia or hereditary spherocytosis, may first be detected during an aplastic crisis when the reticulocyte count is low. Unless the underlying disorder is recognized, the physician may be misled by this finding. Furthermore, because the reticulocyte count is expressed as a percentage of total red cells, often it must be corrected for the degree of anemia. The easiest way to make this correction is to multiply the reticulocyte count by the reported hemoglobin or hematocrit (HCT) divided by a normal hemoglobin or hematocrit:

$$\text{Reticulocyte count} \times \frac{\text{HCT (pt)}}{\text{HCT (nl)}}$$

For example, a reticulocyte count of 5% in a child with severe iron deficiency anemia and a hematocrit of 6% is not elevated when corrected for the degree of anemia (5% × 6%/33% = 0.9%).

The MCV varies with age, necessitating the use of age-adjusted normal values (Table 59.4). In addition, the measured MCV represents an average value. If microcytic and macrocytic red cells are present in the peripheral blood as, for example, in a patient with combined iron deficiency and B_{12} deficiency, the MCV may remain normal. Therefore, the peripheral smear should be examined carefully to determine whether the MCV reflects a single population of red cells of uniform size or two or more populations of distinctly different size. The red cell distribution width (RDW) is elevated in the presence of increased variation in red cell size.

	MCV (fl)	
Age (yr)	Median	Lower Limit[a]
0.5–2	77	70
2–5	79	73
5–9	81	75
9–12	83	76
12–14:		
Female	85	77
Male	84	76
14–18:		
Female	87	78
Male	86	77

[a]Third percentile. fl, femtoliters.

Table 59.4. Age-Related Values for Mean Corpuscular Volume

As shown in Figure 59.1, the reticulocyte count and MCV help in the initial classification of anemia but leave the physician with broad categories of disease, rather than specific diagnoses. In many instances, the history and physical examination, when coupled with these laboratory measurements, permit identification of a particular disorder. Additional laboratory studies and careful examination of the peripheral smear often are required, however, and can be performed readily when the patient is in the ED. The application of these procedures to diseases that are commonly encountered or that are associated with unusually severe anemia is discussed next.

Increased Reticulocytes and Low Mean Corpuscular Value

The thalassemia syndromes associated with moderate or severe anemia can be recognized by the distinctive abnormalities of red cell morphology. In Cooley's anemia (b-thalassemia major), the red cells generally are small but vary markedly in size and shape. Many cells appear to contain little or no hemoglobin; the central pallor extends to the cell membrane. Nucleated red cells, basophilic stippling, and polychromasia reflect active erythropoiesis. The parents of an affected child usually have a low MCV characteristic of thalassemia trait.

Children with HbS-b-thalassemia often have microcytic red cells, although the alterations of red cell morphology are not as dramatic as in Cooley's anemia. Sickled forms are often but not always present. Target cells are common. The solubility tests are positive because of the presence of HbS. Hemoglobin electrophoresis reveals HbS and reduced (less than 50%) or absent HbA.

Increased Reticulocytes and Normal Mean Corpuscular Value

Most membrane disorders can be readily identified by the characteristic changes in red cell shape that lend their names to the diseases (e.g., spherocytosis, elliptocytosis, stomatocytosis). When the diagnosis of a membrane disorder is uncertain, examination of the parents' peripheral smears may be helpful because, in many cases, the inheritance pattern is autosomal dominant.

Abnormalities of red cell morphology are less striking in erythrocyte enzymatic defects. Blister cells and cells with asymmetric distribution of hemoglobin may be found, however, during episodes of active hemolysis in G6PD deficiency. If transfusion is necessary, a pretransfusion sample should be saved for assay of specific enzymes.

The reticulocyte count usually is markedly elevated in autoimmune hemolytic anemia but may be normal or only slightly elevated during the first days of the disease. In rare instances, reticulocytopenia persists. Spherocytes usually are present on the peripheral smear. Clumping of red cells from agglutination may be seen. This agglutination sometimes causes a falsely elevated MCV because the electronic counter measures the volume of red cell couplets or triplets. The direct Coombs test is positive in 90% of cases. Patients with a negative Coombs test present a challenging diagnostic problem because the initial findings may be similar to those in hereditary spherocytosis.

The recognition of homozygous sickle cell disease usually is accomplished by the finding of sickled red cells on the peripheral smear. Rarely, however, such cells are absent, even during an acute illness. Target cells are commonly found in sickle cell disease but are more prominent in HbSC. Solubility tests are positive. Hemoglobin electrophoresis reveals the presence of the abnormal hemoglobin(s) and the absence of HbA. This confirmatory test takes less than 30 minutes to complete and should be performed when important therapeutic decisions depend on the result.

Red cell fragments are found in those diseases characterized by microangiopathic anemia. In HUS or TTP, thrombocytopenia is present, renal or neurologic function is usually impaired, and thrombotic complications may be present. The platelet count also is low in DIC, and clotting studies are abnormal. If intravascular hemolysis is severe, as in anemia associated with certain artificial cardiac valves, hemosiderin may be detected in the urinary sediment.

Low, Normal, or Slightly Elevated Reticulocytes and Low Mean Corpuscular Value

In severe iron deficiency anemia, red cells are markedly microcytic and show substantial variation in size and shape. Elongated red cells (pencil forms) are common. Platelets are often increased. As discussed previously, the erythrocyte protoporphyrin concentration usually is increased in iron deficiency, although values are lower than those found in severe lead poisoning.

Anemia is uncommon in lead poisoning but, when present, resembles the anemia of iron deficiency in its red cell morphology. Basophilic stippling is found in a small percentage of cases. The erythrocyte protoporphyrin is markedly elevated, and the rapid measurement of this compound helps the physician in the ED to distinguish severe lead poisoning, which requires hospitalization and intensive chelation, from iron deficiency, which usually can be treated on an outpatient basis.

Low, Normal, or Slightly Elevated Reticulocytes and Normal or Elevated Mean Corpuscular Value

With the exception of mild macrocytosis, red cell morphology usually is normal in childhood disorders of bone marrow or stem cell failure. Thrombocytopenia and neutropenia are present in aplastic anemia and Fanconi's anemia. Although the platelet count and white count occasionally may be low in patients with Diamond-Blackfan syndrome, the red cells are most severely affected. Erythropoiesis is most severely affected in TEC and acquired pure red cell aplasia, although neutropenia may accompany the former disorder.

The clinical features at the onset of acute leukemia may closely resemble those of aplastic anemia. Examination of a bone marrow aspirate is sometimes required to distinguish these disorders. This procedure rarely is performed in the ED.

Therapy, such as corticosteroids, which might interfere in the interpretation of the bone marrow aspirate, should be withheld until a definitive diagnosis has been made.

As discussed, children with hemolytic disorders may escape detection until pallor is noted during an aplastic crisis when the reticulocyte count is similar to that found in primary disorders of red cell production. An underlying hemolytic disease such as sickle cell anemia or hereditary spherocytosis usually can be recognized during anaplastic crisis, however, by finding characteristic red cells on the peripheral smear. In the autosomal dominant disorders of the red cell membrane, the presence of abnormal erythrocytes in the peripheral blood of one of the parents may support the diagnosis. Solubility tests for HbS or hemoglobin electrophoresis should be performed to detect sickling disorders.

The MCV usually is increased in megaloblastic anemias unless other nutritional disorders are present. Hypersegmentation of the polymorphonuclear leukocytes is characteristic. In severe or long-standing megaloblastic anemia, neutropenia and thrombocytopenia also may be found. In such cases, the findings in the peripheral blood may be similar to those of aplastic anemia or even acute leukemia; examination of the bone marrow and measurement of specific nutrients (B_{12}, folic acid) are necessary to distinguish these disorders.

Suggested Readings

Beutler E. The common anemias. JAMA 1988;259:2433–2437.

Nardone DA, Roth KM, Mazur DJ, McAfee JH. Usefulness of physical examination in detecting the presence or absence of anemia. Arch Intern Med 1990;150:201–204.

Oski FA, Brugnara C, Nathan DG. A diagnostic approach to the anemic patient: differential diagnosis of anemia. In: Nathan DG, Orkin SH, eds. Hematology of Infancy and Childhood. 5th ed. Philadelphia: WB Saunders, 1998:375–384.

Pearson HA. Anemia in the newborn: a diagnostic approach and challenge. Semin Perinatol 1991;15:2–8.

CHAPTER 60

Polydipsia

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Polydipsia, or excessive thirst, is an uncommon complaint in children. Although fluid consumption varies greatly among individuals, pathologic conditions exist when excessive drinking of fluids interferes with daily life or is accompanied by bizarre behavior, such as drinking from a toilet bowl. Polydipsia is routinely accompanied by polyuria (see Chapter 75). Other accompanying symptoms depend on the underlying cause.

PATHOPHYSIOLOGY

The sensation of thirst and subsequent fluid intake is influenced by complex mechanisms that involve the hypothalamus, extracranial thirst receptors, and kidneys. As water is lost from the body, thirst centers in the hypothalamus are stimulated by an increase in serum osmolality. In response to signals from the hypothalamus, the pituitary gland releases an antidiuretic hormone that causes reabsorption of water in the collecting ducts of the kidney. In addition to physiologic controls of thirst, cortical involvement and social conditioning also play a role and may be responsible for the wide variability in fluid consumption.

DIFFERENTIAL DIAGNOSIS

Diabetes mellitus (DM) is the single most common cause of polydipsia (Table 60.1). Additional prominent symptoms of DM include weight loss and polyuria. Other common causes of polydipsia include sickle cell anemia and diabetes insipidus (Table 60.2). In sickle cell anemia, the chronic sickling of cells in the medulla of the kidney results in a limited ability to concentrate urine and mild polydipsia. In diabetes insipidus, a wide variety of lesions in the hypothalamus and neurohypophysis results in a deficiency of antidiuretic hormone. Also, a rare inherited form of nephrogenic diabetes insipidus exists that may be autosomal dominant or X-linked recessive. In instances in which the cause of diabetes insipidus cannot be readily determined, patients are diagnosed as idiopathic. These patients need frequent reevaluations because many are later diagnosed with intracranial tumors.

Diabetes mellitus	Infection
Electrolyte imbalances	Aneurysm
Hypercalcemia	Intraventricular hemorrhage
Hypokalemia	Hereditary
Bartter's syndrome	Drugs
Catecholamine excess	Methylxanthines
Pheochromocytoma	Diuretics
Neuroblastoma	Lithium
Ganglioneuroma	Renal causes
Cystinosis	Renal tubular acidosis
Diabetes insipidus (antidiuretic hormone deficient)	Nephrogenic diabetes insipidus
Craniopharyngioma	Sickle cell trait
Pituitary adenoma	Sickle cell diseases
Histiocytosis	Interstitial nephritis
Head trauma	Obstructive uropathy
Sarcoidosis	Primary polydipsia
Leukemia	Psychogenic polydipsia
	Neurogenic polydipsia

Table 60.1. Causes of Polydipsia

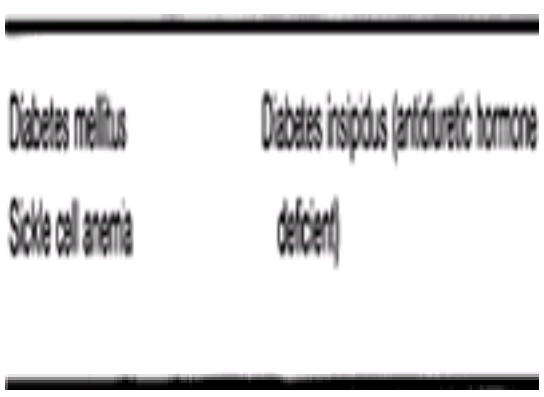

Table 60.2. Common Causes of Polydipsia

Less common metabolic and endocrine causes of polydipsia include electrolyte imbalances, catecholamine excess, and cystinosis. Primary renal causes of hyposthenuria include interstitial nephritis, renal tubular acidosis, medullary cystic disease (nephrophthisis), and obstructive uropathy. In nephrogenic diabetes insipidus, the renal tubule is unresponsive to antidiuretic hormone. Patients with nephrogenic diabetes insipidus usually have onset of symptoms in infancy and present with recurrent episodes of dehydration, fever, failure to thrive, and psychomotor retardation. Pharmacologic causes of polyuria and polydipsia include methylxanthines and diuretics. In addition, chronic lithium therapy may result in nephrogenic diabetes insipidus.

Primary polydipsia is diagnosed when the ingestion of water is in excess of that needed to maintain water balance. It can be caused by an inappropriate psychologic thirst drive (psychogenic polydipsia or compulsive water drinking) or by hypothalamic damage that alters thirst but not antidiuretic hormone release (neurogenic polydipsia).

Most children with polydipsia have serious but nonacute problems. Potential life-threatening conditions may develop, however, in certain circumstances (Table 60.3). Patients with diabetes insipidus or nephrogenic diabetes insipidus may develop severe dehydration if water is withheld for prolonged periods. Conversely, urgent management of hypernatremia is usually unnecessary if patients are able to drink and may be harmful if it is of chronic duration. Diabetic ketoacidosis may be an initial presentation of patients with DM and can result in extreme electrolyte and acid-base imbalances. Patients with primary polydipsia who overload their kidneys' ability to excrete free water may present with hyponatremic seizures. Many of the brain lesions that cause diabetes insipidus can become life-threatening. In fact, diabetes insipidus often is seen in dying patients with severe brain injury.

Diabetes insipidus (antidiuretic hormone deficient)	Diabetes mellitus
Nephrogenic diabetes insipidus	Primary polydipsia

Table 60.3. Life-Threatening Causes of Polydipsia

EVALUATION AND DECISION

When evaluating a child with polydipsia, the physician should seek information from the parent that would characterize the quantity of fluid taken each day and whether the child has used any unusual methods to satiate thirst. A history of nocturnal polydipsia and polyuria is helpful because most children with psychogenic polydipsia do not wake in the middle of the night for fluids. A medical history should include questions on growth and development and past episodes of severe dehydration. Inquiries should be made about known causes of polydipsia such as sickle cell disease, DM, chronic kidney disorders, head trauma, and medications (Fig. 60.1). The physical examination should include a careful evaluation for known systemic and intracranial causes of diabetes insipidus.

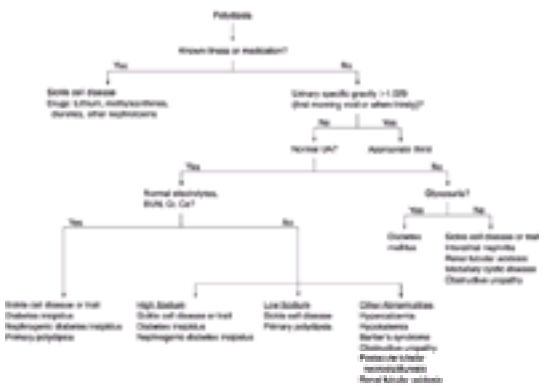

FIGURE 60.1. Diagnostic approach to a child with polyuria.

If the history and physical examination are not revealing, a urinalysis should be obtained. In almost all cases of polydipsia, the urine specific gravity will be low (less than 1.010). A specific gravity greater than 1.020 should represent appropriate thirst. If the urinalysis is abnormal, DM, sickle cell disease or trait, or an intrinsic renal disorder should be suspected. If the urinalysis is normal, electrolytes, calcium, and renal function tests may reveal conditions associated

with electrolyte imbalances. Patients with diabetes insipidus or nephrogenic diabetes insipidus may have hypernatremia if they are examined when dehydrated. A hemoglobin electrophoresis may be needed to determine whether the patient has sickle cell disease or trait. However, patients with sickle cell disease usually have the diagnosis confirmed before the development of tubular dysfunction and polydipsia. Computed tomography (CT) and magnetic resonance imaging scans may be necessary to diagnose intracranial abnormalities.

Patients suspected of having primary polydipsia, diabetes insipidus, and nephrogenic diabetes insipidus require further testing that can be dangerous. These tests should be done under controlled settings and usually are inappropriate in the emergency department.

Patients with primary polydipsia should respond to a water deprivation test by increasing their urine gravity and osmolality. Patients with diabetes insipidus and nephrogenic diabetes insipidus should have rapid weight loss while continuing to excrete urine with a low specific gravity. They may get severely dehydrated if the weight loss is in excess of 3 to 5%. Constant observation needs to be maintained during the water deprivation test to ensure that patients do not covertly consume water and to prevent severe dehydration. A trial of intranasal desmopressin (DDAVP) should distinguish between diabetes insipidus and nephrogenic diabetes insipidus because patients with antidiuretic hormone-deficient diabetes insipidus will respond to the exogenous hormone.

Unfortunately, even these tests are fraught with some inaccuracies. Patients with primary polydipsia who have chronic overhydration and diminished capacity to concentrate urine may have a blunted response to water deprivation. In addition, patients with diabetes insipidus and nephrogenic diabetes insipidus may produce a hypertonic urine if the glomerular filtration rate is decreased as severe dehydration ensues. Radioimmunoassay for antidiuretic hormone can be helpful in confusing cases.

Suggested Readings

Horev Z, Cohen AG. Compulsive water drinking in infants and young children. Clin Pediatr 1994;11:209–213.

Kern KB, Meislin HW. Diabetes insipidus: occurrence after minor head trauma. J Trauma 1984;24:69–72.

Kohn B, Norman M, Feldman H, et al. Hysterical polydipsia. Am J Dis Child 1976;130:210–212.

Robertson GL. Differential diagnosis of polyuria. Ann Rev Med 1988;39: 425–442.

Stevko RM, Balsley M, Segar WE. Primary polydipsia—compulsive water drinking. J Pediatr 1968;73:845–851.

Zerbe RL, Robertson GL. A comparison of plasma vasopressin measurements with a standard indirect test in the differential diagnosis of polyuria. N Engl J Med 1981;305:1539–1546.

CHAPTER 61

Rash—Eczematous

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Pediatricians often use the terms *eczema* and *atopic dermatitis* interchangeably. To dermatologists, *eczema*, which means “to boil over,” is synonymous with *dermatitis*. Eczema is used to describe a complex of signs and symptoms, including erythema, edema, vesiculation, scaling, and pruritus. This broader use of the term is more appropriate because many disease processes and exposures may cause eczematous rashes (see also Chapter 99). The distinct forms of eczema are labeled by cause, pattern, or associated conditions such as atopy. Physical findings may reflect acute or chronic changes. Acute eczema consists of erythema, edema, exudation, clustered papulovesicles, scaling, and crusting. Chronic eczema is characterized by lichenification (thickened skin with accentuated skin markings), hyperpigmentation or hypopigmentation, and signs of excoriation. Histologic examination usually does not suggest a specific type of dermatitis. Classification and diagnosis of a particular process relies on the history and the pattern of the eczematous process.

DIFFERENTIAL DIAGNOSIS

Atopic Dermatitis

Atopic dermatitis (see Chapter 99) is by far the most common cause of an eczematous rash in children (Table 61.1 and Table 61.2). It is a chronic or relapsing condition characterized by pruritic eczematous eruptions and occurs in up to 10% of the population. Affected persons usually have a personal or family history of allergic rhinitis, hay fever, or asthma. One-half to three-quarters of patients have the onset of symptoms before age 6 months, with up to 90% developing symptoms by 5 years of age. Dry conditions (e.g., the dry weather of winter) or the drying caused by frequent bathing often lead to exacerbations. Stress, sweating, and exposure to environmental allergens (e.g., pollens or dust mite antigen) may also precipitate flares in some individuals. Some infants and young children experience worsening symptoms with exposure to certain foods (e.g., milk protein, peanuts, eggs). A United Kingdom Working Party has established diagnostic criteria for atopic dermatitis (Table 61.3).

Atopic dermatitis	Autoeczematization (Id)
Contact dermatitis	Exfoliative dermatitis*
Allergic	Photoallergic reactions
Irritant	Dermatophyte infections
Nummular eczema	Scabies
Asteotxtic eczema	Molluscum contagiosum
Dyshidrotic eczema	Eczematous rashes associated with
Seborrheic dermatitis	systemic illnesses
Lichen simplex chronicus	

*Condition that is potentially life-threatening.

Table 61.1. Differential Diagnosis of Eczematous Rash

Atopic dermatitis	Dermatophyte infections
Contact dermatitis	Scabies
Allergic	Molluscum contagiosum
Irritant	Pityriasis rosea

Table 61.2. Common Causes of Eczematous Rash

The diagnosis of atopic dermatitis is established when a history of an itchy skin condition exists and at least three of the following criteria are met: • History of involvement of the skin creases such as folds of elbows, behind the knees, front of ankles, or around the neck (including cheeks in children under 10 years of age) • A personal history of asthma or hay fever (or family history of atopic disease in children under 4 years of age) • A history of general dry skin in the last year • Visible flexural eczema (or eczema of the cheeks/forehead and outer limbs in children under 4 years of age) • Onset before the age of 2 years (not used if the child is less than 4 years of age)

Table 61.3. UK Working Party Diagnostic Criteria for Atopic Dermatitis

With acute flares of the disease, lesions are poorly demarcated, erythematous, scaly, and often weepy and crusted. Chronic lesions are poorly defined, thickened, hyperpigmented, and often excoriated. Distribution varies by age. Infants have lesions on the cheeks, trunk, diaper area, and extensor surfaces of the extremities. Children show involvement of the feet and flexor areas such as the antecubital and popliteal fossae and the neck. In adolescents and adults, flexor areas, hands, and feet are usually involved. Xerosis (dry skin), ichthyosis vulgaris (inherited fishlike scaling), keratosis pilaris (chicken-skin appearance caused by cornified plugs in the upper hair follicles), infraorbital eyelid folds (Dennie-Morgan sign), hyperlinear palms, pityriasis alba (scaly hypopigmented patches), and follicular accentuation may be seen in some individuals. Superimposed bacterial (*Staphylococcus aureus* or group A streptococcus), fungal, and viral infections (eczema herpeticum caused by herpes simplex virus) are common and may produce severe exacerbations and associated systemic signs.

Particularly severe or persistent symptoms should prompt the clinician to consider an underlying systemic disorder associated with eczematous eruptions (see the following).

Contact Dermatitis

Contact dermatitis (see Chapter 99) is an inflammatory reaction of the skin caused by an allergic stimulus or primary irritant. The reaction may be of an acute, subacute, or chronic nature. Acute eruptions have intense pruritus, severe erythema, edema, vesicles, and erosions with serous discharge and crusting. A sharp demarcation between involved and unaffected skin usually exists. Subacute reactions have mild erythema, dry scale, less vesiculation, and mild thickening of the skin. Chronic exposures may result in lichenification, fissures, scales, excoriations, and hyperpigmentation. Vesicles are rare.

Allergic Contact Dermatitis

Allergic contact dermatitis is caused by a classic delayed hypersensitivity reaction (type IV). Repeated exposure to the inciting substance causes an allergic sensitization. The eruption is delayed after the initial exposure for up to 7 to 10 days. Repeated exposures can cause the rapid appearance of an acute dermatitis (within 12 hours). Rhus dermatitis, caused by an oleoresin in the sap of poison ivy, poison oak, or poison sumac plants, is the most common cause of allergic contact dermatitis in the United States. The allergen is found in the leaves, roots, and twigs of the plants. Delayed exposure may occur because of contact with clothing, gloves, tools, or even pets that have had contact with the plants. Burning of plants leads to aerosolization of the allergen and may cause a widespread and severe outbreak on exposed skin surfaces. Other plants, flowers, pollens (especially ragweed), clothing, shoes, metals (e.g., nickel in jewelry), cosmetics, adhesive tape, and latex-containing products can also cause an allergic contact dermatitis.

Allergic contact dermatitis is rare in infants because of their impaired ability to react to allergens. By age 3 to 8 years, children react to allergens in a fashion similar to adults. The distribution, shape, and pattern of the rash, as well as a history of possible exposures, may elucidate the cause. Linear eruptions are usually seen, especially with plant contact. Shoe dermatitis is likely to cause an eruption limited to the dorsal toes and instep of the foot. Airborne processes (e.g., smoke containing rhus oleoresin) cause a problem on exposed surfaces, including eyelids, whereas, a photoallergic contact dermatitis involves sun-exposed areas (e.g., rash resulting from use of a sunscreen that contains para-aminobenzoic acid).

Irritant Contact Dermatitis

A primary irritant dermatitis is a nonallergic reaction of the skin caused by a single exposure or a series of brief contacts with an irritating substance. Strong soaps and detergents, saliva, urine, stool contents, fiberglass particles, and bubble baths are common causes in children. Occlusive diapers that promote prolonged exposure of the skin to urine and feces are a major contributor to diaper dermatitis (see Chapter 99).

Nummular Eczema

The term *nummular eczema* is derived from the Latin word for coin. Coin-shaped plaques that are erythematous and contain tiny vesicles, crusts, and at times, excoriations manifest this chronic condition. Lesions usually occur on the extensor surfaces of the hands, arms, and legs. They may be single or multiple and are often symmetric in distribution. Nummular eczema seems to be related to dry skin and irritation rather than to atopy but can be seen in atopic individuals. Central clearing of the lesions may result in an appearance similar to dermatophyte infections. Because of their round shape, lesions may be mistaken for impetigo or granuloma annulare.

Asteatotic Eczema

Asteatotic eczema, also called winter eczema, xerotic eczema, and eczema cracquele, is a pruritic condition in which the skin is dry and cracked with red fissures and some scale. The skin has the appearance of cracked porcelain. The most common sites are the extensor legs, dorsal hands, and extensor forearms. The condition tends to occur in adolescents and older persons during the winter and is associated with overbathing and low humidity.

Dyshidrotic Eczema

Dyshidrotic eczema, also called pompholyx, involves the hands and feet. Patients develop the sudden onset of pruritic, tiny, clustered, deep-seated vesicles that look like tapioca. When the condition persists, scaling, lichenification, and painful fissures occur. Lesions appear on the palms, soles, and lateral fingers. The process may be acute, chronic, or persistent and may be provoked by stress. It is associated with hyperhidrosis, although no clear evidence supports that sweating plays a role in the pathogenesis. Approximately 50% of patients have an atopic background. When it involves the feet, it may be confused with tinea pedis.

Seborrheic Dermatitis

Seborrheic dermatitis (see Chapter 99) is a problem of infants, adolescents, and adults and is characterized by nonpruritic, erythematous, greasy, yellow or salmon-colored plaques in regions of the body that have high concentrations of sebaceous glands. These “seborrheic” regions include the scalp, face (nasolabial folds, eyebrows, eyelids, sideburns, beard), postauricular areas, axilla, groin, and presternal area. The scalp (cradle cap) or diaper area is usually involved first in infants between 2 and 12 weeks of age. The rash may spread to the face, trunk, and neck. It usually clears by 8 to 12 months of age and then recurs after the onset of puberty. Although common in infancy, only about 10% of patients will have recurrence in later life. When the rash is particularly severe, associated with petechiae or systemic signs or symptoms, or particularly recalcitrant to the usual therapies in infancy, an underlying systemic illness or immunodeficiency should be considered (see the following). Severe seborrheic dermatitis has been reported as an early sign of acquired immunodeficiency syndrome (AIDS) in adolescents infected with human immunodeficiency virus (HIV).

Lichen Simplex Chronicus

Lichen simplex chronicus, previously called circumscribed neurodermatitis, refers to a chronic, localized lesion that results from repeated rubbing and scratching. It may occur in any location but has a predilection for the sites that are easily reached such as the arms, legs, ankles, neck, and the anogenital area. It is rare in young children but fairly common in adolescents and adults. It may occur in a preexisting area affected by atopic, seborrheic, or contact dermatitis or psoriasis. Typical lesions are single or multiple oval plaques from 5 to 15 cm in size. The skin is reddened and slightly edematous. Chronic lesions usually consist of well-demarcated areas of dry, thickened, scaly, hyperpigmented or hypopigmented plaques. Marked pruritus occurs.

Autoeczematization

Autoeczematization occurs in the presence of an initial active eczematous rash, such as an allergic contact dermatitis, an irritant dermatitis, or a stasis dermatitis. The patient later develops a more extensive eczematous eruption as a result of autosensitization or autoeczematization. A specialized form of this process is seen with dermatophyte infection, in particular tinea capitis, and is called a dermatophytid or *id reaction.*

Photoallergic Reactions

Photoallergic reactions may occur after systemic or topical administration of various drugs or chemicals that absorb radiant energy, primarily in the ultraviolet A (UVA) range. These reactions may manifest as acute, subacute, or chronic dermatitis in sun-exposed areas. Common agents implicated include phenothiazines, sulfonamides, thiazides, sunscreen components such as para-aminobenzoic acid (PABA), and some fragrances.

Infectious Causes of Eczematous Rashes

Eczematous rashes can result from primary skin infections caused by multiple organisms. Fungi, viruses, and parasites

are the most common causes in children. Pityriasis rosea is thought to result from a viral infection.

Dermatophyte Infections

Skin infection caused by a dermatophyte, also called tinea or ringworm, may cause eczematous lesions. The typical lesion of tinea corporis is an annular, erythematous, scaling plaque. These pruritic, circular plaques may be clinically indistinguishable from nummular eczema. A raised vesicular or pustular border may lead to a correct diagnosis of tinea corporis. Likewise, tinea pedis and tinea manuum may be eczematous and vesicular and may mimic dyshidrotic eczema of the feet or hands. Tinea pedis and tinea manuum certainly do occur in prepubertal children, although not as commonly as in adults. Tinea capitis classically presents with scaly, discrete patches of hair loss with black dot hairs and occipital adenopathy. However, it may appear identical to seborrheic dermatitis of the scalp (Fig. 61.1) with greasy yellow scale and should be ruled out before making the diagnosis of seborrhea of the scalp, especially in any child under age 12 who has a scaly scalp.

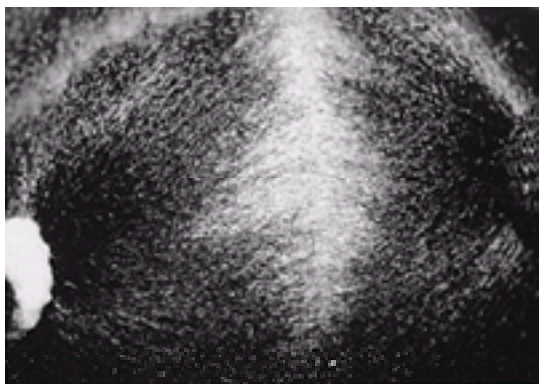

FIGURE 61.1. Seborrheic dermatitis-like tinea capitis in a child.

Scabies

The eruption of scabies (see Chapter 99) is polymorphic with papules, vesicles, nodules, excoriations, crusts, and eczematous plaques. Only a small percentage of patients have the classically described linear tracts or burrows. A particularly severe form of this infestation is termed Norwegian scabies and is characterized by heavy crusting and hyperkeratosis. This is usually seen in immunosuppressed patients. Infants, with their immature immune systems, often have similarly severe infestations; the severity may be related to a delay in diagnosis of scabies in infants and the use of topical steroids in a mistaken attempt to treat an atopic process.

Infants and young children tend to have lesions on the palms, soles, face, and scalp. The lesions may become generalized. Older children and adults are more likely to have involvement of the fingerwebs, flexural regions, breasts, and genital area. Visualization of the mites from skin scrapings under low power on the microscope may confirm this diagnosis; however, a high false-negative rate exists.

Molluscum Contagiosum

The characteristic lesion of molluscum (see Chapter 99) is not eczematous, but rather, is a dome-shaped, waxy papule with a central umbilicated plug. Often, patients develop a surrounding area of dermatitis that may represent a delayed hypersensitivity reaction to a viral antigen. At times, the distinctive papules are barely noticeable within larger eczematous plaques.

Pityriasis Rosea

Pityriasis rosea (see Chapter 99) is common eruption that occurs in epidemics primarily in the spring and autumn. Patients may develop the characteristic herald patch first, then an extensive papulosquamous eruption primarily on the trunk. Lesions may have a “Christmas tree” distribution on the back. In African-American children, an “inverse distribution” of the lesions with involvement of the proximal extremities, inguinal and axillary areas, and the neck is often present, whereas the trunk is relatively spared. Adolescent patients may exhibit a similar and indistinguishable rash as a manifestation of secondary syphilis (see Chapter 84).

Diaper Dermatitis

Diaper dermatitis (see Chapter 99) is possibly the most common cutaneous problem of infancy and young childhood. However, it is not a specific diagnosis, but rather, a group of disorders provoked by the moist, occluded, irritated environment of the diaper region (Table 61.4). Often, a combination of the disorders exists in one patient. The following sections represent the various types of diaper dermatitis.

Irritant dermatitis	Psoriasis
Friction dermatitis	Seborrheic dermatitis
Intertrigo	Letterer-Siwe disease
Candida	Acrodermatitis enteropathica

Table 61.4. Causes of Diaper Dermatitis

Irritant Contact Dermatitis

Irritation from stool and urine, chemicals, soaps, heat, moisture, and sweating may lead to erythematous plaques with minimal scale on the buttocks, perineum, and lower abdomen with sparing of the creases.

Friction Dermatitis

Friction dermatitis is seen on the inner thighs, genitals, buttocks, and abdomen as a mild erythema with a shiny surface caused by chafing in areas of friction.

Intertrigo

Intertrigo results from rubbing and irritation by diapers in a hot climate or from wearing excessive clothing. Erythematous, macerated, exudative plaques appear in the inguinal and intergluteal folds.

Candida

Monilial diaper dermatitis is characterized by beefy red plaques with a fine white scale along the periphery and pinpoint satellite papules and pustules (Fig. 61.2). Candidal diaper dermatitis usually begins in the perianal area. It spreads to involve the perineum and inguinal folds and, in severe cases, extends to the buttocks, upper thighs, lower abdomen, and lower back.

FIGURE 61.2. The erythematous plaques and satellite papules of candidiasis.

Psoriasis

Psoriatic diaper rashes demonstrate bright red, well-demarcated plaques, often with dry, silvery scales (Fig. 61.3). A family history of psoriasis and further cutaneous evidence of psoriasis, such as scalp involvement, nail dystrophy or pitting, intergluteal erythema, and postauricular erythema, is often present. In the diaper area, psoriasis probably flares because of the isomorphic response in which psoriasis appears in sites of epidermal injury.

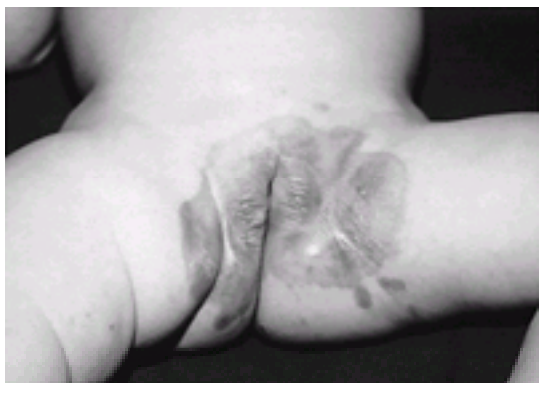

FIGURE 61.3. Diaper area psoriasis with well-demarcated bright red plaques.

Seborrheic Dermatitis

When the diaper area is involved with seborrheic dermatitis (see Chapter 99), erythema and a greasy yellow scale are seen, especially in the creases. Similar lesions in the axilla and scalp are useful signs to aid in diagnosis.

Letterer-Siwe Disease

Letterer-Siwe disease, the infantile form of Langerhans' cell histiocytosis, often presents with a diaper rash. It is seborrheic in distribution with yellow crusted plaques, infiltrative and hemorrhagic papules, and vesicles.

Acrodermatitis Enteropathica

Acrodermatitis enteropathica, resulting from an inherited problem with zinc absorption, demonstrates bright red plaques in periorificial regions, including the diaper area. The plaques have a serpiginous, erosive border and look much like candida or psoriasis. Patients have associated alopecia, diarrhea, and failure to thrive. A similar eruption has been reported in some infants with cystic fibrosis.

Eczematous Rashes Associated with Systemic Illnesses

Several systemic illnesses may include eczematous rashes as one of their manifestations (Table 61.5). When a patient presents with an eczematous rash that is particularly recalcitrant to treatment, especially severe, or associated with systemic signs or symptoms, these diagnoses should be considered. A patient with severe atopic disease or seborrhea in the absence of a family history may be manifesting a more serious process. Fever, failure to thrive, diarrhea, and recurrent infections may be clues to the presence of an underlying process.

Exfoliative dermatitis	Hyperimmunoglobulinemia E syndrome
HIV infection	Letterer-Siwe disease
Wiskott-Aldrich syndrome	

Table 61.5. Systemic Illnesses Associated with Eczematous Rashes

Exfoliative Dermatitis

Exfoliative dermatitis, or exfoliative erythroderma, is an inflammatory condition of the skin in which generalized erythema and scaling exist. It may be idiopathic or a manifestation of underlying dermatologic or systemic disease (Table 61.6). Medication reactions may also produce a severe bullous disorder: toxic epidermal necrolysis (TEN), in which entire regions of the epidermis may become denuded (see Chapter 99). Some of the more common types of dermatitis (atopic, contact, seborrheic) may cause an exfoliative dermatitis, as well as the more unusual dermatoses such as pityriasis rubra pilaris (a diffuse, salmon-colored papulosquamous eruption) and pemphigus foliaceus (a superficial blistering disorder). When exfoliative dermatitis is seen in an infant, an immune deficiency should be excluded (Fig. 61.4). These immunocompromised infants often have diarrhea, recurrent infections, and failure to thrive. They may have various immune problems, including defective yeast opsonization, impaired neutrophil mobility, elevated serum IgE, and hypogammaglobulinemia. In the past, the term *Leiner's syndrome* was used to describe many of these cases. Exfoliative erythroderma may be the cutaneous presentation of other immune disorders such as Wiskott-Aldrich syndrome, Omenn's syndrome, severe combined immunodeficiency syndrome (SCIDS), HIV infection, and graft-versus-host disease. An exfoliative dermatitis is the only eczematous process that may be potentially life-threatening in the acute phase because of temperature instability, fluid losses through the skin, high-output heart failure, pneumonia, and sepsis.

Atopic dermatitis
Seborrheic dermatitis
Contact dermatitis
Psoriasis
Pityriasis rubra pilaris
Pemphigus foliaceus
Inherited ichthyoses
Cutaneous T cell lymphoma
Lymphoma and leukemia
Medications (sulfonamides, penicillins, cephalosporins, phenytoin, barbiturates)
Immunodeficiency (graft-versus-host disease, Ormenn's syndrome, Wiskott-Aldrich syndrome, severe combined immunodeficiency syndrome, agammaglobulinemia, human immunodeficiency virus infection)
Letterer-Siwe disease
Phenylketonuria

Table 61.6. Causes of Exfoliative Dermatitis

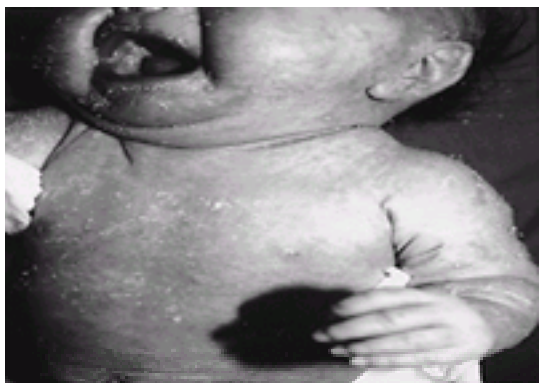

FIGURE 61.4. Infant with exfoliative erythroderma and immunodeficiency.

HIV Infection

About half of children with HIV infection have a “seborrheic looking” dermatitis. Unlike seborrhea, this often persists past age 6 months. Severe, atopic-appearing rashes may also be seen. The diaper area is the most often affected. Children with HIV infection may also have recurrent bacterial infections, diarrhea, failure to thrive, lymphadenopathy, and hepatosplenomegaly, as well as developmental delay. In addition, dermatitis resulting from infectious causes (e.g., bacterial infections, molluscum contagiosum, scabies) may be particularly severe or difficult to treat.

Wiskott-Aldrich Syndrome

Wiskott-Aldrich syndrome is an X-linked, recessive disorder in which children have marked thrombocytopenia, eczematous rashes, and immunodeficiency. These patients often develop petechial or purpuric rashes. They have recurrent infections, often caused by encapsulated organisms. Patients may also have autoimmune disorders. Eczematous rashes are usually severe and recalcitrant to therapy.

Hyperimmunoglobulin E (IgE) Syndrome

Hyperimmunoglobulin E syndrome, or Job's syndrome, is characterized by extremely high IgE levels, repeated cutaneous infections, and chronic dermatitis. The rash usually resembles atopic dermatitis. A personal or family history of atopy usually exists, as well as recurrent skin infections (usually staphylococcal or streptococcal), extreme elevation of the serum IgE, impaired neutrophil chemotaxis, and peripheral blood eosinophilia. A subset of patients, usually women, have a tendency to develop large, cold, chronic, and recurrent staphylococcal abscesses of skin and bone, which cause severe scarring and deformity.

Letterer-Siwe Disease

Letterer-Siwe disease, the infantile form of histiocytic disease, often presents as a seborrheic-appearing rash in the diaper area, scalp, postauricular, and axillary regions. The eruption is usually more severe than usual cases of seborrhea, is resistant to therapy, and tends to recur. Cutaneous nodules and purpura suggest this diagnosis. Hepatosplenomegaly, lymphadenopathy, anemia, thrombocytopenia, and osseous lesions are associated findings.

EVALUATION AND DECISION

The most important points in reaching an accurate diagnosis of an eczematous rash include the distribution of the lesions, the patient's age, and the duration of the disease.

Generalized

If a patient presents with a generalized eczematous process, literally red and scaly from head to toe, this suggests an exfoliative dermatitis or erythroderma (Fig. 61.4 and Fig. 61.5B). This condition is unusual in children and has multiple causes. It can be a manifestation of an underlying dermatologic process, a drug reaction, or a systemic illness. Skin biopsy is often necessary to distinguish the cause. If erythroderma is present in infancy, an immune dysfunction should

be considered, especially if the patient has diarrhea, recurrent infections, or failure to thrive.

FIGURE 61.5. A. Diagnostic approach to the child with an eczematous process. **B.** Diagnostic approach to the child with an exfoliative dermatitis.

Extensive but Not Generalized

Eruptions that may be extensive but with some areas of noninvolved skin include atopic dermatitis, seborrheic dermatitis, scabies, autoeczematization reactions, pityriasis rosea, or contact dermatitis (Fig. 61.5A). A family or personal history of atopy, a history of flares and remittance, extreme pruritus, as well as a distribution compatible with the patient's age may lead toward a diagnosis of atopic dermatitis. No specific laboratory tests aid in diagnosis. The diagnosis of seborrheic dermatitis would be made only in infancy and after puberty. Features that help distinguish seborrhea from atopic dermatitis in infancy include minimal pruritus; salmon-colored, greasy plaques; and predominant involvement of the scalp and intertriginous regions. Scabies may look much like atopic dermatitis. However, a history of acute onset, other family members or close contacts with recent onset of a pruritic eruption, and evidence of a polymorphous eruption might aid in this diagnosis. Occasionally, children with scabies have a chronic rash that has been diagnosed as atopic dermatitis, without a history of rash in contacts. One should always be suspicious of scabies, especially if a child aged 3 years or older presents with the recent onset of an eczematous, pruritic rash. Id reactions can be extensive. A history of an initial contact dermatitis or scaly scalp (tinea capitis) followed by a widespread eczematous rash suggests an autoeczematization reaction. Id reactions tend to be worse in areas near the initial rash. An allergic or irritant contact dermatitis could be extensive, depending on the exposure. Eruptions that are unusually severe, persistent, recalcitrant to treatment, or associated with systemic signs should prompt consideration of an underlying systemic illness.

Localized

If an eczematous process is localized to one or a few areas, contact dermatitis, nummular dermatitis, asteatotic eczema, lichen simplex chronicus, photoallergy, scabies, molluscum, or a dermatophyte infection should be considered. The diagnosis of a contact dermatitis is made based on the appearance and distribution of the dermatitis and aided by a history of contact with an allergen. It is often difficult for patients to determine the allergen because of the delayed onset. Therefore, it is important for the physician to suggest some of the more common agents such as poison ivy and nickel. Linear lesions from a plant rubbing against the skin suggest poison ivy exposure. Patch testing may be useful if the allergen is unclear. Photoallergy is suggested by a dermatitis in sun-exposed areas. A raised, slightly vesicular, scaly border implies tinea corporis. However, it is often helpful to perform a potassium hydroxide (KOH) preparation with all these conditions because tinea corporis may mimic any of them. A chronic, pruritic, lichenified plaque suggests lichen simplex chronicus, although an irritant or allergen could play a role in this condition. Eczema within the setting of dry skin, especially on the extensor extremities, suggests asteatotic eczema. In a patient with onset of localized eczematous plaques, one should closely inspect the skin for the tiny, dome-shaped papules of molluscum contagiosum.

Scalp

When the physician evaluates a child with scaly scalp, the child's age and character of the scale can be most helpful (Fig. 61.1 and Fig. 61.6). Newborns often have cradle cap, which can be a manifestation of seborrheic dermatitis, psoriasis, or rarely, atopic dermatitis. It is important to look carefully at the remainder of the infant's skin to see if greasy yellow or bright red plaques are present elsewhere. Cradle cap often persists for several months and may last up to 2 years. In any child with a scaly scalp between 2 years of age and puberty, tinea capitis must be excluded, even in the absence of hair loss. In teenagers, tinea capitis is less likely and seborrhea is again possible. Psoriasis can occur at any age and is characterized by a thick yellow scale that adheres to the hair.

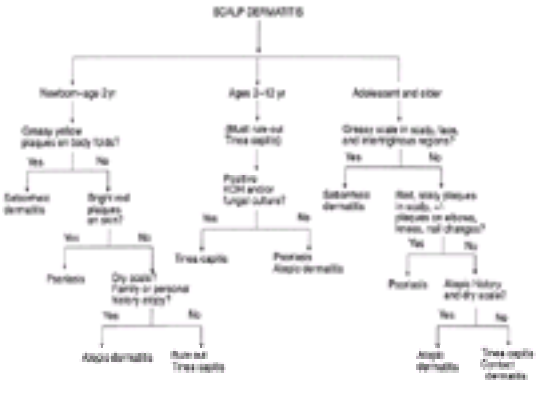

FIGURE 61.6. Diagnostic approach to the child with a scaly scalp.

Hands and Feet

The possibilities for diagnosis when an eczematous process involves the hands and/or feet are psoriasis, dyshidrotic eczema, contact dermatitis, or tinea pedis/manuum. A thorough family history, history of exposure, and a KOH preparation all may be useful.

Diaper Area

In evaluating diaper dermatitis, it is important to pay close attention to the morphology of the lesions (Fig. 61.3 and Fig. 61.7). A bright red, well-demarcated plaque with some scale could be psoriasis, candida, or acrodermatitis enteropathica. One should look at the rest of the skin to see if other sites are involved that suggest psoriasis. A KOH preparation or Gram stain may be useful to look for the spores and pseudohyphae of monilial infection. Hair loss, diarrhea, and a perioral eruption could mean acrodermatitis enteropathica, which could be confirmed by performing serum zinc levels. The most important diagnosis to exclude is Letterer-Siwe disease (Fig. 61.8). A persistent, erosive, or hemorrhagic diaper rash that is unresponsive to treatment is suspicious. One should examine for hepatosplenomegaly, scalp, and gingival involvement. If the rash is more subtle, pink, and scaly, it could be caused by irritation from urine or stool or by friction. Intertrigo would involve the folds and could be macerated or infected secondarily. Greasy, yellow plaques with cradle cap suggest seborrheic dermatitis.

FIGURE 61.7. Diagnostic approach to diaper dermatitis.

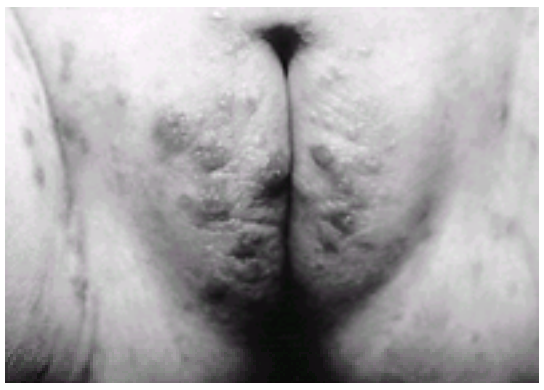

FIGURE 61.8. Persistent vesicles and nodules in a patient with Letterer-Siwe disease.

Suggested Readings

Camassa F, Fania M, Ditano G, et al. Neonatal scabies. Cutis 1995;56: 210–212.

Ghali FE, Steinberg JB, Tunnessen WW. Acrodermatitis enteropathica-like rash in cystic fibrosis. Arch Pediatr Adolesc Med 1996;150:99–100.

Hogan PA. Atopic dermatitis. Med Aust J 1996;164:736–741.

Hoppe JE. Treatment of oropharyngeal candidiasis and candidal diaper dermatitis in neonates and infants: review and reappraisal. Pediatr Infect Dis J 1997;16:885–894.

Hurwitz S. Clinical Pediatric Dermatology. Philadelphia: WB Saunders, 1981.

Kolmer HI, Platts-Mills TAE. Atopic dermatitis: new knowledge and new approaches. Hosp Pract (Off Ed) 1995;30:63–72.

Mimouni K, Mukamel M, Zeharia A, et al. Prognosis of infantile seborrheic dermatitis. J Pediatr 1995;127:744–746.

O'Brien JM. Common skin problems of infancy, childhood, and adolescence. Primary Care 1995;22:99–115.

Sampson HA, McCaskill CC. Food hypersensitivity and atopic dermatitis: evaluation of 118 patients. J Pediatr 1985;107:669–675.

Weiss SJ, Schuval SJ, Bonagura VR. Eczema and thrombocytopenia in an 8-month old infant boy. Ann Allergy Asthma Immun 1997;78: 179–182.

Weston WL, Lane AT, Morelli JG. Color Textbook of Pediatric Dermatology. 2nd ed. St. Louis: Mosby, 1996.

Williams HC, Burney PGJ, Pembroke AC, et al. The UK Working Party's diagnostic criteria for atopic dermatitis. III. Independent hospital evaluation. Br J Dermatol 1994;131:406–416.

CHAPTER 62

Rash—Maculopapular

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Maculopapular rashes are common in pediatric practice, and children often present to the emergency department (ED) for their evaluation and treatment. Before beginning a discussion of specific causes of maculopapular rashes, it is convenient to define the clinical characteristics of these rashes. A *papule* is a small, solid, mostly elevated lesion that is usually smaller then 1 cm in diameter. *Macules* are circumscribed flat lesions that differ from surrounding skin because of their color. Both papules and macules may have any size, shape, or color. Commonly, a rash may have papular and macular components, which leads to the term *maculopapular rash.*

The causes of maculopapular rashes are diverse (Table 62.1) and range from benign to life-threatening (Table 62.2). Common causes include viral exanthems, contact dermatitis, insect bites, and scabies (Table 62.3). The diagnostic approach to these disorders is based on the presence or absence of fever, characteristic clinical appearance, location, and chronicity (Fig. 62.1). Some of these conditions have very characteristic clinical appearances (Table 62.4); however, manifestations of these illnesses can be sufficiently variable that a proportion of the cases are difficult to diagnose.

Infectious	*Fungal*
Viral	Tinea versicolor
Roseola infantum	*Other Infections*
Rubeola	Rocky Mountain spotted fever
Rubella	Mycoplasma (15% of cases)
Erythema infectiosum (fifth disease)	*Etiology Uncertain but Thought to Be Viral*
Varicella (early manifestations before bullae)	Pityriasis rosea
Epstein-Barr virus (10–15% of cases have macular or maculopapular rash)	Kawasaki disease
Molluscum contagiosum (papules)	Papular acrodermatitis
Dengue	**Noninfectious**
"Nonspecific" viral	*Bites and Infestations*
Enterovirus	Insect bites
Echovirus	Scabies
Coxsackie virus	*Miscellaneous*
Adenovirus	Drug reaction
Bacterial	Allergic contact dermatitis
Scarlet fever	Irritant contact dermatitis
Syphilis	Papular urticaria
Disseminated gonorrhea	Erythema multiforme
	Guttate psoriasis
	Pityriasis lichenoides
	Lichen nitidus

Table 62.1. Maculopapular Rash: Etiologic Classification

Rocky Mountain spotted fever	Dengue fever
Kawasaki disease	Rubeola
Erythema multiforme	

Table 62.2. Potentially Life-Threatening Illnesses Associated with Maculopapular Rash

Generalized Rash	Scarlet fever
Nonspecific viral disease	Pityriasis rosea
Enteroviruses	*Localized Rash*
Adenoviruses	Contact dermatitis
Roseola infantum	Irritant dermatitis
Erythema infectiosum (fifth disease)	Scabies
Hand-foot-mouth disease	

Table 62.3. Common Disorders Associated with Maculopapular Rash

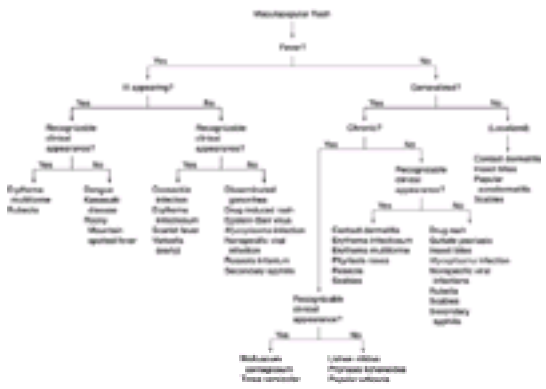

FIGURE 62.1. Diagnostic approach to maculopapular rash.

Rubeola	Tinea versicolor
Erythema infectiosum (fifth disease)	Pityriasis rosea
Hand-foot-mouth disease (coxsackie virus A 16)	Roseola infantum
Molluscum contagiosum	Insect bites
Scarlet fever	Erythema multiforme

Table 62.4. Maculopapular Rashes That Often Have Characteristic Clinical Appearances

DIFFERENTIAL DIAGNOSIS

Presence of Fever

The potentially life-threatening maculopapular rashes (Table 62.2) are all acute illnesses most commonly associated with fever and significant systemic symptoms. Hence, most patients with these illnesses will appear toxic. Erythema multiforme and rubeola have recognizable clinical appearances, whereas Kawasaki disease, Rocky Mountain spotted fever, and dengue fever require a high level of clinical suspicion. Other, less severe, febrile illnesses associated with maculopapular rashes are listed in Figure 62.1.

Potentially Life-Threatening Illnesses

Erythema Multiforme

EM is believed to result from an immune-mediated acute hypersensitivity reaction to exposure to a sensitizing antigen (see Chapter 99). Common offenders include drugs, especially trimethoprim–sulfasoxazole, cefaclor, and phenytoin; foods, especially nuts and shellfish; and infections by any number of viral, bacterial, protozoal, or fungal organisms. Herpetic and *Mycoplasma pneumoniae* infections rank among the most common infectious causes.

The rash of EM is characterized by diffuse erythematous macules with central clearing often called a target or iris lesion. Lesions may also include erythematous papules, macules, urticarial raised lesions, vesicles, and/or bullae. The distribution is most commonly symmetric and may be noted anywhere on the body with a predilection for the hands and feet, including palms and soles. Lesions may appear in isolation or as a more confluent rash. Based on severity, patients are classified as having EM minor or EM major/Stevens-Johnson syndrome. EM minor is characterized by cutaneous skin involvement alone or mucosal involvement that is limited to one surface (usually the mouth) and minimal systemic symptoms. EM major is characterized by extensive skin and mucosal involvement associated with significant systemic symptoms, including fever, chills, and malaise. Skin involvement can progress to sloughing with significant extravascular

fluid losses. Conjunctivitis and keratitis are a common feature of the major form and can lead to permanent corneal scarring. Pulmonary, cardiac, and renal involvement may occur in especially severe cases.

Treatment is predominantly supportive. Potentially inciting drugs should be immediately discontinued. If pruritus is a feature, antihistamines may provide some relief. Oral topical applications of 1:1 mixtures of diphenhydramine: Kaopectate may provide pain relief from oral involvement. For more severe cases of the disease, patients may require fluid support and/or narcotic oral pain medications. Systemic steroids in the dosage range of 1 to 2 mg/kg per day are of unproven benefit. Patients with ocular involvement should undergo ophthalmologic examination.

Kawasaki Disease

Kawasaki disease is a well-described illness of unknown cause assumed to be infectious in origin because of its epidemiologic and clinical presentation (see Chapter 101). The diagnosis is based on an unremitting fever of at least 5 days' duration and four of the five following features: 1) rash; 2) nonexudative bulbar conjunctivitis with limbal sparing; 3) red cracked lips, strawberry tongue, and erythematous oropharynx; 4) erythema, swelling, and/or induration of peripheral extremities; and 5) a solitary unilateral cervical lymph node of greater than 1.5 cm diameter.

The most commonly associated rash is a generalized pruritic urticarialike exanthem with raised erythematous plaques; however, the rash may also present with an erythematous maculopapular, morbilliform, scarlatiniform, or erythema marginatumlike pattern. The exanthem may be fleeting or persist for 2 to 3 days. During the later stages of the acute phase, periungual desquamation and peeling of the palms, soles, or perineal area develops. Laboratory tests often show a persistently elevated erythrocyte sedimentation rate (ESR) and a markedly elevated platelet count (greater than 750,000/mm^3). The acute phase is usually self-limited but requires accurate diagnosis to prevent the development of coronary artery aneurysms that occur in approximately 20% of cases without therapy. Patients with fever and fewer than four of the previously listed clinical features can be diagnosed with atypical Kawasaki disease if coronary artery disease is detected. Of those patients who develop aneurysms, a small percentage go on to develop heart failure, valvular regurgitation, or myocardial infarction.

No specific laboratory tests are available for diagnosis. Therapy consists of anti-inflammatory agents, specifically high-dose intravenous immunoglobulin (IVIG) and aspirin. IVIG and aspirin initiated within 10 days of the onset of fever decrease the prevalence of coronary artery aneurysms. Therapy should be initiated as soon as the diagnosis is strongly suspected, even if begun more than 10 days after onset of illness. A dosage schedule of 2 g/kg as a single dose of IVIG is recommended. Aspirin is given initially at a high dosage (80 to 100 mg/kg per day divided four times daily) for its anti-inflammatory effect. It can be decreased to 3 to 5 mg/kg per day in one dose after fever has subsided to reduce the likelihood of spontaneous coronary thrombosis. Aspirin is usually continued in patients without evidence of coronary aneurysms until the platelet count and ESR return to normal and may be continued indefinitely for those with coronary artery abnormalities. Echocardiograms should be obtained initially and then at 3 and 8 weeks after onset of illness.

Measles (Rubeola)

Measles was one of the most common viral exanthems before the advent of the vaccine (see Chapter 84). The illness is caused by coming in direct contact with droplets from a person infected with the measles virus—an RNA-containing paramyxovirus. The incubation period is 10 to 14 days. In its classic form, measles has a highly characteristic natural history. Prodromal symptoms are cough, fever, coryza, and conjunctivitis. Two to three days after the onset of the prodrome and 12 to 24 hours before onset of the exanthem, pathognomonic Koplik's spots occur in the mouth. Most typically, they occur on the buccal mucosa opposite the molars as pinpoint white lesions on a red base. However, Koplik's spots may be seen on any of the mucosal surfaces of the oral cavity except the tongue.

The measles exanthem begins on the head as reddish maculopapules and spreads downward during the next 4 to 5 days. Within 1 to 2 days of the onset of a rash on any body part, the discrete maculopapular lesions coalesce to produce the confluent phase of the rash. Hence, within 2 to 3 days of onset, the rash on the face becomes confluent, whereas the rash on the lower extremities still consists of individual maculopapules.

There are two variants of typical measles based on altered immune status of the host. Modified measles occurs in children who have received immune serum globulin after exposure to measles. Measles may still occur, but the incubation will be delayed up to 21 days. The symptoms, although following the usual progression, will be milder. Atypical measles may occur in children previously immunized (almost always with killed vaccine) who have incomplete immunity. Atypical measles is characterized by absence of prodrome, peripheral to central spread of the rash, and variability of individual lesions, which may be macules, hemorrhagic vesicles, or petechiae. Because killed measles vaccine has not been in use for more than three decades, atypical measles is unlikely to occur in children.

Rocky Mountain Spotted Fever

Rocky Mountain spotted fever is caused by *Rickettsia rickettsii* transmitted by the bite of a tick (see Chapter 84). Although initially confined to the Rocky Mountain states (hence its name), confirmed cases have been reported from all parts of the United States with varying ticks as vectors. Rocky Mountain spotted fever is associated with a fatality rate of 5% with antimicrobial treatment and 13 to 40% without such therapy. The primary determinants in patient outcome are early diagnosis and treatment.

The rash of Rocky Mountain spotted fever begins on the third or fourth day of a febrile illness as a maculopapular eruption on the extremities, most commonly the wrists and ankles. Over the next 2 days, the rash becomes generalized by spreading centrally to involve the back, chest, and abdomen. Initially, the rash consists of erythematous macules that blanch on pressure; they then become more confluent and purpuric. Notably, the hemorrhagic rash remains more peripherally distributed with involvement of the palms of the hands and the soles of the feet. The severity of the rash is

proportional to the severity of the disease.

All patients with Rocky Mountain spotted fever have some degree of vasculitis that is the basis for many of the associated systemic symptoms. An overall toxic appearance is common. Systemic symptoms include fever; headache; myalgia; conjunctivitis; periorbital, facial, or peripheral edema; disseminated intravascular coagulation or purpura fulminans; shock; seizures; myocarditis; and heart failure.

Diagnosis is most commonly presumptive, based on clinical presentation with a history of potential tick exposure. The causative organism is not routinely cultured, and complement fixation tests are often negative in the acute phase; however, direct immunofluorescent antibody testing on a punch biopsy is available in some locales. The diagnosis can be confirmed by complement fixation tests in the second or third week of the disease. Some reference laboratories are now offering polymerase chain reaction (PCR) testing.

Therapy with doxycycline or chloramphenicol is highly effective and usually curative when given early in the course of disease. Therapy is continued until the patient is afebrile for at least 2 to 3 days, which usually equals about 7 to 10 days of antibiotic therapy.

Dengue Fever

Dengue fever, a biphasic febrile illness caused by several arthropodborne dengue viruses, is seen in tropical and subtropical areas of almost all continents (including areas of Puerto Rico and the Caribbean basin). Initial constitutional symptoms include sudden onset of high fever, severe headache, myalgia, arthralgia, and abdominal pain. During the course of fever that lasts 2 to 7 days, back and leg pain may be severe, hence the disease's nickname of “breakbone” fever. A hemorrhagic vasculitis can develop in some infections that may lead to shock and death.

Two distinct rashes may be seen, which coincides with the disease's biphasic fever pattern. The first rash is a generalized, transient, macular rash that blanches under pressure and is seen within the first 24 to 48 hours of the onset of systemic symptoms. The second rash coincides with or occurs 1 to 2 days after defervescence and is a generalized morbilliform or maculopapular rash, sparing the palms and soles.

Diagnosis is based on clinical suspicion and potential exposure based on the virus's geographic distribution. Treatment is supportive and may require fluid support and pain control.

Causes of Other Maculopapular Rashes Associated with Fever

Among the other illnesses that are not life-threatening but associated with fever are coxsackie infections, erythema infectiosum, scarlet fever, and early varicella. Harder to diagnose are rashes associated with Epstein-Barr virus, *Mycoplasma* infections, roseola infantum, disseminated gonorrhea, secondary syphilis, nonspecific viral eruptions, and drug-induced rash. It is particularly important to consider the diagnoses of disseminated gonorrhea and secondary syphilis in sexually active or potentially abused children.

Coxsackie Infections

Coxsackie infections of groups A and B (multiple types) can all cause maculopapular exanthems. The classic exanthem of coxsackie A16 infection, also appropriately called hand–foot–mouth disease, is common and easily recognized. Infections may occur in epidemics, most commonly in the late summer or early fall. Multiple infected members within a household are common.

Coxsackie A16 infection begins with a prodrome of low-grade fever, anorexia, mouth pain, and malaise, followed within 1 to 2 days by an oral enanthem and then shortly thereafter by a maculopapular exanthem. The oral lesions begin as small red macules, most often located on the palate, uvula, and anterior tonsillar pillar, which evolve into small vesicles that ulcerate and heal over a 1- to 6-day period. The exanthem begins as maculopapular lesions that develop into small crescent or football-shaped vesicles on an erythematous base. These vesicles, which may be pruritic or mildly tender, are usually located on the dorsal and lateral aspects of fingers, hands, and feet but may develop on the buttocks, arms, legs, and face. The lesions either reabsorb over 2 to 7 days or ulcerate and scab.

The other types of coxsackievirus all cause similar or even indistinguishable exanthems, which may more commonly involve the face, trunk, and proximal extremities. Often, children with these exanthems will be diagnosed with nonspecific viral infections. Other symptoms attributed to coxsackie infection include aseptic meningitis and less commonly myopericarditis, encephalitis, or paralysis.

Diagnosis is usually made clinically, although the virus can be easily cultured. The virus is commonly shed for weeks. Coxsackie infections are self-limiting, so no specific treatment is necessary. IVIG with high antibody titer may be considered for immunocompromised patients or in life-threatening neonatal infections.

Erythema Infectiosum (Fifth Disease)

Erythema infectiosum is a benign disease caused by parvovirus B19, the same virus that can cause aplastic crises in patients with sickle cell anemia. For the normal, nongravid host, fifth disease is of no consequence, with the only systemic symptom being fever in 15 to 30% of cases. On the face is a characteristic, intensely erythematous, “slapped cheek” rash. In addition, a symmetric maculopapular, lacelike rash is seen on the arms then trunk, buttocks, and thighs. In its acute phase, the rash usually lasts only for a few days but can wax and wane in intensity with environmental changes (e.g., exposure to heat or sunlight) for weeks and sometimes months. No specific diagnosis or therapy is necessary. For a chronic infection in an immunodeficient patient, IVIG therapy should be considered. Because parvovirus

is associated with fetal anemia, congestive heart failure, and hydrops, exposed pregnant women should be referred to their physicians to discuss possible parvovirus antibody testing.

Scarlet Fever

Scarlet fever is caused by phage-infected group A streptococcus that makes an erythrogenic toxin. This disease is still seen with regularity but does not appear to be any more serious than group A streptococcal infection without rash. Scarlet fever is most commonly associated with streptococcal pharyngitis but may occur in association with pyoderma or an infected wound.

The diagnosis of scarlet fever can be made in a child with signs and symptoms of pharyngitis who has a fine, raised, generalized maculopapular rash. The skin has a coarse or sandpapery feel on palpation. Typically, there is sparing of the circumoral area, leading to circumoral pallor. There usually is a bright erythema of the tongue and hypertrophy of the papillae, leading to the term strawberry tongue. Pastia's lines, bright red, orange, or even hemorrhagic lines, can occasionally be seen in the axillae or antecubital fossa. The rash generally lasts 3 to 5 days, followed by brownish discoloration in association with peeling of the skin. The peeling may range from small flakes to entire casts of the digits. A rapid streptococcal test or throat culture should be sent to confirm infection. Various antibiotic regimens provide effective treatment (see Chapter 84).

Varicella (Chickenpox)

Although varicella is an easily recognizable vesiculobullous eruption, on occasion, the earliest phase can be confusing. The initial skin manifestations of varicella virus infection are small, red macules. Some of them remain as macules, but most progress to the characteristic papules and then umbilicated, tear-shaped vesicles. The earliest lesions appear on the chest and spread centrifugally, but there are many exceptions to the pattern of spread. Occasionally, a child with mild chickenpox may have only a few scattered macules with only one or two progressing to the more typical vesicular lesions of chickenpox. Of children receiving varicella vaccine, 7 to 8% may develop a mild maculopapular or varicelliform rash within 1 month of vaccination (see Chapter 84).

Epstein-Barr Virus

Between 5 and 15% of patients with Epstein-Barr viral infection, otherwise known as infectious mononucleosis, will have an erythematous maculopapular eruption. Infection in young children is usually inapparent, nonspecific, or so mild that diagnosis is not sought. The older patient between 15 to 25 years more commonly presents for evaluation. In addition, 50 to 100% of patients with infectious mononucleosis receiving concurrent ampicillin will develop a maculopapular rash.

The illness begins insidiously with headache, malaise, and fever, followed by sore throat, membranous tonsillitis, and lymphadenopathy. Splenomegaly is common. The exanthem occurs within 4 to 6 days as a macular or maculopapular morbilliform eruption most prominent on the trunk and proximal extremities. An enanthem consisting of discrete petechiae at the junction of the hard and soft palate occurs in approximately 25% of patients.

Diagnosis is often presumed clinically but may be supported by a positive heterophile antibody (Monospot) test or confirmed by serology. The heterophile antibody test is less sensitive in children younger than 4 years of age. The illness is most commonly self-limited, requiring no therapy. Corticosteroids may be considered for patients with particularly severe tonsillitis (see Chapter 84).

Mycoplasma Infections

Infections with *Mycoplasma pneumoniae* may cause maculopapular rashes in up to 15% of cases. The classic clinical presentation is of a child with malaise, low-grade fever, and prominent cough. The cough is initially nonproductive but may become productive particularly in older children and may persist for 3 to 4 weeks. Physical examination may show bilateral rales. Roentgenographic examination of the chest, if abnormal, most commonly shows diffuse nonspecific infiltrates.

Diagnosis can be suggested by serum cold hemagglutinins, which are present in more than 50% of cases by the beginning of the second week. If further confirmatory studies are needed, acute and convalescent serum sera should be assayed for specific mycoplasmal antibodies by complement fixation or immunofluorescence. Erythromycin or one of the newer macrolides (clarithromycin or azithromycin) is the treatment of choice (see Chapter 84).

Roseola Infantum

Roseola infantum, also called exanthem subitum or 6th disease, has recently been attributed to herpes simplex virus (HSV)-6. The illness is characterized by the onset of a maculopapular rash following a 3- to 4-day febrile illness. The fever is characteristically high. The rash is widely disseminated, appearing as discrete, small, pinkish macules that rarely coalesce beginning on the trunk and then extending peripherally. The occurrence of the rash within 24 hours of defervescence rather than the morphologic appearance of the rash leads to the correct diagnosis. The rash can appear very similar to that seen in measles, but the child with roseola is well appearing and no longer febrile. Diagnosis is made clinically and care is supportive.

Disseminated Neisseria gonorrhea

Disseminated *Neisseria gonorrhoeae* should be considered in sexually active or potentially abused children, especially if associated with a history of vaginal or penile discharge (see Chapter 84). A distinct minority of patients develop disseminated gonorrhea infection through hematogenous spread. Disseminated gonorrhea may cause a range of

cutaneous lesions, including small erythematous papules, petechiae, or vesicle–pustules on a hemorrhagic base. These cutaneous lesions usually develop on the trunk but may occur anywhere on the extremities. An etiologic diagnosis can be established by demonstration of the organism on Gram stain of the skin lesion, positive blood culture, or positive culture of oral or genital sites. Based on resistance patterns, recommended current therapy is ceftriaxone 50 mg/kg per day (maximum 1 g/day) for 7 days. Concomitant sexually transmitted diseases should be sought and treated empirically.

Secondary Syphilis

One needs a high level of suspicion when viewing rashes in sexually active (or potentially abused) children to make the diagnosis of secondary syphilis, caused by the spirochete *Treponema pallidum* (see Chapter 84). Manifestations of secondary syphilis usually occur 6 to 8 weeks after the appearance of the primary lesion, which may have gone unnoticed. The exanthem extends rapidly and is usually pronounced, lasting for only hours or persisting for several months.

The rash is characterized by a generalized cutaneous eruption, usually composed of brownish, dull-red macules or papules that range in size from a few millimeters to 1 cm in diameter. They are generally discrete and symmetrically distributed, particularly over the trunk, where they follow the lines of cleavage in a pattern similar to pityriasis rosea. Papular lesions on the palms and soles as well as the presence of systemic symptoms such as general malaise, fever, headaches, sore throat, rhinorrhea, lacrimation, and generalized lymphadenopathy help differentiate secondary syphilis.

Acquired syphilis is sexually contracted from direct contact with ulcerative lesions of the skin or mucous membranes of an infected individual. Diagnosis may be presumed after a positive nontreponemal test, such as the VDRL slide test, rapid reagin (RPR) test, or the automated reagin test (ART). Diagnosis should be confirmed by a treponemal test such as the fluorescent treponemal antibody absorption (FTA-ABS) test, the microhemagglutination test for *T. palladium* (MHA-TP), or the *T. palladium* immobilization (TPI) test. Definitive diagnosis may also be made by identifying spirochetes by microscopic dark field examination or direct fluorescent antibody tests of lesion exudate or tissue. Penicillin is the treatment of choice unless contraindicated, in which case tetracycline, doxycycline, ceftriaxone, or erythromycin may be substituted. Length of therapy should be based on duration and stage of infection. Concomitant sexually transmitted diseases should be sought and treated empirically.

Nonspecific Viral Exanthems

Many times, a specific diagnosis cannot be made even after considering such factors as exposure history, history of preceding illness, description of eruption, time and site of onset, character of initial lesion, progression, distribution patterns, and occurrence of mucosal lesions. This should not be surprising, given the large number of viruses that can be associated with macular or maculopapular eruptions. A number of enteroviruses and adenoviruses can cause a macular or maculopapular eruption. There is little to distinguish the rash caused by one of these viruses from that of another, based on the location and morphology, with the exception of coxsackie A16 infection, which causes hand–foot–mouth disease. One usually arrives at the diagnosis of nonspecific viral exanthem in a child in whom other diagnoses have been excluded and who may have signs of associated illness or systemic features such as fever. Specific etiologic diagnosis, if required, can be determined by viral isolation and/or a rise in diagnostic titer.

Drug-Induced Rash

Multiple drugs can cause maculopapular rashes in susceptible patients. Most commonly, these rashes have an abrupt onset, are generalized, and may be accompanied by systemic signs such as fever, arthralgia, lymphadenopathy, and hepatomegaly. It is often difficult to distinguish drug eruptions from viral exanthems. This is specially true because the emergency physician is often faced with a child who recently was started on one or several medications (often including an antibiotic) who now presents with the emergence of a rash associated with or following a viral-type illness.

The diagnosis of drug eruption depends on a carefully obtained history, including the duration and frequency of all medications taken by the child during the week preceding the onset of the rash. The presence of eosinophilia suggests, but does not confirm, the diagnosis. Often, the final diagnosis is left to the intuition of the physician. In the case of a severe eruption, the potentially offending drug should be discontinued. In milder cases, which more closely resemble nonspecific viral exanthems, a physician may opt to continue therapy as long as the rash does not worsen. The disadvantage of simply discontinuing any potentially offending drug is that the patient is often labeled as “allergic” to the drug for life. In addition, reactions may be caused by preservatives or dyes in a drug preparation and not by the drug itself.

Illnesses Associated with Maculopapular Rashes without Fever

Maculopapular rashes associated with nonfebrile illnesses tend to be benign. Erythema infectiosum, EM, *Mycoplasma* infections, roseola infantum, secondary syphilis, and nonspecific viral exanthems, which in mild cases may not be associated with fever, have been previously discussed. In approaching the acute afebrile disorders associated with maculopapular rash, it is useful to distinguish between those that cause generalized eruptions and those that cause localized ones. Disorders not usually associated with fever but that cause generalized eruptions include rubella, guttate psoriasis, and pityriasis rosea. Disorders that cause mostly local eruptions include papular acrodermatitis (Gianotti-Crosti syndrome), contact dermatitis, insect bites, and scabies. Some of the maculopapular rashes not associated with febrile illnesses are chronic entities, allowing their duration to help with their diagnosis; examples include lichen nitidus, molluscum contagiosum, papular urticaria, pityriasis lichenoides (Mucha-Habermann disease), and tinea versicolor. Molluscum contagiosum, pityriasis rosea, tinea versicolor, and forms of contact dermatitis also present with clinically recognizable rashes.

Generalized Eruptions Associated with Afebrile Illnesses

Guttate Psoriasis

About one-third of cases of psoriasis begin in the first two decades of life among individuals with a genetic predisposition. The guttate form is even more likely to occur in younger age groups. The rash is characterized by multiple small discrete round or oval macules or papules (up to 1 cm in diameter) with a loosely adherent scale. The lesions develop predominantly on the trunk, but the face and scalp may be involved. The distal extremities, palms, and soles are usually spared. The lesions of guttate psoriasis are not as hyperkeratotic as other types of chronic psoriatic plaques and may respond better to standard psoriasis therapy.

Pityriasis Rosea

Pityriasis rosea is a benign, self-limiting condition that most commonly affects older children and adolescents, although it can occur at younger ages. The cause is unknown but is likely to be viral (see Chapter 99).

Pityriasis rosea follows a characteristic clinical course. The initial lesion, the herald patch, is an oval-shaped plaque that occurs in about 80% of cases. The center of the lesion is flat, whereas the borders are raised, red, and scaly. The herald patch can occur anywhere on the body but is most commonly seen on the trunk, neck, or proximal extremities. The herald patch is often mistaken for tinea corporis. One to two weeks later, a more generalized, sometimes pruritic, rash erupts. The rash is most dense on the trunk, neck, and proximal limbs. The face and distal extremities are relatively spared but may be involved in younger children. Individual lesions are erythematous papulosquamous ovals that often resemble smaller versions of the herald patch. The orientation of the long axis of the ovals tends to conform to the skinfold lines of the trunk, giving characteristic “Christmas tree” pattern of distribution when looked at on the patient's posterior trunk. Atypical distributions (predominantly peripheral) and forms of the individual lesions (papules, vesicles, pustules, urticarial or purpuric lesions) can occur.

Rubella

In a classic case of rubella, the rash, as with measles, begins on the head and spreads downward. The progression occurs over 2 to 3 days, and typically, the rash is entirely gone by the fourth day. The rash always remains macular and never becomes confluent, an important distinguishing characteristic. One-third of all rubella virus infections are clinically silent (i.e., they have no exanthem). A rubella rash may show extensive variation in location, progression, and duration, at times disappearing within 12 hours or being localized to one part of an extremity without any progression. Unlike measles, in which systemic toxicity and fever are the rule, fever is uncommon. Associated symptoms and complaints in rubella include joint pain in about 25% of cases and adenopathy (most commonly suboccipital, postauricular, and cervical). Arthralgia that occurs with a viral exanthem is relatively specific for rubella. Diagnosis is based on clinical presentation, and treatment is supportive.

Localized Eruptions Associated with Afebrile Illnesses

Contact dermatitis, insect bites, papular acrodermatitis, and scabies usually have a localized distribution; however, in extensive cases, all may appear as a more generalized eruption.

Contact Dermatitis

Contact dermatitis may be produced by either a local exposure to a primary irritating substance or by an acquired allergic response to a sensitizing substance (see Chapter 99). When the dermatitis results from a nonallergic reaction of the skin, it is termed an irritant contact dermatitis; when it results from a delayed hypersensitivity to a contact allergen, it is termed an allergic contact dermatitis. Although distinct in etiology, both reactions usually have a localized distribution of the rash, which often assumes the pattern of the irritating or sensitizing agent, and there is generally a sharp demarcation between involved and uninvolved areas of skin. Involved areas are erythematous with variable numbers and combinations of macules, papules, vesicles, and/or bullae.

Irritant dermatitis arises from contact with primary irritating agents such as detergents, soaps, acids, alkalis, or rough sheets/clothes. This disorder is commonly seen in infancy, when the skin is relatively thin and susceptible to mechanical or chemical irritation. Allergic contact dermatitis, typified by rhus dermatitis (e.g., poison ivy, poison oak) or nickel dermatitis (from jewelry or wristwatches), occurs most commonly in older children.

Diagnosis depends on obtaining a thorough history of exposure to probable offending allergens and the presence of a characteristic localized pattern of rash. Treatment for both types of contact dermatitis includes reducing exposure to offending irritants, providing topical or systemic antipruritic agents, and for more severe cases, providing topical or systemic steroids.

Insect Bites

Virtually all children experience insect bites. Mosquitoes, fleas, and bedbugs are the most common offenders. Diagnosis depends on the season, the climate, exposure to animals, and distribution and appearance of the lesions. In temperate climates, mosquito bites occur exclusively in the warmer months of the year, whereas flea and bedbug bites occur year round as a result of indoor exposure. Often, a series of bites occurs in groups, causing a maculopapular appearance. Local reactions can be extensive and take several days to resolve. Care is aimed at minimizing discomfort with topical or systemic antihistamines and/or topical steroids.

Papular Acrodermatitis (Gianotti-Crosti Syndrome)

Papular acrodermatitis is an eruption of unclear cause that has been associated with hepatitis B and other viral infections in young children. Of affected children, 85% are less than 3 years old. The eruption may follow a low-grade fever or mild upper respiratory symptoms.

The eruption consists of flesh-colored papules that occur anywhere on the body but often concentrate on the extensor surfaces of the arms, legs, and buttock. Lesions are particularly prominent over the elbows and knees. The rash usually lasts 2 to 8 weeks and then disappears. No treatment is needed for the cutaneous eruption; however, a subset of patients with cutaneous lesions develop generalized lymphadenopathy and hepatosplenomegaly. These children should be evaluated for hepatitis. Follow-up in 2 weeks is recommended for patients with only cutaneous involvement to rule out the development of hepatitis.

Scabies

Scabies is a contagious infestation of the *Sarcoptes scabiei* female mite that selects a favorable body site, burrows beneath the stratum corneum, and deposits eggs along the way. In older children and adults, the usual sites of infestation are the anterior axillary lines, the areolae, the lower part of the abdomen, buttocks, genitals, wrists, interdigital webs, and ankles. In young children, the lesions are usually more diffuse and may also occur on the palms, soles, scalp, and neck (see Chapter 99).

The pathognomonic primary lesion may be visible as a linear, gray–brown, threadlike burrow a few millimeters in length, with a central black dot (the mite). The more usual lesions are erythematous papules that may be excoriated and possibly secondarily infected because of intense pruritus. On occasion, generalized urticarial or “id” reactions develop.

Diagnosis is usually based on clinical suspicion, although definitive confirmation can be made by identifying the adult mite on microscopic examination of a scraping of suspicious burrows. The treatment of choice in children is the topical application of permethrin 5% cream, which may be repeated in 2 weeks if necessary. Pruritus often persists for several weeks after the mites have been eliminated. It is advisable to treat close family members or personal contacts with or without evidence of infestation. Because mites are unable to survive away from their human hosts or at high temperatures, clothing, bedding, and stuffed animals should be laundered in hot water (greater than 50°C, or 120°F) or stored away for several days in plastic bags.

Chronic Eruptions Associated with Afebrile Illnesses

Chronic eruptions are defined as those that are usually present for a minimum of 2 weeks.

Lichen Nitidus

Lichen nitidus is a relatively rare, benign skin disorder that occurs most often in preschool and school-age children. It is thought to perhaps be a variant of lichen planus. The eruption consists of groups of tiny, shiny, flesh-colored papules. The lesions commonly occur in lines of local trauma (Köbner's phenomenon) and are most often seen on the trunk, abdomen, forearm, and genitalia. There is no known effective treatment, and the eruption can last for years.

Molluscum Contagiosum

Molluscum contagiosum is caused by a viral infection and consists of discrete flesh-colored papules, usually 2 to 3 mm in diameter, with umbilicated centers (see Chapter 99). Axillary lines of the trunk, the abdomen, genital region, inner aspect of the thighs, and the face are the most common sites of presentation, although any nonhairy surface may be involved. Usually, a child will have approximately 10 scattered lesions; however, on occasion, some may have many more. The lesions tend to persist anywhere from 2 weeks to $1\,{}^{1}/_{2}$ years and may be spread by autoinoculation. Spread can occur between individuals involved in contact sports. The lesions are asymptomatic, with the exception of a minority of patients who develop an inflammatory reaction. Treatment, when deemed necessary, involves techniques such as liquid nitrogen or curettage that minimize scarring and discomfort.

Papular Urticaria

Papular urticaria, a benign condition seen most commonly in young children, is manifested by a chronic or recurrent papular eruption caused by a sensitivity reaction to insect's bites. The lesions are usually papules with a central punctum that may rest on a urticarial base. The lesions are most commonly seen in the warm months, when exposure to insects is most intense. Diagnosis is usually made clinically. Treatment is aimed at minimizing exposure to insect bites and providing therapy with simple sedation, topical calamine, or topical corticosteroid to minimize pruritus.

Pityriasis Lichenoides (Mucha-Habermann Disease)

Pityriasis lichenoides, or Mucha-Habermann disease, is a relatively rare disorder of unknown cause that can appear in childhood and young adulthood. There are two forms: acute and chronic. The acute disease is characterized by a macular, papular, or papulovesicular rash that often is distributed most heavily on the trunk and upper arms. The lesions occur in successive crops rapidly evolving into vesicular, necrotic, and even purpuric lesions. Lesions may leave pocklike scars. Resolution occurs spontaneously but may take several weeks to months and recurrences may occur. Parents may describe these recurrences as “he keeps getting the chickenpox.” The more chronic form may evolve from the acute form or may arise de novo and often lasts for several years. There is no established therapy.

Tinea Versicolor

Tinea versicolor is a superficial skin disease caused by the fungus *Pityrosporum orbiculare* (see Chapter 99). Although adolescents and young adults are most commonly affected, the disorder can occur at any age. The distribution of scaly macular lesions is patchy and occurs most commonly over the upper trunk and proximal arms. Occasionally, the face and other areas of the body can become involved. In summer, affected areas are relatively hypopigmented compared with unaffected skin because the organism blocks the normal tanning of sun-exposed skin. In winter, the affected areas often are relatively darker than unaffected skin because the fungus causes a mild erythema. This phenomenon of variable coloration of the affected skin gives the disease its name.

The diagnosis often is made by recognition of the characteristic rash. Wood's light examination in a darkened room produces a reddish-brown fluorescence. Microscopic examination of scrapings will demonstrate characteristic hyphae and spores in grapelike clusters (“spaghetti and meatballs” appearance). Treatment consists of selenium sulfide shampoos weekly for 3 weeks and then monthly for 3 months. Infection may recur.

EVALUATION AND DECISION

In approaching a child with a maculopapular exanthem, the initial steps are to take a history and to fully examine all cutaneous surfaces. The most important historical features include the duration of the rash (acute or chronic), initial distribution, extent of spread (generalized or localized), ill contacts (including sexual partners of adolescent patients), and any associated systemic symptoms, including fever. The physical examination should include a careful systematic inspection of all mucocutaneous surfaces, with special attention paid to involvement of the oropharynx, palms and soles, extensor or flexor surfaces, scalp, and trunk.

For patients who do not appear ill, certain exanthems will have distinctive patterns that will immediately strike the examiner and make the diagnosis readily apparent. EM, rubella, coxsackie infections, erythema infectiosum, scarlet fever, varicella, molluscum contagiosum, tinea versicolor, pityriasis rosea, and roseola all have recognizable clinical appearances. Many of these illness have characteristic distributions or associated signs and symptoms that aid in their diagnoses. If the pattern of the rash does not evoke an immediate response from the examiner, a more methodical approach is indicated, as outlined in Figure 62.1.

For patients with maculopapular rash who appear particularly ill, the potential diagnoses of rubeola (measles), EM, Kawasaki disease, Rocky Mountain spotted fever, and dengue fever should spring to mind.

Acutely Ill-Appearing Patients

Rubeola and EM both have characteristic rashes that are often associated with oral involvement. Patients with rubeola may have a history of an ill contact and several days of cough, coryza, conjunctivitis, and escalating fever. EM may present with a history of the recent introduction of a medication. Kawasaki disease should be considered in children who have been febrile for more than 5 days and who have or have had conjunctivitis, red lips/strawberry tongue, a solitary enlarged cervical lymph node, and a rash. Clues to the possibility of Rocky Mountain spotted fever or dengue fever may be obtained from a travel history or known cases within the geographic location. Patients with Rocky Mountain spotted fever may have a history of a tick bite, and the hemorrhagic rash characteristically remains more peripherally distributed involving the palms and soles. Dengue fever should be considered in patients with a biphasic fever pattern and musculoskeletal pain.

Other Generalized Febrile Eruptions

An acute, generalized febrile maculopapular exanthem usually is the result of a nonspecific viral or streptococcal (scarlet fever) infection. The disorders that are seen in acutely ill-appearing patients, discussed previously, may present as milder versions and should be considered as possible causes in less acutely ill febrile children with generalized eruptions.

Other viral and bacterial infections may require a higher index of suspicion and confirmatory studies.

Nonspecific viral exanthems most characteristically consist of multiple, closely spaced small papules. The finding of pharyngitis, a strawberry tongue, or intensely erythematous lines in the antecubital fossae points to scarlet fever; however, a throat culture or rapid screening test for streptococcal infection should still be obtained.

Coxsackie infections, erythema infectiosum, and early varicella should be able to be diagnosed based on their clinical appearance. It should be remembered that the eruption of varicella initially is maculopapular; however, close inspection usually reveals a few vesicles by the time the child is brought to medical attention.

The final considerations in febrile patients with generalized maculopapular rash are Epstein-Barr virus infections (infectious mononucleosis), *Mycoplasma* infections, roseola infantum, disseminated gonorrhea, and secondary syphilis. The exanthem of infectious mononucleosis should be suspected in the child or, more commonly, in the adolescent who has streptococcal negative pharyngitis and/or history of taking ampicillin or a closely related antibiotic. For children with nonspecific viral symptoms with prominent cough, *Mycoplasma* infection may be the diagnosis. Roseola infantum should be considered in the child who develops maculopapular rash after fever has defervesced. Last, disseminated gonorrhea and secondary syphilis should be considered in sexually active adolescents and appropriate tests sent for confirmation.

Generalized Afebrile Eruptions

Although nonspecific viral illnesses that cause rash are more often than not associated with fever, a minority of children with viral exanthems remain afebrile. Often, no specific diagnosis is possible. Again, the appearance of a diffuse rash in

an infant or toddler immediately after the defervescence of a high fever indicates the clinical diagnosis of roseola. Similarly, pronounced posterior occipital lymphadenopathy in an unvaccinated child suggests rubella. If a child is taking any medications, drug rash must be considered. Because a drug reaction is difficult to exclude initially, consideration for discontinuing medications is warranted in severe cases. Also common, pityriasis rosea is distinguished by its characteristic predominantly truncal distribution along the skin folds. Rarely does guttate psoriasis present acutely with a diffuse maculopapular eruption.

Localized Eruptions

The most common causes for acute, localized maculopapular eruptions are contact dermatitis and insect bites. Contact dermatitis may be caused by irritation or allergy. History may be helpful in establishing a diagnosis, as in the case of a child who returns from camp with an allergic dermatitis on the arms and legs (rhus dermatitis or poison ivy) or of a teenager who gets an irritant dermatitis of the wrist after wearing a new watch. Irritant reactions are usually exclusively maculopapular, whereas allergic eruptions may become vesicular or eczematous and may also have a characteristic linear appearance. The papules of insect bites usually are isolated lesions, as opposed to the confluent rash seen in contact dermatitis. In temperate climates, insect bites occur most commonly in the summer, but the possibility of bedbugs or fleas should not be overlooked during the colder months. Scabies is a relatively common and potentially difficult diagnosis. Linear lesions and involvement of the web spaces are characteristic, but often, a diagnostic scraping or presumptive therapy is indicated. Gianotti-Crosti syndrome is a rare disorder that produces primarily an eruption limited to the distal extremities. Any of the causes of localized eruptions may appear more generalized in extensive or severe cases.

Chronic Eruptions

Chronic maculopapular eruptions are usually, and more appropriately, seen by physicians in settings other than the ED. However, parents may become acutely concerned about a real or perceived change in a chronic eruption; thus, the emergency physician should be familiar with the more common disorders.

The most commonly seen of the chronic maculopapular eruptions are papular urticaria, molluscum contagiosum, and tinea versicolor. Papular urticaria is most common in warm weather but may occur at any time of the year; the characteristic lesions have an urticarial wheal around a central papule. The papules of molluscum contagiosum have an easily recognizable umbilicated central core. Tinea versicolor consists of hypopigmented and hyperpigmented areas, predominantly on the trunk. This diagnosis can be confirmed by microscopy or culture. Although uncommon, secondary syphilis needs to be considered in any sexually active patient and a serologic test performed as indicated.

Suggested Readings

American Academy of Pediatrics. In: Peter G, ed. 1997 Red Book: Report of the Committee on Infectious Disease. 24th ed. Elk Grove Village, IL: American Academy of Pediatrics, 1997.

Draelos ZK, Hansen RC, James WD. Gianotti-Crosti syndrome associated with infections other than hepatitis B. JAMA 1986;256:2386– 2388.

Hurwitz S. Clinical Pediatric Dermatology: A Textbook of Skin Disorders of Childhood and Adolescence. Philadelphia: WB Saunders, 1993.

Feign RD, Cherry JD, eds. Textbook of Pediatric Infectious Diseases. 4th ed. Philadelphia: WB Saunders, 1998.

National Vaccine Advisory Committee. The measles epidemic. JAMA 1991;266:1547–1552.

Resnick SD. New aspects of exanthematous diseases of childhood. Dermatol Clin 1997;15(2):257–266.

Shulman ST, De Inocencio J, Hirsch R. Kawasaki disease. Pediatr Clin North Am 1995;42(5):1205–1221.

CHAPTER 63

Rash—Papular Lesions

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Physicians are often confronted by parents who are concerned about “bumps” on their child's skin surface. Most parents will be comforted by a physician's reassurance and recommendations for treatment. Obviously, therapeutic interventions cannot be made until a definitive diagnosis is made. The following algorithm is used to help practitioners diagnose varying papular lesions (Fig. 63.1 and Table 63.1).

FIGURE 63.1. Approach to the diagnosis of papular lesions.

Granuloma annulare (common)	Molluscum contagiosum (common)
Insect bites (common)	Pyogenic granuloma (common)
Juvenile xanthogranuloma	Spitz nevus
Lichen nitidus	Warts (common)
Mastocytomas, urticaria pigmentosa	Xanthomas
Milia (common)	

Table 63.1. Papular Lesions

PAPULES WITH A CHARACTERISTIC CLINICAL APPEARANCE

Many conditions can be diagnosed on sight. The experienced eye can easily distinguish milia from molluscum contagiosum (MC) and warts from the uncommon xanthoma. Papules caused by bites are localized to exposed surfaces (face and extremities). Several clues make the process of separating these entities from one another easier (see section on noncharacteristic clinical appearance).

The distinction between flat warts and xanthomas is more subtle. At times, it is impossible to tell the difference. Because warts are caused by the human papillomavirus, a scratch through any of the lesions may inoculate the virus in the scratch line. This produces flat-topped papules in a linear distribution (a quasi-Köbner's phenomenon—that is, appearance of the primary eruption located elsewhere on the body in the site or sites of trauma). Therefore, linearly arranged yellow, tan, or flesh-colored flat-topped papules should arouse suspicion of the presence of flat warts, especially if the lesions are distributed on the face, back of the hands, and knees—favorite sites for flat warts. Xanthomas are unusual during childhood. When present, however, they usually are associated with elevations of serum lipids. A lipid profile can be helpful in distinguishing these two flat-topped papules from one another. If all else fails, the help of a dermatologist experienced in caring for children can be sought. This is true for any of the entities discussed in this chapter. Skin biopsy is a valuable tool available to the dermatologist and may be required to differentiate many of the entities that are discussed here.

Milia

Milia are 1- to 2-mm firm, white papules. They are produced by retention of keratinous and sebaceous material in follicular openings (orifices from which vellus hairs grow). Newborns often have milia on their face. Fortunately, they disappear by the age of 1 month. Milia can be seen in scars after burns and in healed wounds in patients with epidermolysis bullosa. Persistent milia may be a manifestation of the oral–facial–digital syndrome or hereditary trichodysplasia. Because lesions that are not associated with syndromes disappear spontaneously, no therapy is indicated.

Molluscum Contagiosum

For additional information about MC, see Figure 99.42.

Warts

For additional information about warts, see Chapter 99.

Xanthomas

Papules, plaques, nodules, and tumors that contain lipid are called xanthomas. These lesions can appear on any skin surface and often are associated with disturbances of lipoprotein metabolism (Table 63.2).

Type	Age of Onset	Clinical Presentation	Inheritance
I	Early childhood	Eruptive xanthomas Abdominal pain Hepatosplenomegaly Creamy plasma	AR
IIa	Early childhood	Tendinous xanthomas Xanthelasmas Tuberous xanthomas Corneal arcus	AD
IIb	Early childhood	Same as IIa	AD

Table 63.2. Hyperlipidemias

The most interesting, but the most rare, of hyperlipidemias that arise in the pediatric (infancy to adolescence) age group are type I hyperlipidemias. Fifty percent of patients present with episodic abdominal pain that may be acute at times. Malaise, anorexia, fever, and leukocytosis may be present. The cause of the pain is unclear; however, pancreatitis and splenic infarcts have been hypothesized. Eruptive xanthomas occur in more than 50% of the patients. These are 1- to 4-mm yellow papules that appear in crops on the face, extremities, and buttocks. Their sudden appearance causes significant concern. Hepatosplenomegaly also is commonly present, as are lipemia retinalis and creamy plasma. Patients are found to have increased chylomicrons, slightly elevated cholesterol, and significantly elevated triglycerides. Secondary diseases include pancreatitis and diabetes.

The homozygous form of type IIa hyperlipidemia is seen in children. An elevated low-density lipoprotein (LDL), significantly elevated cholesterol, and mildly elevated triglycerides characterize this disorder. Tendinous and tuberous xanthomas and xanthelasmas are seen clinically. The plane xanthomas (xanthelasmas) may be misinterpreted as flat warts when seen in skin areas away from the eyelids. Secondary disorders include hypothyroidism and nephrotic syndrome. Patients die of atherosclerotic coronary artery disease in their twenties and thirties.

Insect Bites

For additional information about insect bites, see Chapter 99, Mosquitoes and Fleas.

PAPULES WITH A NONCHARACTERISTIC CLINICAL APPEARANCE

When the diagnosis is not obvious, the algorithm presented in Figure 63.1 must be used.

Presence of White or Translucent Core

Milia and MC have white cores. Sidelighting and a magnifying glass may be needed to see the core within the central portion of the papule in MC. Therefore, it is essential to sidelight all papules about which one is not sure. The obvious white core in milia fills the entire papule rather than a small central portion of the papule (as in MC). The other differentiating point is that milia are hard and beady white; MC are more fleshy.

Absence of White or Translucent Core

Rapid Growth of Hemangiomalike Lesions

Hemangiomas generally present within the first month of life. Two lesions that may mimic hemangiomas generally manifest after this period. These lesions are the pyogenic granuloma and Spitz nevus. They are differentiated by the fact that the Spitz nevus has a red, smooth, dome-shaped surface, as opposed to the crusted granular surface of a pyogenic granuloma. The last differentiating point is the common occurrence of bleeding of pyogenic granulomas following minor trauma. Juvenile xanthogranulomas (JXGs) can be red and appear suddenly. They are firm rather than spongy (like a hemangioma), and when blanched, the underlying color usually is yellow.

Spitz Nevus

Spitz nevi appear suddenly between 2 and 13 years of age. Preferred sites of growth include the cheek (15%) (Fig. 63.2), shoulder, and upper extremities. The lesion has a pink to red surface because of numerous dilated blood vessels. Pressure produces blanching of this pink to red color. The lesions can reach a size of 1.5 cm in diameter but are completely benign. Because the histologic appearance of these lesions can be confused easily with a malignant melanoma, an experienced histopathologist should interpret the findings. Most clinicians recommend that Spitz nevi be removed surgically.

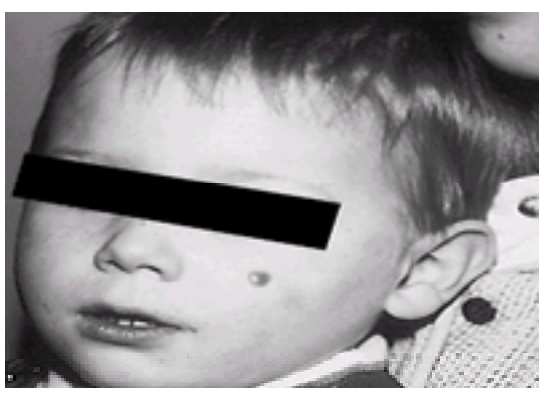

FIGURE 63.2. Red papule that appeared 2 months ago and grew rapidly to this size (Spitz nevus).

Pyogenic Granuloma

For additional information about pyogenic granulomas, see Figure 99.38.

Juvenile Xanthogranuloma

JXG can be confused with urticaria pigmentosa or xanthomas. Numerous yellow or reddish-brown papules appear on the face (Fig. 63.3) and upper trunk in the first year of life. The number of lesions may increase until the child is 18 months to 2 years of age. Serum lipid levels are normal, and the Darier's sign (urtication after scratching—see mastocytoma) is negative. The lesions disappear spontaneously after 2 years of age; therefore, intervention is totally unnecessary.

FIGURE 63.3. Blanching erythematous papule on the ala nasae of this child. Biopsy showed this lesion to be a JXG.

NO HEMANGIOMALIKE LESION: YELLOW, TAN, OR BROWN PAPULE

The yellow, tan, and brown papules include the lesions seen in urticaria pigmentosa (a single, large lesion is called a mastocytoma), flat warts, xanthomas, insect bites, and JXGs.

A first step to differentiate the various papules from one another is to scratch them. If urtication (hiving) occurs (Darier's sign) within a short period (3 to 5 minutes), the lesion must contain mast cells (i.e., a mastocytoma or urticaria pigmentosa). Make sure to scratch normal skin to rule out the presence of dermographism. The latter condition will produce a false-positive Darier's sign. When no urtication occurs, blood should be drawn to check lipid levels. If lipid levels are normal, the next step is to differentiate two of the entities (i.e., flat warts and JXGs). Flat warts tend to be grouped, are flat-topped, and can be autoinoculated in scratch lines (pseudo-Köbner's phenomenon). Lesions characteristic for JXGs are not flat-topped, tend to be singular in number (or when multiple are scattered about), and do not demonstrate the Köbner's phenomenon. (This condition is discussed in greater detail later.) The JXG lesions also may look like xanthomas. Unlike xanthomas, however, abnormal lipid levels do not occur with JXGs.

Mastocytoma, Urticaria Pigmentosa

Parents who bring children with mastocytomas or lesions of urticaria pigmentosa to the physician generally describe a single yellow–tan–brown lesion that was present at birth (mastocytoma) or multiple pigmented papules that erupt during the first year of life (urticaria pigmentosa). One important clue is a history of these lesions becoming red (Fig. 63.4), hivelike, or blistered. The lesions may ooze and form crusts much like impetigo; however, they do not respond to antibacterial preparations.

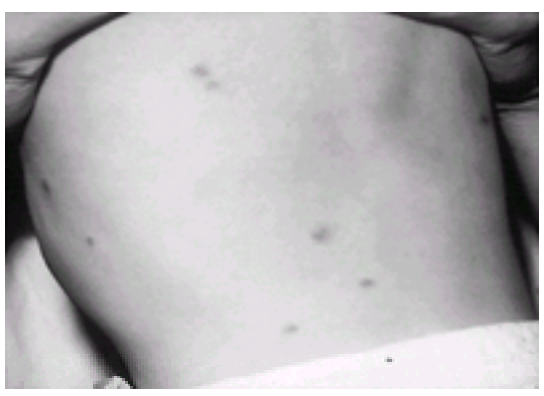

FIGURE 63.4. Erythematous papules representative of urticaria pigmentosa. They will urticate with scratching (Darier's sign).

Physical examination of the lesion provides the next clue. The surface has a peu d'orange appearance at times. Some papules are very yellow and are easily mistaken for xanthomas. When they are tan to brown, they are thought to be raised moles. The clincher is finding a positive Darier's sign (histamine-induced erythema, swelling, and urtication secondary to scratching and subsequent degranulation of mast cells).

Table 63.3 lists medications and physical stimuli that cause mast cell degranulation and histamine and/or prostaglandin D_2 release. These agents should be avoided.

Medications	Physical Stimuli
Alcohol	Rubbing of the skin
Aspirin	Extremes of water temperature
Opiates	
Polymyxin B	
Procaine	
Scopolamine	

Table 63.3. Medications and Physical Stimuli to Be Avoided in Patients

When massive amounts of mediators are released, generalized flushing, persistent diarrhea, or hypotension may ensue. Children with these symptoms require therapy directed against histamine and prostaglandin D_2. The H_1-receptor antagonists (chlorpheniramine) and H_2-receptor antagonists (cimetidine) may be required. In addition, indomethacin is required to inhabit prostaglandin biosynthesis.

Fortunately, with aging, the skin is no longer reactive, and most of the lesions disappear completely.

Juvenile Xanthogranulomas

For additional information about JXGs, see the previous section in this chapter on p. 522.

Warts and Xanthomas

For additional information about warts and xanthomas, see previous sections of this chapter on p. 519.

LESIONS THAT ARE NOT YELLOW, TAN, OR BROWN: FLESH-COLORED LESIONS

Three entities may present as flesh-colored papules: lichen striatus, lichen nitidus, and flat warts. When the papules are arranged linearly, streaming down an extremity or across the face or neck, lichen striatus should be considered. If the papules are not arranged linearly but are tiny pinpoint, flesh-colored papules, lichen nitidus should be considered, especially if a Köbner's phenomenon is present. Flat warts may be flesh-colored.

Lichen Striatus

Lichen striatus is an asymptomatic eruption of unknown cause. The flat-topped papules are arranged linearly and may be confluent. The lesions are flesh-colored to erythematous in Caucasians and hypopigmented in African-Americans. The eruption follows the long axis of an extremity (Fig. 63.5) or may involve any other part of the skin surface (especially the face). Because the eruption resolves spontaneously in 2 years, no treatment is necessary.

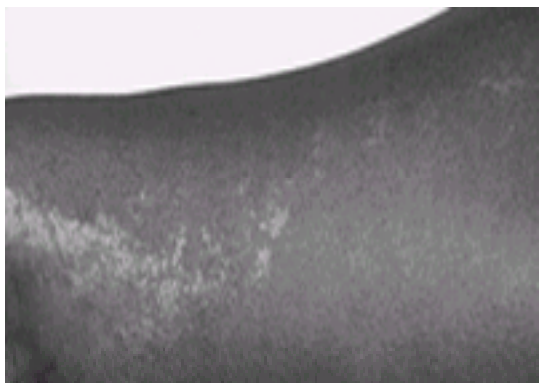

FIGURE 63.5. Hypopigmented papules running linearly along the long axis of the arm in a child with lichen striatus.

Lichen Nitidus

Lichen nitidus is characterized by tiny, pinpoint, flat-topped, flesh-colored papules (Fig. 63.6). The papules often are grouped and are found in scratch lines (i.e., the Köbner's phenomenon). Although any skin surface may be involved, the trunk and genitalia are common sites. The lesions persist for variable periods and generally do not respond to therapy.

FIGURE 63.6. Tiny flesh-colored to hypopigmented papules in a child with lichen nitidus. Note that the papules are arranged linearly (Köbner's phenomenon).

Flat Warts

See previous section about flat warts.

NON-FLESH-COLORED LESIONS

Lichen striatus can be composed of hypopigmented or erythematous papules arranged linearly. Red papules not arranged linearly and concentrated on exposed surfaces usually indicate the presence of insect bites. JXGs can be yellow or reddish-brown. They can be hypopigmented and brown–orange in African-American children. Xanthomas may be yellow or yellow–red. As discussed previously, the serum lipid levels are normal in patients with JXG and elevated in children with xanthomas.

NONCHARACTERISTIC PAPULES

Papules Arranged in Circles—Looks Like Tinea Corporis without Any Scaling

Granuloma Annulare

Granuloma annulare is thought to be an idiosyncratic response to trauma. The location of the changes (i.e., the shins, forearms, back of hands, ankles, and dorsum of the feet) seems to confirm this hypothesis. This skin change may begin as a flesh-colored or violaceous papule that clears centrally as the margins advance, or it may appear as a group of papules arranged in a ringlike configuration (Fig. 63.7). The central portion of the lesion is dusky or hyperpigmented. The key point on physical examination is the lack of scaling. This physical finding distinguishes granuloma annulare from tinea corporis and cannot be stressed enough. The border is firm on palpation, unlike tinea corporis. The rings can be 5 cm in diameter or larger.

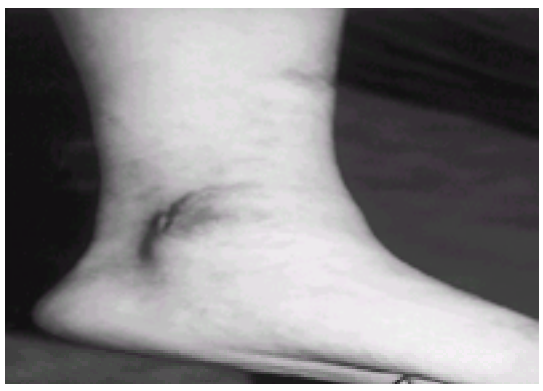

FIGURE 63.7. Note erythematous to violaceous papules arranged in a circle in this child with granuloma annulare.

A potassium hydroxide test would be definitive in ruling out tinea corporis. It would be difficult to obtain scales from a granuloma annulare lesion using this procedure. It is important to diagnose this entity correctly because one can reassure parents that three-quarters of lesions clear spontaneously within a 2-year period. Recurrences are common until children outgrow this tendency. Too often, treatment for tinea corporis is instituted unnecessarily.

If this algorithm has not helped in making a diagnosis, a consultant should be called. Many entities can be difficult to differentiate. A dermatologist often can resolve the matter, however, with a skin biopsy sent for histologic review.

Suggested Readings

Dillman AM, Miller RC, Hansen RC. Multiple pyogenic granulomata in childhood. Pediatr Dermatol 1991;8:28.

Felner EI, Steinberg JB, Weinberg AG. Subcutaneous granuloma annulare: a review of 47 cases. Pediatrics 1997;100:965.

Hernandez-Martin A, Baselga E, Drolet BA, et al. Juvenile xanthogranuloma. J Am Acad Dermatol 1997;36:355.

Lapins NA, Willoughby C, Helwin EB. Lichen nitidus: a study of 43 cases. Cutis 1978;21:634.

Maher-Wiese VL, Marmer EL, Grant-Kels JM. Xanthomas and the inherited hyperlipoproteinemias in children and adolescents. Pediatr Dermatol 1991;7:166.

Stein DH. Mastocytosis: a review. Pediatr Dermatol 1986;3:365.

Wells RS, Smith MA. The natural history of granuloma annulare. Br J Dermatol 1963;75:199.

CHAPTER 64

Rash—Papulosquamous Lesions

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PAPULOSQUAMOUS ERUPTIONS

Of skin conditions seen in a pediatric dermatology clinic, 10% are papulosquamous (i.e., have a papular and scaling component). The algorithm contained in this chapter should be used as a guide to differentiate these disorders (Table 64.1 and Fig. 64.1). Each key point that distinguishes one disease from another is discussed here.

Acrodermatitis enteropathica[a]	Pityriasis rosea (common)
Drug eruption, papulosquamous (common)[a]	Pityriasis rubra pilaris
Lichen nitidus	Psoriasis (common)
Lichen planus	Reiter's syndrome
Nummular eczema (common)	Seborrheic dermatitis
Parapsoriasis	Syphilis, secondary (common)

[a]Potentially life-threatening.

Table 64.1. Papulosquamous Skin Disorders

FIGURE 64.1. Algorithm to distinguish papulosquamous lesions.

Presence or Absence of Pruritus

The initial symptom that should be considered is pruritus. Pruritus is absent in the six conditions listed in Table 64.2. Palmar involvement is prominent in secondary syphilis and papulosquamous drug eruptions but is rare in pityriasis rosea. A positive rapid plasma reagin test (RPR) helps differentiate syphilis from pityriasis rosea and a drug eruption. Pityriasis rosea begins with a herald patch, followed by a truncal eruption in a “Christmas tree” distribution. Finally, the Köbner's phenomenon separates lichen nitidus from other entities.

Drug eruption—papulosquamous	Parapsoriasis
Lichen nitidus	Pityriasis rosea
Nummular eczema	Secondary syphilis

Table 64.2. Nonpruritic Papulosquamous Skin Disorders

Conditions That Lack Pruritus

Drug Eruption—Papulosquamous

The diagnosis of a drug eruption is based on the history of current or recent intake of a medication and the disappearance of the eruption after discontinuation of the medication. Drug eruptions may mimic lichen planus, pityriasis rosea, pityriasis rubra pilaris, psoriasis, seborrheic dermatitis, and syphilis (Table 64.3). The cutaneous manifestations of a drug eruption that mimics one of the previously described disorders, however, will be atypical (e.g., lack of a herald patch and typical truncal distribution in a pityriasis rosea look-alike drug eruption or lack of a violaceous color and feathery white buccal changes in a lichenoid drug eruption). Remember that drug eruptions may or may not itch and may or may not have palmar and plantar involvement.

Lichen Planus		*Pityriasis Rosea*
β-Blockers	Isoniazid	Barbiturates
Captopril	Naproxen	β-Blockers
Carbamazepine	α-Penicillamine	Captopril
Chloral hydrate	Phenytoin	Gold
Diazoxide	Spironolactone	Griseofulvin
Furosemide	Tetracyclines	Ketotifen
Gold		Metronidazole
Griseofulvin		Penicillin
Hydrochlorothiazide		Tripelennamine
Pityriasis Rubra Pilaris		*Psoriasis*
β-Blockers		Antimalarials
Seborrheic Dermatitis		β-Blockers
Contraceptives with progesterone (derived from 19-nortestosterone)		Lithium
Testosterone		? Nonsteroidal anti-inflammatory drugs
Syphilis		
Any drug		

Table 64.3. Eruptions That May Mimic Various Papulosquamous Lesions

Lichen Nitidus

Lichen nitidus is a common disorder of children, seen especially in African-Americans. There is a 4:1 male:female predominance. Lichen nitidus involves the abdomen, genitalia (shaft and glans), and extremities with tiny, pinpoint, sharply demarcated, flat-topped, flesh-colored papules. Often, these lesions are closely grouped and are linear. Linear grouping of lesions is caused by the KöAdobner's phenomenon (Table 64.4) (appearance of the primary lesion at sites of trauma), which often occurs in lichen nitidus (Fig. 64.2). The lesions generally are nonpruritic. The course is variable, and the cause is unknown. Therapy is not warranted.

Lichen nitidus
Lichen planus
Psoriasis

Table 64.4. Conditions That Feature the Köbner's Phenomenon

FIGURE 64.2. Note flesh-colored papules in this African-American patient with lichen nitidus. The Köbner's phenomenon also is seen (papules arranged linearly in a scratch).

Nummular Eczema (Xerosis)

For more information about nummular eczema, see Chapter 99.

Parapsoriasis

Parapsoriasis is an uncommon pediatric skin condition. When it occurs, however, the course is chronic and, on rare occasions, may progress to cutaneous lymphoma. The appearance of this eruption is easily mistaken for nummular eczema, psoriasis, tinea corporis, or a lichenoid change. Small oval scaling, erythematous (Fig. 64.3) to yellow–brown macules are concentrated on the trunk. The skin lesions are asymptomatic, and the patient feels healthy.

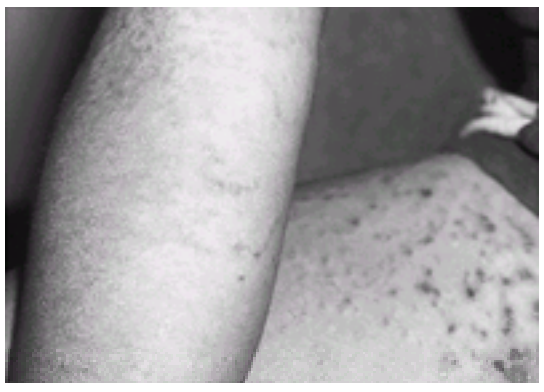

FIGURE 64.3. Red, scaling lesions on arm of child with parapsoriasis.

Treatment is unnecessary because the disease is asymptomatic. Topical steroids may be helpful but may not clear the skin changes. The eruption clears spontaneously after varying periods.

Pityriasis Rosea

For more information about pityriasis rosea, see Chapter 99.

Secondary Syphilis

The secondary phase of syphilis is a great mimicker. Therefore, one must suspect this condition to make the correct diagnosis. The eruption may be localized to the trunk, palms, and soles, as well as to any other skin surface. Other clues should be sought by history and physical examination (a primary chancre, condyloma lata, or white mucous patches on the tongue, buccal, and labial surfaces). Generalized lymphadenopathy usually is present. It may be difficult to differentiate secondary syphilis and pityriasis rosea; however, Table 99.10 (see Chapter 99) may be helpful in separating the two entities clinically.

A positive RPR or fluorescent treponemal antibody (FTA) makes the diagnosis. Remember, a false-negative RPR can occur with antibody excess (the prozone phenomenon). Therefore, dilution of the specimen by the laboratory should be requested if the presence of syphilis is highly suspected. This simple maneuver will result in a positive test for the presence of syphilis.

Color of the Skin Eruption and Pruritus

The eye can discern subtle differences in color. A pruritic papulosquamous eruption that does not look erythematous should suggest four disorders. First, a violaceous (bluish-red) or purple appearance generally indicates lichen planus or a lichenoid drug eruption. However, tones of yellow or salmon (orange–red) suggest the presence of seborrheic dermatitis or an unusual disorder called pityriasis rubra pilaris (PRP). The latter two diseases can be differentiated by looking for yellow thickening of the palms and soles (i.e., in PRP) or knowing that seborrheic dermatitis occurs before 6 months of age or after puberty. Lichen nitidus is obvious when tiny, discrete, flesh-colored papules (white papules in

African-Americans) are found, with some arranged linearly (Köbner's phenomenon).

Violaceous or Yellow- or Salmon-Colored (Orange–Red) Eruptions or Flesh-Colored (Nonerythematous) Eruptions

Lichen Planus

Lichen planus is seen occasionally in pediatric patients as a chronic, pruritic, reddish-blue (violaceous) to purplish eruption. Two to three percent of cases occur in patients younger 20 years old.

The eruption generally involves the flexors of the wrist, forearms, and legs, especially the dorsum of the foot and ankles. The lesions appear as small, violaceous, shiny, flat-topped, polygonal papules (Fig. 64.4). The surface of these papules may have white crosshatching, called Wickham's striae. Lesions may occur in sites of trauma or injury (Köbner's phenomenon). The scalp may be involved, often resulting in a scarring alopecia, called pseudopelade. It is important to examine the buccal mucous membrane for a reticulated or lacelike pattern of white papules or streaks. This finding is characteristic for lichen planus. The nails are often pitted, dystrophic, or ridged (pterygium nails). The lesions in lichen planus can be vesicular or bullous. Hypertrophic and linear lesions occur but are less common. Persistent, severe, postinflammatory hyperpigmentation is common in African-Americans. In two-thirds of patients, the lesions clear within 8 to 15 months. The cause of the disorder is unknown. Topical therapy with steroids can be helpful.

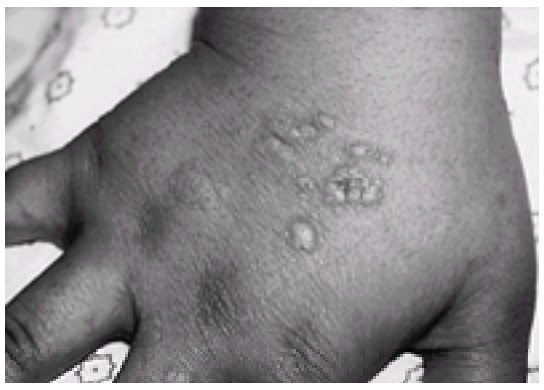

FIGURE 64.4. Note child with red–blue color of flat-topped papules representative of lichen planus.

Seborrheic Dermatitis

For more information about seborrheic dermatitis, see Chapter 99.

Pityriasis Rubra Pilaris

Pityriasis rubra pilaris is characterized by follicular papules and yellow–orange skin that surrounds islands of normal skin. Of patients with PRP, 30% are children.

The onset of the disease is gradual, beginning in the scalp and spreading to involve the face and ears. Acuminate follicular papules with keratotic plugs occur on the back of the fingers, side of the neck, and extensors of the extremities. The skin is generally salmon-colored and scaling. As the eruption progresses, it surrounds islands of normal skin. Yellow thickening of the palms and soles is characteristic (Fig. 64.5). Three subtypes have been described: the familial form has its onset in infancy and childhood; a localized type is found in 60% of cases; and the acquired form occurs in persons more than 15 years of age. The cause of this disease is unknown, although it is a disorder of keratinization. The condition responds to vitamin A and its derivatives.

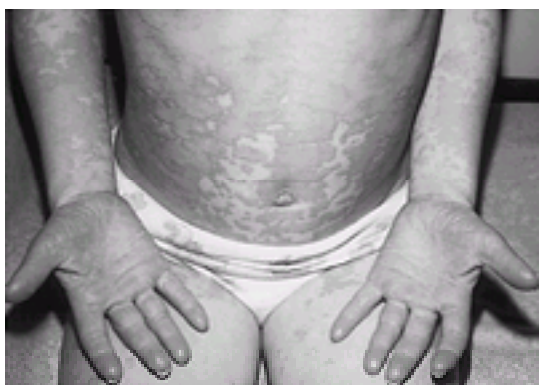

FIGURE 64.5. Hyperkeratosis of palms and islands of normal skin seen in this child with PRP.

Differentiating the Pruritic, Red Papulosquamous Lesions

Pityriasis rosea also should be included in this part of the algorithm. Because the rash of pityriasis rosea may or may not

itch, the first differentiating point is to inquire about a history of a herald patch and look for the characteristic Christmas tree distribution. If one or both is present, a diagnosis of pityriasis is made. If not, one must look for the Köbner's phenomenon. The Köbner's phenomenon is defined as the appearance of the existing rash in areas of traumatized skin (e.g., in scratches, abrasions, blistered sunburns). This phenomenon is discussed further in this chapter under Psoriasis.

Another clue to the presence of psoriasis is the finding of abnormal nails (e.g., nail pits, flaking, thickened nails). Reiter's syndrome and acrodermatitis enteropathica may also manifest nail abnormalities. These entities can be differentiated from one another by a history or finding of uveitis and/or urethritis (Reiter's syndrome) or diarrhea and/or hair loss (acrodermatitis enteropathica).

Psoriasis

Psoriasis is a chronic papulosquamous disease that makes up 4% of all skin disorders encountered in children. The female:male ratio is 2:1. There is a predisposition for involvement of the scalp, perineum, and the extensor surfaces of the body, particularly the elbows and knees.

One-third of adults with psoriasis experience onset of disease in childhood or adolescence. Psoriasis occurs in 12% of children before the age of 10 years. The major human leukocyte histocompatibility antigens (HLA) are important genetic markers of psoriasis. Of these, HLA-B13 and HLA-BW17 are associated with early onset of disease. The HLA-B13 antigen is associated with a history of antecedent streptococcal infections. The HLA-BW17 antigen is more commonly identified with extensive skin involvement and a strong familial history.

Psoriasis occurs in three forms during childhood: guttate, erythrodermic, and pustular. Any or all of these types may develop with silvery scales into the chronic, plaque-type psoriasis (Fig. 64.6). When a scale is removed, pinpoint areas of bleeding occur on the surface (Auspitz sign). Guttate psoriasis is the most common form, occurring in childhood. Guttate or droplike erythematous papules are scattered over the body. The characteristic silvery scale is only minimally expressed, and the lesions may appear quite red. This form often is preceded by a streptococcal infection. Infants often have involvement in the diaper area as well. Erythrodermic psoriasis is less common and more severe. Onset may be abrupt or gradual, with a diffuse erythema and severe desquamation. In the growing child, there may be associated failure to thrive. Pustular psoriasis is rare and the least commonly occurring form of psoriasis seen in children. Various sized sterile and superficial pustules develop on an erythrodermic background.

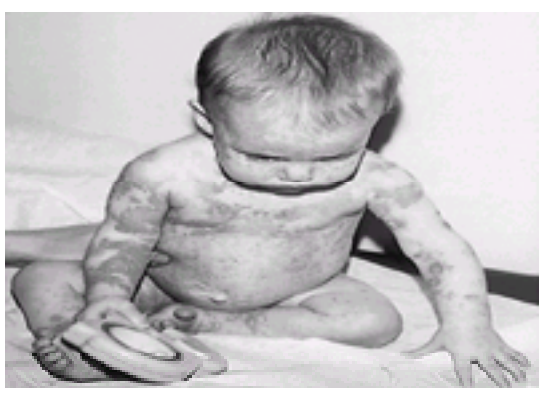

FIGURE 64.6. Plaque-type psoriasis in this infant. Note scaling in scalp.

Characteristically small, pitted lesions are seen on the nails in 25 to 50% of patients in all forms of the condition. Eighty percent of children have scalp involvement, especially at the hair margins. A small number of patients develop arthritis between 9 and 12 years of age; some develop it before the onset of the skin eruption. The distal interphalangeal joints of the hands and feet are involved most often. Therapy with topical agents, including steroids, tar derivatives, emollients, and ultraviolet light, help slow the turnover rate of the epidermis.

Reiter's Syndrome

The skin changes seen in Reiter's syndrome look much like those in psoriasis. A symmetric arthritis of major joints, uveitis, and urethritis complete the syndrome. Although most cases occur in young adult men, on occasion, the syndrome will be seen in adolescents. Only 10 cases have been reported in children under 12 years of age. Ninety percent of patients are HLA-B27 positive. A postinfectious cause has been hypothesized.

The palms and soles are the major sites of involvement. Yellow, scaly, hyperkeratotic lesions appear on an erythematous base in those locations. The skin lesions may begin as macules, vesicles, or pustules. The palmar plantar changes have been called keratoderma blennorrhagicum. The scalp and penis also are characteristically involved with psoriasiform lesions. Erythema and superficial ulcerations may be present in the mouth. Abnormalities of the nails (e.g., dystrophy, onycholysis) are common. (See the section on Reiter's syndrome for therapy.)

Acrodermatitis Enteropathica

Acrodermatitis enteropathica is characterized by skin rash, diarrhea, and alopecia. The condition is caused by poor absorption of zinc, with plasma zinc levels under 50 µg/dL. The disease generally begins around 9 months of age (1 week to 20 months).

The infant usually presents with a psoriasiform diaper eruption, followed by similar involvement of periorificial (eyes, ears, nose, and mouth) and acral skin. The skin often is eroded and crusted. These changes may be confused with a severe candida infection (Table 64.5) or impetigo. Because of involvement of the digits and periungual tissues, nail dystrophies often are present. Hair is lost from the scalp, brows, and lashes. The children are irritable, photophobic, and apathetic. Growth retardation is common. Rapid growth occurs with zinc sulfate or gluconate 5 mg/kg per day.

Differential Diagnosis	
Mucocutaneous candidiasis	Histiocytosis X
Biotin deficiency	Multiple carboxylase deficiency
Essential fatty acid deficiency	Psoriasis
Glucagonoma syndrome	Seborrheic dermatitis

Table 64.5. Acrodermatitis Enteropathica

If this algorithm does not lead to a clear diagnosis, many of the entities can be clarified with a skin biopsy or referral to a dermatologist.

Suggested Readings

ACRODERMATITIS ENTEROPATHICA

Moynahan EJ. Acrodermatitis enteropathica: a lethal inherited human zinc-deficiency disorder. Lancet 1974;2:399.

LICHEN NITIDUS

Lapius NA, Willoushbis C, Helwis EB. Lichen nitidus: a study of 43 cases. Cutis 1978;21:634.

PARAPSORIASIS

Lambert WE, Everett MA. The nosology of parapsoriasis (Review). J Am Acad Derm 1981;5:373–395.

PITYRIASIS ROSEA

Parsons JM. Pityriasis rosea update: 1986. J Am Acad Dermatol 1986; 15:159–167.

PITYRIASIS RUBRA PILARIS

Gelmetti C, Shiuma AA, Cerri D, et al. Pityriasis rubra pilaris in childhood: a long-term study of 29 cases. Pediatr Dermatol 1986;3:446–451.

PSORIASIS

Farber EM, Mullen RH, Jacobs HA, et al. Infantile psoriasis: a follow-up study. Pediatr Dermatol 1986;3:237–243.

Rasmussen JE. Psoriasis in children. Dermatol Clin North Am 1986; 4:99–106.

REITER'S SYNDROME

Vergnani RJ, Smith RS. Reiter's syndrome in a child. Arch Ophthalmol 1974;91:165.

SYPHILIS

Morbidity and Mortality Weekly Report; 1998, Guidelines for treatment of sexually transmitted diseases. US Department of Health and Human Services, Atlanta, 1997.

CHAPTER 65

Rash—Purpura

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Blood in the skin or mucosal membranes is referred to as purpura. The sudden, unexplained appearance of purpura in a child is a finding that is disturbing to parents and may prompt an immediate visit to the local emergency department (ED) or physician. This finding also is particularly disturbing to physicians because it may be the presenting sign of many diseases, some benign and others life-threatening, some treated easily and others requiring complex therapy.

When the onset of purpura is accompanied by massive hemorrhage or by bleeding in a critical site such as the central nervous system (CNS), the patient is easily recognized as being dangerously ill and appropriate measures are taken rapidly. When purpura is the only presenting complaint, however, the patient should still be considered as having the potential for life-threatening sequelae of disordered hemostasis. The cause of purpura should be established as rapidly as possible because early treatment may lead to a more favorable outcome. Fortunately, an understanding of the pathophysiology of purpura, a careful history and physical examination, and an appropriate laboratory evaluation usually establish the cause of the bruising. This chapter presents the initial assessment and differential diagnosis of children with purpura. The management of emergency situations associated with these symptoms is discussed in Chapter 87.

PATHOPHYSIOLOGY

Purpura can be subdivided on the basis of its appearance into petechiae and ecchymoses. Petechiae are small (less than 3 mm in diameter), reddish-purple, macular lesions. Ecchymoses are larger lesions that often are tender and, when severe, may be raised above the level of the skin surface. Purpuric lesions do not blanch, a characteristic that distinguishes them from vascular dilation and vascular anomalies. Under normal conditions, a purpuric lesion resolves in a predictable manner. The purple color gradually fades to golden brown as hemosiderin is formed. The golden brown color may then take as long as 6 weeks to resolve. This progressive and characteristic resolution of purpura often is helpful in determining the time and cause of the injury that resulted in purpura. For example, ecchymoses in various stages of resolution may be the major diagnostic finding in cases of child abuse with repeated assaults.

Complex mechanisms maintain vascular integrity and stop the flow of blood when a blood vessel is damaged. Vitamin C and other factors that affect collagen synthesis are required for normal formation of connective tissue within the vessel walls. When a blood vessel is injured, vasoconstriction and retraction usually occur immediately and decrease the flow of blood to the affected area. Facilitated by von Willebrand factor, platelets adhere to the subendothelium of the damaged wall and, in response to the exposed subendothelial collagen, release adenosine diphosphate (ADP). This release reaction causes platelet aggregation at the site of the injury and the formation of a platelet plug that is responsible for primary hemostasis. A decrease in the number of circulating platelets or an intrinsic or secondary alteration in platelet metabolism and aggregation may disrupt this early phase of hemostasis and result in localized or disseminated purpura.

The intrinsic pathway of coagulation also is activated by the exposed collagen (Fig. 65.1). A sequence of enzymatic reactions, beginning with the binding of factor XII to the exposed subendothelium, leads to the formation of a fibrin clot at the site of the injury. A related pathway (extrinsic pathway) is activated by tissue thromboplastin and contributes to the development of the fibrin clot. Factors XII, XI, IX, and VIII contribute exclusively to the intrinsic pathway, whereas factor VII is involved only in the extrinsic pathway. Factors X, V, II (prothrombin), and I (fibrinogen) are shared by both pathways. Defects in the clotting sequence interfere with the formation of a normal clot behind the platelet plug (secondary hemostasis). As is the case in platelet disorders, alterations in the coagulation pathway may be caused by intrinsic abnormalities of the clotting factors or by abnormalities resulting from systemic diseases.

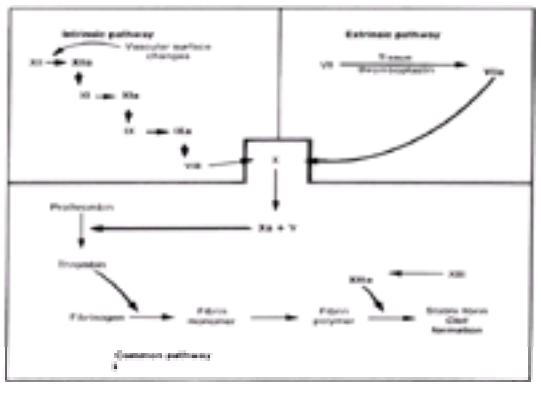

FIGURE 65.1. A simplified version of the coagulation “cascade.” An abnormality in the extrinsic pathway results in a prolonged PT. An abnormality in the intrinsic pathway results in a prolonged PTT. An abnormality in the common pathway results in prolongation of both the PT and the PTT.

Disruption of the normal hemostatic mechanism at any point may result in purpura. Although the pathophysiology is complex and foreboding to most physicians when presented in detail, a basic understanding of altered hemostasis enables the physician to categorize the wide variety of purpuric disorders. With this categorization in mind, the physician becomes more efficient in obtaining a history, performing a physical examination, and selecting the appropriate laboratory tests.

DIFFERENTIAL DIAGNOSIS

A purpuric eruption may result from loss of vascular integrity, thrombocytopenia, disorders of platelet function, or deficiencies of clotting factors. Myriad disorders may cause purpura, but only a few are particularly common (Table 65.1), including trauma, infections, Henoch-Schönlein purpura (HSP), and idiopathic thrombocytopenic purpura (ITP).

Disruption of Vascular Integrity	*Platelet Deficiency or Function Disorders*
Trauma	Idiopathic thrombocytopenic purpura
Viral infections	Sepsis
Henoch-Schönlein purpura	Drug-associated disorders
Rickettsial infection	*Factor Deficiencies*
	Hemophilia

Table 65.1. Common Causes of Purpura

Loss of Vascular Integrity

Purpura may be caused by numerous disorders that disrupt vascular integrity (Table 65.2). The most common cause of purpura from vascular injury in children is trauma. Most active children have bruises, particularly on the anterior aspect of the lower extremities. Occasionally, however, a parent will bring a child to a physician with the complaint that the child bruises unusually easily. A thorough history and physical examination often are sufficient to distinguish the patient who requires further evaluation from the patient who requires only reassurance about normal childhood bruising from trauma. The child's level of activity should be correlated with the degree of bruising. Ecchymoses that might be acceptable in a child who enjoys climbing trees would be most surprising in a child who spends most of his or her leisure time reading. In addition, bruising on an area of the body rarely exposed to trauma (e.g., chest, abdomen, back) or bruising out of proportion to the degree of trauma should be evaluated further. Large, raised ecchymoses rarely are seen in the absence of significant trauma that is easily recalled. A history of sudden onset of excessive bruising without an associated change in activity also suggests an underlying disorder. Even when the history indicates that the ecchymoses can be attributed to repeated episodes of minor trauma, the finding of numerous bruises may be a clue to a subtle neurologic disorder that causes unusual clumsiness.

Trauma: accidental; child abuse[a]
Infection: viral exanthems, infectious mononucleosis, bacterial endocarditis,[a] rickettsial disease,[a] streptococcal infection
Drugs and toxins[a]
Henoch-Schönlein purpura
Vitamin C deficiency
Letterer-Siwe disease
Ehlers-Danlos syndrome
Miscellaneous: acute glomerulonephritis, rheumatic fever, collagen vascular diseases

[a]Conditions that may be life-threatening.

Table 65.2. Causes of Purpura in Children Secondary to Disruption of Vascular Integrity

Foremost in the mind of a physician who cares for children must be the consideration that bruising is caused by child abuse. Suspicion should be raised if the child has presented in the past with unexplained bruising or suspicious injuries; if explanations of the bruises are inconsistent; if bruises are confined to the buttocks, back, or face; if bruises conform to the shape of a belt or cord; or if bruises are in various stages of resolution (see Chapter 128).

Purpura can be the initial manifestation of numerous infectious processes. The purpura may result from a disruption of the vascular integrity by the infecting agent or from the body's reaction to the agent, an infection-induced thrombocytopenia, or disseminated intravascular coagulation (DIC) initiated by the septic process. The latter two disorders are discussed later in this chapter. Capillary damage that results in petechiae or ecchymoses can occur with the viral exanthems and is especially common with rubeola. The child with infectious mononucleosis, bacterial endocarditis, a rickettsial infection, or a streptococcal infection can present with purpura in the absence of coagulation or platelet abnormalities. Rocky Mountain spotted fever should be strongly considered when the patient is from an area in which the disease is endemic or when there is a history of tick exposure, especially in the months of April through October.

The most serious infection that can cause purpura is meningococcemia, and this disorder always must be considered in a child with purpuric lesions. The rapidity with which meningococcemia can progress may warrant the institution of antibiotic therapy in any moderately ill child with purpura until results of cultures are available. Purpura fulminans is a particularly severe form of bleeding caused in part by loss of vascular integrity and may accompany meningococcemia as well as septicemia from other organisms, scarlet fever, varicella, and rubeola. This disorder often is found in association with DIC and is characterized by the sudden onset of large ecchymoses and rapid development of gangrene of the extremities.

Numerous drugs and toxins can cause purpura as a result of increased capillary fragility or vasculitis. The parents of a child with purpura should be questioned closely regarding the recent use of any medications, including over-the-counter drugs and “home remedies.” Drugs that have been implicated include the sulfonamides, iodides, belladonna, bismuth, mercurial compounds, the penicillins, phenacetin, and chloral hydrate. Corticosteroid treatment can cause the appearance of benign purpura, especially striated purpuric lesions just above the buttocks. The lesions resolve with discontinuation of corticosteroid therapy. The appearance of these lesions in a child not taking corticosteroids should cause the physician to search for endogenous corticosteroid production, as in Cushing's disease. Vitamin C deficiency also can present with purpura that ranges from scattered petechiae to substantial ecchymoses, particularly on the lower extremities. Scurvy is rare in the United States but can be seen in patients who receive hyperalimentation with inadequate vitamin C supplementation or in patients with iron overload. The purpuric lesions of scurvy heal rapidly after the administration of vitamin C.

Purpura that results from an aseptic vasculitis within the corium may be the presenting symptom of HSP. The purpuric lesions often are accompanied by pink or brownish-pink macules or maculopapules that later may develop central areas of hemorrhage. They tend to coalesce and usually are located on the lower extremities, buttocks, and lower back. The platelet count is normal in uncomplicated HSP, as are the prothrombin (PT) and partial thromboplastin (PTT) times. Anaphylactoid purpura that resembles HSP also can accompany acute streptococcal infections, rheumatic fever, acute glomerulonephritis, and collagen vascular disorders (see Chapter 101).

Rare disorders of childhood that may be associated with purpura from loss of vascular integrity include Letterer-Siwe disease and Ehlers-Danlos syndrome. Letterer-Siwe disease is a histiocytic disorder with brown, crusted vesiculopapular skin lesions that often are purpuric. Petechiae may also be present. The pathogenesis of purpura in this disorder is uncertain but may be related to the widespread histiocytic infiltration. Ehlers-Danlos syndrome is an unusual defect in collagen synthesis. The altered blood vessel architecture causes capillary hemorrhage; rupture of major blood vessels may occur.

Platelet Disorders

Thrombocytopenia

Thrombocytopenia in childhood may come from shortened platelet survival, decreased platelet production, or platelet sequestration (Table 65.3). However, certain illnesses and drugs may cause thrombocytopenia by more than one mechanism. For example, thrombocytopenia that accompanies viral or bacterial infections may come from decreased platelet production, antiplatelet antibody formation, or the presence of DIC. Similarly, thrombocytopenia associated with sulfonamide therapy may come from diminished platelet production or immune-mediated platelet destruction. Consequently, detailed investigation may be required to determine the mechanism of thrombocytopenia and the appropriate treatment.

Table 65.3. Causes of Childhood Purpura Secondary to Platelet and Coagulation Abnormalities

Increased Platelet Destruction

The most common form of thrombocytopenia in childhood is ITP. This immunologic disorder usually is characterized by the acute onset of petechiae and ecchymoses, although symptoms occasionally occur more gradually. Epistaxis occurs in 10 to 20% of cases. Other bleeding manifestations are much less common. Although ITP occurs in children of all ages, most cases are seen between the ages of 2 and 6 years.

ITP may have no antecedent illness or may follow a mild viral illness by 2 to 6 weeks. The disorder is associated with human immunodeficiency virus (HIV), infectious mononucleosis, cytomegalovirus (CMV) infection, rubeola, mumps, and varicella. A similar relationship between ITP and rubeola and rubella immunization has been observed. ITP may be the first manifestation of a systemic immunologic disorder such as systemic lupus erythematosus (SLE). A careful search for this association is particularly important in older children. The association of ITP with autoimmune hemolytic anemia or neutropenia is called Evans syndrome.

The physical examination of the child with ITP reveals few abnormalities other than purpura. Enlargement of the spleen occurs rarely. At the onset of purpura, the platelet count usually is less than 20,000/mm ^{3}. In the absence of prolonged bleeding or antibodies to other hematologic elements, the hemoglobin concentration and white blood cell (WBC) count are normal. A bone marrow aspirate is sometimes performed to assess megakaryocyte production because the clinical presentation of aplastic anemia or acute leukemia rarely may be indistinguishable from ITP. A bone marrow aspirate is particularly important if treatment with corticosteroids is contemplated because this therapy may obscure the diagnosis of acute leukemia, thereby delaying appropriate therapy and adversely affecting the outcome. Other laboratory studies that may be performed include an antinuclear antibody (ANA) titer as a screening test for collagen vascular disorders and a reticulocyte count and direct Coombs test to detect immune hemolytic anemia.

Thrombocytopenia from shortened platelet survival may be caused by fibrin deposition and platelet consumption as found in DIC and also in hemolytic uremic syndrome (HUS), which is characterized by a microangiopathic anemia and uremia. In this, pallor, purpura, and signs of renal failure usually follow a prodrome of abdominal pain and diarrhea. Thrombotic thrombocytopenic purpura (TTP) is a disorder that resembles HUS in its hematologic aspects but more commonly occurs in adults than in children. However, this disorder has been described in the pediatric age group and may be particularly difficult to distinguish from HUS, although neurologic rather than renal symptoms are more prominent in TTP.

Infants with Wiskott-Aldrich syndrome, an X-linked recessive immunodeficiency disorder, may develop thrombocytopenic purpura beginning in the newborn period. Shortened platelet survival in this disease comes from an intrinsic platelet abnormality. The survival of transfused donor platelets is normal in children with Wiskott-Aldrich syndrome, whereas survival of autologous platelets is shortened. Numerous drugs have been reported to cause thrombocytopenia by the formation of platelet antibodies with resultant increased platelet destruction. The drugs that cause thrombocytopenia that are most commonly used in children include sulfa compounds (including trimethoprim–sulfamethoxazole), valproic acid, phenytoin, acetazolamide, carbamazepine, and quinidine.

Decreased Platelet Production

Diseases associated with decreased amounts of functional bone marrow also may present with thrombocytopenia and purpura. Most notable in this group are the leukemias, neuroblastoma, histiocytosis, and osteopetrosis. Decreased platelet production also may be the result of abnormalities of development of the hematopoietic stem cell (aplastic anemia, Fanconi's anemia, and thrombocytopenia and absent radii), ineffective megakaryocyte development (megaloblastic anemias), or rarely, absence of a humoral factor (presumed thrombopoietin deficiency). Although pancytopenia often is present at the time of diagnosis of bone marrow disorders such as leukemia and aplastic anemia, thrombocytopenia may precede notable alterations in other elements in the peripheral blood.

Numerous drugs have been associated with thrombocytopenia because of decreased platelet production. Any drug capable of causing general bone marrow suppression can produce thrombocytopenia (most antibiotics, anticonvulsants, thiazide diuretics, and the like). Drugs that specifically inhibit megakaryocyte production include the chlorothiazides, estrogenic hormones, ethanol, and tolbutamide.

The circulating platelets in disorders of platelet production usually are older and metabolically less active than those found in most diseases of shortened platelet survival. Consequently, spontaneous purpura often appears at a platelet count of 25,000 to 40,000/mm^3 in leukemia or aplastic anemia but is rare after the first week of ITP unless the platelet count is less than 20,000/mm^3.

Platelet Sequestration

Splenomegaly that comes from numerous causes (e.g., portal hypertension, storage diseases) can result in sequestration of platelets and thrombocytopenia. The spleen is markedly enlarged and very firm in these disorders. Purpura that comes from platelet sequestration alone is rare because the platelet count usually does not fall below 40,000/mm ^{3}. Bleeding may occur, however, when the platelet sequestration is associated with liver disease and clotting abnormalities. Platelet sequestration and consumption also can occur in large hemangiomas (Kasabach-Merritt syndrome).

Disorders of Platelet Function

A clinical picture similar to that seen with thrombocytopenia can occur with a normal platelet count in the presence of a qualitative or functional platelet abnormality. These disorders can be congenital or acquired and, when congenital, can present in infancy with prolonged oozing from venipuncture sites or the umbilical cord, ecchymoses, and petechiae. Glanzmann's thrombasthenia is an autosomal-recessive disorder in which the platelet count is normal but the bleeding time is prolonged, clot retraction is poor, and platelet aggregation and adhesion are absent. Other inherited abnormalities of platelet metabolism (storage pool disease, aspirinlike defect, Bernard-Soulier syndrome) may be associated with purpura, although bleeding is generally not as severe as it is in Glanzmann's thrombasthenia. In addition to shortened platelet survival, platelet dysfunction is found in Wiskott-Aldrich syndrome.

Acquired platelet dysfunction with purpura can occur in the presence of uremia or liver dysfunction and also can be caused by certain medications. Aspirin is the best known of the drugs that cause platelet dysfunction; a single dose of aspirin can alter platelet function for as long as 9 to 10 days. Platelet dysfunction also has been associated with antihistamines, propranolol, phenothiazines, glycerol, guaifenesin, and carbenicillin. These drugs interfere with the release of endogenous ADP and inhibit platelet aggregation and adhesion. Unlike aspirin, however, they cause few, if any, clinical problems.

Factor Deficiencies

Purpura can be the presenting symptom of a congenital or acquired deficiency of coagulation factors. The most commonly encountered congenital deficiencies are von Willebrand's disease, hemophilia A (factor VIII deficiency), and hemophilia B (factor IX deficiency, Christmas disease). Although the latter two disorders have an X-linked recessive mode of inheritance, the de novo appearance of coagulopathy is not uncommon, particularly in children with severe hemophilia A (factor VIII activity less than 1%). Therefore, a family history of affected males may be helpful in establishing the diagnosis of hemophilia, but the absence of such a history does not eliminate this diagnostic possibility.

Congenital Deficiencies

Children with hemophilia often are detected when they develop purpura either spontaneously or after mild trauma. The diagnosis of hemophilia also should be entertained in newborns who develop excessive bleeding after circumcision and in infants with prolonged bleeding from lacerations of the lip, tongue, or frenulum. Prompt recognition of the disorder at this early age allows careful surveillance, appropriate treatment, and early genetic counseling for parents.

Coagulation tests in children with hemophilia A and B reveal a prolonged PTT and normal PT. The bleeding time is usually normal. Specific factor assays will define the particular abnormality. Special care should be taken in establishing the diagnosis of factor IX deficiency in the young infant because the low factor IX levels found in normal infants in the first few days of life may overlap with the factor IX levels found in mild hemophilia B.

Less common congenital factor deficiencies that may cause purpura in children include fibrinogen and factors II (prothrombin), V, VII, X, XI, and XIII. As in hemophilia, specific factor assays will identify the particular abnormality. Alterations in fibrinogen function (dysfibrinogenemias) also are associated with purpura. Fibrinogen levels determined by clotting assay usually are reduced moderately in these disorders.

Von Willebrand's disease is a common bleeding disorder caused by an alteration that adversely affects platelet function as well as clotting. The severity of this autosomal-dominant disorder is extremely variable among affected persons. Although some patients may have spontaneous purpura, others remain asymptomatic and are discovered only after the diagnosis of von Willebrand's disease in a close relative leads to laboratory investigation of other family members. Occasionally, von Willebrand's disease is uncovered when an acquired alteration of hemostasis is superimposed on the inherited abnormality. For example, bruising occurs very easily after aspirin ingestion in many patients with von Willebrand's disease. As in other disorders that affect platelet function, bleeding from mucosal surfaces (epistaxis, menorrhagia) is prominent in von Willebrand's disease. The laboratory abnormalities in von Willebrand's disease are variable and may fluctuate from week to week in the same patient. In its classical form, the disease is characterized by prolongation of the bleeding time, increased PTT, decreased levels of factor VIII coagulant activity and von Willebrand factor antigen, and diminished aggregation of normal platelets when ristocetin is added to the patient's plasma (von Willebrand's or ristocetin cofactor activity). In practice, however, only one or two of the laboratory abnormalities may be found. Indeed, several determinations may be required to detect an abnormality or to confirm the diagnosis of von Willebrand's disease.

Acquired Deficiencies

Causes of acquired deficiencies of clotting factors include DIC, liver disease, vitamin K deficiency, circulating anticoagulants, uremia, and cyanotic congenital heart disease. DIC is a potential complication of infection (bacterial, viral, or rickettsial), extensive burns, severe trauma, malignancies (especially acute promyelocytic leukemia), snake and insect bites, shock, and heat stroke. In DIC, the intravascular consumption of clotting factors may cause purpura because of factor depletion and, in severe cases, may lead to widespread, rapidly progressing purpuric lesions (purpura

fulminans) associated with thrombosis or emboli. Although other signs of serious illness usually are present in the child with purpura caused by DIC, fever and purpura may be the only significant findings in the early stages of severe bacterial infections such as meningococcemia. Further investigations and appropriate therapy should proceed rapidly in such instances.

Laboratory abnormalities in DIC include one or more of the following: a decreased platelet count, prolonged PT and PTT, decreased fibrinogen level, and elevated fibrin split products. A microangiopathic anemia with red cell fragmentation also may be present.

Coagulopathies caused by severe hepatocellular disease or vitamin K deficiency can present with some of the same clinical and laboratory findings as DIC. A comparison of the laboratory values in these three disorders is shown in Table 65.4. Hepatocellular disorders that may cause purpura include Reye syndrome, Wilson's disease, and toxic or infectious hepatitis. Vitamin K deficiency may be associated with malabsorption and chronic diarrhea. Purpura caused by warfarin (Coumadin) therapy or ingestion can resemble vitamin K deficiency clinically. Hemorrhagic disease of the newborn that comes from decreased vitamin K stores at birth can be prevented by the administration of vitamin K routinely following delivery. This important step in normal newborn care may be overlooked, however, when other problems develop in the delivery room, and a careful review of the records may be necessary to ensure that the young infant with purpura actually received vitamin K.

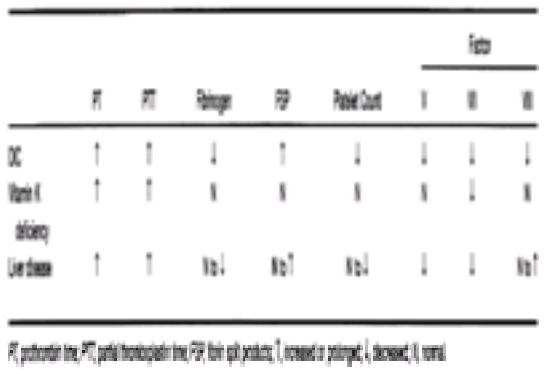

Table 65.4. Comparison of Laboratory Values in Disseminated Intravascular Coagulation (DIC), Liver Disease, and Vitamin K Deficiency

Circulating anticoagulants in children are associated with viral infections, malignancies, and collagen vascular disorders. They usually are characterized by a prolonged PTT that fails to correct with the addition of normal plasma. Because most acquired inhibitors in children, particularly lupus anticoagulants, are not associated with increased bleeding, the identification of an inhibitor in a patient with purpura should not preclude an investigation of other coagulation abnormalities.

Numerous coagulation abnormalities have been demonstrated in vitro in patients with renal disease. However, bleeding is most commonly related to altered platelet function rather than to defects in the fluid phase of coagulation. Abnormalities that resemble those found in DIC have been associated with cyanotic congenital heart disease. Although the coagulopathy seems to be related to the degree of polycythemia, the underlying mechanism remains unidentified.

EVALUATION AND DECISION

The evaluation of a child with purpura must combine speed and skill. The diagnostic approach is outlined in Figure 65.2. Purpura can be the initial sign of a life-threatening meningococcal infection, requiring immediate treatment, or the first sign of child abuse, requiring patient, thorough investigation. The initial approach should be dictated by the general appearance of the child and the presenting vital signs. A well-appearing child with purpura and normal vital signs can be approached with less urgency than a febrile, lethargic child with purpura or a child with hemophilia, purpura of the neck, and respiratory compromise.

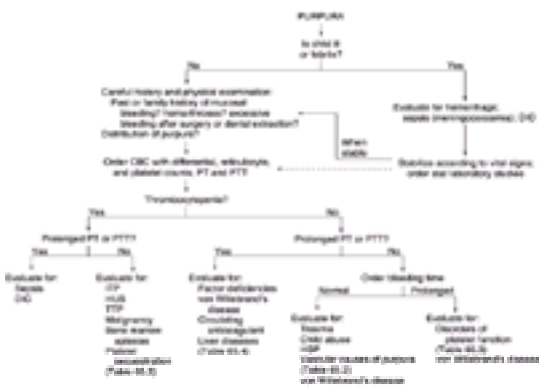

FIGURE 65.2. The approach to the child with purpura. *CBC,* complete blood count; *PT,* prothrombin time; *PTT,* partial thromboplastin time; *DIC,* disseminated intravascular coagulation; *ITP,* idiopathic thrombocytopenic purpura; *HUS,* hemolytic uremic syndrome; *TTP,* thrombotic thrombocytopenic purpura; *HSP,* Henoch-Schönlein purpura.

If the child with purpura appears well or if the more seriously ill child has been given appropriate emergency care, the evaluation of the purpura can proceed in an orderly fashion. The recent and past medical history should be reviewed carefully with the parents and child. Acute onset of purpura after a recent viral illness or immunization is consistent with an acquired disorder such as ITP or a circulating anticoagulant. Recurrent purpura since infancy, however, suggests an inherited abnormality of platelets or clotting factors. Specific inquiries about past surgeries, dental extractions, or significant trauma should be made because the absence of bleeding under these conditions would be unusual in most inherited disorders of even moderate severity. When previous bleeding has occurred, the site of bleeding may be helpful in establishing the alteration in the hemostatic mechanisms. Hemarthroses, a common problem in severe hemophilia, rarely are associated with platelet abnormalities. Conversely, petechiae and subconjunctival hemorrhages are commonly found in children with platelet disorders but occur rarely in hemophilia.

The family history should be reviewed for purpura or bleeding disorders. A positive family history in male relatives on the maternal side suggests factor VIII or factor IX deficiency. A history of bleeding or bruising in numerous family members of both sexes suggests a condition with dominant inheritance such as von Willebrand's disease. As noted earlier, however, a negative family history does not preclude the diagnosis of von Willebrand's disease or hemophilia. A careful review of systems also should be obtained to evaluate underlying conditions such as uremia, hepatic disease, congenital heart disease, and malabsorption that might be associated with a coagulopathy.

The child should be examined carefully to assist in the diagnosis of the specific bleeding disorder and to evaluate hidden areas of hemorrhage. Particular attention should be paid to the skin. The distribution of purpura should be noted. Purpura on the lower extremities and buttocks suggests HSP, and purpuric lesions on the palms and soles often are seen with rickettsial infections. When the purpuric lesion has an unusual shape, such as a folded cord, child abuse should be suspected. Complete neurologic assessment is mandatory when there is suspicion of head trauma in the face of a bleeding diathesis. The eyes should be examined for the presence of conjunctival, scleral, or retinal hemorrhage. The presence of lymphadenopathy or hepatosplenomegaly should be sought. Lymphadenopathy may be present in certain malignancies (leukemias) or viral infections (infectious mononucleosis, CMV) that can present with purpura. Hepatomegaly may signal an underlying hepatic disorder that can cause a coagulopathy. Splenomegaly can be seen in infectious mononucleosis, leukemia, hepatic disease, and the storage diseases. Inflammation or synovial thickening of the large joints is consistent with the hemarthroses seen in hemophilia.

The laboratory approach to a child with purpura also is influenced by the initial presentation and history. Every child who presents with purpura should have a complete blood count (CBC) with a differential and platelet count, a PT, and a PTT. In most hospitals, these tests can be done quickly so that therapy can be chosen or modified on the basis of the results. A decreased hematocrit or hemoglobin concentration may indicate past or present blood loss or bone marrow failure or replacement. The WBC count can give information regarding the possibility of sepsis or leukemia. If sepsis is suspected, the smear should be examined for the presence of toxic granulation, vacuolization, or Döhle bodies. Atypical lymphocytes are seen with many viral infections, especially mononucleosis. Causes of abnormal screening coagulation studies (platelet count, PT, PTT, bleeding time) are outlined in Table 65.5. Depending on the results of these initial tests and the clinical impression derived from the history and physical examination, a more sophisticated laboratory evaluation can be undertaken. If emergency therapy is required before the cause of purpura is known, pretreatment plasma should be saved for later investigation of disorders such as von Willebrand's disease or other inherited factor deficiencies.

Platelet Count (normal 150,000–500,000/mm³)
Decreased: Increased platelet destruction, decreased platelet production, platelet sequestration, some platelet function disorders (see Table 65.3)
Prothrombin Time (normal range may vary between laboratories)
Prolonged: Disseminated intravascular coagulation; vitamin K deficiency; warfarin ingestion; deficiencies of factors II, V, VII, X; abnormalities of fibrinogen; liver disease; renal disease; congenital heart disease
Activated Partial Thromboplastin Time (normal range may vary between laboratories)
Prolonged: Disseminated intravascular coagulation; von Willebrand's disease; deficiencies of factors II, V, VIII, IX, X, XI, XII; abnormalities of fibrinogen; vitamin K deficiency; heparin therapy or sample contamination; liver disease; congenital heart disease
Fibrinogen (normal >150 mg/100 mL)
Decreased: Disseminated intravascular coagulation, liver disease, L-asparaginase therapy, dysfibrinogenemia, afibrinogenemia
Fibrin Split Products (normal <1:20)
Increased: Disseminated intravascular coagulation, liver disease
Bleeding Time (modified Ivy): (normal <8 min, 30 sec)
Prolonged: Idiopathic thrombocytopenic purpura (early) and other thrombocytopenias, von Willebrand's disease, platelet function disorders

Table 65.5. Tests Commonly Used in the Initial Evaluation of Purpura or Suspected Bleeding Disorders

The emergency management of children with purpura is discussed in detail in Chapter 87. However, the general principles are straightforward. When purpura is associated with a serious underlying disorder such as meningococcemia, treatment of that disorder usually is the first priority. Treatment of the coagulopathy is based on the degree and site of bleeding and the actual hemostatic defects. In primary disorders of hemostasis, appropriate replacement therapy is used when the specific alteration is known. When the disease has not been fully defined, broad treatment with one or more blood products may be required while further laboratory studies are performed. In all instances, the standard measures for general emergency care should be used fully during the evaluation and treatment of the purpuric disorders.

Suggested Readings

Bachman F. Diagnostic approach to mild bleeding disorders. Semin Hematol 1980;17:292–305.

Blanchette VS, Sparling C, Turner C. Inherited bleeding disorders. Clin Hematol 1991;4:292–332.

Ewenstein BM. Von Willebrand's disease. Ann Rev Med 1997;48:525–542.

Furie B, Limentani SA, Rosenfield CG. A practical guide to the evaluation and treatment of hemophilia. Blood 1994;84:3–9.

Lusher J. Approach to the bleeding patient. In: Nathan DG, Orkin SH, eds. Hematology of Infancy and Childhood, Philadelphia: WB Saunders, 1998:1574–1584.

McClure PD. Idiopathic thrombocytopenic purpura in children: diagnosis and management. Pediatrics 1975;55:68–74.

Rao K. Congenital disorders of platelet function. Hematol Oncol Clin North Am 1990;4:65–86.

Werner EJ. Von Willebrand disease in children and adolescents. Pediatr Clin North Am 1996;43:683–707.

CHAPTER 66

Rash—Urticaria

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Urticaria is a common cutaneous vascular reaction experienced by nearly 20% of the population at some time during their lives. The cause remains unknown in most cases. Urticaria usually is acute and transient, but if it persists for more than 6 weeks, it is called chronic. Although chronic urticaria sometimes is associated with physical agents or systemic illnesses (e.g., viral hepatitis, juvenile rheumatoid arthritis [JRA], systemic lupus erythematosus [SLE], lymphoma), the cause remains unknown in more than 75% of cases.

Urticarial lesions appear as erythematous papules or wheals from edema in the upper dermis with a surrounding flare of erythema caused by vasodilation. They are pruritic, multiple, and of varying size and shape. Individual lesions are transient, usually lasting 12 to 24 hours or less. They often appear suddenly, resolve almost completely, and may reappear. The cutaneous distribution varies, but lesions secondary to physical agents generally are concentrated in those areas of direct stimulation (i.e., dermographism). Lesions of angioedema often are nonpruritic and involve deeper dermal and subcutaneous tissues with swelling that may involve the lips and eyelids.

Urticaria may be accompanied by angioedema and is associated with systemic symptoms from direct visceral involvement or from symptoms secondary to the release of circulating chemical mediators. The respiratory, cardiovascular, and gastrointestinal (GI) systems may be involved, resulting in a potential life-threatening reaction. Signs and symptoms may include hoarseness, stridor, shortness of breath, wheezing, and general respiratory distress (from laryngospasm and bronchospasm), as well as hypotension, nausea, vomiting, diarrhea, and abdominal pain.

PATHOPHYSIOLOGY

Urticaria is characterized by superficial dermal edema, vasodilation and transudation of fluid and red blood cells, dilated lymphatics, and a mononuclear perivascular infiltrate. Angioedema occurs when these changes involve the deeper portion of the dermis and subcutaneous tissue. Urticaria and angioedema may occur independently or in association. The release of histamine and various other vasoactive and chemotactic substances from mast cells and basophils appears to play a central role in the pathogenesis.

DIFFERENTIAL DIAGNOSIS

As shown in Table 66.1, urticaria may be classified on the basis of the mechanism responsible for its formation or, if unknown, as idiopathic.

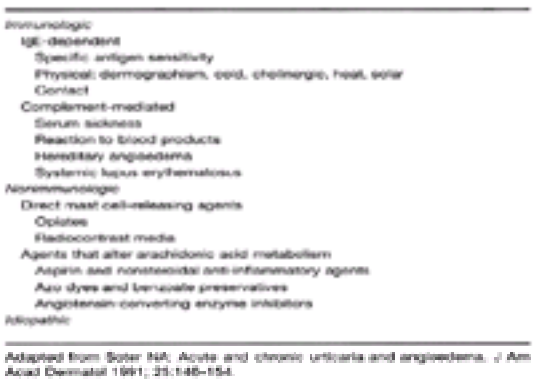
Immunologic
IgE-dependent
Specific antigen sensitivity
Physical: dermographism, cold, cholinergic, heat, solar
Contact
Complement-mediated
Serum sickness
Reaction to blood products
Hereditary angioedema
Systemic lupus erythematosus
Nonimmunologic
Direct mast cell-releasing agents
Opiates
Radiocontrast media
Agents that alter arachidonic acid metabolism
Aspirin and nonsteroidal anti-inflammatory agents
Azo dyes and benzoate preservatives
Angiotensin-converting enzyme inhibitors
Idiopathic
Adapted from Soter NA. Acute and chronic urticaria and angioedema. J Am Acad Dermatol 1981; 31:146–154.

Table 66.1. Classification of Urticaria/Angioedema

Multiple factors—both immunologic and nonimmunologic—are capable of initiating the release of these mediators that result in the histopathologic findings described. The type I hypersensitivity reaction that involves the interaction of an antigen with a mast cell–bound or basophil-bound IgE with release of histamine represents the most common immunologic mechanism. However, type III (immune complex) reactions also can stimulate mediator release through activation of the complement system. Examples include urticaria seen in association with viral hepatitis, infectious

mononucleosis, serum sickness, SLE, and some reactions to blood products. Nonimmunologic causes of urticaria include direct mast cell–releasing agents (e.g., opiates, radio contrast media) and agents that presumably alter arachidonic acid metabolism (e.g., aspirin, nonsteroidal anti-inflammatory agents, azo dyes). Angiotensin-converting enzyme (ACE) inhibitors are believed to enhance bradykinin synthesis.

Genetic factors are important in several relatively rare causes of urticaria and angioedema, including hereditary angioedema, familial cold, and localized heat urticaria. The most common causes of urticaria are listed in Table 66.2. Although idiopathic urticaria is the most common form, it is a diagnosis reached mainly by exclusion. Any variety of urticaria that involves the airway or cardiovascular system is potentially life-threatening.

Foods	*Insect bites*
Peanuts	Hymenoptera venom
Eggs	*Infections*
Chocolate	Hepatitis
Shellfish	Streptococcus
Milk	Infectious mononucleosis
Strawberries	Upper respiratory infection
Food dyes and preservatives	*Physical agents*
Drugs	Cold
Penicillin	Heat
Cytokines	Dermographism
Radiocontrast media	Latex
Aspirin and nonsteroidal anti-inflammatories	
Angiotensin-converting enzyme inhibitors	

[a]Essentially all may be life-threatening if accompanied by systemic reaction (see text).

Table 66.2. Common Causes of Urticaria/Angioedema[a]

EVALUATION AND DECISION

Urticaria is diagnosed by its characteristic appearance and is only rarely confused with erythema multiforme, certain vasculitides (e.g., Henoch-Schönlein purpura), urticaria pigmentosa, or infectious exanthems.

Following clinical recognition, the patient should be evaluated for the presence of an associated systemic reaction that involves cardiopulmonary compromise (outlined in Fig. 66.1). After stabilization, evaluation for a specific cause should begin with a thorough history and physical examination. The cause often remains unknown; however, Table 66.1 and Table 66.2 outline the general classifications and most common identifiable causes of urticaria. In the context of acute onset, the patient must be questioned about specific precipitants, including drugs, foods, and Hymenoptera stings. Febrile patients must be examined for clinical findings suggestive of viral and streptococcal infection, mononucleosis, and hepatitis. Latex allergy is uncommon in the general population, but health care workers and children with spina bifida appear to be at risk for latex allergy. These patients may experience urticaria, conjunctivitis, bronchospasm, and anaphylaxis after contact with or inhalation of latex antigens. Patients with chronic urticaria must be questioned about exposure to parasites or hepatitis, as well as about a family history of urticaria and must be examined for findings suggestive of collagen vascular disease. Laboratory tests generally are not helpful or necessary in the evaluation of acute urticaria.

FIGURE 66.1. Algorithm for the evaluation of urticaria.

Laboratory tests that may be useful in the evaluation of chronic urticaria include complete blood count (CBC) with differential, erythrocyte sedimentation rate (ESR), urinalysis, monospot, antinuclear factor (ANA), and liver function tests. Decreased levels of C_1 esterase inhibitor are found in hereditary angioedema. Stool for ova and parasites should be sent if eosinophilia is present or if symptoms are consistent with this diagnosis. Provocative tests may be tried cautiously if certain physical urticarias are suspected.

If the cause of chronic urticaria cannot be determined or if a severe systemic reaction occurs, referral to an allergist should be considered after initial treatment is instituted.

MANAGEMENT

The initial management of urticaria follows assessment of the patient for a systemic reaction with cardiopulmonary

compromise (i.e., stridor, wheezing, hypotension) (see Chapter 92). If a systemic reaction is present, airway, breathing, and circulation (ABC) should be stabilized. Medications that may be used include oxygen, epinephrine (1:1000) 0.01 mL/kg subcutaneously (which may be repeated in 15 to 20 minutes, maximum dose, 0.3 ml), diphenhydramine 1 mg/kg intravenously/intramuscularly, and volume resuscitation followed by vasopressors (e.g., dopamine) if there is no response. Systemic corticosteroids (i.e., methylprednisolone 1 to 2 mg/kg) are slow to work and inconsistent in their effect. They may suppress the appearance of new urticarial lesions but are indicated only in severe, inadequately controlled cases in which an infectious origin has been excluded.

Although any precipitating factor may result in urticaria accompanied by a systemic reaction, cold, cholinergic and solar urticaria, and hereditary angioedema have been associated with severe attacks. Mortality form hereditary angioedema has been reported to be as high as 30% and generally results from airway obstruction. Danazol, an attenuated androgen, is the preferred long-term prophylactic treatment. Acute attacks often require careful airway management, infusions of fresh-frozen plasma or concentrates of partly purified C_1 esterase inhibitor, and supportive care.

The general management of the more common presentation of urticaria without systemic involvement consists of removing or avoiding the inciting agent (if it can be identified) and providing symptomatic relief with antihistamines of the H_1 class. The two most commonly used oral medications are hydroxyzine (Atarax, 2 mg/kg per day) and diphenhydramine (Benadryl, 5 mg/kg per day) each in three to four divided daily doses for at least 3 to 5 days. Cyproheptadine (Periactin, 0.5 mg/kg per day) also has been found to be effective. If severe pruritus, a rapidly progressing urticarial rash, or angioedema is present, more rapid relief may be achieved with epinephrine (1:1000) 0.01 mL/kg subcutaneously (maximum dose 0.3 mL) and diphenhydramine 1 mg/kg IM. Prolonged sympathetic effect can be maintained with Sus-Phrine 0.005 mL/kg subcutaneously. Recurrent or persistent urticaria sometimes responds to cimetidine (Tagamet, 20 to 30 mg/kg per day) in four divided oral doses. Following or concurrently with treatment, an effort should be made to determine the cause (Table 66.1 and Table 66.2) so that the patient can avoid the precipitating agent.

Suggested Readings

Cahaly RJ, Slater JE. Latex hypersensitivity in children. Curr Opin Pediatr 1995;17:671–675.

Charlesworth EN. Urticaria and angioedema: a clinical spectrum. Ann Allergy Asthma Immunol 1996;76:293–306.

Cicardi M, Agostoni A. Hereditary angioedema (Editorial; Comment). N Engl J Med 1996;334:1666–1667.

Cicardi M, Bergamasni L, Marasini B, et al. Hereditary angioedema: an appraisal of 104 cases. Am J Med Sci 1982;284:2–9.

Czarnetzki BM. Mechanisms and mediators in urticaria. Semin Dermatol 1987;6:272–285.

Howard R, Frieden IJ. Papular urticaria in children. Pediatr Dermatol 1996;13:246–249.

Kulp-Sorten CL, Callen P. Urticaria, angioedema, and rheumatologic disease. Rheum Dis Clin North Am 1996;22:95–115.

Landwehr LP, Boguniewicz M. Current perspectives on latex allergy. J Pediatr 1996;128:305–312.

Sams WM Jr. Solar urticaria; studies of the active serum factor. J Allergy Clin Immunol 1985;45:295–301.

Sorter NA. Acute and chronic urticaria and angioedema. J Am Acad Dermatol 1991;25:146–154, 1991.

Volcheck GW, Li JT. Exercise-induced urticaria and anaphylaxis. Mayo Clin Proc 1997;72:140–147.

CHAPTER 67

Rash—Vesicobullous

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Basic to all vesicobullous (blistering) disorders is the disruption of cellular attachments. Blister formation, therefore, follows intracellular degeneration, intercellular edema (spongiosis), or damage to the anchoring structures associated with the basement membrane (half-desmosomes, basal lamina, anchoring fibrils). The location of these changes, as seen in Table 67.1, can help the physician ascertain a specific diagnosis. When histologic information is not readily available or nondefinitive, however, the historical and clinical features of the case must be relied on.

Type of Blister	Site of Formation	Disease
Subcorneal blister	Subcorneal	Impetigo
Blister from intracellular degeneration	Upper epidermis	Bullous congenital ichthyosiform erythroderma Epidermolysis bullosa of hands and feet Friction blisters
Spongiotic blister Viral blister	Intraepidermal Intraepidermal	Incontinentia pigmenti Variola Herpes simplex Varicella-herpes zoster
Blister from degeneration of basal cell	Subepidermal	Epidermolysis bullosa simplex Lichen planus Lupus erythematosus
Blister from degeneration of basement zone	Subepidermal	Epidermolysis bullosa, dystrophic type Urticaria pigmentosa Bullous pemphigoid Dermatitis herpetiformis Erythema multiforme, dermal type Drug-induced TEN

SSSS, staphylococcal scalded skin syndrome; TEN, toxic epidermal necrolysis.

Table 67.1. Pathologic Diagnosis of Vesicobullous Eruptions

Such an approach is outlined in Figure 67.1. The key features used in this algorithm to distinguish the various entities are a characteristic clinical appearance, chronicity and/or presence at birth, associated fever or systemic illness, distribution of lesions, and the child's age. The diagnosis of vesicobullous lesions in children younger than 1 month of age is not discussed in this chapter. Figure 67.1 also outlines the frequency and potential severity of these diseases.

FIGURE 67.1. The diagnostic approach to the child with vesicobullous lesions. *TEN,* toxic epidermal necrolysis; *EM,* erythema multiforme; *HSP,* Henoch-Schölein purpura; *SSSS,* staphylococcal scalded skin syndrome; *SLE,* systemic lupus erythematosus.

CHARACTERISTIC CLINICAL APPEARANCE

Many times, the appearance of a rash is so characteristic that a diagnosis becomes obvious. Such is the case with the conditions listed in Table 67.2.

Rhus dermatitis	Urticaria pigmentosa
Other contact dermatitides	Herpes simplex
Erythema multiforme	Varicella
Herpes zoster	

Table 67.2. Vesicobullous Rashes with Characteristic Clinical Appearance

Linear or geometric areas of vesiculation are the best clues to the presence of allergic contact dermatitis (see Chapter 99). The shape of the dermatitis provides the information that helps identify the offending agent. A history of playing in a shrubbed area, camping, hiking, or being near burning leaves is helpful. Because children brush against poison ivy leaves, vesicles often are in a line and on exposed surfaces (e.g., the face, extremities). A round group of vesicles on the back of the wrist would point to contact sensitivity to nickel contained in the metal case of a wristwatch.

Dermatomal distribution of vesicles or bullae usually indicates the presence of herpes zoster. On rare occasions, in infants, the same appearance may represent herpes simplex infections. A positive Tzanck smear indicates the presence of the herpes virus. Viral cultures or rapid slide tests using monoclonal antibodies are necessary to differentiate herpes simplex from herpes zoster.

Target or iris lesions are pathognomonic of erythema multiforme. The lesion has a dusky center that may blister and has successive bright red bordering rings. At times, a doughnut-shaped blister occurs.

Pigmented lesions that blister after stroking or trauma (Darier's sign) indicate the release of histamine from a mast cell collection. This collection may be isolated (mastocytoma of the wrist) or generalized (urticaria pigmentosa). Blistering of such lesions generally occurs only until 2 years of age. After this time, only urtication occurs.

A delicate “tear drop” vesicle is characteristic of varicella (chickenpox). Lesions usually begin on the upper trunk and neck. A progression through papules, vesicles, and crusts occurs rapidly (6 to 24 hours). All stages are present in an area at any given time. Mucous membranes are involved. Fever and malaise usually are present but are variable.

DURATION

If there is no characteristic clinical appearance, the duration of the rash must be considered. If it has been present for 4 weeks or more, it should be considered chronic. Rashes that come and go but take not more than 4 weeks to disappear completely are not considered chronic.

Chronic Rash (Duration 4 Weeks or More)

If the blistering disease has been present since birth (congenital), consider the diagnoses listed in Table 67.3.

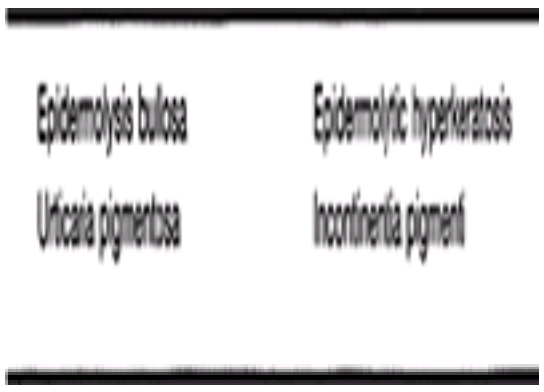

Table 67.3. Congenital Blistering Diseases

Epidermolysis Bullosa Syndromes

Blisters usually occur in areas predisposed to trauma or friction (Fig. 67.2). See Table 67.4 for differentiation of the various types.

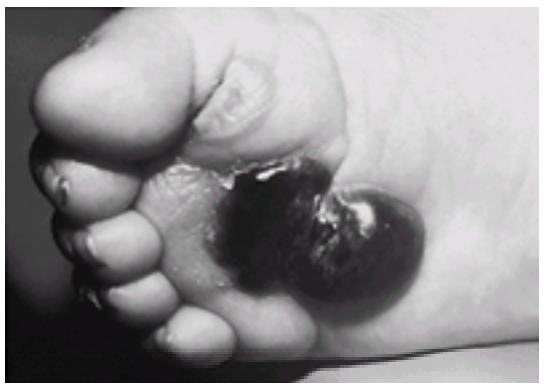

FIGURE 67.2. Infant with epidermolysis bullosa simplex.

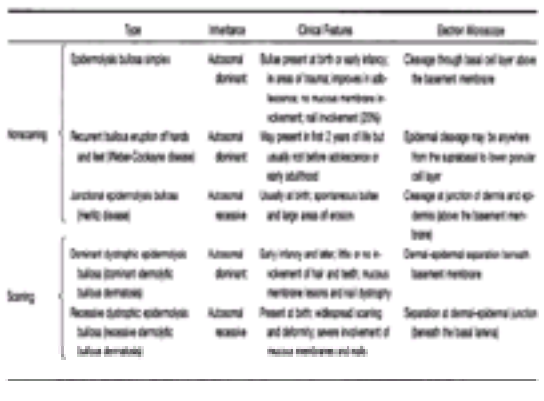

Table 67.4. Epidermolysis Bullosa Syndromes

Urticaria Pigmentosa

Mast cell disease (mastocytoma or urticaria pigmentosa) may cause blistering up until 2 years of age. The pigmented solitary lesion most often occurs on the arm near the wrist (Fig. 67.3). Lesions may be generalized. When a pigmented lesion feels infiltrated, the physician should think of its cause. Gentle mechanical irritation of such lesions causes urtication or blistering (Darier's sign).

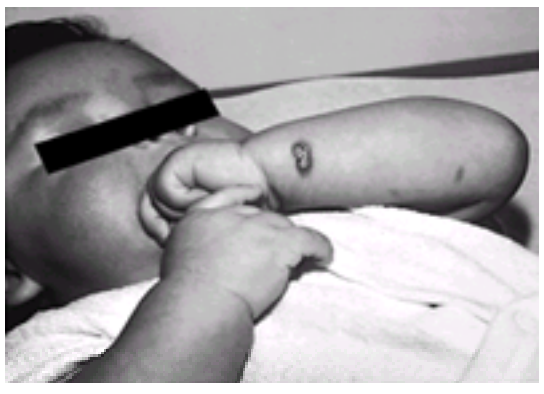

FIGURE 67.3. Infant with mastocytoma that has blistered because of trauma.

Epidermolytic Hyperkeratosis (Congenital Bullous Ichthyosiform Erythroderma)

Epidermolytic hyperkeratosis, an autosomal-dominant trait, is categorized under the ichthyotic syndromes. Children with this problem have recurrent bullous lesions during infancy and childhood. The skin has a background of erythema, scales, and peels. The flexures are always affected, as are the palms and soles.

Incontinentia Pigmenti

Incontinentia pigmenti, a rare condition, occurs almost exclusively in females. Inflammatory bullae erupt in crops in a linear distribution for the first several weeks to months of life (Fig. 67.4). These affected areas then go on to a warty stage. Finally, swirl-like pigmentation occurs but not necessarily in the areas previously involved with warty or blistering lesions. During the vesicobullous stage, a high degree of peripheral eosinophilia occurs (18 to 50%).

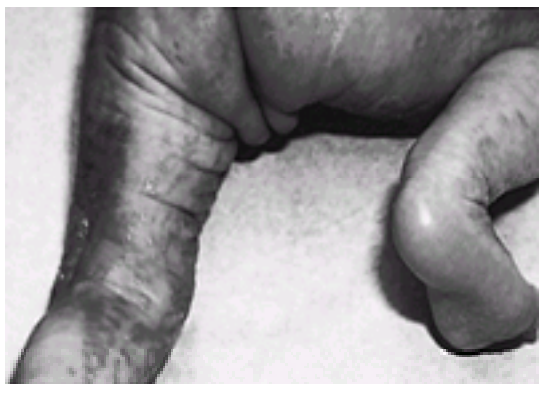

FIGURE 67.4. Linear arrangement of lesions (blisters in some cases) in a child with incontinentia pigmenti.

If the blistering is noncongenital, chronic bullous dermatosis of childhood (Fig. 67.5), dermatitis herpetiformis, and bullous pemphigoid should be considered. These conditions can be differentiated as outlined in Table 67.5.

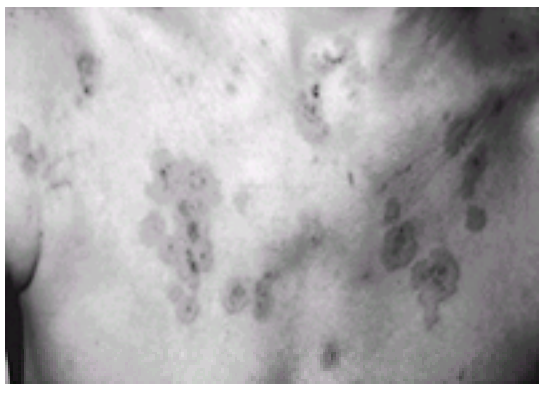

FIGURE 67.5. Chest of patient with chronic bullous dermatosis of childhood. Notice the resemblance to erythema multiforme.

	Bullous Disease of Childhood	Bullous Pemphigoid	Dermatitis Herpetiformis
Type of lesions	Large, tense, clear bullae; annular plaques with active vesicular borders	Large, tense bullae	Grouped papulovesicles, bullae or urticarial lesions
Distribution	Scalp, lower trunk, genitals, buttocks, inner thighs	Trunk and flexor surfaces of extremities	Back, buttocks, scalp, extensor surfaces of extremities, often symmetric
Pruritus	None to severe	Mild	Intense
Mucous membrane involvement	No	Yes	No
Duration	Months to years	Months to years	Months to years
Immunofluorescence	+ or -	+	+
	Linear IgA basement membrane	Linear IgG on basement membrane	Granular IgA at tips of dermal papilla
	(- circulating IgA)	(- circulating IgG)	of uninvolved perilesional skin
Treatment	Sulfapyridine or dapsone	Corticosteroids	Sulfapyridine or dapsone

Table 67.5. Noncongenital Chronic Blistering Disease

Child Who Is Ill

When the blistering lesions occur acutely, it must be determined whether the child is febrile or ill. Conditions that cause such systemic findings with associated blisters include those listed in Table 67.6.

Chickenpox	Herpes simplex
Hand-foot-mouth	Staphylococcal scalded skin syndrome
Viral (nonspecific) + other	Systemic lupus
Drug reaction (TEN, EM)	Atypical measles
Henoch-Schönlein purpura	Smallpox
Nonspecific vasculitis	

TEN, toxic epidermal necrolysis; *EM*, erythema multiforme.

Table 67.6. Blistering Diseases Associated with Fever and/or Systemic Illness

Varicella (Chickenpox)

For more information about varicella, see the section immediately preceding.

Hand–Foot–Mouth Disease

Caused by coxsackievirus A16, hand–foot–mouth disease is fairly characteristic. Vesicles are present on the palms, on the soles, and in the mouth. Other parts of the body may be involved. Fever, malaise, and abdominal pain may be present.

Viral (Nonspecific) and Other Causes

Vesicles have been described in association with other coxsackievirus types (A4, A5, B1, B4), echovirus, reovirus, and *Mycoplasma pneumoniae* infections. These children usually are ill.

Drug Reactions

The presence of vesicles or bullae may indicate a drug reaction. Involvement of palms, soles, mucous membranes, or the presence of target lesions are other possible clues that this problem exists. Therefore, the intake of prescribed and over-the-counter preparations must be investigated.

Drug-induced toxic epidermal necrolysis (TEN) may be associated with blisters. Histology that shows separation of dermis from epidermis excludes the staphylococcal-induced problem. Also, the staphylococcal scalded skin syndrome (SSSS) rarely occurs in children older than 6 years of age. A drug reaction should be considered in children older than 6 years old. The histology of SSSS demonstrates separation just below the stratum corneum.

Children with severe drug reactions may be very toxic. High fevers, malaise, joint problems, and the like can occur.

Henoch-Schölein Purpura

Children with Henoch-Schönlein purpura (HSP) may have blisters because of the severe involvement of blood vessels (vasculitis) in the typical distribution that occurs in this condition. Associated systemic problems include arthritis, abdominal pain, kidney disease (hematuria and/or proteinuria), and seizures.

Nonspecific Vasculitis

Children with vasculitic blisters, at times hemorrhagic, may be sick with fever, malaise, and other symptoms. Some go on to well-defined collagen vascular disease, whereas others smolder, with no diagnosis ever being made.

Herpes Simplex

Primary infection with this virus may cause fever and regional lymphadenopathy. The first encounter for young children usually is herpetic gingivostomatitis. Vesicles involve the lips and the rest of the mouth. These children often are uncomfortable and commonly refuse to eat or drink.

Herpes progenitalis may produce fever and local lymphadenopathy as well. Characteristic clusters of vesicles occur on an erythematous base. Often, erosions or ulcerations evolve on the vulva or penis. Diagnosis can be confirmed by a Tzanck smear that shows aggregates of multinucleated giant cells (see Chapter 99) or a rapid slide test (immune-specific immunofluorescent antibody placed on cells scraped from the blister base).

Systemic Lupus Erythematosus

Although not characteristic, bullous lesions can occur in systemic lupus erythematosus (SLE). Multisystem involvement suggests the diagnosis. Laboratory confirmation, which may include a skin biopsy and lupus band test, in conjunction with the complete clinical picture, is necessary for diagnosis.

Palm and Sole Involvement

If the child is not ill, the physician should search for blisters on the palms and soles. The child's age helps differentiate some disorders (Table 67.7).

Child <3 Years Old	*Any Age*
Scabies	Drug reaction
Acropustulosis of infancy	Friction blisters or burns
Syphilis (transiently at birth)	Dyshidrotic eczema
Adolescent or Older	Vasculitis (e.g., Henoch-Schönlein purpura)
Tinea pedis or manus	Frostbite
"Id" reaction	
Epidermolysis bullosa of hands and feet	

Table 67.7. Acute Vesicobullous Diseases Involving Palms and Soles

CHILD YOUNGER THAN 3 YEARS OLD

Scabies

Infants and very young children can have vesicobullous lesions on the palms (Fig. 67.6), soles, head, and face. It is important to not be misled by this distribution and appearance. Generally, the mother is infested as well and exhibits the typical appearance of this disorder.

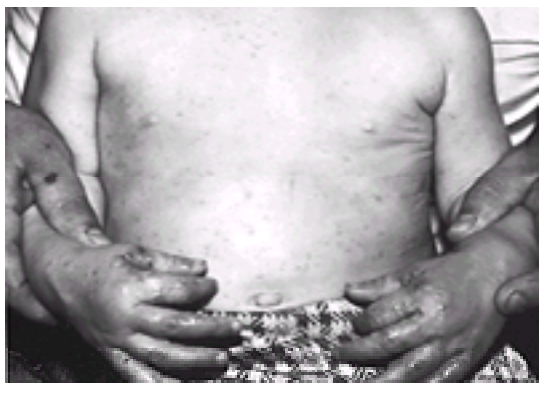

FIGURE 67.6. Blisters on hands of child infested with scabies.

Acropustulosis of Infancy

The appearance of pruritic vesicopustules between 2 and 10 months of age on the palms and soles (Fig. 67.7) in African-American children suggests acropustulosis of infancy. Vesicles often involve the lateral aspects of the fingers, palms, and soles. This condition was commonly diagnosed as dyshidrotic eczema in the past. Cyclic eruptions occur every 2 to 3 weeks, lasting 7 to 10 days. Spontaneous disappearance occurs at 2 to 3 years of age.

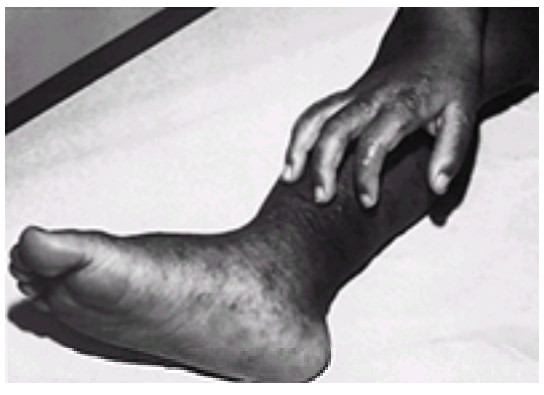

FIGURE 67.7. Note vesicles and pustules on child with acropustulosis of infancy.

Syphilis

Congenital syphilis may produce transient blisters on the palms and soles immediately after birth. “Snuffles,” rhagades, condyloma lata, and violaceous to reddish-brown macules should be sought on the palms and soles. Hepatosplenomegaly is often present. Osteochondritis is an early and common sign. Severe tenderness of a limb may cause pseudoparalysis of Parrot. The serologic test for syphilis is always positive in children with clinical manifestations.

ADOLESCENT OR OLDER

Tinea Pedis or Manus

Certain organisms that cause tinea pedis or manus (e.g., Trichophyton mentagrophytes) induce a severe inflammatory reaction on the hands and feet. Vesicobullous lesions erupt on the palms, instep, or medial aspect of the foot. A potassium hydroxide (KOH) preparation confirms the presence of hyphae in either location.

“Id” Reaction

If an adolescent's palms have blisters, the physician should look at the feet. Patients with tinea pedis may have allergic reactions to dissemination of antigen. A KOH preparation of the lesions on the palms will be negative for fungus.

Epidermolysis Bullosa of the Hands and Feet

For more information, see Epidermolysis Bullosa Syndromes in this chapter (Table 67.4).

ANY AGE

Drug Reaction

For more information about drug reactions, see Child Who Is Ill in this chapter.

Friction Blisters or Burns

Blistering on the palms and soles appears after trauma to the skin. The trauma often is related to a new activity (e.g., golfing, rowing, football) or to new, possibly poorly fitted, shoes.

Occasionally, accidental burns or burns secondary to child abuse are seen. Abused children may have had cigarette burns or have had their feet dipped in scalding water.

Dyshidrotic Eczema (Pompholyx)

A recurrent rash with episodes of vesicles that involve the palms, soles, and lateral aspects of the fingers is called dyshidrotic eczema. On occasion, large bullae occur. The problem generally is bilateral. Often, there is a personal or family history of atopy. A KOH preparation or fungal culture of scrapings from the palms or soles generally is negative.

Vasculitis

Vasculitis, also called HSP, may involve the palms and/or soles. See the section Child Who Is Ill in this chapter for more information.

Frostbite

Fingers, toes, feet, nose, cheeks, and ears are affected by extreme cold. After exposed areas are damaged by the cold temperature, symptoms occur on rewarming. Erythema, swelling, and burning pain occur at first, followed by vesicles and bullae (at times hemorrhagic [Fig. 67.8]) within 24 to 48 hours.

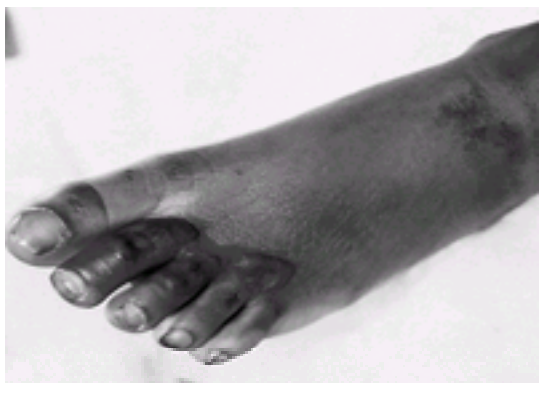

FIGURE 67.8. Frostbite. Child played in the snow for a prolonged period on a cold day wearing sneakers.

Extremities

If there is no involvement or minimal involvement of the palms and soles and the rash is concentrated on the extremities, insect bites, vasculitis, burns, frostbite, and bullous impetigo should be considered.

INSECT BITES

Insects generally bite exposed skin surfaces. Therefore, heaviest involvement occurs on the head, face, and extremities. Mosquito bites occur in the warm weather months, whereas flea bites occur during the whole year. Historical information includes contact with pets, camping trips taken, and involvement in outdoor activities. When blisters are present, the more characteristic urticarial papules usually are present in other locations. If not, confusion with bullous impetigo is easily ruled out with a Gram stain (negative for bacteria).

VASCULITIS

Concentration of hemorrhagic bullae on the extremities and buttocks indicates HSP. The lower extremities are the area most often involved because of settling of immune complexes, cryoglobulins, and so forth in that location.

BURNS

Exposed areas are commonly involved. Children accidentally rub against hot objects, causing burns and blistering. In cases of child abuse, children are burned intentionally with cigarettes (often mistaken for lesions of impetigo) or other heated objects. At times, children are submerged in scalding water. Usually, both lower extremities are involved.

BULLOUS IMPETIGO

In bullous impetigo, *Staphylococcus aureus* usually is present in pure culture. The bullae initially are filled with a clear fluid that rapidly becomes cloudy. The lesions tend to spread locally. Regional lymph nodes usually are not enlarged.

LABORATORY EVALUATION

If there is no clear idea about what caused the blister, the laboratory tests described next can be helpful.

Gram Stain

The Gram stain of fluid from an intact blister will be positive in impetigo and in a secondarily infected lesion. It will be negative, however, in all other conditions.

Tzanck Smear

Multinucleated giant cells will be present on a Tzanck smear (see Fig. 99.7) of material scraped from the base of an intact, freshly opened vesicle caused by herpes simplex, herpes zoster, and varicella.

Rapid Slide Test (Direct Immunofluorescence)

Fluorescent-tagged monoclonal antibody is placed on cells scraped from the blister base and can differentiate herpes simplex virus (HSV)-1, HSV-2, or varicella-zoster virus. Results can be available in 1 to 2 hours.

Bacterial or Viral Cultures

Occasionally, cultures help confirm an etiologic diagnosis when Gram stain, Tzanck smears, and direct immunofluorescence (DIF) are negative or indeterminate.

Skin Biopsy

For perplexing cases undiagnosed by clinical and/or simple laboratory evaluation, dermatologic consultation and skin biopsy are required.

On histologic examination, many characteristic changes can be found that lead to a definitive diagnosis (Table 67.1). Lichen planus, SLE, TEN, SSSS, and vasculitis are some of the diseases that can be identified by histologic studies.

If the picture on histology is compatible with erythema multiforme, DIF should be considered. DIF will be negative in erythema multiforme but will be positive in bullous pemphigoid (linear immunoglobulin G [IgG] on basement membrane), dermatitis herpetiformis (granular IgA at tips of dermal papillae of uninvolved perilesional skin), and chronic bullous disease of childhood (linear IgA on basement membrane). DIF can be negative in chronic bullous disease of childhood

(CBDC).

Indirect immunofluorescence can be done to test for circulating antibodies. Circulating IgG is found in bullous pemphigoid; circulating IgA is found in CBDC.

Suggested Readings

Dromy R, Raz A, Metzker A. Infantile acropustulosis. Pediatr Dermatol 1991;8:284.

Eady RA, Tidman MJ. Diagnosing epidermolysis bullosa. Br J Dermatol 1983;108:621.

Eichenfield LF, Honig PJ. Blistering disorders in childhood. Pediatr Clin North Am 1991;38:959.

Fine JD, Bauer EA, Briggaman NA, et al. Revised clinical and laboratory criteria for subtypes of inherited epidermolysis bullosa. J Am Acad Dermatol 1991;24:119.

Frieden IJ. The dermatologist in the newborn nursery: approach to the neonate with blisters, pustules, erosions, and ulcerations. Curr Prob Dermatol 1992;4:123.

Honig PJ. Bites and parasites. Pediatr Clin North Am 1983;30:563.

Weston JA, Weston WL. The overdiagnosis of erythema multiforme. Pediatrics 1992;89:806.

CHAPTER 68

Respiratory Distress

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Respiratory distress is one of the most common chief complaints of children seeking medical care. It accounts for nearly 10% of all pediatric emergency department visits and 20% of visits of children less than 2 years of age. Twenty percent of patients admitted to the hospital and 30% of those admitted to intensive care units are admitted for respiratory distress. Primary respiratory processes account for approximately 5% of deaths in children less than 15 years of age and 20% in infants. In addition, respiratory distress contributes substantially to deaths in patients with other primary processes. Respiratory arrest is one of the five leading causes of death in pediatric patients, along with congenital anomalies, trauma, neoplasm, and cardiac disease. Respiratory distress results from interruption of the respiratory or ventilatory pathway. The cause of respiratory distress may be within the respiratory system or within organ systems that control or impact on respiration. Respiratory failure is caused by an inability to meet metabolic demands for oxygen (O_2) or by inadequate carbon dioxide (CO_2) elimination. Young children are at particular risk of respiratory distress because of their respiratory anatomy and physiology. Rapid evaluation and aggressive treatment of respiratory distress, as well as anticipation and prevention of impending respiratory distress and failure, are essential to optimize outcome. Respiratory distress is usually reversible, but failure to treat the condition may result in cardiac arrest with long-term neurologic sequelae or death.

PATHOPHYSIOLOGY

Respiration is a complex multisystem process. The primary goals of respiration are to meet metabolic demands for oxygen and to eliminate carbon dioxide. Secondary functions include acid-base buffering, host defense, and hormonal regulation. The upper airway or conducting zone, which includes the nose, nasopharynx, oropharynx, larynx, trachea, major bronchi, and terminal bronchioles, serves as a conduit for air movement. The lower airway, or respiratory zone, consists of the acini and interstitium. Each acinus originates from a terminal bronchiole and includes respiratory bronchioles, alveolar ducts, sacs, and alveoli. The interstitium, which consists of the alveolar walls and interstitial septa, is the fibrous structural framework of the lower airway. Exchange of O_2 and CO_2 between the lungs and the blood occurs at the alveolocapillary membrane and depends on adequate and appropriately matched ventilation and perfusion.

Control of respiration is mediated by central and peripheral neural mechanisms. Respiration is an intrinsic brainstem function of the respiratory centers of the medulla. The dorsal respiratory group produces rhythmic inspiration while the ventral respiratory group controls expiration. Respiration is modulated by impulses within the brain and between the brain, respiratory system, blood, cerebrospinal fluid, and peripheral tissues. In the pons, the apneustic center increases the duration and depth of inspiration while the pneumotaxic center shortens duration and depth of respirations. Central chemoreceptors in the medulla respond to changes in the pH of cerebrospinal fluid. Peripheral chemoreceptors in the carotid and aortic bodies respond to changes in O_2, CO_2, and pH in arterial blood. In the airways, lungs, and chest wall, stretch, juxtacapillary, and irritant reflex mechanoreceptors respond to lung volume, changes in pulmonary microvasculature, chest wall muscle activity, and environmental irritants. Respiration is further influenced by the cerebellum, which alters respiration with postural change; by the hypothalamus, which controls respiration on a moment-to-moment basis; by the limbic system, which modulates respiration in response to emotion; and by the motor cerebral cortex, which controls volitional respiratory activity, including hyperventilation and hypoventilation and speech. Afferent information is transmitted to the brain primarily by the vagus (cranial nerve [CN] X) from the aortic body and mechanoreceptors, the glossopharyngeal (CN IX) from the carotid body, and the spinal motor neurons from muscle proprioceptors. Efferent impulses are transmitted from the brain via the vagus and spinal nerves to the larynx, trachea, bronchi, bronchioles, and acini; the glossopharyngeal to the pharynx; the hypoglossal (CN XII) to the tongue; and the spinal accessory (CN XI) to accessory muscles. Impulses are also transmitted via spinal motor neurons in the anterior

spinal horn to the cervical nerves (C2–C4), the phrenic nerve (C3–C5), and the intercostal nerves (T1–T12), which innervate accessory muscles, the respiratory diaphragm, and intercostal muscles, respectively. The muscles and bones of the chest wall provide structural support and, along with the muscles of the abdomen, impact on lung excursion, and thus movement of air in and out of the lung. Cardiovascular and lymphatic drainage maintain the fluid balance of the lung and thus impact on gas exchange.

Respiratory distress results from dysfunction or disruption of the respiratory/ventilatory pathway and/or systems that control or modulate respiration. Respiratory failure is the inability to meet the metabolic demand for O_2 (hypoxia) or the inability to eliminate CO_2 (hypercapnia). Criteria for defining respiratory failure vary widely; one set of criteria is presented in Table 68.1. Hypoxia can be categorized based on mechanism. Hypoxic or arterial hypoxemia is the most common type of hypoxemia. It results from an inability to deliver adequate oxygen to the blood. Most often, this type of hypoxia results from hypoventilation secondary to airway obstruction, central respiratory depression or impairment, neuromuscular or skeletal insufficiency, or restricted lung expansion. Other causes of hypoxemic hypoxia include low atmospheric P_{O_2} (e.g., high altitude), diffusion impairment (e.g., pulmonary edema, pulmonary fibrosis, acute respiratory distress syndrome [ARDS], oxygen toxicity), anatomic or physiologic shunt (e.g., atelectasis, pneumonia, abnormal pulmonary blood flow), or increased metabolic demand (e.g., exercise, systemic illness). Anemic hypoxia is the result of inability of the blood to deliver adequate oxygen to tissues as a result of decreased hemoglobin oxygen carrying capacity. It is caused by inadequate red cell number (decreased production, increased destruction, loss), low erythrocyte hemoglobin concentration (anemia), abnormal hemoglobin, carboxyhemoglobin, or methemoglobin. Hypokinetic, ischemic, or stagnant hypoxia also results in an inability of the blood to transport oxygen to the tissues. This type of hypoxia is caused by decreased blood flow to a localized area secondary to compromised cardiac output (e.g., cardiac failure), poor tissue perfusion (e.g., shock), sludging (e.g., polycythemia), or obstructed flow (e.g., vascular obstruction). Histotoxic hypoxia results from inability to metabolize oxygen at the tissue level as a result of inactivation of metabolic enzymes by a chemical such as cyanide. Hypercapnia is caused by inadequate alveolar ventilation (e.g., central nervous system [CNS] depression, spinal cord injury, neuromuscular disease, diaphragmatic dysfunction), ventilation-perfusion imbalance with relative hypoventilation (e.g., restrictive airway disease, pulmonary embolism), or increased CO_2 production (e.g., metabolic/endocrine disturbance). Hypercapnia often contributes to respiratory failure as a result of hypoxemia and is less commonly the primary cause.

Clinical	*Laboratory*
Tachypnea, bradypnea, apnea, irregular respirations	Pa_{O_2} <60 mm Hg in 60% O_2[b]
Pulsus paradoxus >30 mm Hg	$Paco_2$ >60 mm Hg and rising[c]
Decreased or absent breath sounds	pH <7.3
Stridor, wheeze, grunting	Vital capacity <15 mL/kg
Severe retractions and use of accessory muscles	Maximum inspiratory pressure <=25 cm H_2O
Cyanosis in 40% O_2[b]	
Depressed or heightened level of consciousness, decreased response to pain	
Weak to absent cough or gag reflex	
Poor muscle tone	

[a]Respiratory failure is likely if two clinical findings and one laboratory finding exist.
[b]Excluding cyanotic heart disease.
[c]Without underlying pulmonary disease.

Table 68.1. Criteria for Respiratory Failure[a]

Infants are at increased risk of respiratory distress compared with children and adults because of anatomic and physiologic differences (Table 68.2). These differences result in greater risk of airway obstruction, less efficient respiratory effort, limited respiratory reserve, and dysfunction of CNS respiratory control.

Difference	Consequence
Nose: infants <4 months obligate nose breathers	Nasal congestion may result in significant respiratory distress
Larynx: higher (C3–C4 vs. C6); funnel-shaped, narrowest at cricoid ring; softer, more elastic	More difficult to intubate Collapses more easily, particularly with head obstruction (i.e., Bernoulli's principle—as the velocity of flow through a collapsible tube increases, the pressure that holds the tube open decreases)
Trachea: one-third diameter of adult at birth, shorter	Poiseuille's law—resistance varies inversely with 4th power of the radius; 1 mm thickening decreases cross-sectional diameter 25% in adult, 75% in child;
	More difficult to intubate/maintain proper depth
Alveoli: elastic fibers less well developed	Alveoli collapse more easily, results in ventilation-perfusion mismatch
Lungs: lower functional residual capacity	Reserve small, therefore limited protection when ventilation is interrupted, Pa_{O_2} decreases more rapidly
Respiratory control apparatus immature—reflexes that inhibit respiration, particularly waning diaphragm reflex, which responds to stretch of lung, are very strong; central nervous system processing of information markedly affected by sleep state, cold, drugs, other metabolic derangements	Apnea or inability to respond appropriately to mechanical respiratory obstruction or increased metabolic demand
Chest wall: more compliant; intercostal muscles immature; ribs more horizontal; diaphragm flatter, fatigues; during rapid eye movement (REM) sleep intercostal muscle movements become uncoordinated	Chest wall more compliant Diaphragm does more work but is less effective

Table 68.2. Anatomic/Physiologic Differences Infant/Child and Adult Airway

DIFFERENTIAL DIAGNOSIS

Establishing a diagnosis for respiratory distress in part depends on localizing the source of the distress to a particular organ system. Respiratory distress may result directly from a disturbance of the upper or lower respiratory system. It may also be caused by inability of the central or peripheral nervous system to interpret or process respiratory requirements, or of the musculoskeletal system to perform the work of breathing. Alternatively, disease or dysfunction of other organ systems may indirectly result in respiratory disturbance by compromising respiratory system function or by stimulating

compensatory respiratory mechanisms (Table 68.3, Table 68.4 and Table 68.5). Treatment of the underlying cause is essential for definitive treatment of the respiratory distress.

Table 68.3. Causes of Respiratory Distress

Neonate	Infant/Child
Nasal obstruction	Peritonsillar abscess
Congenital airway anomalies	Croup
Transient tachypnea	Tracheitis
Respiratory distress syndrome	Foreign body
Meconium aspiration	Bronchiolitis
Pneumonia	Asthma
Sepsis	Allergy
Congenital heart disease	Pneumonia
	Fever
	Sepsis
	Gastroenteritis/dehydration

Table 68.4. Most Common Causes of Respiratory Distress

Foreign body	Pericardial tamponade
Tension pneumothorax	Epiglottitis

Table 68.5. Most Common Acute Life-Threatening Causes of Respiratory Distress

Respiratory System

Conditions may be congenital or acquired. They may be caused by upper or lower airway obstruction or by disorders of the parenchyma or interstitium. Upper airway obstruction is common in infants and young children in part because of their airway anatomy and physiology (see Chapter 72). Manifestations of upper airway obstruction include nasal flaring, stertor or snoring, gurgling, drooling, dysphagia, aphonia, hoarseness, stridor, retractions, and paradoxical chest/abdominal wall movement. In neonates, the common causes include nasal obstruction, congenital upper airway anomalies (particularly laryngotracheomalacia), and congenital or postintubation subglottic stenosis. Common causes for acquired upper airway obstruction in infants and children include adenotonsillar hypertrophy, peritonsillar abscess, croup, foreign body, retropharyngeal abscess, tracheitis, and airway edema from trauma or allergic reaction. Epiglottitis, although less common, is one of the most life-threatening causes of respiratory distress and is a true emergency. The incidence of epiglottitis has declined significantly since routine immunization against *Haemophilus influenzae* B, the pathogen that was responsible for at least 75% of cases. Epiglottitis should be suspected in children who have abrupt onset of fever, dysphagia, drooling, muffled voice, labored respirations, and stridor. Children appear toxic and anxious and assume a sniffing position with protruding jaw and extended neck. These children are at risk of abrupt onset of respiratory arrest from obstruction. Peritonsillar and retropharyngeal abscess may present with symptoms similar to epiglottitis but have more gradual onset. Croup or laryngotracheobronchitis is the most common cause of upper airway obstruction in children 3 months to 3 years of age. Croup causes subglottic narrowing and is characterized by a barky cough, inspiratory stridor, and hoarseness that are worse at night. Viral croup, most often caused by parainfluenza, has an insidious onset following several days of upper respiratory infection symptoms with normal temperature or low-grade elevation. Spasmodic or allergic croup has acute onset, usually with wakening during the night, in a child who was well before going to sleep. Children with recurrent or prolonged croup may have an underlying fixed or functional airway abnormality, most commonly subglottic stenosis or hemangioma. Children with chronic stridor, particularly those less than 2 years of age,

are also likely to have an underlying congenital anomaly. Foreign body aspiration, which has a peak age of occurrence 1 to 5 years, may cause obstruction of the upper or lower airway and is a leading cause of accidental death in toddlers. A history of abrupt onset of choking or gagging is suggestive. Drooling, dysphagia, and stridor suggest an upper airway foreign body, whereas unilateral wheeze, particularly first-time wheeze with acute onset, suggests lower airway position. Presentation, particularly with lower airway foreign body, may be delayed by days to weeks from time of aspiration. Other common causes of lower airway obstruction involve inflammation and bronchospasm and include asthma, allergy, and bronchiolitis. Wheeze, most often diffuse, is usually a predominant feature of these conditions (see Chapter 80). Asthma may be triggered by infection, exercise, environmental irritants, and/or stress. Allergy, usually accompanied by coryza, congestion, mucosal edema, and/or rash, may be in response to environmental exposures, food, or medications. Bronchiolitis, most often caused by respiratory syncytial virus, presents with wheeze in children less than 2 years of age. These conditions cause airway obstruction by decreasing airway lumen secondary to bronchospasm, edema, or thickening of the wall of the lumen. Other causes of lower airway obstruction include filling of the airway lumen by excessive secretions (e.g., from inflammation, infection, toxin such as organophosphate) or aspirated fluids and decreasing of lumen diameter due to loss of radial traction of the airway wall as with emphysema and masses.

Disorders of the alveoli and interstitium involve pus or fluid collection, collapse, and structural or functional abnormality. Alveolar and interstitial disease is characterized by tachypnea, cough, grunting, crackles, rhonchi, wheeze, and decreased and/or asymmetric breath sounds with or without fever. In neonates, transient tachypnea of the newborn and meconium aspiration are common causes. Pneumonia is one of the most common causes of lower airway disease in neonates, infants, and children. Findings are more likely to be localized in the setting of bacterial pneumonia, whereas patients with viral and atypical pneumonias, such as *Mycoplasma,* chlamydia, and pertussis, tend to have diffuse peribronchial, interstitial processes. Less commonly, aspiration, hemorrhage, and pulmonary edema cause fluid collection in the acini and interstitium. Atelectasis, or airway collapse, resulting from loss of air from the pulmonary parenchyma, often occurs secondary to other processes, including pneumonia, particularly viral; bronchospasm; and inadequate lung expansion, most often resulting from pain, neuromuscular disease, or inactivity. Structural and/or functional abnormalities include bronchopulmonary dysplasia, hyaline membrane disease or respiratory distress syndrome, bronchiectasis (most commonly seen in cystic fibrosis), congenital or acquired emphysema, and pulmonary fibrosis (usually from radiation and chemotherapy).

Nervous System

CNS disturbances may result in hypoventilation or hyperventilation, loss of protective airway reflexes, or airway obstruction from loss of pharyngeal tone. These conditions include CNS malformation, immaturity, infection, degenerative disease, seizures, mass, trauma, and intoxication. Focal neurologic deficits, visual disturbances, pupillary abnormalities, papilledema, abnormal muscle tone, and altered level of consciousness suggest CNS processes. Spinal cord trauma and anterior horn cell disease cause bulbar and respiratory muscle dysfunction, which results in airway obstruction and/or hypoventilation. Peripheral neuromuscular (i.e., peripheral nerve, neuromuscular junction, muscle) disorders result in muscle weakness or paralysis. Physical findings that suggest significant chest wall weakness may include hypotonia, hyporeflexia, muscle weakness, weak cry, hoarse voice, cough, gag, shallow or irregular respiratory pattern, and inability to lift the head or extremities.

Chest Wall/Thoracic Cavity

Musculoskeletal deformity or disease involving the support structures of the chest may severely restrict lung expansion, limiting normal ventilatory efforts or attempts at compensatory ventilation for respiratory dysfunction and other systemic disturbances. Intrathoracic conditions that may produce respiratory distress include air leak and space-occupying lesions, including fluid collections and masses. Air leak, is most commonly caused by pneumothorax or tension pneumothorax, which may be traumatic or spontaneous. Pneumothorax occurs when air enters the pleural space either by chest wall penetration (open pneumothorax) or by rupture of lung through the visceral pleura (closed pneumothorax) and causes collapse of the lung. With tension pneumothorax, air is able to enter but not egress. Pneumothorax, in addition to nonspecific signs of respiratory distress, is suggested by chest wall hyperexpansion, decreased breath sounds, and hyperresonance on the side of the air leak. With tension pneumothorax, there is also jugular venous distension (JVD) and deviation of the trachea and mediastinum away from the air leak. Tension pneumothorax decreases venous return and thus cardiac output. It is therefore life-threatening and must be relieved immediately by thoracentesis. The most commonly occurring space-occupying lesion is pleural effusion. Pleural effusion, which may be caused by infection, inflammation, ischemia, trauma, malignancy, major organ failure, drug hypersensitivity, or venous or lymphatic obstruction, is suggested on physical examination by decreased breath sounds and a pleural rub. Mass lesions include congenital or traumatic diaphragmatic hernia, esophageal anomalies, benign or neoplastic masses, and vascular malformations.

Cardiovascular

Congenital and acquired heart disease may result in respiratory distress from decreased cardiac output, reduced oxygen saturation, and/or congestive heart failure (CHF). Compromised cardiac output, most commonly caused by congenital structural heart defects, cardiac arrhythmias, myocarditis, pericardial effusion, pericardial tamponade, or hypotension, may result in insufficient tissue oxygen delivery to meet metabolic demands. Pericardial tamponade causes decreased cardiac output as a result of compromised cardiac filling. It is recognized on physical examination by Beck's triad of arterial hypotension, JVD, and distant heart sounds. It may be caused by infection, inflammation, trauma, or surgery. Acute tamponade may be immediately life-threatening and must be relieved expeditiously by pericardiocentesis. Cardiac anomalies with right-to-left shunting of deoxygenated blood result in reduced oxygen saturation of blood entering the systemic circulation, hence causing hypoxia with cyanosis. Cardiac defects causing left-to-right shunting result in pulmonary overcirculation, pulmonary venous congestion, and pulmonary edema that directly compromises pulmonary function. In children, congenital heart defects are the most common cause of CHF. Other cardiac causes of CHF include valvular heart disease, myocardial dysfunction, arrhythmias, ischemia, and infarction. Metabolic disturbances, sepsis, fluid overload, and severe anemia may also result in CHF. Pulmonary manifestations of CHF include tachypnea, increased work of breathing, dyspnea on exertion, orthopnea, cough, wheeze, and bibasilar rales. Other manifestations

include poor feeding, failure to thrive, fatigue, tiring with feeds, diaphoresis, edema, tachycardia, weak thready pulses, JVD, displaced point of maximum impulse (PMI), cardiac murmur, gallop, rub, cardiomegaly, and hepatosplenomegaly. Vascular causes of respiratory distress include pulmonary embolism, pulmonary hypertension, and pulmonary arteriovenous fistula.

Gastrointestinal

Abdominal obstruction, perforation of hollow viscous, laceration of solid organs, hematoma, contusion, appendicitis, infection, inflammation, ascites, or mass may result in impaired diaphragmatic excursion secondary to abdominal distension and/or pain. Prolonged shallow respiration may result in pulmonary hypoventilation. Gastroesophageal reflux or vomiting, particularly in children unable to protect their airway, may result in pulmonary aspiration.

Metabolic and Endocrine Disturbances

Metabolic disturbances often manifest as compensatory alterations in respiratory status. Metabolic acidosis results in rapid, deep breathing. Hyperammonemia directly stimulates the respiratory center to produce tachypnea, which results in primary respiratory alkalosis with secondary metabolic acidosis. Metabolic disruption of oxygen metabolism is another cause for respiratory distress. Endocrine disturbances that cause alterations in metabolic rate or chemical imbalances also result in respiratory distress.

Hematologic

Inadequate concentrations of hemoglobin or hemoglobin with decreased oxygen-carrying capacity result in deficient oxygen delivery to tissues. Polycythemia results in sludging of blood and therefore compromised oxygen delivery.

EVALUATION AND DECISION

Triage and Stabilization

Every child with significant respiratory distress must be considered to be at potential risk of respiratory collapse. Airway patency, breathing, and circulation should be rapidly assessed and, if compromised, should be established and optimized immediately (Table 68.6). For the child in respiratory arrest, cardiac arrest, if not already present, is imminent. Cardiorespiratory status should be continuously monitored. A health care provider skilled in airway management and resuscitation should remain with the patient at all times. Evaluation that is stepwise and focused is critical for determining the source and severity of respiratory distress. Anticipation and rapid aggressive management are essential for optimizing outcome. In the child who is alert and otherwise healthy, the position that he or she has naturally assumed is likely to be the one that minimizes respiratory distress and thus should be maintained. A child with significant respiratory distress should not be agitated; he or she should be allowed to remain with the parents. Anxiety increases minute ventilation and adds significantly to the child's oxygen consumption. Any patient thought to have ventilatory compromise should be treated immediately with humidified oxygen at the highest concentration available. Supplemental oxygen provides a small but often crucial margin of safety in ensuring adequate cerebral and myocardial oxygenation. In patients with decreased sensorium or neuromuscular disease, a position to optimize airway patency must be established. Airway devices or assisted ventilation may be necessary. For management of cardiorespiratory arrest, resuscitation efforts must be initiated immediately, as detailed in Chapter 1 and Chapter 2.

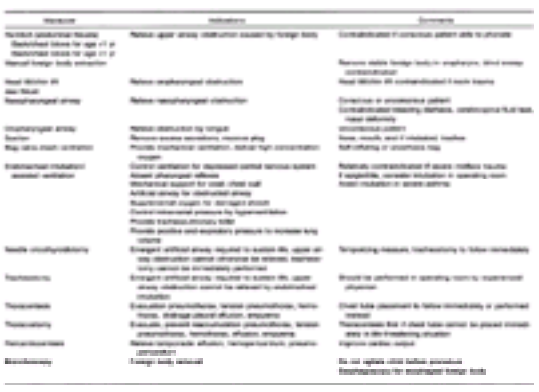

Table 68.6. Life-Saving Maneuvers to Relieve Respiratory Distress

History

A detailed history usually provides important clues to the cause of respiratory distress, but in a critically ill child comprehensive detail should not be obtained at the expense of patient care. A brief history can be obtained while emergent treatment is initiated. Details can follow once the child is stabilized. Information obtained by history should include a description of respiratory and other symptoms, onset and duration of symptoms, possible precipitating factors, exacerbating factors, therapeutic interventions, history of previous similar symptoms, underlying medical conditions, medications, allergies, and immunizations.

Physical Examination

The physical examination should assess the degree of respiratory distress and should identify the site and likely cause of

respiratory distress (Fig. 68.1 and Fig. 68.2). Continuous cardiopulmonary monitoring and frequent assessment are important because respiratory status can change instantaneously. General appearance, level of consciousness, vital signs, respiratory rate, respiratory effort, and adequacy of oxygenation and ventilation give immediate information regarding the severity of respiratory distress and possible sites. Heightened level of consciousness, manifest as restlessness, anxiety, or combativeness, is more likely an early sign of hypoxia, whereas diminished level of consciousness, manifest as somnolence, lethargy, stupor, obtundation, or coma, tends to result from hypercarbia or severe hypoxia. The child's posture may suggest the site of the disturbance. Children with upper airway obstruction tend to assume a sniffing position, an upright sitting posture with neck slightly flexed and head extended. For lower airway obstruction, a tripod position, in which the child is sitting up and leaning forward, may be preferred.

FIGURE 68.1. Approach to the child with respiratory distress. *CNS,* central nervous system; *SpO_2,* percent oxygen saturation; *O_2,* oxygen.

FIGURE 68.2. Approach to the child with respiratory distress (continued). *CNS,* central nervous system.

Vital sign abnormalities provide important clues about the severity of illness and adequacy of compensatory mechanisms. To maintain cardiac output (CO), children are more dependent on increasing heart rate (HR) than on stroke volume (SV) (CO = HR × SV). Tachycardia is one of the early signs of respiratory compromise and is expected because of increased sympathetic tone due to respiratory distress. Bradycardia in a hypoxic child is a late and ominous sign that often signals impending cardiac arrest. Cardiac arrhythmias that compromise cardiac output may result in respiratory distress. Respiratory rate in children varies with age (Table 68.7). Tachypnea is a compensatory mechanism for hypoxia, hypercapnia, and acidosis, and it also occurs with pain, anxiety, and exercise. Although not specific for respiratory distress, tachypnea is one of the findings most consistently present with respiratory distress and is particularly pronounced with lower airway processes. Tachypnea may be the only manifestation of lower respiratory infection in children less than 6 months of age. Bradypnea, or decreased respiratory rate, may reflect central respiratory depression, increased intracranial pressure, diabetic coma, or fatigue of respiratory muscles. It is usually an ominous sign that heralds impending respiratory arrest. Blood pressure is often increased because of anxiety. Pulsus paradoxus, an exaggeration (greater than 10 mm Hg) of the normal decrease in blood pressure during inspiration, correlates well with degree of airway obstruction. Pulses paradoxus is also caused by compromised venous return because of forces on the pericardium that result in decreased cardiac output, particularly during forced inspiration. Hypotension in a child is a late and extremely worrisome finding. It suggests profound shock, significantly decreased cardiac output, and impending cardiorespiratory arrest. Fever results in an increase in the respiratory rate of approximately 3 breaths/minute for each degree centigrade of temperature elevation above normal, due at least in part to an increase in CO_2 production.

Age Group	Breaths/Minute
Neonate	35–50
Older infants/toddlers	30–40
Elementary school age	20–30
Older child/adolescents	12–20

Table 68.7. Normal Respiratory Rates

On inspection, in addition to respiratory rate, one should appreciate depth, rhythm, and symmetry of respirations; the use of accessory muscles; and perfusion. Breathing that becomes progressively more rapid and more shallow results from greater air trapping because airway resistance increases in obstructive lower airway disease. Rapid shallow breathing may also result from chest pain or chest wall musculoskeletal dysfunction. Kussmal's respirations (deep, regular, sighing breaths that may be rapid, slow, or normal in rate) are seen with metabolic acidosis, particularly diabetic ketoacidosis. Cheyne-Stokes respirations (respirations with increasing then decreasing depth alternating with periods of apnea) are seen with CNS immaturity in otherwise normal neonates and infants, particularly during sleep, and with inadequate cerebral perfusion, brain injury, increased intracranial pressure, and central narcotic depression. Biot's, or ataxic, respirations (breaths of irregular depth interrupted irregularly by periods of apnea) suggest CNS infection, injury, or drug-induced depression. Asymmetric chest wall movement and/or expansion suggests unilateral chest wall or thoracic cavity pathology. Nasal flaring and supraclavicular, suprasternal, and subcostal retractions of accessory muscles of respiration usually reflect upper airway obstruction but may occur with lower processes. Intercostal retractions are usually a sign of inadequate tidal volume as a result of lower airway disease. Thoracoabdominal dissociation, also called respiratory alterans or paradoxical breathing, in which the chest collapses on inspiration and abdomen protrudes, is a common sign of respiratory muscle fatigue. Central cyanosis results from reduced ambient oxygen, airway obstruction with impaired oxygenation, alveolar diffusion impairment, cardiac defect with right-to-left shunting, left ventricular heart failure with pulmonary edema, or methemoglobinemia. Cyanosis usually reflects at least 5 g/dL of unsaturated hemoglobin and an oxygen saturation of less than 90%. Cyanosis may not be recognized in severely anemic patients and may be more pronounced in polycythemic patients. Peripheral cyanosis is caused by local vascular changes of the extremities that result in inadequate perfusion or vascular stasis; it is not usually associated with a decrease in systemic oxygen saturation.

Palpation of the chest commonly reveals vibratory rhonchi over the large airways, which suggests fluid in the airway. Tactile fremitus, when increased, suggests bronchopulmonary consolidation or abscess, and when decreased or absent, it suggests bronchial obstruction or space-occupying processes of the pleural cavity. Crepitus on palpation of the chest or neck may reveal subcutaneous emphysema caused by pneumothorax or pneumomediastinum.

Auscultation is particularly useful for localizing the site of respiratory distress (Table 68.8). Stertor, gurgle, dysphonia, aphonia, hoarseness, barky cough, and inspiratory stridor localize the respiratory distress to the upper airway. A lower airway cause is suggested by decreased or asymmetric breath sounds, changes in pitch of breath sounds, expiratory stridor, grunting, and/or adventitial sounds, including crackles, rhonchi, wheeze, rub, bronchophony, egophony, and whispered pectoriloquy. The ratio of inspiratory to expiratory phase of respiration, normally 1:1, is often useful in distinguishing an upper from lower respiratory tract cause of respiratory distress. Respiratory distress from upper airway disease usually results from difficulty of inward air movement. The inspiratory phase is often increased relative to the expiratory phase to between 1:1 and 2:1. Lower airway processes often impede outward air movement and may result in a prolonged expiratory phase with ratios of 1:3 to 1:4. Absence or disappearance of wheeze in a child with continued or worsening respiratory distress may represent severe obstruction and should not be considered reassuring, but rather may herald impending respiratory arrest.

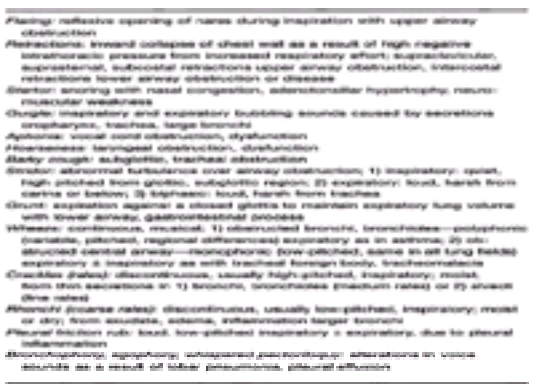

Table 68.8. Localization of Respiratory Distress by Physical Examination Findings

Percussion of the chest may reveal either hyperresonance, suggesting air trapping, or dullness, suggesting an area of consolidation, a mass in the lung or pleural space, or pleural fluid. Air trapping is further suggested by depressed position of the diaphragm. Diaphragmatic excursion can be accessed by measuring the difference between the level of dullness on percussion during full inspiration and full expiration. Poor diaphragmatic excursion may reflect diaphragmatic dysfunction.

The remainder of the physical examination should concentrate on the nervous, cardiac, gastrointestinal, renal, metabolic/endocrine, and hematologic systems, and may reveal pathology of these organ systems that localizes the underlying source of respiratory distress.

Approach

In approaching children with respiratory distress (Fig. 68.1 and Fig. 68.2), the physician should first assess the adequacy

of oxygenation and ventilation and then provide appropriate resuscitation. Patients in extremis (Fig. 68.1) are most likely to have sustained an injury, resulting in conditions such as a tension pneumothorax, flail chest, or cardiac tamponade, or have an obstructed airway, either as a result of aspiration of a foreign body or infection.

For patients with mild to moderate respiratory distress, as well as for those in extremis for whom the most likely diagnoses do not provide an explanation, the physician should proceed with a history, a physical examination, and a determination of oxygen saturation, and deliver supplemental oxygen as indicated. Respiratory distress of any degree calls for an immediate assessment of the airway. Stridor, altered phonation, and/or dysphagia suggests partial obstruction, most likely from infection, anaphylaxis, or foreign body. Assessment of the airway is followed by auscultation of the lungs for rales and wheezes. Children with abnormal auscultatory findings and fever are likely to have pneumonia or bronchiolitis, whereas asthma, bronchiolitis, and foreign body aspiration are common in afebrile patients.

Patients can be further categorized on the basis of tachypnea (Fig. 68.2). Children with rapid respirations and fever are likely to have pneumonia, even in the absence of rales; empyema, pulmonary embolism, and encephalitis are also important considerations. Tachypnea without fever points to trauma, cardiac disease, metabolic disturbances, toxic ingestions, and miscellaneous disorders.

Febrile children without tachypnea may have apnea or bradypnea as late manifestations of CNS infection. In afebrile patients, considerations include the myriad causes of CNS depression, spinal cord injury, neuromuscular disease, and neonatal apnea. Diagnostic tests should be used selectively to rule out diagnoses suggested by history and physical examination (Table 68.9). Nearly all patients with respiratory distress should have oxygenation tested by pulse oximetry. Arterial blood gas, more useful for lower than upper respiratory processes, and chest radiograph are the tests most likely to be helpful in the diagnosis of respiratory failure and determination of its cause.

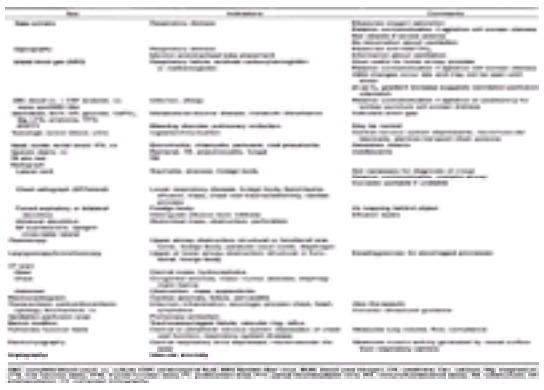

Table 68.9. Diagnostic Studies for Evaluation of Respiratory Distress

Treatment

Regardless of the cause of respiratory distress, aggressive treatment must be initiated immediately to rapidly restore oxygenation and ventilation. Airway patency, if inadequate, must be established. In the patient with decreased sensorium, positioning of the airway by chin lift (contraindicated if neck injury is suspected) or jaw thrust may relieve soft-tissue obstruction of the airway. The oral cavity should be cleared of secretions, vomitus, blood, and visible foreign matter. The unconscious patient may benefit from placement of an oropharyngeal airway or endotracheal intubation. In the alert patient with suspected soft-tissue obstruction of the airway, a nasopharyngeal airway may improve airway patency. Placement of a nasogastric tube to decompress a distended abdomen often improves respiratory effort by allowing full expansion of the lungs. The child in whom airway patency cannot be maintained or adequate ventilation and oxygenation cannot be established likely requires endotracheal intubation. Indications for intubation directly related to respiratory distress include respiratory failure or impending failure, apnea, airway obstruction, inability to handle secretions, and risk of aspiration.

SUMMARY

Respiratory distress is one of the most common chief complaints of children seeking medical care. The causes of respiratory distress are numerous and varied. History and physical examination provide important clues that allow rapid localization of the site of impairment. The underlying cause must be identified and may be within the respiratory system or organ systems that control or impact respiration. Any disorder that causes respiratory distress may be life-threatening. Airway and ventilatory problems not only must be recognized, but also must be anticipated and addressed aggressively. The underlying cause must also be treated. Patients must be monitored continuously and frequently reassessed. Airway, breathing, and circulation must be established and maintained. Diagnostic evaluation of body fluids, radiologic studies, direct visualization, and specialized tests of organ function must be performed prudently so that respiratory status is not further compromised.

Suggested Readings

Battaglia JD. Severe croup: the child with fever and upper airway obstruction. Pediatr Rev 1986;7:227–233.

Custer JR. Croup and related disorders. Pediatr Rev 1993;14:19–29.

Fireman P. The wheezing infant. Pediatr Rev 1986;7:247–254.

Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:85–251, 383–409, 823–901.

Klassen TP. Recent advances in the treatment of bronchiolitis and laryngitis. Pediatr Clin North Am 1997;44:249–261.

Kopelman AE, Mathew OP. Common respiratory disorders of the newborn. Pediatr Rev 1995;16:209–217.

Kravitz RM. Congenital malformations of the lung. Pediatr Clin North Am 1994;41:453–472.

Larson GL. Asthma in children. N Engl J Med 1992;326:1540–1545.

Mancuso RF. Stridor in neonates. Pediatr Clin North Am 1996;43:1339–1356.

Schidlow DV, Callahan CW. Pneumonia. Pediatr Rev 1996;17:300–309.

Vital Statistics of the United States. U.S. Department of Human Services. Public Heath Service. Centers for Disease Control and Prevention. National Center for Health Statistics, 1992. Vol II. Mortality, Part A. Hyattsville, MD: NSHC, 1996.

West JB. Pulmonary Pathophysiology. 5th ed. Baltimore: Williams & Wilkins, 1998.

CHAPTER 69

The Septic-Appearing Infant

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A young infant may be brought to the emergency department (ED) because he or she “just doesn't look right” to the parents. Even inexperienced parents whose first baby is just a few weeks old may notice when their child is unusually sleepy, fussy, or not drinking as well as usual. To the physician in the ED, such an infant may appear quite ill with pallor, cyanosis, or ashen color. There may be notable irritability or lethargy, and fever may or may not be present. The infant may be found to have tachypnea, tachycardia, or both. Hypotension or other signs of poor perfusion also may be apparent.

Generally, an ill-appearing infant, such as the one already described, will be considered immediately to have sepsis and will be managed reflexly. Although this may be the correct approach in most cases, the physician should remember that several other conditions can produce a septic-appearing infant.

The purpose of this chapter is to establish a differential diagnosis for infants in the first 2 months of life who appear quite ill. An approach to the evaluation and management of such an infant is discussed.

DIFFERENTIAL DIAGNOSIS

Numerous disorders (Table 69.1) may cause an infant to appear septic. The most common of these disorders (Table 69.2) include certain bacterial infections and viral syndromes. The remaining disorders, although uncommon, demand diagnostic consideration because they are potentially life-threatening, yet treatable.

Infectious Diseases	Inborn errors of metabolism
Bacterial sepsis	Hypoglycemia
Meningitis	Drugs/toxins—aspirin, carbon monoxide
Urinary tract infection	*Renal Disorders*
Viral infections—Enterovirus, respiratory syncytial virus, herpes simplex	Posterior urethral valves
Pertussis	*Hematologic Disorders*
Congenital syphilis	Severe anemia
Cardiac Disease	Methemoglobinemia
Congenital heart disease	*Gastrointestinal Disorders*
Supraventricular tachycardia	Gastroenteritis with dehydration
Myocardial infarction	Pyloric stenosis
Pericarditis	Intussusception
Myocarditis	Necrotizing enterocolitis
Kawasaki disease	Appendicitis, volvulus
Endocrine Disorders	*Neurologic Disease*
Congenital adrenal hyperplasia	Infant botulism
Metabolic Disorders	Shunt obstruction, infection
Hyponatremia, hypernatremia	Child abuse—intracranial hemorrhage
Cystic fibrosis	

Table 69.1. Differential Diagnosis of the Septic-Appearing Infant

Urinary tract infection	Congestive heart failure
Viremia	Gastroenteritis with dehydration

Table 69.2. Most Common Disorders That Mimic Sepsis

Sepsis

Sepsis (Chapter 84) should always be considered when the emergency physician is confronted with an ill-appearing infant. The signs and symptoms of sepsis may be subtle. The history may vary, and some infants may seem to be ill for several days, whereas others deteriorate rapidly. Likewise, any one or combination of symptoms, such as lethargy, irritability, diarrhea, vomiting, anorexia, or fever, may be a manifestation of sepsis. Fever generally is an unreliable finding in the septic infant; most septic infants younger than 2 months of age will be hypothermic instead. On physical examination, a septic infant may be pale, ashen, or even cyanotic. The skin is often cool and may be mottled because of poor perfusion. The infant may seem lethargic, obtunded, or irritable. There is often marked tachycardia, with the heart rate approaching 200 beats/minute, and tachypnea may be noted (respiratory rate above 50 breaths/minute). If disseminated intravascular coagulopathy (DIC) has developed, there may be scattered petechiae or purpura may be evident. If meningitis is also present, a bulging or tense fontanelle may be found. Likewise, if the infection has localized elsewhere, there may be otitis media, abdominal rigidity, joint swelling, or tenderness in one extremity, or possibly chest findings such as rales. Finally, if the disease process has progressed, the infant may develop shock and may be hypotensive.

The laboratory often is helpful in suggesting a diagnosis of sepsis; however, definitive cultures require time for processing. A complete blood count (CBC) may reveal a leukocytosis or left shift. In addition, a coagulation profile may show evidence of DIC, and blood chemistries may reveal hypoglycemia or metabolic acidosis. If localized infection is suspected, aspiration and Gram stain of urine, joint fluid, spinal fluid, or pus from the middle ear may reveal the offending organism. Similarly, a chest radiograph may show a lobar infiltrate if pneumonia is present. A Gram stain of a petechial scraping may also reveal the responsible organism.

Viral Infections

Overwhelming viral infections may mimic sepsis in the young infant. In one study of *enteroviral infections* in the neonate, it was noted that 25% of infants younger than 1 month of age developed a sepsislike illness. Respiratory distress was present in all of these infants; and hemorrhagic manifestations, including gastrointestinal bleeding and bleeding into the skin, were commonly seen. Seizures often occurred, as well as icterus, splenomegaly, congestive heart failure, and abdominal distension. In this series and in others, the mortality rate for enteroviral infections in neonates was high. This infection is indistinguishable from bacterial sepsis except that bacterial cultures will be negative, whereas viral isolates from stool and cerebrospinal fluid (CSF) may confirm the offending enterovirus.

Epidemics of *respiratory syncytial virus* (RSV) occur in the wintertime, and babies younger than 2 months old may present with apnea or respiratory distress with cyanosis. Those born prematurely or with previous respiratory or cardiac disorders are especially susceptible to apnea. These infants often appear septic, but knowledge of illness in the community and a predominance of wheezing on chest examination may lead to the suspicion of RSV bronchiolitis. Still, some infants develop wheezing later in the course, and thus, the initial diagnosis is difficult. A rapid slide test for RSV, if available, will be quickly diagnostic. Culture for RSV requires several days. A CBC may show a lymphocytosis, but because of stress, a left shift can also be found. Chest radiographs may show diffuse patchy infiltrates and, possibly, lobar atelectasis.

Another viral infection to consider is *herpes simplex,* which usually causes systemic symptoms and encephalitis at 7 to 21 days of life. Neonates present with fever, coma, apnea, fulminant hepatitis, pneumonitis, coagulopathy, and seizures, which are often difficult to control. History of maternal genital herpes should lead to suspicion of systemic herpes infection in the neonate. In most cases, however, the mother is completely asymptomatic. Ocular findings such as conjunctivitis or keratitis may be noted, as well as focal neurologic findings. If vesicular lesions are present on the skin, this infection should be strongly considered. However, they are present in only one-third to one-half of patients. When this infection is suspected, a Tzanck smear or direct fluorescent antibody staining of a scraped vesicle may provide rapid diagnosis. An electroencephalogram (EEG) or computed tomography (CT) scan may also be helpful. The diagnosis is confirmed by culture of a skin vesicle, mouth, nasopharynx, eyes, urine, blood, CSF, stool, or rectum.

Pertussis is another infection to consider when evaluating a very ill infant. Apnea, seizures, and death have been reported in this age group. Parents may report respiratory distress, cough, poor feeding, and vomiting (often posttussive). History of exposure to pertussis may be lacking because the infant usually acquires the disease from older children or adults who have only symptoms of a common upper respiratory infection. Physical examination will distinguish the infection from sepsis if the infant has a paroxysmal cough. The characteristic inspiratory “whoop” after a coughing paroxysm is common in young infants. Auscultation of the chest is usually normal; tachypnea and cyanosis may be present. Initial laboratory studies may not identify the condition. The CBC in young infants may fail to show a marked

lymphocytosis as expected in older patients with pertussis. Likewise, the chest radiograph may not show the typical “shaggy right heart border”; atelectasis or pneumonia may be present. Nasopharyngeal culture for *B. pertussis* is confirmatory. Polymerase chain reaction technique can reliably identify the condition from nasopharyngeal specimens.

Infants with *congenital syphilis* may present in the first 4 weeks of life with extreme irritability, pallor, jaundice, hepatosplenomegaly, and edema. They may have pneumonia and often have painful limbs. Snuffles and skin lesions are common. Although these infants may appear to be ill on arrival in the ED, their histories reveal that they also have been chronically ill. Certainly, if a history of maternal infection is obtained, the diagnosis should be considered. Laboratory tests will be helpful in that radiographs of the infant's long bones may reveal diffuse periostitis of several bones. A serologic test is needed to confirm the diagnosis.

Cardiac Diseases

In addition to infections, cardiac disease should be considered with a very ill infant. An infant with underlying *congenital heart disease* (CHD), such as ventriculoseptal defect, valvular insufficiency, valvular stenosis, hypoplastic left heart syndrome (HLHS), or coarctation of the aorta, may present with congestive heart failure and clinical findings similar to those of an infant with sepsis. There may be tachycardia and tachypnea, as well as pallor, duskiness, or mottling of the skin. Cyanosis is not always present. There may also be sweating or decreased pulses and hypotension caused by poor perfusion. However, a careful history and physical examination may help the physician differentiate CHD with heart failure from sepsis. For instance, a chronic history of poor growth and poor feeding may suggest heart disease. Also, the presence of a cardiac murmur may suggest a structural lesion. Moreover, a gallop rhythm, hepatomegaly, neck vein distension, and peripheral edema may lead one to consider primary cardiac pathology. Intercostal retractions and rales, rhonchi, or wheezing are nonspecific findings and may be present on chest examination in either heart failure or pneumonia. In a young baby, difference between upper and lower extremity blood pressures suggests coarctation of the aorta. If cardiac output is inadequate, however, pulse differences may not be detected. Normal femoral pulses do not exclude a coarctation because the widely patent ductus arteriosis provides flow to the descending aorta.

Laboratory evaluation is essential in establishing cardiac disease as the cause of an infant's moribund condition. A chest radiograph often shows cardiac enlargement and may show pulmonary vascular engorgement or interstitial pulmonary edema rather than lobar infiltrates (as in pneumonia). The electrocardiogram (ECG), may be helpful in revealing certain congenital heart lesions. For instance, in HLHS, the ECG invariably shows right-axis deviation, with right atrial and ventricular enlargement. The ECG often is a nonspecific indicator of cardiac decompensation however, and an echocardiogram is more helpful. Finally, a CBC may be helpful in that the absence of leukocytosis and left shift may make sepsis a less likely consideration. Rarely, an infant with anomalous or obstructed coronary arteries will develop myocardial infarction and appear to be septic initially. Such young infants may have dyspnea, cyanosis, vomiting, pallor, and other signs of heart failure; however, these infants usually have cardiomegaly on chest radiograph. This will prompt the physician to perform an ECG, which usually shows T-wave inversion and deep Q waves in leads I and AVL. Echocardiogram and cardiac catheterization with contrast are needed to confirm the diagnosis.

In addition to CHD, certain *arrhythmias* may cause an infant to appear ill. For instance, a young baby with *supraventricular tachycardia* (SVT) often presents with findings similar to those of a septic infant. This arrhythmia may be idiopathic (50%), associated with CHD (20%), or related to drugs, fever, or infection (20%). Often young infants with SVT go unrecognized at home for 2 days or more because initially, they have only poor feeding, fussiness, and some rapid breathing. As this condition goes untreated, however, the infants will develop congestive heart failure and may present with all the signs of sepsis, including shock. Because fever can be a precipitating cause of the arrhythmia, the condition obviously is confused with sepsis. However, a careful physical examination will make the diagnosis of SVT obvious. Particularly, the cardiac examination will reveal such extreme tachycardia in the infant that the heart rate cannot even be counted. It is usual for the heart rate to exceed 250 to 300 beats/minute in such infants. With this information, laboratory aids can confirm the diagnosis. An ECG will show regular atrial and ventricular beats with 1:1 conduction, whereas P waves appear different than sinus P waves and may be difficult to see at all. They are often buried in the T waves. Moreover, a chest radiograph may show cardiomegaly and pulmonary congestion.

Additional cardiac pathologies to consider include *myocarditis* and *pericarditis*. Pericarditis may be caused by bacterial organisms such as *Staphylococcus aureus;* myocarditis usually results from viral infections such as coxsackie B. In infants, these often are fulminant infections and the baby with such a condition will appear critically ill, with fever and grunting respirations often present. A complete physical examination may help the physician distinguish these conditions from sepsis in that signs of heart failure may be seen and unexplained tachycardia is often present. Also, pericarditis may produce neck vein distension and distant heart sounds if a significant pericardial effusion exists. In addition, a friction rub may be present. Laboratory tests may be helpful in that a chest radiograph will show cardiomegaly and a suggestion of effusion if pericarditis is present. The ECG will show generalized T-wave inversion and low-voltage QRS complexes, especially if pericardial fluid is present. Also, ST-T wave abnormalities may be seen. The echocardiogram will confirm the presence or absence of a pericardial effusion and poor ventricular function in the case of viral myocarditis. The CBC will not distinguish these infections from sepsis because leukocytosis is common and a left shift may be present.

Kawasaki disease with associated coronary artery aneurysms is very rare in young infants and is associated with a poor prognosis. A baby with Kawasaki disease may present with cyanosis and shock. Usually, history reveals prolonged and unexplained fever, rash, and mucous membrane inflammation. The physical examination may distinguish this illness from sepsis if there is a diffuse, raised, erythematous rash or cracked red lips, swollen hands and feet, conjunctivitis, and cervical lymphadenopathy. However, these classic features, found in older infants and children, may be absent in young babies. Routine laboratory studies may not differentiate this condition from sepsis either. A CBC may reveal leukocytosis and/or thrombocytosis. CSF usually shows a pleocytosis, with a lymphocytic predominance. Sterile pyuria is sometimes noted. In some cases, findings consistent with myocardial ischemia or an arrhythmia may be noted on ECG. Normal findings or nonspecific abnormalities are more common. Coronary artery aneurysms may be discovered with an echocardiogram, making the diagnosis highly likely.

Endocrine Disorders

Certain endocrine disorders can also mimic sepsis. For instance, infants with *congenital adrenal hyperplasia* (CAH) may present in the first few days or weeks of life with a history of vomiting, lethargy, or irritability. On arrival, signs of marked dehydration may be present, with tachycardia and possibly hypothermia. The recent history may be revealing in that such infants may have been poor feeders since birth and the symptoms may be progressive over a few days. The physical examination can be extremely helpful in establishing the diagnosis in females if ambiguous genitalia are noted. The laboratory evaluation also is helpful in that the presence of marked hyponatremia with severe hyperkalemia should make CAH a likely diagnosis. Other nonspecific laboratory findings in this disorder include hypoglycemia, acidosis, and peaked T waves or arrhythmias on ECG. Specifically, the finding of elevated 17-hydroxyprogesterone and renin with decreased aldosterone and cortisol in the serum confirms the diagnosis of CAH.

Metabolic Disorders

Various metabolic disorders can also look like sepsis and should be considered in the differential diagnosis. Prolonged diarrhea or vomiting can produce *dehydration, electrolyte disturbances,* and *acid-base abnormalities* such that an infant will appear quite ill. For instance, young infants with diarrhea may develop marked hyponatremia caused by iatrogenic water intoxication. Such infants may appear extremely lethargic, with slow respirations, hypothermia, and possibly, seizures.

A special cause of hyponatremic dehydration to consider is *cystic fibrosis* (see Chapter 96). The history in these cases may not be helpful initially, except that the infant usually gets very ill in hot weather. The mother may report poor intake, poor growth, and increased lethargy. Only with specific questioning might the mother report that the baby's skin tastes “salty” or that the baby had meconium plug syndrome (transient form of distal colonic obstruction secondary to inspissated meconium) as a newborn or prolonged neonatal jaundice. In some cases, pulmonary symptoms such as cough, tachypnea, or pneumonia may have been treated earlier in life. On examination, the dehydrated baby looks much like any other septic infant. However, laboratory tests that show profound hypoelectrolytemia, especially when not accounted for by gastrointestinal losses, should suggest cystic fibrosis. A sweat test will help confirm the diagnosis.

Likewise, dehydrated infants with *hypernatremia* may be lethargic or irritable, with muscle weakness, seizures, or coma. Infants with persistent vomiting may have hypochloremic alkalosis with hypokalemia, and they may appear weak or have cardiac dysfunction (see Chapter 86). In addition, rare inborn errors of metabolism such as *inherited urea cycle disorders* may produce vomiting in young infants, who will then present with lethargy, seizures, or coma resulting from metabolic acidosis, hyperammonemia, or hypoglycemia (see Chapter 98). *Hypoglycemia* also can be secondary to sepsis, certain drugs, or alcohol intoxication. It is thus essential to evaluate the electrolytes, blood sugar, and possibly, plasma ammonia levels in young infants with significant symptoms of gastroenteritis, lethargy, or irritability.

Another metabolic problem to consider is that of *toxins* (see Chapter 88). Obviously, young infants are incapable of accidental ingestions, but well-meaning parents may rarely cause salicylism in their attempts to aggressively treat fever with aspirin (despite current Reye's syndrome warnings). Affected infants can then present with vomiting, hyperpnea, hyperpyrexia, or convulsions and coma. In such cases, the history of medication given is crucial because the physical examination will not distinguish this ill baby from the infant with sepsis. The laboratory evaluation may lead to the suspicion of some metabolic problem because abnormalities of sodium, blood sugar, or acid-base balance often are found. Moreover, hypokalemia can be seen in salicylism as well as in abnormal liver function or renal function studies. An elevated salicylate level in the serum confirms the diagnosis of aspirin poisoning, but in chronic poisoning, the aspirin level may be relatively low despite a fatal course.

Carbon monoxide poisoning may present as an unknown intoxication when families are unaware of a defective heating system in the home. The young baby may have a history of sluggishness, poor feeding, and vomiting. A more careful history generally reveals that other family members are also ill with headache, syncope, or flulike symptoms. Their symptoms may improve after leaving the home environment. The classic “cherry red” skin color may be lacking, and physical examination may reveal only lethargy. Elevation of the carboxyhemoglobin level is diagnostic.

Renal Disorders

A young infant may also appear extremely ill because of renal failure or dysplasia. Such renal failure could be caused by *posterior urethral valves* that cause bladder outlet obstruction, especially in males. About one-third of those cases are diagnosed in the first week of life, but more than half go undetected for the first few months of life. The parents may give a history of vomiting or poor appetite, or they may say that the baby has not grown well or that the infant's abdomen appears swollen. On physical examination, hypertension or an abdominal mass (hydronephrosis) may be detected, as well as urinary ascites. Laboratory tests will elucidate the diagnosis even more. Suprapubic ultrasound may demonstrate the dilated posterior urethra and bladder, strongly suggesting posterior urethral valves. A voiding cystourethrogram (VCUG) should be obtained; this will show a dilated posterior urethra, hypertrophy of the bladder neck, and trabeculated bladder. The serum creatinine and blood urea nitrogen may be markedly elevated. Of course, urosepsis is a possible complication of posterior urethral valves.

Hematologic Disorders

It is also important to consider hematologic disorders when confronted with a critically ill infant. Any infant with severe *anemia* caused by aplastic disease, hemolytic process, or blood loss can look quite ill (see Chapter 87). In addition to anemia, disorders of hemoglobin such as *methemoglobinemia* can cause an infant to appear toxic. Although the chronic forms are uncommon inherited disorders of hemoglobin structure or enzyme deficiency, transient methemoglobinemia in infants occasionally is caused by environmental toxicity from oxidizing agents, such as nitrates found in some specimens of well water. This intoxication presents in very young infants with cyanosis, poor feeding, failure to thrive, vomiting,

diarrhea, and then lethargy. In other patients, the oxidant stress is less obvious. Methemoglobinemia has been described in infants with gastroenteritis and acidosis. Often, the associated diarrhea is severe, and it has been thought that the infectious agent that causes the diarrhea or the secondary metabolic acidosis may produce an oxidant stress that leads to methemoglobin formation. On examination, such infants have been described as toxic and lethargic, with hypothermia, tachycardia, tachypnea, and hypotension. They often appear mottled, cyanotic, or ashen. One key to the diagnosis of methemoglobinemia is that oxygen administration does not affect the cyanosis, yet no cardiac problem exists. Also, laboratory tests show a profound acidosis (pH 6.9 to 7.2), yet the Pa_{O_2} is normal despite the cyanosis. Leukocytosis and thrombocytosis are present. The blood itself may appear chocolate brown (most easily noted when a drop of blood on filter paper is waved in the air and compared with a normal control), and methemoglobin levels will be elevated up to 65% (normal 0 to 2%). Hemoglobin electrophoresis will be normal, except in rare cases of Hemoglobin M, as is the glucose-6-phosphate dehydrogenase (G6PD) assay in most cases. Prerenal azotemia may be noted. With appropriate treatment, the methemoglobin level returns to normal. However, death can occur from methemoglobinemia in infants.

Gastrointestinal Disorders

Gastrointestinal disorders can cause an infant to appear acutely ill. *Gastroenteritis,* even without electrolyte disturbances, can lead to profound dehydration. In a very young infant with little reserve, this can quickly lead to lethargy and even shock. Bacterial infections such as salmonella may cause sepsis in a young infant, and viral agents may mimic this. A history of bloody diarrhea may suggest this diagnosis. Stool cultures will diagnose bacterial infections, but a few days are needed for isolation. Viral isolation takes even longer. In the ED, a stool smear may reveal polymorphonuclear leukocytes, suggesting bacterial infection. A CBC with many band forms and a white blood cell (WBC) count in the normal range suggest *Shigella.* Laboratory tests are otherwise not helpful. Fluid resuscitation may improve the infant's appearance and make dehydration the likely diagnosis. However, sepsis cannot often be ruled out in the ED regardless of laboratory studies and initial therapy.

Also, *pyloric stenosis* in the young infant causes severe vomiting. This is most often seen in male infants 4 to 6 weeks old. An infant with pyloric stenosis may present to the ED with significant dehydration and may be lethargic. Usually, no fever is present. A careful history reveals that vomiting is the predominant feature of the illness, and there may be a positive family history for pyloric stenosis. The physical examination may reveal an abdominal mass, or “olive,” in 25 to 50% of cases, which would strengthen the diagnosis of pyloric stenosis. Electrolytes typically show hypochloremia and hypokalemia, and alkalosis is prominent. Plain films of the abdomen, a barium study, or ultrasound of the upper gastrointestinal tract may be needed to confirm the diagnosis.

Another gastrointestinal disorder to consider is *intussusception*. Although this rarely occurs in infants younger than 5 months old, it has been noted in some infants 2 to 3 months old. These infants may present with vomiting, fever, or signs of abdominal pain (e.g., legs drawn up, irritability). The infant may appear to have spasms of pain during which he or she is fretful. This can be followed by apathy and listlessness. Diarrhea may be seen, and if the typical currant jelly stool is noted, the diagnosis of intussusception should be strongly suspected. On physical examination, an abdominal mass may be palpated or bloody stool found on rectal examination. The laboratory may show nonspecific abnormalities such as leukocytosis and possibly anemia on CBC. However, a plain film of the abdomen will show evidence of small bowel obstruction, and a barium enema will show a filling defect usually near the ileocecal valve. A history of colicky behavior and the physical findings point to a gastrointestinal lesion rather than to sepsis.

Several other unusual but important gastrointestinal disorders have to be considered in infants. *Necrotizing enterocolitis* (NEC) occurs in premature infants in the first few weeks of life and can also occur in term infants, usually within the first 10 days of life. A history of an anoxic episode at birth or other neonatal stresses may suggest NEC. These infants are quite ill, with lethargy, irritability, anorexia, distended abdomen, and bloody stools. Radiographs of the abdomen may be helpful and usually show pneumatosis cystoides intestinalis caused by gas in the intestinal wall. Neonatal *appendicitis* is a rare event, but several cases have been reported to closely mimic sepsis. The mortality for this disorder is close to 80%, and perforation obviously worsens the prognosis. Thus, rapid diagnosis is essential. The most common presenting signs include irritability, vomiting, and abdominal distension on examination. There may also be hypothermia, ashen color, and shock as the condition progresses, as well as edema of the abdominal wall, localized to the right flank, and possibly, erythema of the skin in that area. The WBC count may be elevated, with a left shift, and there may be a metabolic acidosis, as well as DIC. Abdominal radiographs may show a paucity of gas in the right lower quadrant, evidence of free peritoneal fluid, or a right abdominal wall thickened by edema. Other unusual gastrointestinal emergencies to consider include *volvulus*, perforation caused by *trauma* from enemas or thermometers, and *Hirschsprung's enterocolitis*.

Neurologic Diseases

Neurologic problems should be considered in the evaluation of a critically ill infant. For instance, an unusual process that produces a sepsislike picture is *infant botulism*. This illness is produced by neurotoxins elaborated by *Clostridium botulinum.* An infant with botulism is often lethargic at presentation to the ED, with a weak cry and, possibly, signs of dehydration. These infants usually are afebrile. A thorough history may help distinguish botulism from sepsis. If constipation has preceded the acute illness, botulism should be seriously considered. The disease also is associated with the ingestion of honey, breast-feeding, a recent change in feeding practices, and a rural environment. The parents may note a more gradual progression with this illness. On physical examination, infants with botulism are notably hypotonic and hyporeflexic and may have increased secretions caused by bulbar muscle weakness. Also, the presence of a facial droop, ophthalmoplegia, and decreased gag reflex are consistent with botulism, whereas they remain unusual findings with a septic infant. Laboratory evaluation (cultures) should rule out bacterial illness. Moreover, abnormal (decreased) pulmonary function tests such as the measurement of maximal inspiratory force and vital capacity lend supportive evidence to the diagnosis of botulism. Finally, specific tests will confirm the diagnosis of botulism. A stool specimen to identify toxins of *C. botulinum* may be diagnostic but requires considerable time for identification. However, electromyography will show decreased muscle action potential with the “staircase” phenomenon in this disease. The

WBC count is normal in botulism.

A young baby with a ventriculoperitoneal shunt in place because of hydrocephalus can develop serious complications that cause the baby to appear extremely ill. *Shunt infection* could present with fever and irritability in a young infant. Abdominal pain or tenderness may be found on examination, as well as erythema or pus around the shunt itself. The definitive diagnosis is made by shunt aspiration under sterile conditions, but other causes of fever, such as meningitis, should be ruled out first. *Shunt obstruction* may result in increased intracranial pressure that causes a young infant to present with a history of lethargy or poor feeding. On examination, the baby may have bradycardia, apnea, coma, opisthotonic posturing, bulging fontanelle, or cranial nerve VI palsy. The shunt may be found to pump poorly. Laboratory tests such as radiographic evaluation of the shunt may be helpful if it shows a disconnection. Otherwise, a CT scan will demonstrate ventricle size and indicate the adequacy of shunt function.

Child Abuse

Intracranial hemorrhage that results from child abuse (see Chapter 128) must be considered in the very ill infant. It must be emphasized that the absence of bruises on an infant does not rule out child abuse. Vigorous shaking of an infant, followed by throwing the baby against a soft surface such as a mattress or sofa, can produce subdural or subarachnoid hemorrhages. The history may or may not be helpful in establishing a diagnosis. The parents may note that the child seemed to be in respiratory distress at home; only a few may admit to shaking the infant. On examination, the infant may appear gravely ill with apnea, bradycardia, hypothermia, bradypnea, and possibly, seizures. However, a careful physical examination may suggest abuse rather than sepsis. For instance, bruises may be present elsewhere on the body. More often, no external evidence of trauma is present. However, respiratory distress without stridor or lower airway sounds may be apparent, leading to the consideration of a central nervous system cause. The head circumference is often at the 90th percentile, and the fontanelle may be full or bulging. Retinal hemorrhages often are found, strongly suggesting trauma or intracranial hemorrhage rather than meningitis. Some neurologic signs may be confused with meningitis, such as nuchal rigidity, irritability or coma, seizures, or posturing. The laboratory is helpful in confirming suspicions of intracranial bleeding. Although the CBC often shows a leukocytosis and thus is confusing, the spinal fluid from a shaken baby usually is bloody. A computed axial tomography (CAT) scan or magnetic resonance imaging (MRI) usually demonstrates a small posterior, interhemispheric subdural hematoma. Such shaken babies have a high incidence of serious morbidity and mortality.

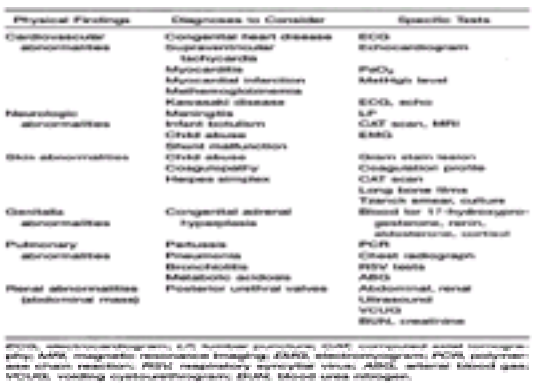
EVALUATION AND DECISION

Any infant who is critically ill in the first few months of life should initially be presumed to have sepsis. Because such illness is a life-threatening situation that may respond to early treatment, it is imperative to stabilize the child rapidly (Fig. 69.1). After airway, breathing, and circulation have been restored, vascular access should be obtained. Unless another diagnosis is immediately obvious, it is best to give intravenous antibiotics while pursuing alternative diagnoses. If time permits, cultures should be sent to the laboratory before giving antibiotics. Use of prostaglandins should be considered if cardiogenic shock is suspected.

FIGURE 69.1. Initial approach to the septic-appearing child.

A complete history should be obtained. It is important to learn of any previous medical problems such as known heart disease or failure to thrive. The time of onset of symptoms, exposure to infection, medications given at home, and specific symptoms noted by the parents must be determined. Next, careful physical examination must be performed because specific findings may lead to a diagnosis other than sepsis (Table 69.3). After the physical examination, a complete laboratory evaluation should be performed. All sick infants should have a blood culture and urine culture obtained by a urethral catheter or suprapubic bladder tap. A lumbar puncture also should be performed unless physical findings point strongly to a diagnosis other than sepsis or the infant is too ill to tolerate the procedure. A chest radiograph is also essential to look for pulmonary infection and to evaluate the heart size. A CBC should be obtained; leukocytosis will add support to a suspicion of sepsis but also may be found in various other disorders, including viral infections, myocarditis, pericarditis, intracranial bleeds, NEC, appendicitis, intussusception, and methemoglobinemia. Because metabolic problems (disturbances in acid-base balance, electrolytes, blood sugar) can result from sepsis or be the primary problem that mimics sepsis, all sick infants should have chemistries to evaluate serum sodium, potassium, chloride, glucose, and bicarbonate. If hyponatremia is found, water intoxication, aspirin toxicity, cystic fibrosis, and CAH should be considered. If there is also a marked hyperkalemia, CAH is most likely. If there is hypochloremic alkalosis or alkalosis alone, then pyloric stenosis, aspirin toxicity, or gastroenteritis should be considered. If there is hypoglycemia, it should be considered secondary to poor glucose reserves in an ill infant or related to drug (aspirin) toxicity, inborn errors of metabolism, CAH, or methemoglobinemia. If the serum bicarbonate is low, this should be confirmed with an arterial blood gas. Then, if acidosis is present, poor perfusion caused by shock should be considered, as well as dehydration,

drug toxicity, methemoglobinemia, appendicitis, CAH, and inborn errors of metabolism, as primary problems.



Physical Findings	Diagnoses to Consider	Specific Tests
Cardiovascular abnormalities	Congenital heart disease	ECG
	Supraventricular tachycardia	Echocardiogram
	Myocarditis	PaO_2
	Myocardial infarction	Met-Hgb level
	Methemoglobinemia	
	Kawasaki disease	ECG, echo
Neurologic abnormalities	Meningitis	LP
	Infant botulism	CAT scan, MRI
	Child abuse	EMG
	Shaken impairment	
Skin abnormalities	Child abuse	Gram stain lesion
	Coagulopathy	Coagulation profile
	Herpes simplex	CAT scan
		Long bone films
		Tzanck smear, culture
Genitalia abnormalities	Congenital adrenal hyperplasia	Blood for 17-hydroxyprogesterone, renin, aldosterone, cortisol
Pulmonary abnormalities	Pertussis	PCR
	Pneumonia	Chest radiograph
	Bronchiolitis	RSV tests
	Metabolic acidosis	ABG
Renal abnormalities (abdominal mass)	Posterior urethral valves	Abdominal, renal Ultrasound
		VCUG
		BUN, creatinine

Table 69.3. Approach to the Septic-Appearing Infant with Characteristic Physical Findings

Finally, if laboratory tests are not revealing for a specific disorder or the patient does not improve quickly as an inpatient receiving antibiotics, stool and CSF isolates for viruses should be considered.

If the physical examination suggests a specific problem, it may be necessary to obtain additional laboratory tests (Table 69.3). For instance, if the examination reveals pallor, cyanosis, or cardiac abnormality (muffled heart sounds, murmur, unexplained tachycardia, or arrhythmia) the physician should consider various cardiac disorders and possibly methemoglobinemia. An ECG, arterial blood to measure PaO_2, and possibly an echocardiogram should then be obtained. If there are unusual neurologic findings, such as a bulging fontanelle, a lumbar puncture should be performed to rule out meningitis, as well as blood studies mentioned previously. The presence of seizures should prompt a CT scan, EEG, and culture and treatment for herpes simplex virus. Also, if marked hypotonia is present, an electromyogram (EMG) may help diagnose botulism. Retinal hemorrhages may suggest an intracranial bleed, and thus a CAT scan, MRI, and lumbar puncture would be valuable studies. Likewise, if there is abdominal distension, rigidity, mass, or bloody stools, this would indicate a gastrointestinal emergency. In such cases, abdominal radiographs, ultrasound, or barium studies would be important diagnostic aids, but a workup for sepsis may still be indicated.

Furthermore, if the physical examination reveals bruises or purpura, further evaluation for child abuse, coagulopathy, and sepsis should be considered. In addition, long bone radiographs, coagulation profile (including platelet count), and Gram stain of the purpura may then be desirable. If vesicular lesions are seen on the skin, a Tzanck smear and culture for herpes should be obtained. If ambiguous genitalia are noted, blood should be drawn for 17-hydroxyprogesterone, renin, aldosterone, and cortisol to rule out CAH (see Chapter 97). Last, if wheezing is detected on chest examination, a nasopharyngeal swab should be sent for rapid slide detection of RSV or for culture of RSV.

SELECTED READINGS

Arvin AM, Probert CG. Herpes simplex virus infections: the genital tract and the newborn. Pediatr Rev 1992:11–16 (Special Edition).

Avner JR, Henretig FM, McAneney CM. Acquired methemoglobinemia: the relationship of cause to course of illness. Am J Dis Child 1990;144:1229.

Baraff LJ, Bass JW, Fleisher GR, et al. Practice guideline for management of infants and children 0 to 36 months of age with fever without source. Pediatrics 1993;82:1–12.

Belfer RA. The emergency department evaluation of the febrile infant and child with HIV infection. The AIDS Reader, 1994;Jan–Feb:8–12.

Byard RW, Edmonds JF, Silverman E, et al. Respiratory distress and fever in a two month old infant. J Pediatr 1991;118:306–313.

Crain EF, Gershel JC. Urinary tract infections in febrile infants younger than eight weeks of age. Pediatrics 1990;86:363.

Feit LR. Keeping up with Kawasaki syndrome. Contemp Pediatr 1995; 12:37–49.

Hall CB. Respiratory syncytial virus: what we know now. Contemp Pediatr 1993;Nov:92–110.

Hampers L, Tunnessen WW. A neonate in extremis. Contemp Pediatr 1995;12:91–93.

Hurst DL, Marsh WW. Early severe infantile botulism. J Pediatr 1993; 122:909–911.

Press S, Lipkind RS. Acute myocarditis in infants: initial presentation. Clin Pediatr 1990;29:73–76.

Saez-Llorens X, McCracken GH. Sepsis syndrome and septic shock in pediatrics: current concepts, terminology, pathophysiology, and management. J Pediatr 1993;123:497–508.

Selbst, SM, Goren E. Parapertussis infection in a neonate. J Fam Pract 1988;26:336–337.

Ward JC. Inborn errors of metabolism of acute onset in infancy. Pediatr Rev 1990;11:205–216.

CHAPTER 70

Seizures

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Seizures are the most common neurologic disorder in childhood and among the more common symptoms that lead to an emergency department (ED) visit. Studies have shown that 4 to 6% of all children will have at least one seizure in the first 16 years of life. These can range from a self-limited, nonrecurring episode to a prolonged, life-threatening event. The pediatric emergency physician must have a fundamental knowledge of all aspects of seizure management, including initial stabilization, determination of cause (differential diagnosis), appropriate definitive treatment, and patient disposition.

BACKGROUND

A *seizure* is defined as a transient, involuntary alteration of consciousness, behavior, motor activity, sensation, and/or autonomic function caused by an excessive rate and hypersynchrony of discharges from a group of cerebral neurons. A *convulsion* is a seizure with prominent alterations of motor activity. *Epilepsy*, or seizure disorder, is a condition of susceptibility to recurrent seizures.

Seizures may be *generalized* or *partial.* Generalized seizures reflect involvement of both cerebral hemispheres. These may be convulsive or nonconvulsive. Consciousness may be impaired and this impairment may be the initial manifestation. Motor involvement is bilateral. Types of generalized seizures include absence (petit mal), myoclonic, tonic, clonic, atonic, and tonic-clonic (grand mal) seizures.

Partial (focal, local) seizures reflect initial involvement limited to one cerebral hemisphere. Partial seizures are further classified on the basis of whether consciousness is impaired. When consciousness is not impaired, the seizure is classified as a *simple* partial seizure. Simple partial seizures may have motor, somatosensory/sensory, autonomic, or psychic symptoms. When consciousness is impaired, the seizure is classified as a *complex* partial seizure. Both simple and complex partial seizures may evolve into generalized seizures (e.g., Jacksonian march).

Status epilepticus is the condition of prolonged seizure activity (more than 20 to 30 minutes) or persistent, repetitive seizure activity without recovery of consciousness in between episodes.

A *postictal* (decreased responsiveness) period usually follows the seizure. During this time, the patient may be confused, lethargic, fatigued, or irritable; also, headache, vomiting, and muscle soreness may occur. In general, the length of the postictal period is proportional to the length of the seizure. For brief seizures, there may be few or no postictal symptoms. Transient focal deficits (e.g., Todd's paralysis) may occur during the postictal period, but one must first rule out a focal central nervous system (CNS) deficit.

PATHOPHYSIOLOGY

The underlying abnormality in all seizures is the hypersynchrony of neuronal discharges. Cerebral manifestations include increased blood flow, increased oxygen and glucose consumption, and increased carbon dioxide and lactic acid production. If a patient can maintain appropriate oxygenation and ventilation, the increase in cerebral blood flow is usually sufficient to meet the initial increased metabolic requirements of the brain. Brief seizures rarely produce any lasting effects. However, prolonged seizures may result in permanent neuronal injury.

Systemic alterations may occur with seizures and result from a massive sympathetic discharge, leading to tachycardia, hypertension, and hyperglycemia. Failure of adequate ventilation, especially in patients in whom consciousness is impaired, can lead to hypoxia, hypercarbia, and respiratory acidosis. Patients with impaired consciousness may be unable to protect their airway and are at risk for aspiration. Prolonged skeletal muscle activity can lead to lactic acidosis, rhabdomyolysis, hyperkalemia, hyperthermia, and hypoglycemia.

DIFFERENTIAL DIAGNOSIS

It is important to remember that a seizure does not constitute a diagnosis but is merely a symptom of an underlying pathologic process that requires a thorough investigation (Table 70.1). Often, no underlying condition is “identified,” and the diagnosis of idiopathic epilepsy is made. However, it is important not to exclude potentially treatable causes prematurely. For instance, seizures that result from metabolic derangements (e.g., hyponatremia, hypoglycemia) are often refractory to anticonvulsant therapy until the abnormality is corrected. Furthermore, every effort should be made to rule out a potentially life-threatening cause of seizures (e.g., intracranial injury or hemorrhage, meningitis, ingestions) before a less serious diagnosis is accepted.

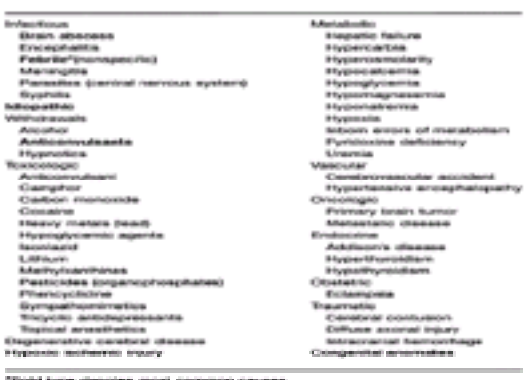

Table 70.1. Etiology of Seizures

Although the diagnosis of a seizure is often made in the ED on the basis of the clinical history, other childhood paroxysmal events are often mistaken for seizure activity (Table 70.2). Every attempt should be made to differentiate these events from seizures to ensure appropriate diagnosis, correct treatment, and accurate prognosis. Each episode or “spell” should be evaluated by examining the preceding events, the episode itself, and the nature and duration of the postictal impairment. If any of these features seem atypical, an alternative diagnosis should be considered.

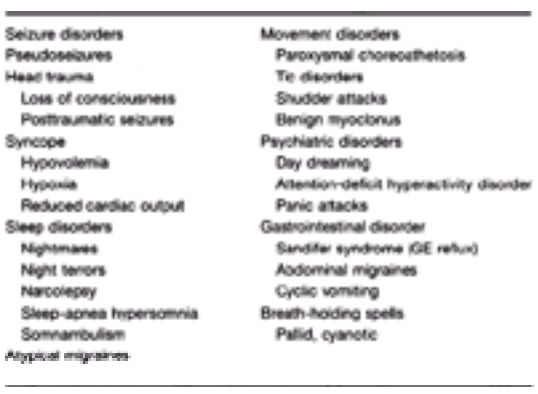

Table 70.2. Differential Diagnosis of Paroxysmal Events

Syncope, or the transient loss of consciousness that results from inadequate cerebral perfusion or substrate delivery, is the most common alternative diagnosis given to patients who present for evaluation of a seizure episode (see Chapter 73). Further complicating matters is the fact that a small percentage of patients with syncope exhibit some sort of convulsive movement. Although vasovagal episodes or orthostatic hypotension are the most common causes for syncope, it is important to evaluate these patients for potential underlying cardiac disease.

Breath-holding spells are common, affecting 4 to 5% of all children (see Chapter 131). They typically present between the ages of 6 and 18 months and disappear by age 5 years. The two types of breath-holding spells—cyanotic and pallid—have common features, including a period of apnea and an alteration in the state of consciousness. Usually, some initiating event (e.g., pain, fear, agitation) triggers the episode. The diagnosis is based on the clinical findings, and the prognosis is excellent.

A variety of movement disorders can mimic seizures. Paroxysmal choreoathetosis is often associated with a positive family history and exacerbated by intentional movement. Tic disorders can be manifested by twitching, blinking, head-shaking, or other repetitive motions. These are usually suppressible and are not associated with any loss of consciousness. Shudder attacks are whole body tremors similar to essential tremor in adults. Benign myoclonus of infancy can look like infantile spasms but is associated with a completely normal electroencephalograph (EEG).

Sleep disorders, such as somnambulism, night terrors (preschool-age children), and narcolepsy (typically in adolescents) can often be diagnosed based on the history alone (see Chapter 131). Infants with gastroesophageal reflux may exhibit torticollis or dystonic posturing (Sandifer syndrome). Atypical migraines and pseudoseizures are often diagnosed after other causes are excluded.

INITIAL STABILIZATION

The first priority in the seizing patient is to address the ABCs (airway, breathing, circulation) (see Chapter 1). An adequate airway is necessary to allow for effective ventilation and oxygenation. Patients with impaired consciousness as part of their seizure are at risk for obstruction (the tongue, oral secretions, emesis), aspiration (loss of protective reflexes), and hypoventilation. Simple maneuvers such as the jaw thrust or suctioning of the oropharynx may improve compromised air flow. The use of adjunctive airways (oral or nasopharyngeal) may also help maintain an adequate airway. In patients who are actively seizing, it may be difficult to insert these adjuncts and the patient may be injured if the intervention is forced. Furthermore, in patients for whom trauma is a possibility, these maneuvers must be undertaken with cervical spine (C-spine) immobilization. In patients in whom the airway remains unstable despite these actions, endotracheal intubation is warranted. When it is necessary to use a muscle relaxant to intubate a seizing patient, one should use the shortest-acting agent possible. The presence of motor activity may be the only clinical manifestation of seizure and a long-acting muscle relaxant will mask the ongoing seizure activity.

The patient's circulatory status must also be closely monitored. Seizures generally cause a massive sympathetic discharge that results in hypertension and tachycardia. Continuous cardiac monitoring and intravenous access should be obtained. Blood samples, including rapid blood glucose testing, should be acquired at this time in an attempt to establish a diagnosis. Peripheral intravenous (IV) access, which is often difficult in the pediatric age group, may be nearly impossible in the actively seizing patient. Intraosseous and/or central venous access may be required in the patient with prolonged seizures.

Once the respiratory and circulatory functions have been assessed and maintained, efforts should be directed at making a diagnosis and stopping any ongoing seizure activity. As long as adequate ventilation and oxygenation are maintained, long-term sequelae are unlikely to result from a transient seizure. The initial increase in cerebral blood flow compensates for any increase in brain metabolic requirements. Consensus management suggests the initiation of anticonvulsant treatment for anyone who has been seizing for more than 10 minutes. This likely represents all patients who are brought to the ED actively seizing.

EVALUATION AND DECISION

History

As a result of the numerous potential causes of seizures, as well as the large number of events that can be mistaken for a seizure, a focused history is important. The parent or caretaker needs to carefully describe the episode and the preceding events. Was there a warning (aura) that the patient was about to have an event? Was there a loss of consciousness, tongue biting, or incontinence? Did the event involve the entire body or only a portion? How long did the event last? How was the patient acting after the event was over?

In addition to the episode itself, the preceding events also are crucial. Was there a history of trauma? toxin exposure or ingestion? fever? other systemic signs of illness (e.g., headache, ataxia, vomiting, diarrhea)? Does the child have an underlying seizure disorder, history of seizures, or other neurologic problems? Is the child taking any anticonvulsants? If yes, was there a recent change in medications or is there a chance that the patient could have a subtherapeutic level? Any other significant medical history (including abnormal developmental history)? Any significant surgical history (including placement of a ventricular shunt)? Family history of seizures? Other medication use? Distant travel history (neurocysticercosis is one of the leading worldwide causes of seizures)?

Physical Examination

With the history, a directed physical examination is performed to look for a possible cause of the seizure. Vital signs, including temperature, need to be obtained. An elevated temperature points to a potential infectious cause. The entire body needs to be examined for evidence of trauma, either as a preceding cause or as a result of falling during the seizure episode. The skin should be examined for rashes or other congenital skin lesions. Dysmorphic features may be associated with other congenital CNS anomalies. Stigmata of underlying hepatic, renal, or endocrinologic disorders should also be noted.

The head should be carefully examined for swelling, deformity, or other signs of trauma. The presence of a ventricular shunt should be noted. The pupils are studied for shape, size, reactivity, and equality. The fundi are examined for the presence of retinal hemorrhages or papilledema. The tympanic membranes are examined for the presence of hemotympanum or for a source of potential infection. The mouth should be examined for evidence of tongue biting.

The neck is assessed for meningeal irritation. If there is a history or other physical signs of trauma, neck immobilization should be maintained until the C-spine can be “cleared.” Examination of the chest, lungs, and abdomen is performed in the usual fashion. The extremities are examined for evidence of trauma, especially as the result of falling during a seizure.

The neurologic examination may be limited by either ongoing seizure activity or a postictal state and may consist solely of the pupillary examination and an assessment of any asymmetric movements (focality). Any abnormal posturing (decerebrate or decorticate) should be noted.

If there is a question of a possible ingestion, the examination is also directed at uncovering a potential toxicologic syndrome (toxidrome) that may suggest a specific class of drugs or toxins that are responsible for the seizure (see Chapter 88). Important variables include temperature, heart rate, blood pressure, pupil size, sweating, flushing, and

cyanosis.

As the patient recovers from the seizure episode, periodic reassessment is needed to assess for any underlying neurologic abnormalities.

Diagnostic Approach (Figure 70.1)

Once it has been determined that a seizure may have taken place, the initial diagnostic evaluation starts with the history and physical. Laboratory, radiologic, and other neurodiagnostic testing (e.g., EEG) are other tools that can be part of the seizure evaluation.

FIGURE 70.1. Diagnostic approach to seizures. The most common causes are in bold type.

Patients with obvious trauma who are seizing should be treated per Advanced Trauma Life Support (ATLS) guidelines (see Chapter 103), with close attention to possible intracranial injury (see Chapter 105).

Often, patients with a known seizure disorder will present to the ED seizing. Patients known or suspected to be taking anticonvulsants should have drug levels evaluated. A subtherapeutic anticonvulsant level is among the most common reasons for patients to present with seizures.

Many different laboratory tests may reveal a cause for a seizure and as a result suggest a potential treatment. A rapid glucose reagent strip should be performed with the initial blood sample. Hypoglycemia is a common problem that can often precipitate seizure activity. If hypoglycemia is documented or a rapid assessment is not available, treatment with 0.25 to 1 g/kg of dextrose is indicated.

A febrile seizure is defined as a seizure caused by a fever, but this is a diagnosis of exclusion. Other infectious etiologies that can be the direct cause of a seizure (e.g., meningitis) must first be ruled out (see Chapter 28 and Chapter 84). Furthermore, infections not involving the CNS may still be the cause of the seizure through the elaboration of fever. Presence of fever or an elevated white blood cell (WBC) count should direct one to look for a potential infectious cause. Blood cultures should be drawn with the initial samples in patients at risk for bacteremia in an effort to identify a specific pathogen. Urinalysis and chest radiographs can also be used to confirm a source of infection.

A lumbar puncture (LP) with analysis of the cerebrospinal fluid (CSF) is the only way to make the diagnosis of meningitis and should be performed when meningitis is being considered. An elevated CSF protein and CSF WBC count and a low CSF glucose are all suggestive of CNS infection. CSF cultures, Gram stain, latex studies, and polymerase chain reaction (PCR) may identify a specific agent. Ideally, CSF cultures should be obtained before antibiotic therapy is initiated. However, in the critically ill or unstable patient, antibiotics should not be withheld until an LP is performed. Furthermore, in cases in which a potential metabolic disease is being considered, CSF lactate, pyruvate, or amino acid determinations can be used to diagnose a specific disorder. In these cases, it is often helpful to collect an extra tube of CSF to be frozen and used for later analysis. In any patient with suspected elevated intracranial pressure (ICP), an LP should not be performed until head imaging can be done.

Electrolyte abnormalities may also cause seizures, with hyponatremia, hypocalcemia, and hypomagnesemia being most common. In general, the routine screening for electrolyte abnormalities in a seizure patient is a low-yield procedure. Unfortunately, seizures caused by electrolyte derangements are often refractory to anticonvulsant therapy, and patients will continue to seize until the underlying abnormality is corrected. Serum electrolytes should be measured in all seizure patients with significant vomiting or diarrhea; with underlying renal, hepatic, neoplastic, or endocrinologic disease; who are taking medications that may lead to electrolyte disturbances; or who have seizures that are refractory to typical anticonvulsant management. One characteristic scenario involves hyponatremic seizures in infants, typically less than 6 months of age, after prolonged feedings of dilute formula (“infantile water intoxication”). Other patients may be evaluated on a case-by-case basis. Intravenous calcium, magnesium, and hypertonic (3%) sodium should be used to treat the appropriate abnormal condition. In the case of hyponatremia, once the seizure activity has been stopped, the rate of sodium correction must be titrated to avoid possible central pontine myelinolysis.

Other chemistries can be helpful in identifying specific organ dysfunction as a cause of the seizure activity or as an assessment of systemic injury. An elevated blood urea nitrogen or creatinine suggests uremia as a potential cause. Elevated liver function tests (transaminases or coagulation times) can be a reflection of hepatic failure. Metabolic acidosis or hyperammonemia can suggest an underlying metabolic disorder. In patients with prolonged seizures, an arterial or venous blood gas can help in assessing adequacy of ventilation, and a creatine kinase (CK) can identify

possible rhabdomyolysis.

Toxicologic screening can also be helpful in the seizing patient because certain ingestions are managed with specific antidotes or treatments. Typically, the clinical scenario is the young child with a possible accidental ingestion or the adolescent after a suicide attempt. In general, the toxicologic screen should be directed at agents known to cause seizures (Table 70.1) or those suggested by a clinical toxidrome.

Radiologic imaging of the seizure patient generally consists of a computed tomographic (CT) scan in the acute-care setting. The following situations should be considered emergent: 1) a patient who has signs or symptoms of elevated ICP, 2) a patient who has a focal seizure or a persistent focal neurologic deficit, 3) a patient who has seizures in the setting of head trauma, 4) a patient who has persistent seizure activity, or 5) a patient who appears ill. Until cervical spine injury is ruled out, it is important to remember to maintain C-spine immobilization in patients for whom head trauma is a concern. Patients with transient generalized seizures in whom a cause of the seizure activity is identified probably do not require any further head imaging studies. Patients with transient generalized seizures in whom no cause is identified and who appear clinically well can have their head imaging performed on a nonemergent basis.

A magnetic resonance imaging (MRI) study has several advantages over a CT scan. MRI is better at identifying underlying white matter abnormalities, disorders of brain architecture, lesions in the neurocutaneous syndromes, lesions in the posterior fossa and the brainstem, and small lesions. In general, it is used in patients on a nonemergent basis.

EEG is an important diagnostic tool in the evaluation of seizure types, response to treatment, and prognosis. It is rarely indicated in the acute-care setting.

Emergency Treatment

Prolonged seizure activity is a true medical emergency. In one series, 88 of 239 patients who had convulsive status epilepticus for more than 1 hour had permanent neurologic sequelae. Thus, following stabilization of the ABCs, further treatment is directed at stopping the seizure activity. Although certain causes of seizures may require a specific treatment, anticonvulsant therapy is initiated simultaneously during the evaluation of the seizing patient (Fig. 70.2). The approach to this subject is detailed in Chapter 83, but some emergency treatment guidelines are reviewed here.

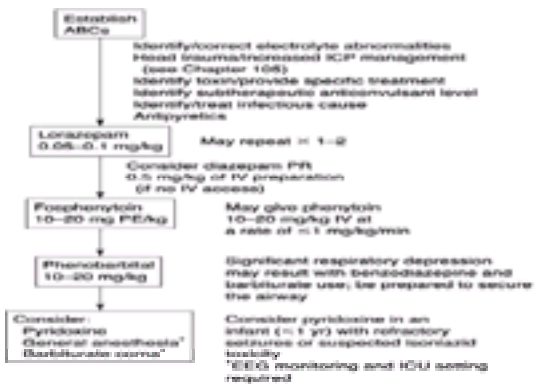

FIGURE 70.2. Management of status epilepticus. *ICP*, intracranial pressure; *PR*, per rectum; *IV,* intravenous; *PE*, phenytoin equivalent; *ICU*, intensive care unit.

The benzodiazepines are the initial drug of choice for the treatment of seizures. Lorazepam (Ativan) has a rapid onset of action (less than 5 minutes) and can be given in the intravenous or intramuscular form. The dose is 0.05 to 0.1 mg/kg with a maximal dose of 4 mg and can be given over 1 to 2 minutes. Its anticonvulsant effects can last for several hours. It may be repeated at 10- to 15-minute intervals, but its effectiveness decreases with successive doses. The major side effects are respiratory depression and sedation (dose dependent), especially when combined with phenobarbital.

Diazepam (Valium) had been the standard initial treatment of seizures for many years before the development of the newer benzodiazepines. Diazepam is similar to lorazepam, but because of its increased lipid solubility, it has a much shorter half-life. Diazepam has an advantage in that it can be given rectally, which is useful when a patient does not have IV access. The IV preparation of the drug should be used when it is given rectally at a dose of 0.5 mg/kg.

Phenytoin (Dilantin) is a second-line agent for the treatment of seizures. The dose is 10 to 20 mg/kg as an initial load. It has several limitations as compared with the benzodiazepines. First, peak CNS concentrations may not be reached until 10 to 30 minutes after its infusion is completed, and thus, it is much slower in onset. Furthermore, it must be administered slowly (no faster than 1 mg/kg per minute) because of concerns of cardiac conduction disturbances, which further lengthens its onset of action. It cannot be given in dextrose-containing solutions.

As a result of the limitations in administration of phenytoin, fosphenytoin (Cerebyx) was created. It is a prodrug whose active metabolite is phenytoin. The drug is dosed as phenytoin equivalents (PE) and the loading dose is 10 to 20 mg PE/kg. The advantages are that it can be given much more rapidly (up to 150 mg PE/minute) and that it may be given in either normal saline or a 5% dextrose–containing solution or intramuscularly.

Phenobarbital (Luminal) is another second-line agent in the treatment of seizures. The loading dose is 10 to 20 mg/kg. Its advantage over phenytoin is that it can be given much more rapidly (100 mg/minute). However, it has an extremely long half-life (up to 120 hours) and a pronounced sedating effect. Furthermore, it can cause significant respiratory depression, especially when given after a benzodiazepine. One must be prepared to intubate a patient who has received both a

benzodiazepine and a barbiturate for the treatment of seizures. It is important to remember that if a patient needs intubation, a muscle relaxant can mask the motor manifestation of seizure activity. With the creation of fosphenytoin, phenobarbital should now be considered a third-line agent.

Paraldehyde was used as a fourth-line agent if the previously discussed therapies had failed to control the seizure activity or when seizing patients had no IV access. It was administered rectally in a corn oil suspension at a dose of 0.3 mL/kg. Its major side effect (besides a foul smell) was metabolic acidosis. Although many centers still have supplies of paraldehyde, it is currently no longer being manufactured.

Pyridoxine deficiency is an uncommon cause of seizures in newborns. One should consider its use in patients less than 1 year of age whose seizure activity is refractory to the other therapies (100 mg). It is also used in the treatment of isoniazid overdose (usual initial dose 70 mg/kg).

If all of the described therapies fail, patients may require general anesthesia to abort the seizures. A variety of agents can be used, such as inhalational anesthetics (e.g., halothane, isoflurane) or large doses of short-acting barbiturates (e.g., pentobarbital). The patient needs both to be intubated (if not already done) and to have continuous EEG monitoring in an intensive care unit. The level of anesthesia should be sufficient to maintain either a flat-line or burst-suppression pattern on the EEG. The anesthesia can be then withdrawn slowly to see if any electrical seizure activity persists.

SPECIAL CONSIDERATIONS

Febrile Seizures

Febrile seizures are the most common convulsive disorder in young children, occurring in 2 to 5% of the population (see Chapter 83). A consensus statement by the National Institutes of Health defines febrile seizures as a seizure occurring between 3 months and 5 years of age that is associated with a fever but without evidence of intracranial infection or other defined cause.

Febrile seizures can be of any type, but most commonly, they are generalized tonic-clonic seizures. They are usually self-limited and last for only a few minutes. Febrile seizures are classified as simple febrile seizures, which last less than 15 minutes and are generalized, and complex febrile seizures, which are prolonged, recur within 24 hours, and are focal. Simple febrile seizures (85%) are much more common. A familial tendency toward febrile seizures exists, but the exact inheritance is uncertain.

After the first febrile seizure, approximately 33% of patients will have at least one recurrence and about 9% will have three or more episodes. The younger the patient is at first presentation, the greater the likelihood of recurrence. Most recurrences (75%) will also happen within 1 year. The exact risk of developing epilepsy after a febrile seizure is unknown, but most studies indicate that it is less than 5%. Risk factors for developing epilepsy after a febrile seizure include abnormal development before the episode, a family history of afebrile seizures, and a complex first febrile seizure.

The treatment of a patient who presents during a febrile seizure is identical to that for other seizure types. The primary goal is the establishment of a clear airway; secondary efforts are then directed at termination of the seizure. However, because most febrile seizures are brief in duration, the typical patient who presents for evaluation of a febrile seizure is no longer seizing upon arrival to the ED. In those instances, if the history is consistent with a simple febrile seizure, the patient has no stigmata of a CNS infection, and the patient's neurologic examination is completely “normal” (the patient may be postictal or slightly hyperreflexive), further evaluation for the cause of the seizure is unnecessary. However, the evaluation should focus on the possible cause of the fever.

It is important to note that typical signs of meningitis may be absent in patients less than 12 to 18 months of age. Furthermore, seizure may be the first presentation of meningitis. Thus, one should strongly consider an LP in all patients less than 12 months who present with a simple febrile seizure, and one should maintain a low threshold to perform one in patients 12 to 18 months of age. LP is recommended in patients with complex febrile seizures or a concerning physical or neurologic evaluation.

Patients who have a simple febrile seizure may be safely discharged to home. Parents should be reassured that febrile seizures are common and that most patients have no further episodes. They need to be cautioned that a recurrence may happen and should be given simple instructions on what to do should another seizure occur. Furthermore, parents should understand that patients with recurrent febrile seizures may still contract meningitis and may require clinical evaluation for this possibility. They can also be instructed on the proper use of antipyretics, even though no studies have been conducted to document that this is effective in reducing the recurrence rate. Finally, any identified source of the fever should be properly treated.

Suggested Readings

Barron TF, Hunt SL. A review of the newer antiepileptic drugs and the ketogenic diet. Clin Pediatr 1997;36:513–521.

Berg AT, Shinnar S, Hauser WA, et al. A prospective study of recurrent febrile seizures. N Engl J Med 1992;327:1122–1127.

Dichter MA, Brodie MJ. New antiepileptic drugs. N Engl J Med 1996;334:1583–1590.

Dodson WE. Medical treatment and pharmacology of antiepileptic drugs. Pediatr Clin North Am 1989;36:421–433.

Golden GS. Nonepileptic paroxysmal events in childhood. Pediatr Clin North Am 1992;39:715–725.

Hirtz DG. Febrile seizures. Pediatr Rev 1997;18:5–8.

Lowenstein DH, Alldredge BK. Status epilepticus. N Engl J Med 1998;338:970–976.

Mizrahi EM. Seizure disorders in children. Curr Opin Pediatr 1994; 6:642–646.

Nelson KB, Ellenberg JH. Prognosis in children with febrile seizures. Pediatrics 1978;61:720–727.

Pellock JM. Seizures and epilepsy in infancy and childhood. Neurol Clin 1993;11:755–775.

Shields WD. Status epilepticus. Pediatr Clin North Am 1989;36:383–393.

Verity CM, Butler NR, Golding J. Febrile convulsions in a national cohort followed-up from birth. I. Prevalence and recurrence in the first five years of life. Br Med J 1985;290:1307–1310.

Vining EP. Pediatric seizures. Emerg Med Clin North Am 1994;12:973–988.

CHAPTER 71

Sore Throat

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Sore throat refers to any painful sensation localized to the pharynx or the surrounding areas. Because children, particularly those of preschool age, cannot define their symptoms as precisely as adults, the physician who evaluates a child with a sore throat must first define the exact nature of the complaint. Occasionally, young patients with dysphagia (see Chapter 53) that results from disease in the area of the esophagus or with difficulty swallowing because of a neuromuscular disorder will verbalize these feelings as a sore throat. Careful questioning usually suffices to distinguish between these complaints.

Although a sore throat is less likely to portend a life-threatening disorder than dysphagia or the inability to swallow, this complaint should not be dismissed without a thorough evaluation. Most children with sore throats have self-limiting or easily treated pharyngeal infections, but a few have serious disorders, such as retropharyngeal or lateral pharyngeal abscesses. Even if the reason for the complaint of sore throat is believed to be an infectious pharyngitis, several different organisms may be responsible. Symptomatic therapy, antibiotics, anti-inflammatory drugs, or surgical intervention may be appropriate at times. Most children experience no adverse consequences from misdiagnosis and inappropriate therapy, but a few may develop local extension of infection; chronically debilitating illnesses, such as rheumatic fever; or life-threatening airway obstruction.

DIFFERENTIAL DIAGNOSIS

Infectious Pharyngitis

Infection is the most common cause of sore throat and usually is caused by respiratory viruses, including adenoviruses, coxsackie A viruses, or parainfluenza virus (Table 71.1, Table 71.2 and Table 71.3). Several of the respiratory viruses produce easily identifiable syndromes, including hand–foot–mouth disease (coxsackievirus) and pharyngoconjunctival fever (adenovirus). These viral infections are closely followed in frequency by bacterial infections caused by group A streptococcus *(Streptococcus pyogenes).* In the winter months during streptococcal outbreaks, as many as 30 to 50% of episodes of pharyngitis may be caused by *S. pyogenes*. The only other common infectious agent in pharyngitis is the Epstein-Barr virus, which causes infectious mononucleosis. Although infectious mononucleosis is not often seen in children under 5 years of age (Fig. 71.1), it cannot be considered rare even during these early years of life. More commonly, however, it affects the adolescent. An additional consideration in adolescents with an infectious mononucleosis-like syndrome is human immunodeficiency virus (HIV).

Infectious pharyngitis	Other Causes
Respiratory viruses	Herpetic stomatitis
Group A streptococci	Irritative pharyngitis
Epstein-Barr virus (infectious mononucleosis)	Foreign body
Human immunodeficiency virus	Peritonsillar abscess
Neisseria gonorrhoeae	Retropharyngeal and lateral pharyngeal abscesses
Anaerobic bacteria	Epiglottitis
Group C and G streptococci (?)	Kawasaki disease
Arcanobacterium haemolyticum (?)	Chemical exposure
Mycoplasma pneumoniae (?)	Psychogenic pain
Chlamydia pneumoniae (?)	Referred pain
Francisella tularensis	
Corynebacterium diphtheriae (diphtheria)	

Table 71.1. Differential Diagnosis of Sore Throat in the Immunocompetent Host

Infectious pharyngitis
Respiratory viruses
Group A streptococci
Epstein-Barr virus
Irritative pharyngitis

Table 71.2. Common Causes of Sore Throat

Retropharyngeal and lateral pharyngeal abscesses
Epiglottitis
Tonsillar hypertrophy (severe) with infectious mononucleosis
Diphtheria
Peritonsillar abscess

Table 71.3. Life-Threatening Causes of Sore Throat

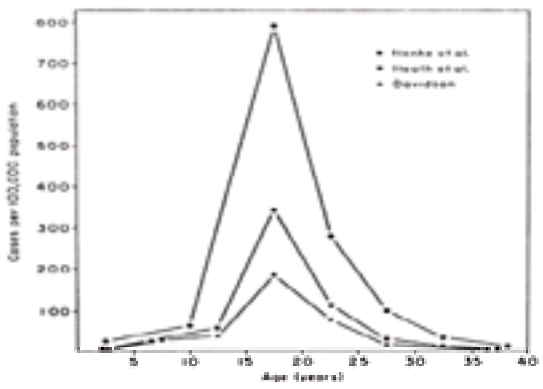

FIGURE 71.1. Incidence by age of infectious mononucleosis in three large studies.

Other organisms produce pharyngitis only rarely; these include *Neisseria gonorrhoeae, Corynebacterium diphtheriae, Francisella tularensis*, and anaerobic bacteria. *N. gonorrhoeae* may cause inflammation and exudate but more often remains quiescent, being diagnosed only by culture. Diphtheria is a life-threatening but seldom encountered cause of infectious pharyngitis, characterized by a thick membrane and marked cervical adenopathy. Oropharyngeal tularemia is rare and should be entertained only in endemic areas among children who have an exudative pharyngitis that cannot be categorized by standard diagnostic testing and/or persists despite antibiotic therapy. Although unusual, mixed anaerobic infections should be considered in the ill-appearing adolescent with a severe pharyngitis because these organisms occasionally lead to sepsis (Lemierre's disease). Other bacteria—group C and G streptococci, *Arcanobacterium hemolyticum, Mycoplasma pneumoniae*, and *Chlamydia pneumoniae*—have been implicated as agents of pharyngitis in adults, but in childhood, their roles remain unproved and their frequency is unknown.

Irritative Pharyngitis/Foreign Body

Drying of the pharynx may irritate the mucosa, leading to a complaint of sore throat. This condition occurs most commonly during the winter months, particularly after a night's sleep in a house with forced hot-air heating. Occasionally, a foreign object such as a fishbone may become embedded in the pharynx.

Herpetic Stomatitis

Stomatitis caused by herpes simplex usually is confined to the anterior buccal mucosa but may extend to the anterior tonsillar pillars. Particularly in these more extensive cases, the child may complain of a sore throat.

Peritonsillar Abscess

A peritonsillar abscess may complicate a previously diagnosed infectious pharyngitis or may be the initial source of a

child's discomfort. This disease is most common in older children and adolescents. The diagnosis is evident from visual inspection, augmented occasionally by careful palpation. These abscesses produce a bulge in the posterior aspect of the soft palate, deviate the uvula to the contralateral side of the pharynx, and have a fluctuant quality on palpation.

Retropharyngeal and Lateral Pharyngeal Abscesses

Retropharyngeal abscess is an uncommon cause of sore throat, usually occurring in children less than 4 years of age. Although most children with this disorder appear toxic and have respiratory distress, a few complain of sore throat and dysphagia without other manifestations early in the course. A soft-tissue radiographic examination of the lateral neck demonstrates the lesion readily, whereas direct visualization often is impossible. Unfortunately, even limited flexion of the neck during the radiograph may cause a buckling of the retropharyngeal tissues that resembles a purulent collection. The physician must insist on a film with the neck fully extended before hazarding an interpretation. If the diagnosis remains uncertain despite adequate radiographs, a computed tomography (CT) scan should be obtained.

Lateral pharyngeal abscesses manifest in a fashion similar to retropharyngeal infections but occur less often. High fever is a common symptom, and both trismus and swelling below the mandible may be seen. To confirm the diagnosis, a CT scan is appropriate.

Epiglottitis

The incidence of epiglottitis, a well-appreciated cause of life-threatening upper airway infection, has declined significantly since the introduction of vaccination against *Haemophilus influenzae* type b. This disease manifests with a toxic appearance, high fever, stridor, and drooling. In every reported series of cases, sore throat appears on the list of symptoms. Although rarely this may be the primary complaint in a child, other more striking findings almost always predominate. Epiglottitis should be excluded easily as a diagnosis in the patient with a sore throat who is without stridor and appears relatively well.

Kawasaki Disease

Kawasaki disease is characterized by high fever along with at least four of the five following findings: 1) conjunctivitis, 2) mucositis, 3) peripheral erythema and/or edema, 4) truncal rash, and 5) cervical adenopathy. The mucositis most commonly involves the lips, but occasionally pharyngitis may be a prominent feature.

Chemical Exposure

Certain ingestions, such as paraquat and various alkalis, may produce a chemical injury to the mucosa of the pharynx. Usually, these findings occur in the setting of a known ingestion and are accompanied by lesions of the oral mucosa. In addition, several systemic inflammatory conditions (Behçet's syndrome and Stevens-Johnson syndrome) may involve the pharynx.

Referred Pain

Occasionally, pain from inflammation of extrapharyngeal structures is described as arising in the pharynx. Examples include dental abscesses, cervical adenitis, and occasionally, otitis media.

Psychogenic Pharyngitis

Some children who complain of a sore throat have no organic explanation for their complaint after a thorough history and physical examination and a throat culture. In these cases, the physician should consider the possibility of anxiety, at times associated with frequent or difficult (globus hystericus) swallowing.

Pharyngitis in the Immunosuppressed Host

Immunosuppressed hosts may develop pharyngitis from any of the previously discussed causes. In addition, these patients exhibit a particular susceptibility to infections with fungal organisms, such as *Candida albicans.*

EVALUATION AND DECISION

The history and physical examination should focus on findings seen with systemic illnesses causing pharyngitis and the appearance of the oral cavity. A medical history of an immunosuppressive disorder or missed immunizations raises the specter of unusual infections. A sudden onset is most characteristic of epiglottis.

Fever, either historical or measured, points to an infection or, less commonly, Kawasaki disease. Toxicity and/or respiratory distress occurs with infections leading to respiratory obstruction, such as peritonsillar, retropharyngeal, and lateral pharyngeal abscesses; epiglottitis; diphtheria; and infectious mononucleosis with severe tonsillar hypertrophy. Conjunctivitis suggests pharyngoconjunctival fever (adenovirus) or Kawasaki disease; generalized adenopathy occurs with infectious mononucleosis and human immunodeficiency virus (HIV); and a rash is seen with scarlet fever (group A streptococcus), Kawasaki disease, and infectious mononucleosis, particularly after the administration of amoxicillin.

The tendency of most clinicians is to assume that one of the common organisms is the cause of pharyngitis in the child with a sore throat. Before settling on infectious pharyngitis, however, the emergency physician should first consider several more serious disorders (Fig. 71.2). Conditions that have immediate life-threatening potential include epiglottitis, retropharyngeal and lateral pharyngeal abscesses, peritonsillar abscess, severe tonsillar hypertrophy (usually as an exaggerated manifestation of infectious mononucleosis), and diphtheria. Generally, stridor and signs of respiratory

distress accompany the complaint of sore throat in epiglottitis and retropharyngeal abscess. Drooling and voice changes are common in children with these two conditions, as well as in patients with peritonsillar abscess and severe infectious tonsillar hypertrophy. In cases of epiglottitis or retropharyngeal abscess that are not clinically obvious, a lateral neck radiograph, obtained under appropriate supervision, is confirmatory. Peritonsillar abscess and tonsillar hypertrophy are diagnosed by visual examination of the pharynx. Diphtheria is rarely a consideration except in unimmunized children, particularly those from underdeveloped nations.

FIGURE 71.2. Diagnostic approach to the child with sore throat.

The next phase of the evaluation of the child with a complaint of sore throat hinges on a careful physical examination, particularly of the pharynx (Fig. 71.2). The appearance of vesicles on the buccal mucosa anterior to the tonsillar pillars points to a herpetic stomatitis or noninfectious syndromes, such as Behçet's or Stevens-Johnson syndrome (erythema multiforme). Uncommonly, a small, pointed foreign body, most commonly a fishbone, becomes lodged in the mucosal folds of the tonsils or pharynx; usually, the history suggests the diagnosis, but an unanticipated sighting may occur in the younger child. Significant asymmetry of the tonsils indicates a peritonsillar cellulitis or, if extensive, an abscess. Clinically, the diagnosis of an abscess is reserved for the tonsil that protrudes beyond the midline, causing the uvula to deviate to the uninvolved side. Kawasaki disease produces a systemic syndrome with a prolonged fever and other characteristic findings that are usually more prominent than the pharyngeal involvement.

The remaining organic diagnoses, once those already discussed have been eliminated by history, physical examination, and occasionally, imaging, include referred pain, irritative pharyngitis, and infectious pharyngitis. Sources of referred pain (otitis media, dental abscess, and cervical adenitis) usually are identified during the examination. Irritative pharyngitis, seen most commonly during the winter among older children who live in homes with forced hot-air heating, produces minimal or no pharyngeal inflammation. It often is transient, appearing on arising and resolving by midday.

Infectious pharyngitis (Fig. 71.3) evokes a spectrum of inflammatory responses that range from minimal injection of the mucosa to beefy erythema with exudation and edema formation. The three relatively common causes are streptococci, respiratory viruses, and infectious mononucleosis (Fig. 71.3). In a few cases, a viral pharyngitis that results from coxsackievirus infection will be self-evident on the basis of vesicular formation in the posterior pharynx or involvement of the extremities (hand–foot–mouth syndrome). Such patients require only symptomatic therapy. A small number of additional patients will have signs of infectious mononucleosis: large, mildly tender posterior cervical lymph nodes; diffuse lymphadenopathy; and/or hepatosplenomegaly. In these children, the physician should obtain a white blood cell (WBC) count with differential and a slide test for heterophil antibody (Fig. 71.4) in an effort to confirm the clinical diagnosis, thereby guiding therapy and discussion of prognosis. Some children, especially those less than 5 years of age, will not have the characteristic lymphocytosis or heterophil antibody response and will require repeated testing or specific serologic assays for antibodies to Epstein-Barr virus. In the rare child with an unusual history, the physician must pursue diagnoses such as gonococcal pharyngitis (sexual abuse, oral sex) or diphtheria (immigration from an underdeveloped nation, lack of immunization).

FIGURE 71.3. Diagnostic approach to infectious pharyngitis in the immunocompetent child.

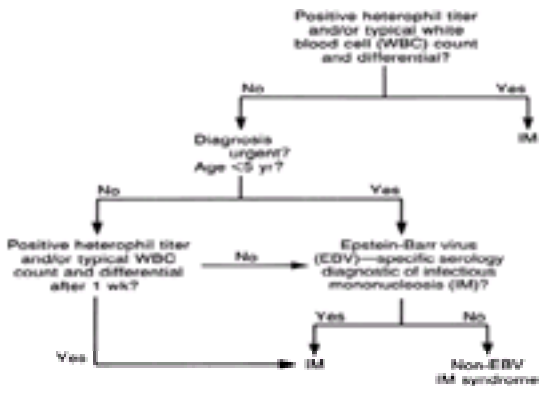

FIGURE 71.4. Diagnostic approach when findings are clinically suggestive for mononucleosis.

Ultimately, most children will have a mildly to moderately inflamed pharynx but no specific etiologic diagnosis based solely on the history and physical examination. Although certain symptoms and signs favor streptococcal infection, none is conclusive. Thus, obtaining a rapid test (latex agglutination or optical immunoassay) for group A streptococcus, followed by a culture, if negative, is prudent. Rapid tests are most helpful when positive because specificity of the tests is high; however, a negative test does not exclude streptococcal infection reliably, although some authorities would be satisfied with a negative optical immunoassay alone. With the recent reported rise in the incidence of rheumatic fever, the accurate diagnosis of streptococcal pharyngitis assumes increasing importance. Generally, symptomatic therapy suffices in the patient with a negative rapid test, although the physician may elect to initiate therapy with penicillin while awaiting the results of the throat culture in selected cases with highly suggestive clinical features.

Suggested Readings

Bisno AL, Gerber MA, Gwalteny JM, et al. Diagnosis and management of streptococcal pharyngitis: a practice guideline. Clin Infect Dis 1997;25:574–583.

Breese BB. A simple scorecard for the tentative diagnosis of streptococcal pharyngitis. Am J Dis Child 1977;131:514.

Davidsohn RJL. A survey of infectious mononucleosis in the Northeast Regional Hospital Board area of Scotland, 1960–1969. J Hyg (Camb) 1970;68:393.

Eichenwald H. Respiratory infections in children. Hosp Pract 1976;11:4, 81–90.

El-Daher NT, Hijazi SS, Rawasdeh NM, et al. Immediate vs. delayed treatment of group A beta-hemolytic streptococcal pharyngitis with penicillin V. Pediatr Infect Dis J 1991;10:126.

Fleisher GR, Lennette ET, Henle G, et al. Incidence of heterophile antibody responses in children with infectious mononucleosis. J Pediatr 1979; 94:723.

Heath CW, Brodsky AL, Potolsky AI. Infectious mononucleosis in a general population. Am J Epidemiol 1972;95:46.

Henke CE, Kurland LT, Elveback LR. Infectious mononucleosis in Rochester, Minn, 1950 through 1969. Am J Epidemiol 1973;98:483.

Kaltwasser G, Diego J, Welby-Sellenriek PL, et al. Polymerase chain reaction for *Streptococcus pyogenes* used to evaluate optical immunoassay for the detection of group A streptococci in children with pharyngitis. Pediatr Infect Dis J 1997;16:748–753.

Komaroff AL, Aronson MD, Pass TM, et al. Serologic evidence of chlamydial and mycoplasmal pharyngitis in adults. Science 1983;222:927.

Lieu TA, Fleisher GR, Schwartz JS. Clinical evaluation of a latex agglutination test for streptococcal pharyngitis: performance and impact on treatment rates. Pediatr Infect Dis J 1988;7:847.

Lieu TA, Fleisher GR, Schwartz JS. Cost-effectiveness of rapid latex agglutination testing and throat culture for streptococcal pharyngitis. Pediatrics 1990;85:246.

Veasey LG, Weidmeier SE, Ormond GS, et al. Resurgence of acute rheumatic fever in the intermountain area of the United States. N Engl J Med 1987;316:421.

Wald ER, Green MD, Schwartz B, et al. A streptococcal scorecard revisited. Pediatr Emerg Care 1998;14:109–111.

Webb KH. Does culture confirmation of high-sensitivity rapid streptococcal tests make sense? Pediatrics 1998;101:E2.

Weisner PJ, Tronca E, Bonin P, et al. Clinical spectrum of pharyngeal gonococcal infection. N Engl J Med 1973;288:181.

CHAPTER 72

Stridor

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Stridor, although a relatively common occurrence, can be frightening to both child and parents. The presence of stridor necessitates a complete and careful evaluation to determine the cause of this worrisome and occasionally life-threatening symptom. This chapter presents the causes of stridor and provides the emergency practitioner with guidelines for initial evaluation and management.

PATHOPHYSIOLOGY

Stridor is an externally audible sound associated with respiration. It is produced by turbulent air flow through large airways. It occurs when a normal respiratory volume of air moves through narrowed airways, which results in the normal laminar flow becoming turbulent. Stridor thus signifies partial airway obstruction.

DIFFERENTIAL DIAGNOSIS

Stridor may occur in a wide variety of disease processes affecting the large airways from the nares to the bronchi but most often arises with disorders of the larynx and trachea (Table 72.1). For the purposes of differential diagnosis, it is helpful to categorize the common causes of stridor as acute or chronic in onset and to further divide acute onset into febrile and afebrile causes (Table 72.2). In addition, life-threatening causes of stridor must be considered during the earliest phases of evaluation (Table 72.3).

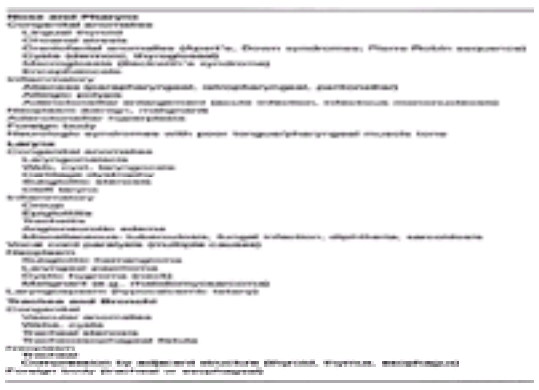

Table 72.1. Causes of Stridor by Anatomic Location

Table 72.2. Common Causes of Stridor

Usually Febrile	Usually Afebrile
Epiglottitis	Foreign body
Retropharyngeal abscess	Angioneurotic edema
Tracheitis	Neck trauma
	Neoplasm (compressing trachea)
	Thermal or caustic injury

Table 72.3. Life-Threatening Causes of Stridor

Stridor with Acute Onset in the Febrile Child

Laryngotracheitis (croup) is by far the most common cause of stridor in the febrile child. However, it is important to consider other diagnoses such as retropharyngeal abscess, epiglottitis, and tracheitis (see Chapter 84). Although rare, these diseases are life-threatening causes of inspiratory stridor. History and physical examination are important tools in determining which patients with inspiratory stridor need further evaluation.

Laryngotracheitis affects children most often between 6 to 36 months of age but is seen throughout childhood. The illness begins with upper respiratory tract symptoms and fever, usually ranging from 38° to 39°C (100.4° to 102.2°F). Within 12 to 48 hours, a barky, “seal-like” cough and inspiratory stridor are noted. Supraclavicular and subcostal retractions may be present. Symptoms are aggravated by crying and ameliorated by cool mist or nebulized epinephrine. Most children appear only mildly or moderately ill.

Tracheitis closely resembles croup except that affected patients generally appear toxic, tend to be older (4 to 6 years), and do not respond as well to cool mist or nebulized epinephrine. Dysphagia is common, and drooling may be present. The verbal child may complain of anterior neck pain or a painful cough. Less severe cases can more closely resemble croup.

Epiglottitis may be divided into disease caused by *Haemophilus influenzaen* or that caused by other pathogens. Patients with *H. influenzae* epiglottitis appear toxic and are febrile. Respiratory distress and a tripod stance are characteristic; drooling is usually present. *H. influenzae* epiglottitis is associated with sudden airway compromise that can be precipitated by manipulation of the oropharynx. Therefore, children in whom *H. influenzae* epiglottitis is strongly suspected should have the airway inspected under controlled conditions.

Epiglottitis caused by pathogens other than *H. influenzae* differs in many important ways from disease caused by *H. influenzae.* It is much more common in adults, has a slower onset, and is more likely to be associated with difficulty swallowing or sore throat. Last, the risk of airway compromise is also less than with epiglottitis caused by *H. influenzae.* However, any child with epiglottitis should be managed as if he or she has disease caused by *H. influenzae.*

The clinical picture of a retropharyngeal abscess is similar to epiglottitis except symptoms appear more gradually. In addition to drooling and stridor, meningismus caused by muscular irritation by the abscess may be present. Physical examination may rarely reveal midline swelling of the pharynx.

Stridor with Acute Onset in the Afebrile Child

A foreign body in either the trachea or esophagus may produce stridor. There may be a history of choking on food or a small object. Physical examination varies, depending on location of the foreign body.

Ingestion of either caustic or hot substances may result in injury to the airway or hypopharynx. Symptoms of airway compromise may be delayed for as long as 6 hours. Drug abuse is yet another potential source of injury: thermal epiglottitis has been reported after inhalation of crack smoke, a screen from a crack pipe, and a marijuana cigarette.

Other causes include spasmodic croup, angioneurotic edema, and trauma (see Chapter 112).

Chronic Stridor

The age at onset narrows the differential diagnosis. Stridor noted shortly after birth is most likely caused by a structural defect. This type of stridor tends to slowly worsen and is severe only when the infant is stressed such as during crying. Laryngomalacia is the most common cause of congenital stridor. Stridor associated with laryngomalacia is positional and is ameliorated by placing the infant in the prone position. Other congenital causes of stridor include laryngeal webs, laryngeal diverticula, vocal cord paralysis, subglottic stenosis, tracheomalacia, and vascular anomalies such as a double aortic arch or a vascular sling. Stridor in infants has also been reported to be associated with gastroesophageal reflux.

Stridor in older children may be caused by papillomas or neoplastic processes. Patients with papillomas generally present between 2 to 4 years of age with complaints of hoarseness and stridor. Neoplastic processes causing tracheal compression can also lead to stridor in the older child.

Psychogenic, also called functional, stridor is an uncommon cause of stridor in the older child. Cases have been reported

in adolescents, with the youngest age being 10 years old. Adolescent girls are diagnosed three times more often with this condition than are males. More than 50% of patients meet diagnostic criteria for a psychiatric disorder. Characteristically, stridor improves when the patient is unaware that he or she is being observed, and it may clear with cough. The diagnosis can be confirmed only by direct laryngoscopy in the symptomatic patient when the vocal cords are noted to be adducted during inspiration.

EVALUATION AND DECISION

The first priority is to ensure that the airway is adequate by assessing level of consciousness, color, perfusion, air entry, breath sounds, and work of breathing, including respiratory rate, nasal flaring, and retractions. Resuscitative measures should be instituted as necessary (see Chapter 5). The child may then be evaluated systematically. In the child with acute onset of stridor, history should focus on associated symptoms such as fever, duration of illness, drooling, rhinorrhea, and history of choking (Fig. 72.1). Immunization status should be verified, particularly *H. influenzae* vaccination. In the case of a child with chronic stridor, important historical points include onset and progression of stridor, as well as ameliorating and aggravating factors.

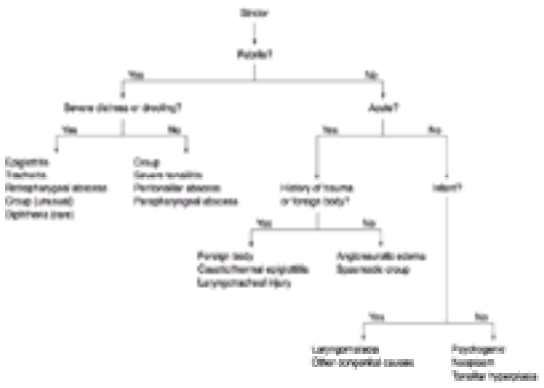

FIGURE 72.1. Diagnostic approach to stridor.

Several characteristics of stridor such as associated phase of respiration, pitch, and length of respiratory phase can help determine the level of obstruction. Inspiratory stridor occurs with obstruction of the extrathoracic trachea, biphasic stridor when both extrathoracic and intrathoracic trachea are involved, and expiratory stridor when only the intrathoracic trachea is involved. The pitch of the stridor also helps determine location. Laryngeal and subglottic obstructions are associated with high-pitched stridor. In contrast, obstruction of the nares and nasopharynx results in lower-pitched snoring or snorting sounds also called stertor. Because the passage of saliva and the flow of air are impeded in pharyngeal obstruction, these patients often have a gurgling quality of breathing. Last, the relative length of inspiratory and expiratory phase may be helpful. In children with laryngeal obstruction, the time of inspiration is greatly increased, whereas expiration tends to be prolonged in bronchial obstruction. Both inspiration and expiration times are increased in patients with tracheal obstruction.

Physical examination should include careful examination of the nares and oropharanyx with particular attention to increased secretions, drooling, visible mass, and abnormal phonation. Regional findings such as adenopathy, neck masses, meningismus, trauma, or bruising should also be sought. Position of comfort should be noted. Children with airway obstruction at the level of the larynx and above usually hyperextend their heads upon their necks and lean forward (“sniffing” position) in an effort to straighten their upper airway and maximize air entry. This posture does not help relieve more distal obstruction. Quality of the voice should be noted as normal, hoarse (vocal cord paralysis, papilloma), weak (neuromuscular disorder), or aphonic (laryngeal obstruction by a foreign body). Response to therapies, such as nebulized racemic epinephrine (croup), should be noted.

Emergency management of the child with stridor depends on its severity and its likely cause. Oxygen, humidified air, nebulized racemic epinephrine, corticosteroids, laryngoscopy, intubation, and even emergency cricothyroidotomy or tracheostomy all have specific roles in the emergency department (ED) management of stridor, depending on its cause (see Chapter 112 and Chapter 121).

Febrile Child

In the febrile child with stridor, the onset is generally acute and the most likely, as well as concerning, diagnostic possibilities are croup, epiglottitis, and bacterial tracheitis. Radiographs of the neck may be helpful in evaluation and should be considered if the practitioner suspects a diagnosis other than croup. If epiglottitis is strongly suspected, a lateral neck radiograph should be obtained in the ED or the child should be taken to the operating room to have direct visualization of the epiglottis under controlled conditions. Abnormal findings on a lateral neck radiograph include increased prevertebral width (retropharyngeal abcess), swollen epiglottis or aryepiglottic folds (epiglottitis), and irregular tracheal borders or stranding across the trachea (tracheitis). Radiographic findings consistent with croup are a narrowed subglottic area on anteroposterior view (the “steeple sign”) and ballooning of the hypopharynx, best appreciated on the lateral view. Airway films must be interpreted with care because they are subject to artifact. To properly interpret the prevertebral space, the lateral neck radiograph must be taken with the patient's head extended and during inspiration. Normal tracheal buckling, which is seen during expiration in a young child, may be misinterpreted as tracheal mass lesion or deviation from an extrinsic mass (Fig. 72.2).

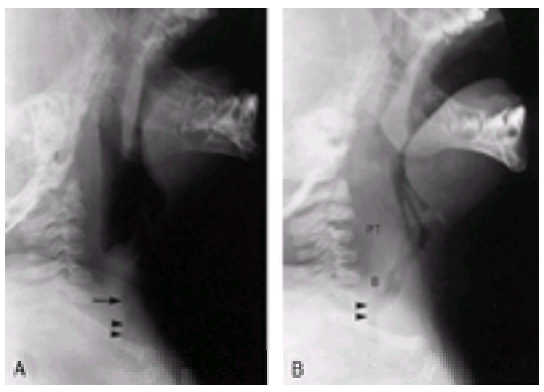

FIGURE 72.2. Inspiratory **(A)** and expiratory **(B)** lateral neck radiographs of a child with upper airway obstruction secondary to a granuloma *(arrow)* in the upper trachea. Note ballooning of the pharynx during inspiration **(A)** and narrowing of the trachea *(arrowheads)* below the level of obstruction. On expiration **(B)** note the normal pharyngeal lumen and dilation *(arrowheads)* of the trachea distal to the obstruction. The "bunching up" of the pharyngeal tissues *(PT)* and the buckling of the trachea **(B)** are normal findings on expiratory films.

Afebrile Child

In the afebrile child with acute onset of stridor, the duration of stridor, the likelihood of foreign body aspiration, and the child's age are all key elements to consider. Emergent otolaryngologic or surgical consultation should be obtained in a child with evidence of airway obstruction if either aspirated foreign body or trauma are likely causes of stridor. Angioneurotic edema, an autosomal-dominant trait, is characterized by rapid onset of swelling without discoloration, urticaria, or pain. Symptoms may occur in affected patients as young as 2 years of age but usually are not severe until adolescence; they may be precipitated by trauma, emotional stress, or menses. A C_1 esterase inhibitor level should be considered if angioneurotic edema is suspected.

A child with chronic stridor generally does not require an extensive evaluation in the ED unless significant respiratory distress is present. The infant with chronic stridor who is otherwise well should be referred back to the private pediatrician or to an otolaryngologist. Once a neoplastic cause is deemed unlikely, the older child with chronic stridor should be referred to otolaryngology for evaluation, including direct visualization of the vocal cords.

Figure 72.1 presents an algorithm that summarizes the ED diagnostic approach to the child with stridor.

Suggested Readings

Athreya B, Silverman BK. Pediatric Physical Diagnosis. Norwalk, CT: Appleton-Century-Crofts, 1982:124–136.

Donnelly BW, McMillan JA, Weiner LB. Bacterial tracheitis: report of eight new cases and review. Rev Infect Dis 1990;12:729–735.

Handler SD. Diagnosis and management of stridor in children. In: Cummings CW, Fredrickson JM, Harker LA, et al, eds. Otolaryngology-Head and Neck Surgery. St Louis: Mosby 1986:2219–2228.

Holinger LD. Etiology of stridor in the neonate, infant and child. Ann Otol Rhinol Laryngol 1980;89:397–400.

Kissoon N, Kronick JB, Frewen TC. Psychogenic upper airway obstruction. Pediatrics 1988;81:714–716.

Kulick RM, Selbst SM, Baker MD, et al. Thermal epiglottitis after swallowing hot beverages. Pediatrics 1988;81:441–444.

Kuppersmith R, Rosen DS, Wiatrak BJ. Functional stridor in adolescents. J Adol Health 1993;14:166–171.

Mayo-Smith MF, Spinale JW. Thermal epiglottitis in adults: a new complication of illicit drug use. J Emerg Med 1997;15:483–485.

Mayo-Smith MF, Spinale JW, Donskey CJ, et al. Acute epiglottitis in adults: an 18-year experience in Rhode Island. Chest 1995;108: 1640–1647.

O'Hollaren MT, Everts EC. Evaluating the patient with stridor. Ann Allergy 1991;67:301–305.

Potsic WP, Bilaniuk LT, Wetmore RF. Diagnostic uses of computed tomography in otolaryngology. Audiology 1984;9:29–41.

Quinn-Bogard AL, Potsic WP. Stridor in the first year of life. Clin Pediatr 1977;16:913–919.

Simon NP. Evaluation and management of stridor in the newborn. Clin Pediatr 1991;30:211–216.

CHAPTER 73

Syncope

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From the Greek *synkoptein*, meaning “to cut short,” syncope is defined as the temporary loss of consciousness and postural tone, resulting from an abrupt, transient, and diffuse reversible disturbance of cerebral function. Often, the terms *fainting* or *blackout* spells are used. Pathophysiologically, syncope can be explained as a sudden reduction in delivery of substrates such as O_2 or glucose to the brain. Most transient altered consciousness events in children include seizures, syncopes, or hysteric episodes (“fits, faints, or fakes”) and the approach to diagnosis of the former is to exclude the latter two.

Syncope during childhood is generally a benign, isolated event, although in few instances, it can represent potentially life-threatening causes. A thorough history and physical examination, with attention to clues provided by the premonitory signs and symptoms, are usually sufficient to provide the cause of syncope in most cases. An overall incidence in the pediatric population of 0.1 to 0.5% has been reported. Syncope occurs predominantly in teenagers, with a peak incidence occurring between 15 and 19 years. Females were evaluated more commonly than males. It is estimated that at least 50% of all individuals will have a least one syncopal episode during their adolescence. Syncopal events account for 1 to 3% of all emergency room visits, and in this setting, the patient has usually regained consciousness by the time of initial assessment by a physician.

DIFFERENTIAL DIAGNOSIS

Pathophysiologically, all causes of transient and abrupt onset of alterations of consciousness can be best categorized in three broad groups (Table 73.1): 1) true syncope reflecting any mechanism that causes a transient decrease in substrate delivery to the brain (e.g., O_2, glucose, blood); 2) all seizures; and 3) hysterical pseudoloss of consciousness. Most children and adolescents who faint have orthostatic syncope, vasovagal episodes, or breath-holding spells (Table 73.2). This is in marked contrast to adults, who often have cardiovascular disease. Studies report obtaining a definite cause of the syncopal episode in 27% of all pediatric cases, of which migraines were found in 11%, seizures in 8%, and cardiac causes in 6%. Most cases were classified as vasovagal episodes.

1. Transient decrease in substrate delivery to the brain: syncope	3. Hysterical pseudoloss of consciousness
2. Seizures	

Table 73.1. Pathophysiologic Classification of Transient, Paroxysmal, Altered Consciousness

Table 73.2. Common Causes of Syncope

The goal in evaluating syncopal episodes must be to accurately identify the common benign events from the occasional warning signs of serious pathology. The causes of true syncope may be classified into three etiologic categories: *autonomic* (vasovagal), *cardiovascular,* and *metabolic.* Table 73.3 lists the major causes of syncope and conditions that mimic it.

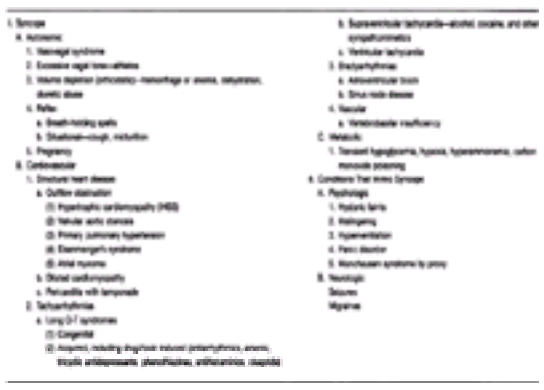

Table 73.3. Classification of Syncopal Episodes

Vasovagal Syncope

Vasovagal syncope is also called neurocardiogenic syncope, vasodepressor syncope, or fainting spell. It is by far the most common cause of fainting in children and adolescents. It accounts for more than 50% of cases of childhood syncope.

The pathophysiologic mechanism of vasovagal syncope is thought to be caused by an exaggerated Bezold-Jarisch reflex. This reflex is responsible for maintaining blood pressure during orthostatic stress. In patients prone to syncope, the cascade of events begins with a decrease in systemic venous return and therefore decreased preload after prolonged upright posture. Enhanced or compensatory sympathetic activity causes an elevation of circulating catecholamines (particularly epinephrine), which increase left ventricular contractility in a relatively empty ventricle. In response, a negative feedback loop via vagal afferents results in sympathetic withdrawal *(hypotensive vasodepressor response)* and augmented vagal tone *(the bradycardic cardioinhibitory response).* The factors that trigger this abnormal response are still unclear. It is probable that a combination of abnormal catecholamine response to orthostatic or other stress, exaggerated ventricular contraction, diminished ventricular volume from venous pooling in the upright position, and enhanced sensitivity of ventricular mechanoreceptors are all involved in the clinical predisposition to recurrent vasovagal syncope.

Most episodes occur while the patient is standing or during a rapid change from a supine or sitting position to standing. Syncope represents a cascade of signs and symptoms that begin with a brief prodrome or presyncopal phase. This progresses to a brief and sudden stage of unconsciousness that typically lasts 1 to 2 minutes and ends with arousal to a previous level of consciousness within a short period.

A syncopal episode may be triggered by a wide array of emotional events such as pain, fear, and anxiety, which increase circulatory catecholamines in response to a real or perceived threat. The prodromal symptoms may include light-headedness, dizziness, nausea, shortness of breath, diaphoresis, pallor, and visual changes. Physical conditions such as anemia, dehydration, exertion, hunger, pregnancy, and/or concurrent illness can predispose to a syncopal event. Other factors include confinement to enclosed or poorly ventilated spaces and environmental heat. The patient may remain nauseated, pale, and diaphoretic for several hours after the syncopal episode.

A full syncope can be avoided if the patient recognizes the prodromal symptoms and assumes a supine or Trendelenburg position. Prognostically, vasovagal syncope is considered a benign illness. A prophylactic approach is taken to prevent symptoms of presyncope or near-syncope. Patient education must be geared toward rapid symptom recognition, allowing the patient to assume a recumbent position and abort a potential syncopal event. Other preventative measures may include avoidance of dehydration and use of salt-enriched diets during athletic activity or environmental stress.

Several other related forms of autonomic syncope are orthostatic hypotensive syncope and situational syncope related to

micturition, defecation, cough, and swallow. Orthostatic hypotensive syncope is associated with an excessive and prolonged fall in blood pressure on assuming the erect posture from a recumbent position. An unusual and uncommon condition, micturition syncope, follows rapid bladder decompression, in which reduced cardiac return is associated with both postural effects and splanchnic vascular stasis. Underlying medical conditions such as anemia or pregnancy may exacerbate the tendency toward any of these vasovagal events.

Another common, usually benign, pediatric variant of vasovagal syncope is that of breath-holding spells, which occur in two forms (see Chapter 131). *Pallid* breath-holding spells result from vagally mediated cardiac inhibition. *Cyanotic* breath-holding spells involve interplay between hyperventilation, Valsalva maneuver, expiratory apnea, and intrinsic pulmonary mechanisms. In pallid breath-holding spells, an inconsequential injury induced by a sudden emotional stimulus such as pain, fright, or anger provokes one or two short cries, followed by pallor and sudden loss of consciousness. In cyanotic spells, the initial result is followed by vigorous crying and breath-holding in expiration, then loss of consciousness. These events typically occur in children between 6 and 18 months of age.

Cardiac Syncope

Syncope caused by significant cardiac or vascular pathology occurs far less often than autonomic syncope. It does not follow the stimuli typical of vasovagal syncope. Hypercyanotic spells, usually associated with tetralogy of Fallot, can occur with any heart defect associated with intracardiac right-to-left shunting. An increase in obstruction to pulmonary blood flow or a fall in systemic vascular resistance can precipitate such a spell. The most common arrhythmia causing syncope with an apparently normal heart structurally is supraventricular tachycardia (SVT), especially in the context of Wolff-Parkinson-White (WPW) syndrome. Arrhythmias occur more often in structurally abnormal hearts. Many drugs and toxins may induce arrhythmias (Table 74.4).

	Syncope, Vasovagal	Metabolic (e.g., hypoxia, hypoglycemia)	Seizure	Breath-holding
Period of Unconsciousness	Usually seconds	Variable	Minutes or longer	Seconds
Prodrome	Fright, pain, "feels faint"	Confusion, altered mental status, ↑ HR, diaphoresis	Occasional aura	Pain, fright → vigorous cry → apnea → LOC
Incontinence	Absent	Absent	May be present	Absent
Confusion on awakening	Absent or mild	Mild	Marked	Absent
Tonic-clonic movements	Occasionally present, if LOC is prolonged	May occur	Commonly present	Rare, may see 1–2 beats
EEG	Normal	Normal	Often abnormal	Normal

EEG, Electroencephalogram; HR, heart rate; LOC, loss of consciousness.

Table 73.4. Differentiating Syncope from Other “Spells”

The diagnosis of prolonged Q-T syndrome is made by documenting the prolongation of the corrected Q-T (Q-Tc greater than 0.45 seconds) interval by Bazett's formula. The prolongation of the Q-Tc interval results from a prolongation of the refractory period of the ventricular myocardium, which places it at risk for “torsades de pointes,” a malignant form of ventricular tachycardia (VT) (see Chapter 82). Prolonged Q-T syndrome can be congenital or acquired. Syncope occurs because of paroxysmal episodes of rapid VT. Of the congenital forms, the Jervell and Lange-Nielsen syndrome is associated with autosomal-recessive deafness. The Romano-Ward syndrome is associated with an autosomal-dominant trait in families with normal hearing. Recent work has identified the genetic basis of the congenital forms of long Q-T syndrome and relates them to the mutant genes coding for abnormal myocardial K^+ channel and, in some cases, inner ear endolymph proteins. Long Q-T syndromes often present as syncope on exercise or exertion and can also masquerade as a seizure. In acquired forms, the Q-Tc will be prolonged as a result of electrolyte abnormalities (hypokalemia, hypocalcemia), increased intracranial pressure (ICP), or medication use or overdose. Drug exposure may be intentional or accidental. A thorough history should include types of medications available at home, possible environmental exposures, and drug use such as psychotropic medications, including tricyclic antidepressants (TCAs) and phenothiazines. Nonsedating antihistamines such as terfenadine (Seldane) and astemizole (Hismanal) or the promotility agent cisapride may be used alone or in combination with other medications such as erythromycin and ketoconazole, which can inhibit the metabolism of these antihistamines and thus prolong the Q-T interval.

Episodic, complete heart block accompanied by syncope may occur in children and adolescents with baseline abnormalities of cardiac conduction. Children who have undergone surgical repair of ventricular defects, such as in Tetralogy of Fallot, are also at risk.

Hypertrophic cardiomyopathy (IHSS), which is associated with recurrent syncope, is a disease that presents with a thickened left ventricular myocardium, resulting in subaortic stenosis that causes obstruction to ventricular outflow. Patients with severe aortic valvular stenosis present with the classic triad of syncopal episodes, anginal chest pain, and dyspnea on exertion.

Noncardiac Syncope and Disorders That Mimic Syncope

Metabolic causes of syncope include hypoglycemia, which often is associated with pallor, dizziness, and diaphoresis and is unrelated to position. Seizures may occur and unconsciousness may be prolonged and will often require the administration of glucose for recovery. Hypoglycemia may be a component of other childhood disorders that include diabetes mellitus, ketotic hypoglycemia, and hepatic enzyme deficiencies. Other metabolic causes of syncope include hypoxia by itself or in association with mild to moderate carbon monoxide poisoning, which is notorious for producing syncope. Recovery occurs when the child is removed from the offending environment. Hyperammonemia may rarely

cause syncope by direct cytotoxic central nervous system effect.

Hyperventilation is associated with high anxiety and emotional events, during which the patient will complain of shortness of breath, tachypnea, chest pain, paresthesias, and light-headedness. It is believed to result from cerebral vasoconstriction in response to self-induced hypocapnia.

Loss of consciousness often occurs with generalized seizures, which may be difficult to distinguish from vasovagal syncope if the event was not witnessed. Seizures are likely to be preceded by an aura and followed by a prolonged postictal state. Neonatal seizures and complex partial seizures may be subtle and particularly difficult to differentiate from syncope (Table 73.4) (see Chapter 70).

Syncopal migraine is rare. It is usually preceded by an aura and followed by severe occipital headache. Basilar artery migraine accounts for 24% of childhood migraines, most often in adolescent girls. It should be considered in patients with severe paroxysmal headaches.

Syncopelike events caused by hysteria are common in the adolescent patient. Characteristic features of the clinical event are helpful in differentiating hysteria from organic causes of true syncope. Hysterical “syncope” may be associated with hyperventilation, usually occurs in the presence of an audience, and lacks true loss of consciousness. No overt or objective prodromal symptoms such as hypotension or bradycardia are recognized. It may occur when the patient is in the supine position, which is virtually unreported with vasovagal syncope. There may be a peculiar fluttering of the eyes behind half-closed eyelids. The patient describes the event in a calm and indifferent manner and vividly recalls the event, suggesting a lack of complete loss of consciousness.

Drug or toxin exposure may be accidental or intentional and, in addition to precipitating arrhythmias as previously noted, may occasionally cause an acute, transient loss of consciousness or gradual altered mental status changes leading to syncope rather than the typical prolonged alterations in consciousness. Such an effect may be more characteristic of carbon monoxide poisoning or abused volatile inhalants, for example (see Chapter 88). Antihypertensives, b blockers, diuretics, antiarrhythmics, and drugs that decrease cardiac output such as barbiturates, TCAs and phenothiazines may cause syncope. Substances of abuse such as alcohol, sedative-hypnotics, and opiates can cause alterations in consciousness that mimic syncope but are usually more prolonged.

EVALUATION AND DECISION

In the era of rising health care costs and cost containment, one must be mindful of the utility and expense of diagnostic studies used to evaluate syncope. Extensive and expensive testing is usually unnecessary. A thorough history and physical examination will often suggest the diagnosis. One should pay attention to the airway, respiratory effort, and hemodynamic stability. Vital signs, including orthostatic blood pressure and pulse oximetry measurements, must be documented and reviewed. In most cases, a complementary electrocardiogram (ECG) screen is useful to rule out symptomatic arrhythmias. This approach, emphasizing primarily clinical features of the syncopal episode, supplemented by ECG evaluation is outlined in Figure 73.1.

FIGURE 73.1. The diagnostic approach to syncope. *ECG,* electrocardiogram; *EEG,* electroencephalogram.

The goal in evaluating a syncopal child or adolescent is to identify conditions that are associated with a risk of serious injury or are life-threatening. Those potentially life-threatening conditions for which hospitalization is indicated are outlined in Table 73.5.

Presence of cardiovascular disease or abnormal cardiovascular examination—congestive heart failure, arrhythmias	Apnea or bradycardic spells requiring vigorous stimulation
Abnormal ECG—prolonged Q-T interval, tachyarrhythmias, atrioventricular or severe bundle branch blocks	Abnormal neurologic findings—local signs, status epilepticus, signs of meningeal irritation
Chest pain with syncope	Acute toxic ingestions
Cyanotic spells	Orthostatic hypotension resistant to fluid therapy

ECG, electrocardiogram.

Table 73.5. Life-Threatening Causes of Syncope Requiring Hospitalization

A thorough history is the most important part of the evaluation. Parents and relatives often contribute important information to the cause of the syncopal event. A typical vasovagal spell is preceded by a prodromal sign or symptom. Most occur while standing. Stressful situations, emotional upset, and mild physical trauma can trigger such an event.

Syncope occurring during intense physical activity, may identify those patients with potentially fatal conditions. These patients' symptoms may also suggest vagal tone and/or volume depletion caused by dehydration and heat stress. A detailed evaluation should be considered for patients who have syncope during exercise or have a family history of sudden death, myocardial disease, or arrhythmias. A history of palpitations before syncope should alert the physician to the possibility of tachyarrhythmias. Palpitations are also reported in hyperventilation episodes. History is sought regarding medication use, recent food intake, and intercurrent illnesses to consider additional causes of nonvasovagal syncope. A medical history or family history of cardiac or neurodevelopmental disorders is important to elicit in regard to possible cardiac or neurologic causes of syncope. If there are no suggestive historical features of either vasovagal or the more worrisome causes of syncope, it might be prudent to cautiously consider some psychological assessment questions, particularly in well-appearing adolescents.

On physical examination the heart rate and blood pressure should be measured while the patient is supine and standing. In orthostatic hypotension caused by autonomic dysfunction, dehydration, or blood loss, the patient will have an abnormal decrease in systolic blood pressure (greater than 20 mm Hg) or an abnormal increase in heart rate. Palpation of an abnormal apical impulse or peripheral pulses may suggest a structural heart disease. On auscultation, a loud systolic ejection murmur in the midsternum and upper sternum is present in severe aortic stenosis or IHSS, or the diastolic murmur of a rare left atrial myxoma may be heard. A fourth heart sound may be present in hypertrophic cardiomyopathy. The general physical examination should include a careful neurologic examination, auscultation for cervical and carotid bruits, an assessment of hydration status, and consideration of the presence of any toxidromes.

Routine Laboratory Testing

The emergency physician must assess the cardiac status of all children who present with a history of a syncopal episode. While in the emergency department, the patient should be placed on a continuous cardiac monitor for evaluation of heart rate, rhythm, and conduction intervals unless an obvious noncardiac cause is revealed by the clinical examination. An ECG should be included with all initial evaluations for syncope. The rhythm strip would assess the presence of any arrhythmias (SVT, atrioventricular [AV] block, sick sinus syndrome, or prolonged Q-T interval). Particular attention should be paid to the Q-T interval and T-wave morphology for evidence of long Q-T syndrome (see Chapter 82). Q-Tc must be calculated. Voltage criteria to determine ventricular hypertrophy in evaluating obstructive outflow lesions and evidence of preexcitation in WPW syndrome should be sought. The ECG is also helpful in recognizing ectopy and conduction disturbances.

Serum laboratory tests, if indicated, may include a complete blood count, serum glucose, and carboxyhemoglobin determinations. Toxicology screens should be performed in patients suspected of ingestion or illicit drug use. In teenage girls, pregnancy should be ruled out.

Head-Upright Tilt Table Testing

The utility of the head-upright tilt table test in pediatric patients is controversial for the diagnosis of vasovagal syncope. It has emerged as a laboratory method for provoking episodes of neurally mediated (vasodepressor) syncope in susceptible individuals. The patient is placed on a standard tilt table with a foot board. After 15 to 20 minutes of rest, baseline blood pressure and heart rate measurements are done, after which the patient is positioned at an angle of 90 degrees for up to 30 minutes. A test is considered positive if hypotension and bradycardia occur during the tilt. If no syncope occurs, an isoproterenol infusion is started and the patient is then tilted in an attempt to induce syncope.

Such testing may be indicated in the evaluation of children with recurrent unexplained syncope. However, it appears to be more specific (83 to 100%) and less sensitive (43 to 57%) in young patients. Also, caution must be used in interpreting results with isoproterenol because intravascular access and provocative agents may decrease specificity.

Several nonemergent studies or devices may contribute to the diagnostic evaluation in selected patients. An *echocardiogram* may be a helpful diagnostic test for recognition of hypertrophic cardiomyopathy or obstructive disease. *Holter monitoring* is expensive and rarely diagnostic in children and thus is often not included in an initial workup. *Event recorders,* however, are similar in size and appearance and have replaced Holter monitors in many medical centers for the evaluation of children with syncope. A digitally recorded rhythm strip can be transmitted via a telephone line to a receiving and recording device. Lastly, *electroencephalogram (EEG)* is indicated if a seizure disorder is suspected.

THERAPY

Any therapeutic approach to the child with syncope should be individualized based on several issues, including the likely pathologic process, frequency and nature of symptoms, likelihood of recurrence, and risk of adverse outcomes.

If the diagnosis of vasovagal syncope is made, reassurance and education with regard to the benign prognosis of the process is required. Parental and patient education involves learning to identify and avoid precipitating situations. The patient is instructed to recognize prodromal symptoms and then to assume a seated or supine position and elevate his or

her feet to avoid loss of consciousness. Other simple prophylactic measures include avoidance of dehydration, encouragement of salt enriched diets during periods of intense physical activity, and rarely, the use of mineralocorticoids *(fluorocortisone)* to induce salt and water retention.

The patient can be discharged home with home observation instructions that have been explained to the parents or guardians. Follow-up visits with their primary care physician should be arranged and encouraged.

If these measures are unsuccessful, medical management for severe vasovagal events can be directed at breaking the cycle of events that lead to syncope. Pharmacologic therapy with *b-adrenergic blockers* (atenolol or propanolol), which decrease the mechanical stimulation of the cardiac mechanoreceptors, may be indicated. *Disopyramide* acts as an anticholinergic, negative inotropic, and vasoconstrictive agent.

Transdermal scopolamine has been reported to be effective presumably by reducing the vagal tone associated with these episodes. Recently, *sertraline* has been shown to be successful in decreasing the frequency of syncope. Pacemaker implantation for vasovagal syncope is reserved for the rare refractory case that has failed aggressive pharmacologic measures.

Table 73.6 offers guidelines regarding which patients require referral to pediatric subspecialists, such as a cardiologist or neurologist.

Atypical episodes	Syncope associated with abnormal
Recurrent episodes or episodes not resolved with conventional therapy	cardiac history, examination, or ECG
	Family history of sudden death
Exertional syncope	Seizures
Syncope associated with chest pain, arrhythmias, or palpitations	A focal neurologic examination or neurologic abnormality

ECG, electrocardiogram.

Table 73.6. Indications for Subspecialist Referral or Consultation

Suggested Readings

ACC Expert Consensus Document. Tilt table testing for assessing syncope. J Am Coll Cardiol 1996;28:(1)263–275.

Braden DS, Gaymes CH. The diagnosis and management of syncope in children and adolescents. Pediatr Ann 1997;26:7,422–426.

Breningstall GN. Breath-holding spells. Pediatr Neurol 1996;14:2,91–97.

DiMario FJ, Burleson JA. Clinical and laboratory observations. Behavior profile of children with severe breath-holding spells. J Pediatr 1993;122:488–491.

Feit LR. Syncope in the pediatric patient: diagnosis, pathophysiology and treatment. Adv Pediatr 1997;43:469–494.

Hannon DW, Knilans TK. Syncope in children and adolescents. Curr Probl Pediatr 1993;23:358–384.

Oslizlok PC, Allen M, Gillette PC, et al. Recurrent unexplained syncope in young patients: investigation, treatment and outcome. J Am Coll Cardiol 1991;17:333A.

Pratt JL, Fleisher GR. Syncope in children and adolescents. Pediatr Emerg Care 1989;5:80–82.

Splawski I, Timothy KW, Vincent GM, Atkinson DL, Keating MT. Brief reports: molecular basis of the long QT syndrome associated with deafness. N Engl J Med 1997;336(22):1562–1567.

Tanel RE, Walsh EP. Syncope in the pediatric patient. Cardiol Clin 1997;15:2,277–294.

Wiley JF, Gelber ML, Henretig FM, et al. Cardiotoxic effects of astemizole overdose in children. J Pediatr 1992;120:799–802.

CHAPTER 74

Tachycardia/Palpitations

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Palpitations represent a disagreeable perception of the heart beat by the patient. Descriptions commonly given include “pounding,” “fluttering,” “jumping in the chest,” or a sensation of the heart “stopping.” A high degree of variation in the sensitivity of patients to changes in the heart rate (HR) or rhythm exists. Severe symptoms may be expressed by a patient who actually experiences trivial cardiac events. However, the absence of palpitations by history does not rule out the possibility of a life-threatening arrhythmia. The challenge to the emergency physician is to determine which complaint can be managed in the emergency department (ED) and which merits further consideration by a cardiologist.

PATHOPHYSIOLOGY

The heart is innervated primarily by the vagus nerve (cranial nerve X) and the sympathetic ganglion. Cardiovascular reflexes (e.g., vasovagal bradycardia) are transmitted by the vagus nerve. Pain sensation (e.g., related to myocardial ischemia) travels through afferent fibers associated with the sympathetic ganglia. In most patients, the sensation of the heartbeat is not felt. Children with documented arrhythmias, such as supraventricular tachycardia (SVT) and stable ventricular tachycardia, may not complain of any symptoms. Even patients with heart murmurs audible to the unassisted ear can learn to ignore this obvious cue.

Patients with palpitations often relate an indirect perception of increased force of cardiac contraction, tachycardia, or irregular heartbeat. Increased force of the contraction often is detected when the patient is supine. At times, it may be described as a rushing or pounding in the ears, particularly when the ear is pressed against a pillow. This sensation of a large ventricular stroke volume can be produced by caffeine or alcohol consumption, exercise, and emotional arousal. Tachycardia may be detected even in a preverbal child as parents incidentally note a rapid HR when holding their child, or they observe rapid jugular venous pulsations. In this way, asymptomatic SVT may come to medical attention. Noncardiac causes of tachycardia include exercise, fever, anemia, hyperthyroidism, and rarely, pheochromocytoma. Patients with premature contractions and a compensatory pause may describe the feeling that their hearts “flip-flop” or “stop.” Many patients with premature atrial or ventricular contractions notice the subsequent beat after the initial “short” beat because of the increased stroke volume ejected. Other patients may complain of a choking or full sensation in the neck. Jugular venous pulsation associated with right atrial contraction against a closed tricuspid valve (atrioventricular block with or without atrial tachycardia) can present in this way.

True cardiac arrhythmias arise from various mechanisms that are discussed in Chapter 82.

DIFFERENTIAL DIAGNOSIS

Many conditions may produce palpitations (Table 74.1). Most patients with palpitations do not have significant cardiac pathology (Table 74.2), with the exception of patients with SVT and mitral valve prolapse. Many life-threatening conditions can come to medical attention because of abnormal cardiac sensation (Table 74.3). Wolff-Parkinson-White (WPW) syndrome and the prolonged Q-T syndrome are two potentially lethal diseases that may be diagnosed on a resting electrocardiogram (ECG). A patient with palpitations during exercise should also raise the concern of possible hypertrophic cardiomyopathy, SVT, ventricular tachycardia, or myocardial ischemia. Diagnosis of noncardiac causes of life-threatening palpitations, including hypoxemia, hypoglycemia, hyperkalemia, and hypocalcemia, can be made by characteristic ECG changes, serum electrolyte determinations, rapid bedside glucose, and oxygen saturation measurements.

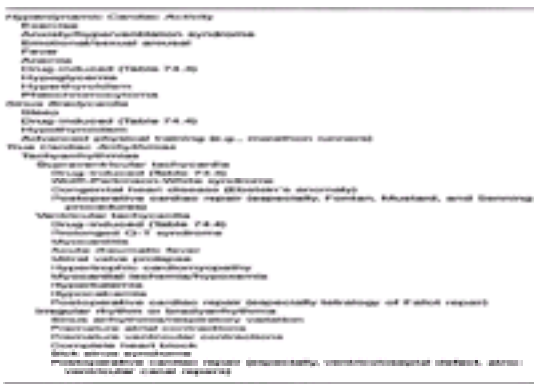

Table 74.1. Differential Diagnosis of Palpitations

Exercise	Supraventricular tachycardia
Anxiety/hyperventilation syndrome	Mitral valve prolapse
Emotional arousal	Premature atrial or ventricular con-
Drug-induced	tractions

Table 74.2. Common Causes of Palpitations

Cardiac	*Noncardiac*
Wolff-Parkinson-White syndrome	Hypoxemia
Prolonged Q-T syndrome	Hypoglycemia
Hypertrophic cardiomyopathy	Hyperkalemia
Congenital heart disease/postoperative cardiac repair	Hypocalcemia
	Pheochromocytoma
Myocarditis/acute rheumatic fever	Drug-induced
Mitral valve prolapse	
Sick sinus syndrome	
Complete heart block	
Myocardial ischemia	

Table 74.3. Life-Threatening Causes of Palpitations

Hyperdynamic Cardiac Activity

Increased HR and contractility are physiologic responses to catecholamine release, like that which may occur with exercise, emotional arousal, hypoglycemia, and pheochromocytoma. Similarly, increased cardiac work accompanies conditions that increase the basal metabolic rate such as fever, anemia, and hyperthyroidism. Sympathomimetic and anticholinergic drugs are among a group of commonly available substances that directly modulate the autonomic nervous system, causing tachycardia, hyperdynamic cardiac activity, and palpitations (Table 74.4).

Sinus or Supraventricular Tachycardia	Nonsedating antihistamines (astemizole, terfenadine)
Ephedrine, pseudoephedrine	Organophosphate pesticides
Amphetamines	Chlorinated hydrocarbons
Cocaine	Digoxin
Albuterol, metaproterenol	Caffeine, theophylline
Antihistamines	Amphetamines
Phenothiazines	Cocaine
Antidepressants	Arsenic
Tobacco	*Bradycardia*
Caffeine, theophylline	β-Adrenergic blockers
Ventricular Tachycardia or Torsades de Pointes	Calcium channel blockers
	Digoxin
Tricyclic antidepressants	Clonidine
Phenothiazines	Sedative/hypnotic agents
Antiarrhythmic agents (e.g., quinidine, procainamide, mexiletine, flecanide, encainide)	Narcotics
	Organophosphate pesticides
Chloral hydrate	

Table 74.4. Drugs That Cause Palpitations/Arrhythmias

Sinus Bradycardia

Low basal metabolic rate associated with hypothyroidism may present with a slow HR and sinus rhythm. Similarly, in the absence of significant sympathetic nervous system input, the HR may slow. This state may be responsible for the sinus bradycardia associated with sleep or with ingestion of drugs such as clonidine, sedative-hypnotics, or narcotics. Advanced physical training results in a highly efficient heart with high ventricular ejection fraction and sinus bradycardia.

True Cardiac Arrhythmias

SVT represents the most common tachyarrhythmia of childhood (see Chapter 82). Possible underlying causes include drug exposure, congenital heart disease, and WPW syndrome. Sympathomimetics in cough and cold preparations are the most common drugs to incite SVT in children. Cardiac lesions associated with SVT include Ebstein's anomaly, repaired dextrotransposition of the great arteries, and single ventricle lesions status post-Fontan operation. Up to 75% of patients with WPW syndrome have a shortened P-R interval or delta wave on resting ECG (see Chapter 82). However, approximately 50% of children with SVT have no physical findings and no ECG abnormalities between episodes. In these patients, descriptions of abrupt onset and rapid termination of palpitations often can be elicited.

Infection, including viral myocarditis and acute rheumatic fever, constitutes one of the most common causes of ventricular tachycardia in children with normal cardiac anatomy. Similarly, ingestion of drugs with quinidinelike effects, such as tricyclic antidepressants, phenothiazines, and antiarrhythmic agents, is a preventable cause of torsades de pointes (polymorphic ventricular tachycardia) and unstable ventricular tachycardia in the otherwise normal child (Table 74.4). Syncope or palpitations associated with exercise may be caused by ventricular tachyarrhythmias that occur in conjunction with hypertrophic cardiomyopathy or myocardial ischemia (usually secondary to congenital anomalies of the coronary arteries). Patients with the prolonged Q-T syndrome have a genetically determined predisposition to fatal ventricular arrhythmias that can be detected by calculation of the corrected Q-T interval on a resting 12-lead ECG (see Chapter 82). Patients who have undergone ventriculotomy for tetralogy of Fallot comprise another group who are at high risk for ventricular arrhythmias as a result of the postoperative development of scarring in the right ventricular outflow tract. Finally, electrolyte disturbances, particularly hyperkalemia, hypocalcemia, and hypomagnesemia, may be causative in a child with palpitations and ventricular tachycardia (see Chapter 86).

Premature atrial contractions produce the most common arrhythmia of childhood, with 50% of normal children experiencing at least one premature atrial contraction per day. Premature ventricular contractions (PVCs) also account for many reports of irregular heartbeat. Although this arrhythmia can herald serious underlying pathology, patients with an unremarkable history, normal physical examination, and unifocal PVCs that disappear with exercise do not require further evaluation. Patients with significant sinus or atrioventricular (AV) node dysfunction as a cause of an irregular or slow heartbeat often have a history of syncope or seizure, slow HR (25 to 50 beats per minute) on examination, a pulmonic flow murmur, or signs of congestive heart failure (CHF). Patients who have undergone intra-atrial repairs (D-transposition of the great arteries and atrial septal defect) are at highest risk for these potentially life-threatening arrhythmias.

EVALUATION AND DECISION

The ill-appearing child with palpitations requires rapid assessment for the presence of hypoxemia, shock, hypoglycemia, or an existing life-threatening arrhythmia. Further evaluation should include measurement of hemoglobin, serum glucose (Dextrostick), serum electrolytes, calcium, and pulse oximetry or arterial blood gas. The presence of heart disease should be assessed by a 12-lead ECG and rhythm strip, followed by continuous monitoring, frequent vital signs, and chest radiographs (Fig. 74.1). Specific arrhythmias should be treated as outlined in Chapter 82.

FIGURE 74.1. A diagnostic approach to palpitations. *ECG,* electrocardiogram; *SVT,* supraventricular tachycardia; *WPW,* Wolff-Parkinson-White; *CHF,* congestive heart failure; *Hgb,* hemoglobin.

The asymptomatic child with palpitations by history also may have an intermittent or continuing arrhythmia. Continuous cardiac monitoring and a resting 12-lead ECG performed while the patient is in the ED increases the likelihood that this abnormality will be detected. Some patients in this category may benefit from Holter or event monitoring, particularly if there is a history of palpitations. Any patient with a history of syncope, congenital heart disease, or particularly, postoperative or exercise-induced palpitations is at greater risk for having a true cardiac arrhythmia as the cause of his or her symptoms. Similarly, the presence of a short P-R interval with the typical delta wave morphology of WPW syndrome or a prolonged corrected Q-T interval (see Chapter 82) indicates the need for further evaluation and consultation by a pediatric cardiologist.

The presence of fever or an upper respiratory infection should prompt the emergency physician to look for signs and

symptoms of myocarditis or acute rheumatic fever. Myocarditis describes inflammation of the muscle wall of the heart. Multiple organisms can cause this pathology, with the most common identified agent being coxsackievirus. Clinical features of this disease are fever, tachycardia out of proportion to activity or degree of fever, pallor, cyanosis, respiratory distress secondary to pulmonary edema, muffled heart sounds with gallop, and hepatomegaly caused by passive congestion of the liver. The ECG findings are nonspecific and include low voltage QRS complexes (less than 5 mm total amplitude in limb leads), “pseudoinfarction” pattern with deep Q waves and poor R wave progression in the precordial leads, AV conduction disturbances that range from P-R prolongation to complete AV dissociation, and tachyarrhythmias such as ventricular tachycardia and SVT. A child with palpitations and clinical findings suggestive of myocarditis requires emergent supportive care (see Chapter 3), echocardiography, and admission to a unit capable of intensive monitoring and rapid treatment of cardiac arrhythmias.

Acute rheumatic fever follows pharyngeal streptococcal infection and is an inflammatory disease that targets the heart, vessels, joints, skin, and central nervous system (CNS). Diagnosis and management of acute rheumatic fever are discussed in Chapter 82.

A detailed history of recent medications or precipitating events may reveal the cause of palpitations in some patients. Ingestion of caffeinated beverages (including soft drinks), cough and cold preparations, and illicit drugs, as well as a smoking history should be ascertained. The patient's emotional state before the onset of palpitations should be discussed to determine the likelihood of anxiety or emotional arousal as the cause of symptoms (see Hyperventilation Syndrome, Chapter 131). The presence of diaphoresis, hypertension, and headache should encourage the consideration of pheochromocytoma, whereas widened pulse pressure and thyroid enlargement suggest hyperthyroidism (see Chapter 97). Anemia may be the cause of symptoms in a patient with pallor (see Chapter 59).

In some patients, an exact cause of palpitations cannot be determined at the time of ED evaluation. Patients with a single episode should have close follow-up arranged with their primary care physicians and should be instructed to return for further evaluation if symptoms recur. Patients with multiple episodes of palpitations deserve further evaluation and consultation with a pediatric cardiologist.

Suggested Readings

Benson DW Jr, Benditt DG, Anderson RW, et al. Cardiac arrest in young, ostensibly healthy patients: clinical, hemodynamic, and electrophysiologic findings. Am J Cardiol 1983;52:65–69.

Corrado D, Thiene G, Nava A, Rossi L, Pennelli N. Sudden death in young competitive athletes: clinicopathologic correlations in 22 cases. Am J Med 1990;89:588–596.

Drucker NA, Newburger JW. Viral myocarditis: diagnosis and management. Adv Pediatr 1997;44:141–171.

Jackman WM, Clark M, Friday KJ, Aliot EM, Anderson J, Lazzara R. Ventricular tachyarrhythmias in the long QT syndromes. Med Clin North Am 1984;68:1079–1109.

Jacobsen JR, Garson A Jr, Gillette PC, McNamara DG. Premature ventricular contractions in normal children. J Pediatr 1978;92:36–38.

Kugler JD, Danford DA. Management of infants, children, and adolescents with paroxysmal supraventricular tachycardia. J Pediatr 1996;129:324–328.

Maron BJ, Roberts WC, McAllister HA, Rosing DR, Epstein SE. Sudden death in young athletes. Circulation 1980;62:218–229.

Monarrez CN, Strong WB, Rees AH. Exercise electrocardiography in the evaluation of cardiac dysrhythmias in children. Paediatrician 1978; 7:116–125.

Nagashima M, Matushima M, Ogawa A, et al. Cardiac arrhythmias in healthy children revealed by 24-hour ambulatory ECG monitoring. Pediatr Cardiol 1987;8:103–108.

Olson KR, Pentel PR, Kelley MT. Physical assessment and differential diagnosis of the poisoned patient. Med Toxicol 1987;2:52–81.

Vetter VL. What every pediatrician needs to know about arrhythmias in children who have had cardiac surgery. Pediatr Ann 1991;20: 378–385.

CHAPTER 75

Urinary Frequency in Childhood

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Urinary frequency is a symptom of several commonly encountered, clinical pediatric problems such as urinary tract infection (UTI), urethritis, vulvovaginitis, diabetes mellitus (DM), drug side effect (with caffeine, theophylline, and diuretics), or psychogenic stress. Moreover, urinary frequency may suggest underlying disease processes with life-threatening potential, such as diabetic ketoacidosis, diabetes insipidus, or congenital adrenal hyperplasia, that require emergent diagnosis and management. Therefore, an organized approach in the emergency department (ED) evaluation of this symptom is important for any clinician who provides acute care to children.

Urinary frequency (pollakiuria) is defined as an increase in the number of voids per day. It is a symptom distinct from polyuria (excretion of excessive amounts of urine). Although the two symptoms can be related, most children who present to the ED with frequency have a normal daily urine output, although the individual voids are frequent and small. Frequency also is distinct from enuresis, which is defined as inappropriate urination at an age when bladder control should be achieved.

PATHOPHYSIOLOGY

More than 90% of newborns void during the first day of life. Infants void between 6 to 30 times each day. Over the next 2 years, the number of voidings per day decreases by about half, whereas the volume of urine produced increases fourfold. Children between the ages of 3 and 5 years average 8 to 14 voids per day. By 5 years of age, the number of voids decreases to 6 to 12 times per day. Adolescents average 4 to 6 voids per day. In the school-aged population, urinary frequency usually is defined as voiding more often than every 2 hours.

Normal bladder mucosa is pressure-sensitive and pain-sensitive. An uncomfortable sensation is produced when urine volume approaches the age-dependent capacity of the bladder. Voiding is initiated by relaxation of the striated muscle of the urinary sphincter and an associated contraction of the smooth muscle of the bladder, resulting in bladder emptying. This mechanism is mediated by sacral nerves II–IV. Uncontrolled, “uninhibited” bladder contractions are the normal mechanism for infant and toddler voiding. Uninhibited (parasympathetic-mediated) bladder contractions do not normally occur after toilet training. By 5 years of age, 90% of children have achieved direct voluntary mastery of the voiding reflex and exhibit the adult pattern of urinary control.

Urinary frequency may be caused by reduced bladder capacity, polyuria, or psychologic stress. The urinary volume per voiding will be low if frequency is related to reduced bladder capacity or psychological stress. Moreover, there will not be associated polydipsia. If frequency is secondary to polyuria, the urine volume per voiding will be normal or high, and polydipsia is usually associated (see Chapter 60).

A reduced bladder capacity may be secondary to inflammation of the bladder, changes in the bladder wall induced by distal obstruction, or extrinsic masses pressing on the bladder. When the bladder is inflamed, its pain/pressure sensitivity threshold is significantly decreased, so less stimuli is necessary to initiate the urge to void.

Distal infravesical obstruction leads to bladder muscle hypertrophy because of the increased effort needed to empty the bladder. This hypertrophied muscle has a higher resting tone so that smaller than normal urine volumes are necessary to initiate the desire to void. A decrease in the size and force of the urinary stream and/or straining to urinate may be noted. Eventually, the bladder muscle fatigues and is unable to empty the bladder effectively. This decompensated bladder has an increased residual urine volume with a resultant decrease in the functional bladder capacity. This large, hypotonic bladder contracts poorly, resulting in small, frequent voids.

Extrinsic extravesical masses that impinge on the bladder may cause frequency by interfering mechanically with normal bladder expansion. Extrinsic masses may also stimulate frequent voiding by causing an irritable focus in the bladder wall.

Normal pediatric values for urine output are useful in determining the presence of polyuria. The traditional definition of polyuria is a urinary output greater than 900 mL/m^2 per day. An infant/toddler up to 2 years of age rarely exceeds 500 mL/day. Children 3 to 5 years of age void up to 700 mL/day. Children 5 to 8 years of age have an approximate maximum volume of 1000 mL/day. Children 8 to 14 years of age void up to 1400 mL/day. When polyuria is the cause of urinary frequency, the urine volume per void generally is greater than 2.0 mL/kg.

Polyuria with dilute urine is classically associated with a decreased production of antidiuretic hormone or with impaired

renal responsiveness to circulating antidiuretic hormone. Polyuria with dilute urine also can be seen when the stimulus for antidiuretic hormone release is absent (e.g., chronic water overloading). In all these situations, the specific gravity of the urine seldom is greater than 1.005, and urine osmolality rarely exceeds 200 mOsm/kg. This contrasts with a normal urinary concentrating ability, which is confirmed by a specific gravity of greater than 1.020.

Polyuria with isotonic or slightly hypertonic urine occurs with an osmotic or solute diuresis. Unlike a water diuresis, both urine flow rate and solute excretion are increased. The urine osmolality is never lower than 300 mOsm/kg. However, the urine specific gravity is variable, ranging from 1.010 when the solute is primarily electrolytes and urea (e.g., renal failure, administration of diuretics) to as high as 1.045 when the solute mass is large (e.g., diabetes mellitus, intravenous contrast).

Psychogenic/emotional stress also may induce urinary frequency. Cystometric studies have documented significant anxiety-related increases in intravesical pressure, usually accompanied by a desire to void.

DIFFERENTIAL DIAGNOSIS

A differential diagnosis of urinary frequency is outlined in Table 75.1. In-depth discussions of many of these subjects can be found in other chapters of this textbook (in particular, see Chapter 54, Chapter 60, Chapter 86, Chapter 97, and Chapter 122). The paragraphs that follow highlight selected topics in the differential diagnosis.

Table 75.1. Differential Diagnosis of Urinary Frequency

Frequency often is associated with UTIs, so this diagnosis always must receive significant consideration in the differential. The importance of accurate diagnosis of a pediatric UTI has assumed even greater significance with current urologic recommendations for radiologic evaluation after the first documented infection in preschool-age children of both sexes.

The term *urethral syndrome* refers to an entity that can be seen in adolescent girls, characterized by acute onset of frequency and dysuria with “insignificant” bacterial counts (less than 10^5/mL). Pyuria is generally, but not absolutely, present. Vaginitis is a common cause of the urethral syndrome. *Chlamydia trachomatis* is also a relatively common etiology. The urethral syndrome also can be associated occasionally with *Neisseria gonorrhoeae*. Evidence supports the causal relationship of low-level bacteriuria and symptomatic disease. Therefore, in the context of the urethral syndrome, after all other causes have been excluded, “significant” bacteriuria may be considered as greater than or equal to 10^2 Enterobacteriaceae/mL.

Irritative vulvovaginitis (e.g., secondary to poor hygiene or bubble baths) is a relatively common cause of frequency, usually associated with dysuria but not with pyuria. Frequency may be secondary to urethral trauma secondary to straddle injuries, catheterization, masturbation, or sexual abuse. Pinworms *(Enterobius vermicularis)* occasionally may cause frequency in young girls. Children with pinworm infestation may or may not present with perineal itching. Pyuria and dysuria usually are absent.

Frequency may be a presenting symptom of a pelvic appendicitis or appendiceal abscess. Potential for significant morbidity is obvious. Associated abdominal pain, by history and examination, should be present. Rectal examination may be abnormal (differential tenderness, mass). Pyuria, microscopic hematuria, and proteinuria (but generally not bacteriuria) may also be present.

Frequency may be secondary to a partial distal urethral obstruction. The urinary stream in the male infant or child with posterior urethral valves usually is nonforceful and nonsustained. Straining to urinate also may be noted. A lower abdominal mass (enlarged bladder) may be palpable.

A neurogenic bladder associated with a spinal cord lesion (e.g., tethered cord) may present with urinary frequency. There may be associated lumbosacral abnormalities (hairy patches, cutaneous dimples or tracts, lipoma, or bony irregularities). Decreased anal tone and lower extremity weakness or reflex abnormalities may be noted. An enlarged bladder may be palpable.

It is well recognized that children with urinary tract dysfunction often have associated constipation. Large fecal accumulations may restrict maximal bladder capacity or directly produce symptoms of frequency by stimulating uninhibited bladder contractions. Resolution of the fecal accumulation decreases frequency symptoms.

Pregnancy should always be considered as a cause of frequent urination in the adolescent girl. A lower abdominal mass may be palpable. Adolescent sexual histories are notoriously unreliable.

Uninhibited bladder contractions (“unstable bladder” syndrome) occur involuntarily in children who have failed to gain complete voluntary control over the voiding reflex. This appears to represent a delay in nervous system maturation. A child who attempts to maintain continence must constrict the voluntary urinary sphincter tightly. If the sphincter is relatively weak, urinary frequency associated with urgency and enuresis may result. Girls may exhibit the so-called curtsey sign, so named because the child squats and attempts to prevent leakage by compressing the perineum with the heel of one foot. This maneuver will usually avoid major incontinence, but generally, small amounts of urine leakage occur. If performed, a screening ultrasound examination would reveal normal (minimal) residual urine volumes. With maturity, spontaneous resolution of uninhibited contractions occurs in most cases. In children with significant mental retardation or behavioral disorders, the infantile pattern of spontaneous bladder contraction may persist. Unstable bladder syndrome may also develop in otherwise normal children who have undergone normal toilet training. If symptoms are persistent, a trial of oxybutynin after urologic consultation may be warranted.

Anatomic anomalies of the urogenital tract may result in chronic leakage of urine. Ectopic ureter would an example of such an anatomic defect.

Uncontrolled DM is a potentially life-threatening condition that can present with frequent urination. Polyuria results from a glucose-induced osmotic diuresis. At initial presentation, polydipsia, polyphagia, Kussmaul's respirations, and weight loss also may be present.

In chronic renal failure and in certain diseases of the renal parenchyma (e.g., renal tubular acidosis, Fanconi's syndrome, and Bartter's syndrome), the renal tubules lose their ability to concentrate urine. This leads to polyuria and frequency, with large volumes of relatively dilute urine. A concentration defect also may occur with sickle cell disease or trait and may be evident as early as 6 months of age.

Hypercalciuria has been reported as a significant noninfectious cause of the “frequency-dysuria syndrome” in the pediatric patient, although Brock has recently challenged this association. Onset of symptoms may present throughout the pediatric age range, generally 2 to 14 years of age. Occasionally, hypercalciuria can present in early infancy, in which irritability is a hallmark symptom. Symptoms often spontaneously resolve within 2 months. There may be a positive family history of calcium urolithiasis. When symptomatic, frequency is almost always associated with dysuria. Hematuria (generally microscopic) and crystalluria are often seen. However, the urinalysis may be normal. If the diagnosis is suspected and symptoms persist, studies of urinary calcium excretion and urologic consultation should be considered.

The salt-losing form of congenital adrenal hyperplasia is a life-threatening, although relatively rare, cause of frequency. Excessive urinary excretion of sodium leads to severe water loss and marked dehydration with associated hyperkalemia and hyponatremia. At the initial presentation in early infancy, however, urinary frequency as a symptom generally is not appreciated. Infant girls may exhibit virilization of the external genitalia. Infant boys may demonstrate increased pigmentation of the external genitalia and/or a relatively enlarged phallus.

Diabetes insipidus (DI) is an uncommon, although life-threatening, cause of frequency seen in the ED. It is clinically characterized by polyuria (with resultant frequency) and polydipsia. It is caused by an inability of the kidneys to concentrate urine. This is related to a deficiency in the hypothalamic production of antidiuretic hormone (central DI) or a renal unresponsiveness to antidiuretic hormone (nephrogenic DI). Some causes of central DI (e.g., septo-optic dysplasia) present in the neonatal period. However, most causes of central DI are acquired (e.g., head injury, brain tumors) and, therefore, can present at any age. The most common type of nephrogenic DI in childhood is the X-linked recessive type, which presents in boys during early infancy. If fluids are not accessible or if the thirst sensation is impaired, hypernatremic dehydration develops. If DI is suspected, oral fluids should not be restricted. The child should be admitted to the hospital for evaluation and treatment under strict medical supervision.

Drugs are a relatively common cause of frequency in childhood. Methylxanthines (theophylline, caffeine) and ethanol inhibit production of antidiuretic hormone. Lithium, chronic hypokalemia, hypercalcemia, and vitamin D are also associated with urinary frequency, interfering with renal responsiveness to antidiuretic hormone. Diuretic agents may cause urinary frequency. These agents represent only a few of the many drugs that can cause urinary frequency as a side effect. Therefore, a detailed pharmacology history should be obtained in the child with urinary frequency.

Frequency may occur secondary to the polyuria of water intoxication. Absence of nocturia and enuresis in the presence of polyuria suggests an excessive fluid intake. The serum sodium and osmolality generally are decreased. Psychogenic water drinking is an extremely unusual diagnosis in young children but may present in adolescents. Water intoxication secondary to Munchausen syndrome by proxy would be an unusual presentation of child abuse.

The “extraordinary urinary frequency syndrome” is a relatively recently described entity but probably represents a common cause of urinary frequency in pediatric primary care settings. Average age of onset is about 6 years (with a range of about 2 to 11 years). Daytime frequency occurs as often as every 5 minutes. Dysuria is not present. Nocturia is present in about half the cases but usually occurs only about 1 to 2 times per night. Polydipsia and polyuria are absent. The physical examination is normal. The urinalysis and other laboratory studies also are normal. If the diagnosis of “extraordinary urinary frequency syndrome” is likely, reassurance and follow-up are indicated. Initial radiologic evaluation and pharmacologic therapy are generally unnecessary. Left untreated, frequent voiding resolves spontaneously, usually in about 2 months. The cause is unclear but often has a psychogenic component, with an apparent “trigger” (e.g., school problems, parental death, sibling illness) identifiable in about 40% of cases. More extensive urologic and psychologic evaluation is warranted if isolated urinary frequency persists for more than 6 months.

As an isolated symptom, frequency would be an atypical presentation of pediatric sexual abuse. However, urinary

frequency may be seen in association with pertinent history or physical findings (e.g., vulvovaginal venereal infection or genital trauma), which would be suggestive of sexual abuse.

EVALUATION AND DECISION

The primary role of the emergency physician in evaluating the child with urinary frequency is to exclude significant underlying pathology that may result in morbidity and to identify treatable conditions. When confronted with a child whose chief complaint is frequent urination, the physician should initially determine whether the criteria for true urinary frequency have been met (refer to the section on pathophysiology for age-related normal values). Additional history should then focus on symptoms related to infection of the urinary tract. Are associated symptoms of dysuria, fever, or flank pain also present? Is there a history of prior UTIs? Questions specifically related to DM also should be included (polyuria, polydipsia, polyphagia, weight loss, family history). The presence or absence of nocturia and enuresis are also important historical points. The urine volume per voiding should be determined (large versus small). Generally, the presence of polyuria (copious volumes of dilute urine) is obvious from the history. The onset and duration of the symptoms and the quality of the urinary stream should be documented.

In addition, other historical features may be pertinent. For example, are there symptoms to suggest central DI (polydipsia, nocturia, intracranial pathology)? Is there a history of poor growth, suggesting renal disease? Is there a family history of sickle cell disease or trait? Is the child taking any medications or drugs (including caffeinated beverages) associated with frequency? Is there a history of chronic constipation, vulvovaginal infection/trauma, or pruritus ani? Are there symptoms of abdominal pain, suggesting the possibility of acute appendicitis or appendiceal abscess? In the young boy, what is the quality of the urinary stream? In an adolescent girl, when was her last menstrual period? Is there a family history of urolithiasis or renal disease?

A complete physical examination should be performed, including an accurate blood pressure measurement. The child's growth parameters should be plotted and the blood pressure should be compared with age-specific normal values to screen for hypertension (see Chapter 35). The abdomen should be palpated carefully for the presence of abdominal masses and/or tenderness. Percussion of the flanks should be performed. The lumbosacral area should be examined closely for anomalies (hairy patches, dimples, tracts). Special attention should be focused on the function of sacral nerves II–IV (anal wink and sphincter tone). Unless the diagnosis is readily apparent, a rectal examination should be performed noting tone, tenderness, masses, and the quality and quantity of stool in the rectal vault. The external genitalia should always be thoroughly examined, meticulously searching for signs of infection, trauma, or anatomic abnormalities. Signs of virilization (in the girl) or hyperpigmentation (in the boy) should be evaluated. A thorough neurologic examination with careful attention to the retinal fundi and visual fields is warranted.

The laboratory evaluation is fairly straightforward. A urinalysis (including specific gravity) and urine culture should be performed in all cases. Caution should be exercised in interpreting pyuria and/or bacteriuria from “bag” or “midstream” urine specimens in the infant or toddler. A confirmatory catheterized specimen should be considered in these cases. If a UTI is confirmed, additional elective radiologic evaluation is recommended for all boys and preschool-age girls. Glycosuria obviously suggests the diagnosis of DM.

If the diagnosis is not apparent at this point, serum chemistries (including electrolytes, glucose, blood urea nitrogen, creatinine, and calcium) should be obtained. A sickle cell preparation also should be considered in the African-American child, and a pregnancy test should be done in the adolescent girl. This workup generally is sufficient for those children who do not have both daytime and nighttime symptoms, or anatomic and/or neurologic abnormalities.

In the child with progressive or worrisome symptoms or signs (e.g., nocturia, persistent dysuria, poor urinary stream, straining to urinate, growth failure, hypertension, fixed low urinary specific gravity, anatomic or neurologic abnormalities), urologic and/or nephrologic consultation is recommended. Additional studies may include a screening ultrasonogram of the urinary tract and abdomen, a voiding cystourethrogram, urinary calcium studies, and possible urodynamic investigation. If the presence of polyuria is in doubt, a 24-hour urine collection may be necessary to establish the diagnosis.

A simplified schematic approach to the evaluation of the child with urinary frequency is outlined in Figure 75.1. This schema provides general guidelines, but it is not all inclusive, and the diagnostic categories are not absolute.

FIGURE 75.1. Evaluation of urinary frequency.

Suggested Readings

Alon U, Warady BA, Hellerstein S. Hypercalciuria in the frequency-dysuria syndrome of childhood. J Pediatr 1990;116:103–105.

Asnes RS, Mones RL. Pollakiuria. Pediatrics 1973;52:615–617.

Bass LE. Pollakiuria, extraordinary daytime urinary frequency: experience in a pediatric practice. Pediatrics 1991;87:735–737.

Brock JW. The frequency and frequency dysuria syndromes of childhood: hypercalciuria as a possible etiology. Urology 1994;44:411–412.

Fernandes E, Vernier R, Gonzalez R. The unstable bladder in children. J Pediatrics 1991;118:831–837.

Fivush B. Irritability and dysuria in infants with idiopathic hypercalciuria. Pediatr Nephrol 1990;4:262–263.

Koff SA, Byard MA. The daytime urinary frequency syndrome of childhood. J Urol 1988;140:1280–1281.

Leung AKC, Robson WLM, Halperin ML. Polyuria in children. Clin Pediatr 1991;30:634–640.

Loening-Baucke V. Urinary incontinence and urinary tract infection and their resolution with treatment of chronic constipation of childhood. Pediatrics 1997;100:228–232.

Robson WLM, Leung AKC. Extraordinary urinary frequency syndrome. Urology 1993;42:321–324.

Watemberg N, Shalev H. Daytime urinary frequency in children. Clin Pediatr 1994;33:50–53.

Zoubek, J, Bloom DA, Sedman AB. Extraordinary urinary frequency. Pediatrics 1990;85:1112–1114.

CHAPTER 76

Vaginal Bleeding

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Vaginal bleeding can be either a normal event or a sign of disease and, when pathologic, can indicate variously a local genital tract disorder, endocrinologic or hematologic disease, or a complication of pregnancy. During childhood, vaginal bleeding is abnormal after the first week or so of life and before menarche. After menarche, abnormal vaginal bleeding must be differentiated from menstruation, and in turn, menstrual bleeding must be categorized as either normal or excessive. *Menstruation* is defined as the spontaneous, periodic shedding of endometrial tissue and blood.

Menstrual patterns during the first 2 years after menarche vary widely, but it is possible to set outside limits. Ninety-five percent of young adolescents' menstrual periods are between 2 and 8 days long. A duration of 10 days or more is abnormal. An occasional interval of less than 21 days from the first day of one menstrual period to the first day of the next is normal for teenagers, but several short cycles in a row are abnormal. Whether the quantity of a patient's menstrual bleeding is normal can be difficult to determine historically. However, it is uncommon for adolescents to soak more than 6 to 8 perineal pads or tampons a day. Normal menstrual bleeding *never* produces an acute fall in hemoglobin or hematocrit.

Because the relative prevalence of disorders that produce vaginal bleeding correlates more strongly with patients' hormonal status than with their chronologic age, the diagnostic approach outlined in this chapter is presented in two sections divided according to patients' menarchal status (Table 76.1).

I. At Any Time	III. After Menarche
A. Trauma	A. Hormonal contraception
B. Tumor	B. Endometritis
II. Before Normal Menarche	C. Dysfunctional uterine bleeding
A. Hormonal	D. Bleeding diathesis
1. Neonatal bleeding	E. Ectopic pregnancy
2. Exogenous estrogen	F. Spontaneous abortion
3. Precocious puberty	G. Placenta previa
B. Nonhormonal	H. Abruptio placentae
1. Urethral prolapse	
2. Genital warts	
3. Lichen sclerosus	
4. Infectious vaginitis	
5. Foreign body	

Table 76.1. Differential Diagnosis of Vaginal Bleeding

VAGINAL BLEEDING DURING CHILDHOOD

Evaluation and Decision

During the patient's general physical examination, the emergency physician should be particularly alert for signs of hormonal stimulation—breast development, pubic hair growth, a dull pink vaginal mucosa, or physiologic leukorrhea. For the initial examination of the genitalia, an infant or child should be placed in a frog-leg position either on the parent's lap or on the examining table (Fig. 94.2A). The physician then gently separates the child's labia majora and inspects the introitus for a bleeding site. A vaginal speculum should not be used. If the vulva is normal, the child should next be placed in the knee–chest position for examination of her vagina (Fig. 94.2B). In this position, the girl is encouraged to relax her abdominal muscles while the examiner gently separates her labia and buttocks. As air enters the vaginal vault, it falls open, allowing the physician to look for a foreign body, using an otoscope without a speculum as a light source. If

no foreign body is seen, the child is returned to the supine position, and a vaginal specimen for culture is obtained, using either a soft plastic medicine dropper or a cotton-tipped swab moistened with nonbacteriostatic saline solution. Finally, if the interior of the vagina could not be seen well but the examiner suspects a firm foreign body or trauma, a rectal examination should be done to palpate the vagina indirectly and to check for lacerations.

VULVAR BLEEDING

The vulva consists of several structures: the labia majora, the labia minora, the clitoris, and the vaginal introitus. A premenarchal girl with the complaint of vaginal bleeding whose vulva looks abnormal may have a vaginal disorder, a vulvar disorder, or both (Fig. 76.1A).

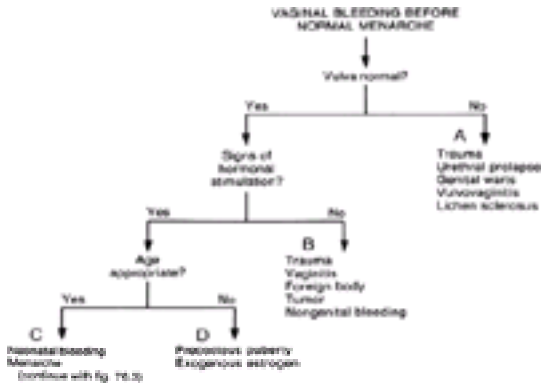

FIGURE 76.1. Diagnostic approach to vaginal bleeding before normal menarche.

Trauma to the vulva often produces lacerations or ecchymoses or both. Any vulvar injury should alert the emergency physician to the possibility of concurrent, potentially serious vaginal or rectal injuries. Vulvar lacerations do not usually bleed excessively, but hematomas can extend widely through the tissue planes, forming large, painful masses that occasionally produce enough pressure to cause necrosis of the overlying vulvar skin. Because even minor periurethral injuries can produce urethral spasm that leads to acute urinary retention, the injured child's ability to void should be checked routinely. The possibility of sexual assault must be considered in the management of every child with a genital injury.

Urethral prolapse (see Chapter 94) is probably the most common cause of apparent vaginal bleeding during childhood. Some patients with urethral prolapse complain of dysuria or urinary frequency, but most have bleeding as their only symptom. A prolapse is diagnosed by its characteristic doughnut shape (Fig. 94.6). The ring of protruding urethral mucosa above the introitus is swollen and dark red with a central dimple that indicates the meatus. When the child is supine, the prolapse is often large enough to cover the vaginal introitus and appears to protrude from the vagina. Bleeding comes from the ischemic mucosa. If the diagnosis is in doubt, one may safely catheterize the bladder through the prolapse to obtain urine. Some patients with small prolapses whose urethral tissue is still pink will improve with the use of sitz baths alone for several days. However, if the prolapsed tissue is dark and necrosing at the time of the patient's first examination or if sitz baths are not effective, elective surgical excision of the prolapsed tissue will be needed within a few days after diagnosis.

Genital warts, like a urethral prolapse, can be recognized by inspection (Fig. 94.12) and can produce bleeding when they are located on the mucosal surface of the introitus or just inside the hymenal ring. Because the presence of such warts in a child indicates that sexual contact may have occurred, cases should be reported to the state child protective services agency (see Chapter 128). Because topical podophyllin can be absorbed through disrupted mucosal surfaces and result in systemic toxicity, bleeding genital warts should be treated surgically.

Vulvar inflammation can be seen in some patients with vaginal bleeding resulting from bacterial or fungal vulvovaginitis. Infections caused by *Shigella* species, group A hemolytic streptococci, *Neisseria gonorrhoeae*, and *Candida albicans* produce vaginal bleeding or bloody discharge in varying proportions of cases. A few children with rectal *Enterobius vermicularis* (pinworm) infestations scratch so vigorously that they excoriate the perineal area and cause bleeding. Pinworm ova can often be discovered by low-power microscopic examination of perianal material that is collected with clear cellophane tape and then attached to a glass slide.

Although bleeding per se is not common, ecchymoses and telangiectasias are frequent clinical manifestations of lichen sclerosus (Fig. 76.2), an uncommon, chronic, idiopathic skin disorder that most often affects the vulva. In this condition, white, flat-topped papules gradually coalesce to form atrophic plaques that involve the vulvar and perianal skin in a symmetric hourglass pattern. Topical treatment with either hydrocortisone or testosterone is helpful in most cases.

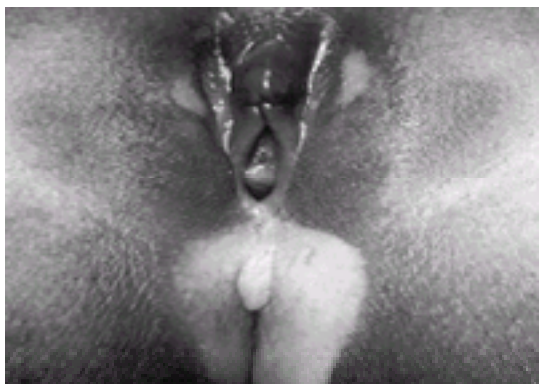

FIGURE 76.2. Figure-of-eight pattern of vulvar and perianal hypopigmentation in a 10-year-old girl with lichen sclerosus.

VAGINAL BLEEDING WITHOUT SIGNS OF HORMONAL STIMULATION

Trauma, infection, and foreign bodies are the most common causes of vaginal bleeding during childhood (Fig. 76.1B).

Vaginal bleeding after trauma indicates a potential emergency. A penetrating narrow object can damage the rectum, bladder, or abdominal viscera without producing much external evidence of injury. Because vaginal lacerations do not always produce a great deal of bleeding or pain, the emergency physician cannot rely on the severity of the patient's symptoms to indicate the extent of the injury. When a child sustains a genital injury, the physician must consider the possibility that it was inflicted during a sexual assault.

If the clinician knows or suspects that trauma has occurred, the girl's abdomen should be evaluated carefully. Lower quadrant tenderness may provide a clue to intra-abdominal injury. The vulva is inspected for bruises, and a rectal examination is performed to identify any lacerations. A general principle of management is that patients with penetrating genital injuries, even apparently minor ones, should undergo careful vaginal examination. This may require conscious sedation or general anesthesia, particularly in young children. Laboratory evaluation of the child with vaginal trauma should include a baseline hemoglobin determination and a urinalysis to screen for hematuria that might indicate urethral or bladder injury.

About half of all patients with *Shigella* vaginitis have bleeding that may be more noticeable than the associated discharge. Most patients do not have concurrent diarrhea. Vaginal infections with group A streptococci, *N. gonorrhoeae,* and *C. albicans* also cause bleeding in some cases. A vaginal culture will provide the diagnosis and guide the selection of an appropriate antibiotic. The manifestations and treatment of vaginal infections in children are discussed in more detail in Chapter 94.

Although a chronic, foul-smelling discharge is generally considered the hallmark of a vaginal foreign body, many girls have intermittent scanty vaginal bleeding alone or with an unimpressive discharge. If a foreign body is strongly suspected but the patient's vagina cannot be visualized when she is placed in the knee–chest position, the patient should receive either gentle vaginal lavage (using saline solution, a 50-mL syringe with the plunger discarded, a red rubber catheter, and gravity) or an examination under conscious sedation or anesthesia. Because the most common foreign body—toilet paper—is not radiopaque, pelvic roentgenography is not likely to be helpful and should be avoided.

Genital tumors are a rare cause of vaginal bleeding. Clear cell adenocarcinoma of the vagina or cervix occurs in about 0.2% of daughters whose mothers took diethylstilbestrol or other estrogen-containing drugs during pregnancy. Widespread publicity about this problem resulted in the abandonment of DES treatment in the early 1970s. Adenocarcinoma unassociated with DES exposure occurs rarely. Vaginal bleeding may be the first symptom of this cancer or of another rare malignancy, rhabdomyosarcoma (sarcoma botryoides). Urethral prolapse is sometimes mistaken for a malignant tumor and should be considered in the differential diagnosis.

Occasionally, a patient with a history of bleeding has no abnormalities and no bleeding at the time of the examination. This history should not be dismissed lightly because most parents are good observers, but the patient's urine and stool should also be checked for blood. Vaginal foreign body and inapparent genital trauma are also in the differential diagnosis.

VAGINAL BLEEDING WITH SIGNS OF HORMONAL STIMULATION

During the first 2 to 3 weeks of life, and late in puberty, hormonal fluctuations produce physiologic vaginal bleeding of uterine origin (Fig. 76.1C). Before female infants are born, high levels of placental estrogen stimulate growth of both the uterine endometrium and breast tissue. As this hormonal support wanes after birth, some infants have an endometrial slough that results in a few days of light vaginal bleeding. The bleeding will stop spontaneously and requires no treatment except reassurance for the parents. Occasionally, an adolescent girl is brought to the emergency department (ED) by her family to confirm their belief that she is having her first menstrual period. In this case, if the adolescent's age and degree of pubertal development are appropriate for menarche (Table 76.2), no further evaluation is necessary.

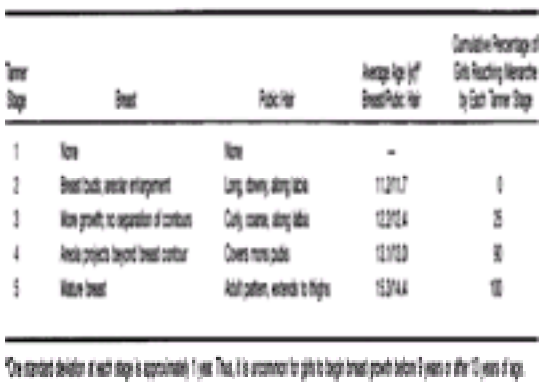

Table 76.2. Chronology of Pubertal Development in Normal Girls

If a girl less than 8 years of age has bleeding that is cyclic or is associated with breast development (thelarche), pubic hair growth (adrenarche), or accelerated linear growth, the various causes of precocious puberty must be given careful consideration in the differential diagnosis (Fig. 76.1D). Such a patient and her parents should be questioned about possible environmental exposure to feminizing hormones (e.g., chronic use of creams or medications containing estrogen). The possibility that a girl early in puberty simply has a nonendocrinologic disorder (foreign body, trauma) must also be considered. If the patient does appear to have precocious puberty, she should be checked in particular for café-au-lait spots (McCune-Albright syndrome) and an abdominal mass (endocrinologically active ovarian tumor or cyst) and referred from the ED to a pediatrician or pediatric endocrinologist for subsequent evaluation and follow-up.

VAGINAL BLEEDING AFTER MENARCHE

Evaluation and Decision

In the discussion to follow, only postmenarchal adolescents with abnormal vaginal bleeding are considered. Accordingly, the first discrimination the emergency physician must make is between those patients whose menstrual bleeding is heavier or more prolonged or more frequent than they would like but is nevertheless normal, and those patients whose bleeding falls outside the limits presented on page 613. In the evaluation of this latter group of patients, it is important to inquire specifically about the patient's menstrual chronology, including her age at menarche, her usual menstrual pattern, and the date of onset of her most recent normal menstrual period. Every patient who has been sexually active, regardless of age, should be asked whether she is or has ever been pregnant. Any current method of contraception should also be ascertained.

Because anovulatory bleeding is almost without exception painless, but approximately 90% of patients with ectopic pregnancy complain of abdominal pain at some time before the diagnosis is established, the physician must inquire carefully about the presence or absence of recent lower abdominal or pelvic pain. Other pertinent historical details include the presence or absence of fainting, fever, easy bruising, sexually transmitted diseases, and trauma.

During the physical examination, the patient's pulse and blood pressure are noted carefully and checked for orthostatic change. If the patient has been injured or is sexually active, a complete pelvic examination is performed. A speculum examination is not necessary for virginal adolescent patients who have not been injured, but a bimanual examination should be carried out routinely because teenagers are not always candid about their sexual activity. If it is more comfortable, bimanual rectoabdominal palpation with the patient in the lithotomy position can be substituted, or the examiner can place one finger intravaginally instead of two.

It is advisable to obtain a rapid qualitative urine pregnancy test early in the evaluation of most postmenarchal adolescents presenting to the ED because of vaginal bleeding (Fig. 76.3). For the occasional parent who finds it difficult to understand the rationale for this test for his or her daughter, it may be helpful to point out that the medical consequences of failing to recognize a pregnant patient can be substantial.

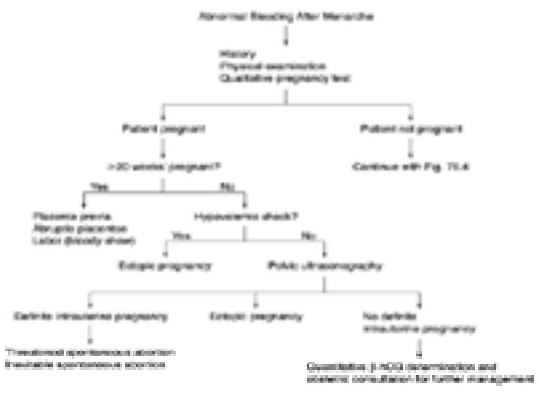

FIGURE 76.3. Diagnostic approach to abnormal uterine bleeding after menarche: pregnant patients.

BLEEDING IN LATE PREGNANCY

If the patient is 20 weeks' pregnant or more by history or abdominal examination, potential causes of bleeding that must be identified promptly are a bloody show during labor, premature separation of the placenta (abruptio placentae), and placenta previa (Fig. 76.3).

Because pelvic examination of a patient with placenta previa can provoke uncontrollable hemorrhage, the emergency care of a patient with vaginal bleeding after the 20th week of pregnancy starts with the management of potential hypovolemic shock (see Chapter 3), rather than with an examination to determine the anatomic site of bleeding. Thus, the patient's vital signs are recorded, the fetal heart rate is monitored, and a large-bore intravenous catheter is put in place. The patient with an apparently normal initial blood pressure is watched carefully nonetheless because her baseline pressure during pregnancy may have been elevated. Initial laboratory evaluation should include determinations of the hematocrit, the platelet count, the prothrombin time, and the partial thromboplastin time to screen for disseminated intravascular coagulation, which may be present in moderate and severe abruption. If the patient continues to bleed while in the ED, volume replacement is initiated while blood is typed and cross matched for use when available. If the patient is stable and the fetus is in no distress, the goal of subsequent investigation is to determine the location of the placenta and to identify the cause of the bleeding if it is not placenta previa. Obviously, an obstetrician should be consulted at the earliest opportunity regarding further ED management of the pregnant patient with second- or third-trimester bleeding.

BLEEDING WITH SHOCK

If the patient with vaginal bleeding is in the first or second trimester of pregnancy and has shock or early signs of cardiovascular instability (pallor, perspiration, vomiting), ruptured ectopic pregnancy must be ruled out. In this case, the treatment of shock and diagnostic measures should be undertaken simultaneously. Pelvic examination is performed, and obstetric consultation should be obtained rapidly. Emergency laparoscopy or laparotomy may be necessary for critically ill patients. If the patient is relatively stable, culdocentesis or transabdominal or transvaginal ultrasonography may help to clarify the diagnosis.

EARLY PREGNANCY

The patient with uterine bleeding before 20 weeks' gestation has a pregnancy that is either intrauterine but complicated or ectopic. In 1992, approximately 2% of all pregnancies in the United States were ectopic. The risk is lower among adolescents than among older women. The case-fatality rate declined dramatically between 1970 and 1986, from 35 to 4.9 deaths per 10,000 ectopic pregnancies.

Vaginal bleeding occurs in 60 to 80% of patients who have ectopic pregnancies. The bleeding is usually light. The timing of the bleeding sometimes leads the patient to consider it a normal menstrual flow, but about 75% of patients report having missed between one and three menstrual periods. Approximately 90% of patients with ectopic pregnancies experience localized or diffuse abdominal pain that may be present only briefly or for longer than a week before the diagnosis is made. The classic triad of amenorrhea, abdominal pain, and abnormal bleeding occurs in only about 70% of patients with ectopic pregnancy. On examination, the uterus is normal in size or only minimally enlarged because it does not contain the embryo.

Transvaginal or transabdominal ultrasonography is used to guide the management of an adolescent patient with vaginal bleeding and a positive qualitative pregnancy test. If sonography shows that the pregnancy is intrauterine and the fetus is viable, threatened abortion is diagnosed. The bleeding is usually light; some patients have uterine cramps. A spontaneous abortion is considered inevitable or incomplete if the fetal heartbeat is not detectable or if tissue fragments have already been expelled from the uterus. The bleeding is usually heavier, and the patient reports painful uterine contractions. Quantitative b-hCG levels can remain well above 0 for as long as 4 to 6 weeks after spontaneous and induced abortions.

Septic abortion is diagnosed if signs of infection—usually fever, disproportionately severe pelvic pain, and leukocytosis—are present during a spontaneous or induced abortion. After an induced abortion, persistent or heavy bleeding may indicate retained products of conception. In a missed spontaneous abortion, the embryo is not expelled from the uterus within 4 weeks of its death. Dark bleeding is often seen. The patient's symptoms of pregnancy may have regressed, the uterus is smaller than it should be according to her menstrual history, and disseminated intravascular coagulation may occur. Although the emergency physician needs to be able to recognize these complications of pregnancy, consultation with an obstetrician is an important component of clinical management for pregnant patients with bleeding.

Sonographic signs suggestive of ectopic pregnancy include a solid or complex adnexal mass, a pelvic mass, particulate fluid in the fallopian tube, an endometrial pseudogestational sac, and cul-de-sac fluid. If sonography fails to demonstrate either a definite intrauterine pregnancy or a clearly ectopic pregnancy, a quantitative b-hCG level should be obtained. On ultrasonographic examination, an intrauterine gestational sac should be visible with an abdominal probe when the b-hCG level is above 6000 mIU/mL and with a transvaginal probe when the b-hCG is above 2000 mIU/mL. A fetal heartbeat is detectable by approximately the fifth week of gestation (3 weeks after conception) on transvaginal ultrasonographic examination and by the sixth or seventh week of gestation on transabdominal ultrasonography. Among patients with vaginal bleeding, no definite intrauterine gestational sac on transvaginal sonography, and a b-hCG level of 2000 mIU/mL or higher, about 40% will miscarry, about 55% have ectopic pregnancies, and only about 5% have normal intrauterine pregnancies.

Serum progesterone measurement can also be helpful in predicting the outcome of pregnancy complicated by vaginal bleeding. The likelihood of fetal viability declines as the progesterone level declines. Approximately 90% of pregnant patients with vaginal bleeding but with progesterone concentrations above 20 ng/mL have normal pregnancy outcomes. However, at a progesterone level below 10 ng/mL, only about 20% of pregnancies will continue normally. Serum

progesterone measurement is not helpful in distinguishing spontaneous abortion from ectopic pregnancy.

Pregnant patients with vaginal bleeding and indeterminate results on transvaginal ultrasonography should be followed carefully, either by admission to hospital or by close outpatient follow-up with serial quantitations of b-hCG. Obstetric consultation should be obtained in developing an appropriate management plan. In a normal pregnancy, between days 5 and 42 after conception and above an initial level of 100 mIU/mL, the b-hCG level doubles approximately every 2 days. A decline in b-hCG on serial measurement or an increase of less than 66% in 48 hours will suggest a nonviable fetus, but cannot differentiate intrauterine from extrauterine pregnancy.

VAGINAL OR CERVICAL BLEEDING

On pelvic examination, only a few patients will prove to have vaginal or cervical bleeding. Patients with bleeding from significant vulvar, vaginal, or cervical lacerations should be referred to a gynecologist. The evaluation and management of victims of sexual assault are discussed in detail in Chapter 94 and Chapter 128. Hymenal tears produced by coitus rarely require any treatment beyond reassurance for the patient. Bleeding genital warts should not be treated with topical podophyllin because toxic amounts of the resin can be absorbed systemically (see Chapter 94). Malignant tumors are a rare cause of vaginal bleeding during adolescence.

Patients are unlikely to be aware of cervical friability or bleeding caused by infection. On examination, however, punctate cervical hemorrhages (a strawberry cervix) can be seen in about 3% of women with trichomonal vaginitis. Cervical bleeding after swabbing and mucopurulent discharge are common manifestations of cervicitis caused by *Chlamydia trachomatis*. Cervical lesions of herpes simplex may also cause a small amount of bleeding.

UNDIAGNOSED UTERINE BLEEDING

Most adolescents with the complaint of vaginal bleeding are not pregnant, and their bleeding is uterine in origin. Each of these patients should receive a complete blood count to screen for anemia and thrombocytopenia. Thrombocytopenia of any cause is the most common coagulation defect responsible for excessive menstrual bleeding. The history and physical examination may now lead the emergency physician to consider as more or less likely each of the several diagnoses discussed below (Fig. 76.4).

FIGURE 76.4. A diagnostic approach to abnormal uterine bleeding after menarche: nonpregnant patients.

ADNEXAL MASS

An adnexal mass in a teenager with abnormal uterine bleeding and a negative pregnancy test may be an ovarian cyst, an abscess, or rarely, a neoplasm. Laboratory investigations should be used to clarify the size, location, and nature of the mass (ultrasonography) and to estimate the likelihood of pelvic infection (leukocyte count, sedimentation rate, cervical tests for gonorrhea and chlamydial infection).

PELVIC TENDERNESS

Pelvic inflammatory disease and ovarian cysts are the likely diagnostic possibilities in a patient with abnormal uterine bleeding and pelvic pain or tenderness. Abnormal bleeding occurs in nearly one-third of patients with pelvic inflammatory disease, probably as a result of endometritis. Pelvic inflammatory disease is discussed in greater detail in Chapter 94. Rarely, an adolescent has spotty midcycle bleeding for 24 hoursor less in association with the transient decline in estrogen level that occurs at the time of ovulation. The unilateral pain of mittelschmerz may accompany this brief bleeding episode.

PAINLESS UTERINE BLEEDING

Most patients with painless abnormal bleeding have dysfunctional uterine bleeding (DUB), a consequence of anovulation. Anovulation is an especially likely diagnosis in patients who experience alternating oligomenorrhea and menometrorrhagia. The most common underlying causes of the anovulation itself are functional immaturity of the hypothalamic–pituitary axis, obesity, and polycystic ovary syndrome (see Chapter 48). The management of DUB is detailed in Chapter 94. Hypothyroidism should be considered as a possible underlying cause of dysfunctional uterine bleeding if the patient has other symptoms or signs of thyroid dysfunction.

Hormonal contraception is an important, common cause of dysfunctional uterine bleeding. Of women who use birth

control pills containing 35 µg or less of estrogen, 5 to 10% will have breakthrough intermenstrual spotting or bleeding, especially during the first 3 months of contraceptive pill use. Breakthrough bleeding is also a common side effect of progestin-only contraceptive pills, injectable medroxyprogesterone, and long-acting progestin implants. Many patients using birth control pills experience estrogen withdrawal bleeding if they forget to take one or several pills.

Less common causes of painless abnormal bleeding include gonococcal and chlamydial endometritis, pharmacologic anticoagulation, and disorders of platelet function (e.g., Glanzmann's thrombasthenia, von Willebrand's disease). Every sexually active patient should be screened for gonorrhea and chlamydial infection. Also to be kept in mind is the possibility, albeit unlikely, of a false-negative urine pregnancy test (as a result of testing too soon after conception, dilute urine, or low hCG production by a blighted or ectopic ovum). The large number of commercially available urine pregnancy tests vary in sensitivity. The ED physician should be aware of the lower limit of detection of the hCG test in use at his or her facility.

Suggested Readings

Brennan DF. Ectopic pregnancy. Part II: diagnostic procedures and imaging. Acad Emerg Med 1995;2:1090–1097.

Claessens EA, Cowell CA. Acute adolescent menorrhagia. Am J Obstet Gynecol 1981;139:277–280.

Dewhurst CJ, Cowell CA, Barrie LC. The regularity of early menstrual cycles. J Obstet Gynaecol Br Commonw 1971;78:1093–1095.

Dowd MD, Fitzmaurice L, Knapp JF, et al. The interpretation of urogenital findings in children with straddle injuries. J Pediatr Surg 1994;29:7–10.

Hill NCW, Oppenheimer LW, Morton KE. The aetiology of vaginal bleeding in children: a 20-year review. Br J Obstet Gynaecol 1989;96:467–470.

Mateer JR, Valley VT, Aiman EJ, et al. Outcome analysis of a protocol including bedside endovaginal sonography in patients at risk for ectopic pregnancy. Ann Emerg Med 1996;27:283–289.

Paradise JE, Willis ED. Probability of vaginal foreign body in girls with genital complaints. Am J Dis Child 1985;139:472–476.

CHAPTER 77

Vaginal Discharge

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Normal infants over 1 month of age and prepubertal girls do not have liquid vaginal secretions. Consequently, any vaginal discharge in a female child is abnormal. However, vaginal discharge in neonates and girls who are pubertal may be either normal or abnormal because during these times, estrogen, either maternal or endogenous, stimulates growth of the vaginal epithelium and secretion of mucus by the paracervical glands. The resulting vaginal discharge consists of desquamated epithelial cells and mucus, is not irritating, and requires no treatment. It is known as physiologic leukorrhea. A vaginal discharge that persists beyond the neonatal period, that occurs during childhood, or that is accompanied by discomfort in a pubertal patient is abnormal and needs to be investigated.

EVALUATION AND DECISION

General Considerations

Although the complaint of vaginal discharge is common among both children and adolescents, the emergency physician should recognize from the outset that this symptom is not necessarily a sensitive or a specific indicator of actual lower genital tract disease. On the one hand, as noted in the definition given previously, an asymptomatic vaginal discharge during the first several weeks of life or after the onset of puberty is a normal occurrence. This physiologic leukorrhea nevertheless may prompt an emergency department (ED) visit by a girl in early puberty concerned about the unexpected change in her body's function. On the other hand, among prepubertal girls, the complaint of vaginal discharge, irritation, itching, or dysuria can indicate a urologic, gastrointestinal (GI), dermatologic, or gynecologic disorder. Thus, the emergency physician must routinely review GI and dermatologic, as well as genitourinary, symptoms when evaluating a girl with the complaint of vaginal discharge. In addition, every child with genital complaints (and her parents) should be asked directly about the possibility that she has experienced sexual contact (see Chapter 128). Although physicians are sometimes reluctant to raise this question, many parents will have considered it already, and some will have asked their daughters before the visit to the doctor.

The physical examination and cultures of any vaginal discharge visible on examination are the emergency physician's best guides to proper management of an infant or child with the complaint of vaginal discharge. For examination of the external genitalia, infants and children should be placed in the frog-leg position either on the parent's lap or on an examining table (see Fig. 94.2A). The genital mucosa of infants and children is normally reddish rather than dull pink, because the epithelium is relatively thin in the absence of estrogenic stimulation. This appearance of the introitus should not be mistaken for inflammation. Children should be examined next in the knee–chest position to check for the presence of a foreign body (see Fig. 94.2B). If the examiner sees a vaginal discharge when the child is in either position, a specimen should be collected for culture after the child has returned to the supine position. The use of a soft plastic medicine dropper or a bladder catheter attached to a 3-mL syringe with butterfly tubing is a fairly comfortable method for aspirating vaginal secretions. If a girl's secretions are minimal, the dropper or catheter can be used instead to instill and then withdraw nonbacteriostatic saline washings for culture. Alternatively, the physician can obtain secretions with a cotton-tipped swab moistened with nonbacteriostatic saline solution, but this method is usually less comfortable for the patient.

If a postpubertal girl with vaginal discharge or other lower genital tract complaints has never had sexual intercourse, a speculum examination is not necessary for her evaluation. Either the frog-leg or the lithotomy position may be used for inspection of the girl's external genitalia and for the collection of specimens for microscopic examination, culture, or both.

Standard speculum and bimanual pelvic examinations are integral components of the evaluation of sexually active adolescents with vaginitis. Because these patients have high rates of sexually transmitted infections, the examiner should routinely obtain vaginal specimens for microscopy and cervical specimens for the diagnosis of *Chlamydia trachomatis* and *Neisseria gonorrhoeae.* In addition, the emergency physician must be alert for signs of pelvic inflammatory disease and pregnancy.

The patient's age and hormonal status should be considered first in the differential diagnosis of vaginal discharge (Fig. 77.1). For a more detailed discussion of the specific vaginal infections mentioned in this section, the reader is referred to Chapter 94.

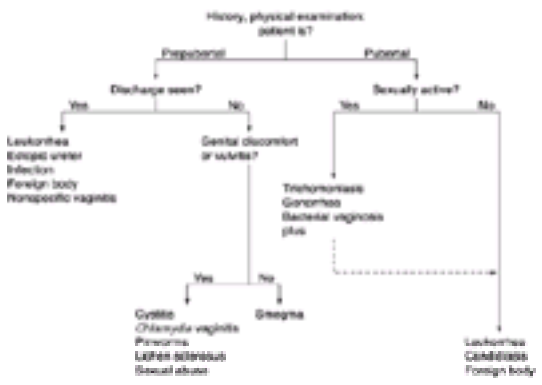

FIGURE 77.1. A diagnostic approach for vaginal discharge.

Infancy and Childhood

Physiologic leukorrhea is a normal vaginal discharge common among female infants during the first 2 to 3 weeks of life. It is clear or white, slippery when fresh, and sticky when dried. Some neonates have associated withdrawal bleeding when maternal estrogenic stimulation of the uterine endometrium wanes. Trichomonal vaginitis should be suspected if an infant's discharge persists beyond the neonatal period. Occasionally, a baby whose mother has trichomonal vaginitis acquires this infection during delivery. Infected infants may be irritable and have a whitish or yellowish thin discharge. Uncommonly, infants have purulent discharge associated with a congenital malformation of the genitourinary tract (e.g., ectopic ureter). A malformation should be suspected if an infant's discharge is accompanied by signs of systemic infection (fever, vomiting, or poor appetite) or if a child with chronic discharge also has had recurrent urinary tract infections (UTIs).

Among older infants and children, a visible vaginal discharge is most likely to indicate a bacterial infection. Cultures for *N. gonorrhoeae* and for other pathogens must therefore be obtained. Children with vaginitis should never be treated with antibiotics on the basis of the character of the discharge, microscopy, or Gram stain. Definitive diagnosis by culture is necessary to guide antibiotic selection and to address the medicolegal or public health questions raised when gonorrhea or *Shigella* is diagnosed.

Gonococcal infection typically produces a whitish to greenish purulent discharge. Bloody discharge occurs in half the cases of *Shigella* vaginitis and is common in vaginitis caused by group A b-hemolytic streptococci. Trichomonal vaginitis occurs virtually exclusively in infants less than 6 months of age and in postpubertal children. Chlamydial infections are nearly always asymptomatic but can produce dysuria, genital discomfort, or a scant mucoid vaginal discharge. Diagnostic tests for *C. trachomatis* vaginal infection should be reserved for children with histories of sexual abuse and for those whose bacterial cultures already have proved negative because the prevalence of infection is low in unselected populations and because false-positive antigen-detection tests are relatively common. As in adults, diabetes mellitus, broad-spectrum antibiotics, and immunodeficiency are risk factors for the development of candidal vaginitis in children.

An intermittently bloody, foul-smelling vaginal discharge is the classic complaint of the patient with a vaginal foreign body. Small wads of toilet paper, the most common foreign bodies, are usually easy to see just inside the vaginal vault on knee–chest examination. The emergency physician must have a high index of suspicion for this diagnosis if the child's vagina cannot be inspected satisfactorily while in the knee–chest position, because intravaginal toilet paper cannot be palpated rectally. Rigid foreign bodies—pencil erasers, pins, beads, nuts—are more likely to be palpable during rectal examination but are uncommon. Gentle vaginal lavage with saline solution can be used to flush out bits of toilet paper. Small round objects sometimes can be removed if the examiner places a finger in the rectum and then applies gentle outward pressure. However, if the object is large or sharp or if simpler maneuvers fail, visualization and removal of a foreign body under conscious sedation or general anesthesia may be required.

If examination of the patient discloses vulvar inflammation, excoriation, or hypopigmentation but little or no vaginal discharge, lichen sclerosus and other dermatologic disorders should be considered in the differential diagnosis. Lichen sclerosus is a chronic, idiopathic dermatitis characterized by atrophy, telangiectasia, and hypopigmentation of the perineal skin, often in a figure-of-eight pattern (see Fig. 76.2). Because severe cases can resemble genital trauma, the physician should take care not to confuse the two. Perineal excoriation or inflammation secondary to pinworm infestation, varicella, or any generalized dermatitis also can be misinterpreted as primary vulvovaginal disease.

Some girls with complaints of vaginal discharge or discomfort will have no abnormality on genital examination. In these cases, diagnostic possibilities include poor perineal hygiene, smegma, masturbatory behavior, sexual abuse, *Chlamydia* vaginitis, and UTI. Girls with poor perineal hygiene will respond well to frequent sitz baths. Genital smegma occurs in girls, as well as in boys, and sometimes is mistaken by parents for a pathologic discharge. It consists of desquamated epithelial cells; is thick, yellow or white, and sticky; and is characteristically located in the interlabial folds and around the clitoral prepuce. Somatic genital discomfort is the presenting complaint of a small number of children who have been sexually abused but are not injured or infected. The task of differentiating abused children from those who display age-appropriate masturbation or genital curiosity can be difficult and often requires consultation with a specialist in child mental health or sexual abuse assessment. The possibility of UTI also should be pursued in girls with genital symptoms but normal physical examinations.

Children with vaginal discharge that cannot be ascribed to any of the conditions just discussed generally are considered to have nonspecific vaginitis. This condition has been attributed to poor perineal hygiene and mechanical or chemical irritants. Accordingly, frequent sitz baths, careful wiping anteroposteriorly after defecation, and the avoidance of

presumed irritants (e.g., ballet tights, bubble bath, sand) are recommended for its treatment. These measures will produce improvement in most patients with nonspecific vaginitis but should not be recommended until after examination has been performed and appropriate cultures obtained.

Adolescence

With the onset of puberty, girls' rising estrogen level promotes the discharge of vaginal mucus and cells. This physiologic leukorrhea persists throughout the reproductive years but is most likely to arouse the concern of girls who are starting puberty and are therefore unaccustomed to its presence. On microscopic examination, the discharge shows only abundant epithelial cells. Culture of a specimen is unnecessary. Postpubertal girls with genital itching are most likely to have candidal vaginitis, even in the absence of any predisposing factor. The associated cheesy discharge may be so scanty that it goes unmentioned by the patient and unnoticed by the clinician. Among postmenarchal patients, birth control pills, broad-spectrum antibiotics, diabetes mellitus, and pregnancy are associated with an increased likelihood of symptomatic vulvovaginal candidiasis. Physical irritants, either mechanical or chemical, should be considered in the differential diagnosis for patients with nonspecific examinations and only polymorphonucleocytes or no abnormality on microscopic examination of the vaginal discharge. Although it is much more common among sexually active adolescents, bacterial vaginosis (see below) occurs occasionally in teenagers who have never had sexual intercourse. A forgotten tampon, the most common intravaginal foreign body, is a rare cause of vaginitis in adolescents.

In girls who have had sexual intercourse, bacterial vaginosis (known previously as nonspecific vaginitis, *Gardnerella* vaginitis, *Haemophilus vaginalis* vaginitis, and *Corynebacterium* vaginitis), trichomoniasis, and gonorrhea are the most likely causes of an abnormal vaginal discharge. Bacterial vaginosis is a clinical syndrome characterized by increased, malodorous vaginal discharge with a pH above 4.5, clue cells but few neutrophils seen on microscopy, and the release of an aminelike odor when potassium hydroxide is added to the discharge. The syndrome is thought to reflect overgrowth of anaerobes and *G. vaginalis* and a relative paucity of lactobacilli in the vaginal ecosystem; it is probably the most common cause of symptomatic vaginal discharge among sexually active adolescents who visit urban EDs. Trichomonal vaginitis is characterized by an increased volume of vaginal discharge associated in some cases with mild pruritus. The discharge is frothy in about 25% of cases. Gonococcal cervicitis or endometritis can produce a noticeable vaginal discharge, but most infected adolescent girls have no lower genital tract symptoms. A pathologic discharge in a girl with gonococcal cervicitis is most likely to be caused by concomitant trichomoniasis or bacterial vaginosis.

Three maneuvers—measurement of the pH of vaginal discharge and microscopic examination of the discharge suspended in 0.5 mL or less of saline solution and in 10% potassium hydroxide—are needed to provide a diagnosis for adolescent patients with vaginitis. To avoid contamination by cervical discharge, specimens should be obtained by swabbing the lateral vaginal wall. Indicator paper should be applied directly to undiluted discharge to measure pH because blood, seminal fluid, lubricating jelly, and saline can falsely elevate the measurement.

Bacterial vaginosis may be diagnosed when three of four conditions are present (Amsel criteria): 1) a homogeneous, whitish discharge; 2) vaginal pH above 4.5; 3) a fishy odor liberated when potassium hydroxide is added to the discharge; and 4) clue cells. Clue cells are squamous epithelial cells with indistinct borders obscured by granular bacteria, best seen at 400 magnification. On Gram stain, long Gram-positive rods (lactobacilli) are scarce, and short Gram-negative and Gram-variable coccobacilli (*Gardnerella, Prevotella, Mobiluncus* species) are abundant. Trichomoniasis is easily diagnosed if motile flagellates are seen on microscopic examination (see Fig. 94.7). However, because microscopy is negative in 20 to 50% of infected women, a rapid DNA probe test or a culture for *Trichomonas* can be helpful for diagnosing patients with suggestive symptoms or signs but negative wet preps. Fungal hyphae are not visible microscopically in over half of patients with symptomatic candidiasis (see Fig. 94.8). It is therefore reasonable to recommend empirical antifungal treatment for patients with vulvovaginal pruritus or burning and a vaginal pH less than 5 if a mechanical or physical irritant cannot be identified and if no alternative diagnosis is identified on microscopy.

A follow-up plan should be outlined clearly for every adolescent patient seen in the ED with a newly diagnosed sexually transmitted infection to identify treatment failures and reinfections, to ascertain whether the patient's sexual partner was treated, to explore the patient's need for contraception, and to carry out other preventive health measures, including Papanicolaou screening and acquired immunodeficiency syndrome (AIDS) counseling.

Suggested Readings

DeMeo LR, Draper DL, McGregor JA, et al. Evaluation of a deoxyribonucleic acid probe for the detection of *Trichomonas vaginalis* in vaginal secretions. Am J Obstet Gynecol 1996;174:1339–1342.

Embree JE, Lindsay K, Williams T, et al. Acceptability and usefulness of vaginal washes in premenarcheal girls as a diagnostic procedure for sexually transmitted diseases. Pediatr Infect Dis J 1996;15:662–667.

Hammerschlag MR, Rettig PJ, Shields ME. False positive results with the use of chlamydial antigen detection tests in the evaluation of suspected sexual abuse in children. Pediatr Infect Dis J 1988;7:1114.

Hillier SL. Diagnostic microbiology of bacterial vaginosis. Am J Obstet Gynecol 1993;169:455–459.

Paradise JE, Campos, JM, Friedman HM, et al. Vulvovaginitis in premenarcheal girls: clinical features and diagnostic evaluation. Pediatrics 1982;70:193–198.

Redmond, CA, Cowell CA, Krafchik BR. Genital lichen sclerosus in prepubertal girls. Adolesc Pediatr Gynecol 1988;1:177–180.

Sobel JD. Vaginitis. N Engl J Med 1997;337:1896–1903.

CHAPTER 78

Vomiting

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Vomiting is defined as the forceful, coordinated act of expelling gastric contents through the mouth. Vomiting may be caused by number of problems in diverse organ systems. Although it often represents a transient response to a self-limited infectious, chemical, or psychologic insult, it also may portend serious infections, metabolic disturbances, or diseases in gastrointestinal (GI), neurologic, or other major organ systems. Thus, an orderly approach to diagnosis is crucial.

Vomiting is a highly complex act, involving coordinated closure of gastric pylorus and glottis; relaxation of stomach, cardioesophageal junction, and esophagus; and vigorous diaphragmatic and abdominal wall muscular contraction. A series of interconnected coordinating centers in the medulla have been identified, with varying responsiveness to afferent signals from diverse areas of the body, including nocireceptors, chemoreceptors, and mechanoreceptors in the pelvic and abdominal viscera and peritoneum, genitourinary system, pharynx, labyrinth, and heart. The chemoreceptor trigger zone (CTZ) in the floor of the fourth ventricle contains chemoreceptors that monitor both blood and cerebrospinal fluid and is probably the key center initiating the emetic response to drugs (especially cytotoxic chemotherapeutic agents) and metabolic aberrations. Recent therapeutic advances arise from an evolving understanding of neurotransmitter activity in these centers. Blocking serotonin (5-hydroxytryptamine) in the CTZ and in vagal nuclei using receptor antagonists (ondansetron hydrochloride or granisetron) has proven to be successful in preventing emesis associated with many chemotherapeutic and radiotherapeutic cancer treatments, as well as emetogenic poisonings.

A related complaint, also often heard in the emergency department (ED), is that of young infants who “spit up.” This refers to the nonforceful reflux of milk into the mouth, which often accompanies eructation. Such nonforceful regurgitation of gastric or esophageal contents is most often physiologic and of little consequence, although it occasionally represents a significant disturbance in esophageal function.

It is convenient to attempt to organize the many diverse causes of regurgitation and vomiting into age-related categories (Table 78.1). Although overlap is considerable, the most common and serious entities tend to fall into such groupings.

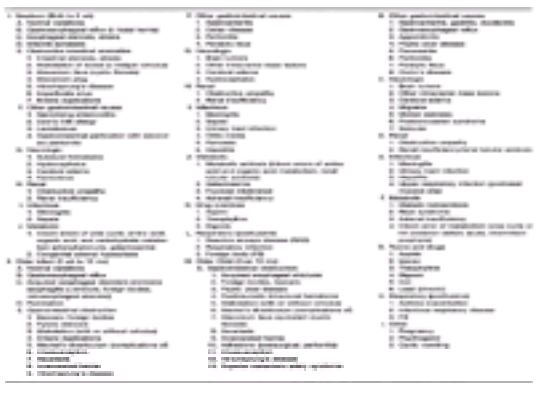

Table 78.1. Vomiting and Regurgitation: Principal Causes by Usual Age of Onset and Etiology

EVALUATION AND DECISION

General Approach

A brief perusal of the long list of causes for vomiting in Table 78.1 serves to emphasize the need for an orderly approach to the differential diagnosis of this symptom. The approach advocated here focuses on three key clinical features: child's *age*, evidence of *obstruction*, and signs or symptoms of *extra-abdominal organ system disease.* Other important points to

consider include *appearance* of the vomitus, *overall degree of illness* (including the presence and severity of dehydration or electrolyte imbalance), and *associated GI symptoms*.

HISTORY

The history should focus on the key elements already listed. The patient's age often is critical because certain important entities (especially those that cause intestinal obstruction) are seen exclusively in neonates, older infants, or children beyond the first year of life. Evidence of obstruction, including symptoms of abdominal pain, obstipation, nausea, and increasing abdominal girth, are sought in addition to vomiting. Other associated GI symptoms may include diarrhea, anorexia, flatulence, and frequent eructation with reflux. The suspicion of significant extra-abdominal organ system disease is raised by neurologic symptoms such as severe headache, stiff neck, blurred vision or diplopia, clumsiness, personality or school performance change, or persistent lethargy or irritability; by genitourinary symptoms such as flank pain, dysuria, urgency and frequency, or amenorrhea; by common infectious complaints such as fever, sore throat, or rash; or by respiratory complaints such as cough, increased work of breathing, or chest pain (Table 78.2 and Table 78.3).

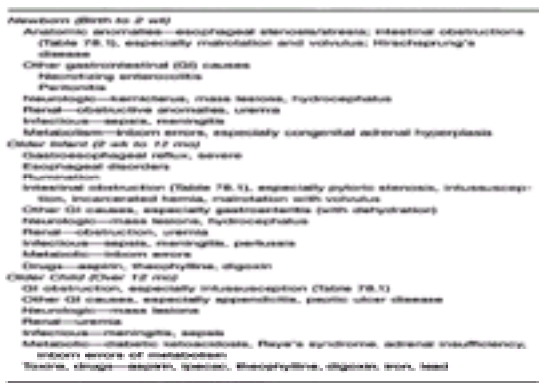

Table 78.2. Life-Threatening Causes of Vomiting

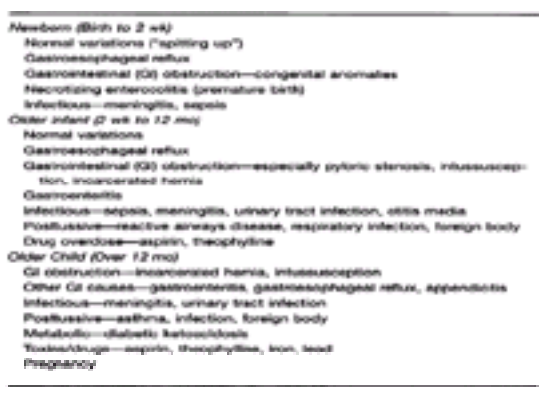

Table 78.3. Common Causes of Vomiting

The appearance of the vomitus (by history and inspection when a specimen is available) often is helpful in establishing the site of pathology. Undigested food or milk should suggest reflux from the esophagus or stomach caused by lesions such as esophageal atresia (in the neonate), gastroesophageal (GE) reflux, or pyloric stenosis. Bilious vomitus suggests obstruction distal to the ampulla of Vater, although it occasionally is seen with prolonged vomiting of any cause when the pylorus is relaxed. Fecal material in the vomitus is seen with obstruction of the lower bowel. Hematemesis usually reflects a bleeding site in the upper GI tract; its evaluation is detailed in Chapter 30.

PHYSICAL EXAMINATION

The physical examination is directed first toward evaluating the overall degree of toxicity. Does the baby look septic? Is there the inconsolable irritability of meningitis? Are there signs of life-threatening dehydration or concern for symptomatic hypoglycemia? Does the child exhibit the bent-over posture, apprehensive look, and pained avoidance of unnecessary movement typical of peritoneal irritation in appendicitis? Next, attention is aimed at the abdomen. Are there signs of obstruction such as ill-defined tenderness, distension, high-pitched bowel sounds (or absent sounds in ileus), or visible peristalsis? A complete physical examination must include a search for signs of neurologic, infectious, toxic/metabolic, and genitourinary causes, as well as an evaluation of hydration status (see Chapter 18).

The diverse nature of causes for vomiting makes a “routine” laboratory or radiologic screen impossible. The history and physical examination must guide the approach in individual patients. Some well-defined clinical pictures demand urgent radiologic workup. For example, abdominal pain and bilious vomiting in a child requires supine and upright plain films and a limited upper GI series for evaluation of congenital obstructive anomalies such as malrotation; or a child with paroxysms of colicky abdominal pain and grossly bloody stools requires immediate flat and upright abdominal films and usually a contrast (air or barium) enema for the likely diagnosis and reduction of intussusception. Other situations require no imaging studies (e.g., a typical case of viral gastroenteritis or a classic history for pyloric stenosis with definite palpation of the pyloric tumor). In many cases, body fluid cultures or serum chemical analyses are essential for making a diagnosis (e.g., meningitis, aspirin toxicity, Reye syndrome, pregnancy) or for guiding management (e.g., degree of metabolic derangement in pyloric stenosis, diabetic ketoacidosis). For most straightforward, common illnesses (e.g.,

gastroenteritis, cold with posttussive emesis), laboratory investigation is unwarranted.

APPROACH TO CHILDREN BY AGE GROUPS

With these introductory concepts in mind, we can approach the differential diagnosis of the principal causes of vomiting on an age-related basis. An algorithm for such an approach that uses the key clinical features previously outlined is illustrated in Figure 78.1.

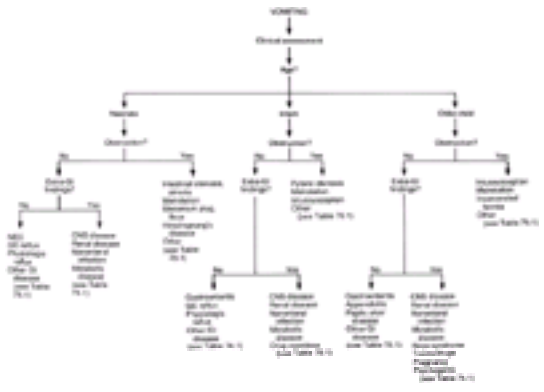

FIGURE 78.1. Differential diagnosis of vomiting. *GI,* gastrointestinal; *NEC,* necrotizing enterocolitis; *GE,* gastroesophageal; *CNS,* central nervous system.

Neonates

A careful history should focus on the perinatal events, onset and duration of vomiting, nature of the vomitus, associated GI symptoms, and the presence of symptoms referable to other organ systems. Newborn babies with the onset of vomiting in the first days of life always should be suspect for one of the common *congenital GI anomalies* that cause obstruction, such as esophageal or intestinal atresia or web, malrotation, meconium ileus, or Hirschsprung's disease. If the vomiting is bilious, bright yellow, or green, an urgent surgical consultation is required. In most cases, a serious and possibly life-threatening mechanical obstruction may be the cause of bilious vomiting. All patients in whom the possibility of GI obstruction is entertained must have immediate flat and upright abdominal films. Other clinical features, such as toxicity, dehydration, and lethargy, usually attest to the length of time of the obstruction and its severity. Except for the later presentations of malrotation, most neonates with a congenital basis for their bowel obstruction will present during their initial nursery stay. Therefore, it is uncommon to see such babies for the first time in the ED. Neonates or infants with malrotation and volvulus may present with abdominal pain (crying, drawing up their knees, poor feeding), with evidence of obstruction (bilious emesis), or an acute abdomen (abdominal distension or rigidity). The diagnosis of malrotation is confirmed by the abnormal radiographic location of the duodenal–jejunal junction (upper GI series) and/or the cecum (contrast enema).

Other serious causes of neonatal vomiting that may present to the ED include *infection*, such as meningitis, sepsis, pyelonephritis, or necrotizing enterocolitis (it should be noted that such serious infections are often not accompanied by fever in the neonate); *increased intracranial pressure* (ICP) related to cerebral edema, subdural hematoma, or hydrocephalus; *metabolic acidosis* or *hyperammonemia* caused by the rare inborn errors of amino acid and organic acid metabolism; and *renal insufficiency* or *obstruction*. Such infants usually appear ill, with associated lethargy and irritability; sometimes fever, a full fontanel, a diminished urinary stream, an abdominal mass, or respiratory signs will suggest the correct cause. Obviously, any ill neonate with vomiting, even in the absence of obstruction, also requires hospitalization and prompt evaluation for sepsis and neurologic, renal, and metabolic disease.

Commonly, however, a young infant in the first 2 to 4 weeks of life who appears entirely well is brought to the ED with the complaint of persistent vomiting. The birth history and perinatal course are unremarkable. The baby has gained weight appropriately (usually 5 to 7 oz/week after the first week of life), is vigorous, and has an entirely normal physical examination. Usually, a close description of the “vomiting” (or even better, a trial feeding in the ED) reveals the problem to be *physiologic regurgitation* or *reflux*; so-called spitting up. This is a common (nearly 20% of infants reflux) and insignificant problem, probably representing some normal variation in the developmental maturation of the lower esophageal sphincter (LES). These infants do not exhibit forceful abdominal contractions but rather reflux milk effortlessly into their mouths, which dribbles out, usually when prone, and often with a burp. The degree of reflux may be increased by improper feeding techniques such as failure to burp the baby, using nipples with holes that are too small, bottle propping, or overfeeding. Observation of a feeding trial and emphasis on good technique suffices for initial management of such babies. Like all newborns, they should be referred for ongoing pediatric care. Most babies outgrow such regurgitation by 6 to 9 months of age, and 95% have resolution of symptoms by 12 months.

Other infants who regurgitate easily may not be managed so easily. Their course may have more significant symptoms of pain, arching, and high volume and frequency of regurgitation, or it may be complicated by distal esophagitis or gastritis, failure to thrive, esophageal–peptic strictures, pulmonary disease, or rarely, apnea or near–sudden infant death syndrome (SIDS) event. Such infants are diagnosed as having *gastroesophageal reflux disease* (GERD), a more severe or pathologic degree of LES dysfunction that is much less common (1:500). Several imaging and physiologic studies may be used to confirm the diagnosis and to correlate a patient's signs and symptoms with episodes of reflux. A 24-hour intraesophageal pH probe is the most sensitive diagnostic test for GE reflux. Based on the patient's history, evaluation of delayed gastric emptying can be done by GE scintiscan, or by an upper GI series to rule out an anatomic cause for the delay. Endoscopy is used to assess suspected complications (esophagitis or stricture); esophageal manometry is primarily a research tool in this disease. Infants with GE reflux should be followed closely by a pediatrician. In

uncomplicated GE reflux, reassurance, postural management, and dietary measures are usually adequate. For more severe symptoms or with complications, additional medical management includes the use of prokinetic agents (cisapride or metoclopramide) and acid blockade (ranitidine or cimetidine). Omeprazole, a proton pump inhibitor, has been useful for resistant or severe esophagitis, but its safety and dosing in young children has not been well established. GERD that is resistant to vigorous medical therapy (continues to cause serious complications) may be considered for surgical fundoplication (see Chapter 93 and Chapter 118).

Older Infant

Infants who present with vomiting after the first few weeks of life may still have intestinal obstruction, but the underlying causes are somewhat different than in the neonate. The important lesions responsible for mechanical obstructions in this age group include congenital hypertrophic pyloric stenosis (HPS), malrotation, intussusception, incarcerated hernia, enteric duplications, and complications of Meckel's diverticulum. Occasionally, other anomalies that might be expected to present in the neonate, such as Hirschsprung's disease, will appear only after several weeks or months of life. In all cases, these conditions have physical findings suggestive of intestinal obstruction and often are specific for the level of obstruction. Having a high index of suspicion for both common and uncommon forms of intestinal obstruction is important to making a timely diagnosis.

The typical infant with *pyloric stenosis* (see Chapter 118) appears in the ED at 4 to 6 weeks of age (95% present by 3 months; rarely after 20 weeks) with a chief compliant of projectile vomiting during or shortly after a feeding. The vomiting in pyloric stenosis is typically crescendo in nature, with increasing frequency and severity over days to weeks. By contrast, vomiting caused by GE reflux tends to be relatively consistent over time; in malrotation, vomiting is sudden in onset and can be episodic. The vomitus is nonbilious, reflecting obstruction at the pylorus, and usually is voluminous, nearly the entire content of the feeding. The infant may become constipated if vomiting has been of sufficient duration. On examination, an olive-sized mass may be palpated (most easily after vomiting has occurred) in the right upper quadrant to the right of the midline and just above the umbilicus. Peristaltic waves may be visualized, moving from left upper to right upper quadrants, again indicating obstruction at the pylorus. Unless the infant is significantly dehydrated, the child usually is vigorous and active, although irritable because of hunger. These infants often develop hypochloremic, hypokalemic metabolic alkalosis, which should be corrected before surgery (see Chapter 86).

The diagnosis of pyloric stenosis is clinical, based on the classic history of projectile, nonbilious emesis, and examination with hyperperistalsis and palpation of a pyloric mass or “olive.” Imaging studies (ultrasound (US) or upper GI series) are not necessary if the history and examination are conclusive. In recent years, however, earlier presentations have resulted in a greater number of infants evaluated before the development of the diagnostic clinical hallmarks, and consequently an increased reliance on imaging studies to confirm the diagnosis. Early diagnosis has been beneficial with a decrease in the proportion of patients with alkalosis, shortened hospital stay, and decreased morbidity. US has become the diagnostic modality of choice with characteristic findings in HPS of a thickened pyloric wall (greater than 3 mm) with a lengthened canal (greater than 15 mm). However, some centers have found upper GI series to be more cost effective because of greater operator experience with the procedure and therefore fewer repeat studies, and the ability to provide information on other origins of nonbilious vomiting. Endoscopy has recently been recommended as an adjunct test for complicated cases in which clinical examination, US, and/or upper GI series were inconclusive. Surgical pylorotomy, the standard treatment for HPS, is scheduled as soon as dehydration and metabolic derangements (if present) have been corrected. Medical management with intravenous or oral atropine has been successful in a recent re-investigation. Still, a larger, well-controlled trial and cost analysis would be necessary before its use is recommended in place of surgical correction.

Between 2 months and about 5 to 6 years of age, the most common cause of obstruction is *intussusception* (see Chapter 118). Most children develop this disorder between 3 months and 2 years of age; the average was 16 months old in a recent large series. Early symptoms usually include paroxysms of colicky abdominal pain and vomiting, suggesting a GI illness. Initially, the infant may appear relatively well between attacks, but some children will fall asleep or seem prostrate at these times. Initially, there may be a normal stool, then occult positive, but usually within 6 to 12 hours, dark maroon blood is passed per rectum; this blood often is mixed with mucus, earning the label of “currant jelly” stool. However, some infants with intussusception may present primarily with lethargy and decreased responsiveness, without striking GI symptoms (so-called neurologic or painless intussusception). Examination of the abdomen usually reveals a somewhat tender, sausage-shaped mass on the right side. The mass may be more easily appreciated by bimanual rectal and abdominal examination, and a test for occult blood may be positive in the absence of gross blood.

Recommendations for the diagnosis and treatment of suspected intussusception include supine and right-side up decubitus radiographs (decubitus to look for free air or mass at the hepatic flexure region; supine for signs of obstruction such as abnormal air–fluid levels, paucity of gas, or mass effect). US sometimes is used instead of radiographs if experienced personnel are available. Reduction is attempted by contrast (air or liquid) enema if perforation (free air on radiograph or peritonitis) or shock is not evident. Recent success rates for reduction in pediatric centers using air or liquid contrast enemas have improved to the range of 80 to 90%, with some centers reporting their increased success with repeated attempts after short intervals (45 to 60 minutes). Open reduction with laparotomy is reserved for patients with perforation or shock at initial diagnosis or when enema reduction is unsuccessful.

Other important causes of obstruction in the older infant include incarcerated inguinal hernia, volvulus, Hirschsprung's disease, or complications related to Meckel's diverticulum. The presence of an incarcerated hernia will be apparent on examination. Volvulus of the bowel is virtually always associated with bilious vomiting. A good clue to diagnosis of Hirschsprung's disease is asking, “Has your child ever had a normal (unstimulated) bowel movement?” (see Chapter 14 and Chapter 118). The obstructive complications of Meckel's diverticulum include intussusception and volvulus and have similar presentations of these types of obstruction related to other causes.

The principal nonobstructive causes of vomiting in the older infant include GI, neurologic, renal, infectious, and metabolic disorders. Nonobstructive GI disturbances are probably the most common cause for vomiting in this age group. *Viral gastroenteritis*, although usually appearing predominantly as diarrhea associated with vomiting, often begins with a

prodromal phase of vomiting alone (see Chapter 84). Physical findings usually are limited to ill-defined and inconsistent abdominal pain and signs of a variable degree of dehydration. Vomiting in older infants also is caused at times by persistent GE reflux, as well as by abdominal disorders uncommon in infancy, such as peptic ulcer disease or appendicitis. Occasionally, vomiting is seen in paralytic ileus related to infection (pneumonia, peritonitis) or electrolyte disorders.

Neurologic causes of vomiting in infancy also include *mass lesions* such as tumor, abscess, and intracranial hematoma (see Chapter 83), as well as *meningitis* and *encephalitis*. There may be evidence of increased ICP: increasing head circumference, bulging fontanel, and split sutures (papilledema is rarely noted during infancy). However, some brainstem tumors cause protracted vomiting by direct effect on the vomiting center without an accompanying increased ICP. Again, it is to be emphasized that meningismus rarely is seen with meningitis in infancy and that signs of increased ICP occur late (see Chapter 84). Early findings include fever, vomiting, lethargy, and irritability, especially the paradoxic irritability of increased crying with parental fondling.

Infections outside the GI and neurologic systems may cause vomiting in infants and, occasionally, in older children. The more important such infections are *otitis media* (OM), *urinary tract infection* (UTI), *respiratory infections*, and *viral hepatitis*. Positive physical findings on otoscopic examination are seen in OM, along with mild irritability, and often, fever (see Chapter 84). UTIs may be surprisingly devoid of localizing signs and symptoms in preschool children (see Chapter 122); nonspecific GI complaints, including vomiting and abdominal pain, fever, irritability, and anorexia, may be the only presenting symptoms. Urinalysis and culture provide the specific diagnosis. Vomiting also is a common event after the paroxysms of coughing seen in infants with pertussis (see Chapter 84). It is a common symptom in the prodromal phase of infectious hepatitis, usually preceding the onset of jaundice (see Chapter 93). Abnormal liver function tests substantiate this latter diagnosis.

Renal and *metabolic disorders* also cause vomiting in the older infant. Renal failure, renal tubular acidosis, or rarely, diabetic ketoacidosis may be seen in this age group. Hypoadrenalism, hepatic failure, Reye syndrome, and inborn errors of metabolism such as galactosemia and fructose intolerance also may present in infancy and may have vomiting as a prominent symptom in an ill-appearing infant.

Occasionally, parental overzealous use of over-the-counter or prescribed drugs in infants will lead to *intoxication.* Drugs that often produce vomiting in excessive doses include aspirin, theophylline, and digoxin; all of these intoxications are easily verified by associated signs and symptoms and specific drug levels. The problem of accidental ingestion is discussed later.

An additional rare cause of regurgitation or vomiting in infants, with onset usually at 6 to 12 months of age, is *rumination* (see Chapter 131). This severe psychiatric disorder of infancy, related to abnormal maternal–infant relationship, may progress to severe failure to thrive and to death. These infants seem to self-induce the reflux, often by gagging themselves, and often appear to partially rechew and reswallow their vomitus.

Older Child

Many of the causes of intestinal obstruction and other important GI diseases described in neonates and older infants, such as volvulus associated with malrotation, Hirschsprung's disease, a meconium ileus “equivalent” in the child with cystic fibrosis, and an incarcerated hernia, occasionally may first appear in the older child. Older children with malrotation and/or volvulus will often have a previous episodic history of vomiting or intermittent colicky abdominal pain. In addition, older children often are subjected to blunt abdominal trauma; persistent vomiting after such injury may reflect obstruction related to a duodenal intramural hematoma or ileus secondary to pancreatitis. Gastroenteritis, as in infants, continues to be the most common cause of vomiting in the older child seen in the ED. Two entities that usually occur in older children, appendicitis and peptic ulcers, are discussed here, although they occur rarely in infancy as well.

Appendicitis (see Chapter 118) in a preadolescent child classically begins with periumbilical, crampy abdominal pain and anorexia, often followed by vomiting. Then the pain shifts to the right lower quadrant and fever may develop. Younger children may deviate from this pattern by exhibiting less specific symptoms early in their illness and a more rapid progression to perforation and generalized peritonitis. As peritoneal irritation becomes well established, the child attempts to minimize any motion to the abdomen. Physical examination usually reveals localized involuntary right lower quadrant guarding and tenderness that, when mild, may be easier to elicit by asking the child to cough or to hop on one foot. In addition, there may be rebound and referred rebound tenderness along with a tender fullness high on the right during rectal examination. Atypical positions of the appendix (e.g., retrocecal, retroileal, pelvic) will be reflected in atypical areas of maximal tenderness, as well as in confusing symptoms such as diarrhea or dysuria (caused by appendiceal inflammation adjacent to colon or ureter/bladder). Pertinent laboratory findings often include leukocytosis with a left shift in the differential count, but a normal white blood cell (WBC) count in an afebrile patient does not rule out appendicitis. The urinalysis usually is normal. Occasionally, in an atypical patient, abdominal radiographs may be helpful in showing a right lower quadrant fecalith, localized obstruction, a mass effect with a paucity of gas in the right lower quadrant, or lumbar spine scoliosis. Appendicitis is a clinical diagnosis, but when the differential diagnosis is difficult or in early or equivocal cases, imaging studies are helpful. US has become the modality of choice in most centers for children with suspected appendicitis. US can be particularly helpful in distinguishing tubo-ovarian pathology or renal pathology from appendicitis. Computed tomography (CT) scan is widely becoming the imaging study of choice in adults; recent studies show excellent sensitivity of right lower quadrant CT with rectal contrast. In children, its use has currently been limited to patients with equivocal US studies or if an abdominal abscess is suspected. Its use has been suggested for patients with an unclear diagnosis who would be admitted to the hospital for observation, with the goal of decreasing therapeutic delays or of averting unnecessary admissions or surgeries.

Vomiting as a symptom of *peptic ulcer disease* in children usually is seen in association with abdominal pain (see Chapter 93). In young children, the pain often is nonspecific and not easily related to meals. In adolescents, the pattern becomes more classically related to food or antacids. There may be hematemesis and/or melena. The abdominal

examination may be normal or reveal mild to moderate epigastric tenderness. A strong clinical suspicion of peptic ulcer disease should be confirmed with an upper GI series or endoscopy. Other inflammatory lesions of the upper GI tract (gastritis, duodenitis, and Crohn's disease) can also cause persistent vomiting.

Genitourinary causes of vomiting in the older child include UTI and obstructive urologic disease. An important additional concern in adolescent girls is early *pregnancy* (see Chapter 48). It is common for such patients to visit the ED with the chief complaint of persistent vomiting (not necessarily only in the morning) for several weeks, and often sexual activity and/or amenorrhea is initially denied. Physical findings at this stage of pregnancy may be subtle. Thus, prolonged vomiting in a postmenarchal girl should be pursued with the appropriate urine or serum gonadotropin assays (see Chapter 48 and Chapter 130).

The important extra-GI infectious diseases of the older child that cause vomiting have been discussed for the most part under the neonatal and infantile headings of this chapter. Serious infections localize symptoms more readily in this older age group. Meningitis usually is accompanied by meningismus after the age of 2 years. Lower urinary infections tend to present with dysuria, frequency, and urgency as children approach school age, and pyelonephritis with fever and lower back pain or tenderness. The toddler or school-age child also may vomit with pharyngeal irritation (pharyngitis, postnasal mucous drip) or have posttussive emesis with persistent or severe cough caused by asthma, respiratory infection, or respiratory foreign body.

Neurologic disease that causes vomiting in the older child again represents (primarily) lesions that cause increased ICP or direct irritation of the medullary vomiting center; they usually lead to papilledema and/or abnormal neurologic findings on examination. One important exception is childhood *migraine* (see Chapter 83). Preadolescent children do not usually present with the classic migraine picture with aura, hemicranial headache, and scotomas. More often, they complain of rare but severe, poorly localized headaches accompanied by nausea and vomiting and followed by sleep. The physical examination between attacks usually is normal. Another common but minor form of vomiting on a neurologic basis (caused by labyrinthine stimulation) would be the propensity to motion sickness.

Metabolic aberrations, including hepatic, renal, and adrenal failure, all may cause vomiting in the older child (as well as during infancy). *Ketoacidosis* presenting for the first time in an as yet undiagnosed diabetic occurs more commonly in older children, especially at school entrance age and later as adolescence begins (see Chapter 97). Vomiting may be the chief complaint of such children, although careful questioning usually uncovers a preceding 3- to 4-week history of polyuria, polyphagia, polydipsia, and at times, weight loss. A fruity breath odor, dehydration, hyperpnea, and varying degrees of altered sensorium typically are present, and a urinalysis and serum glucose determination confirm the diagnosis of diabetic ketoacidosis.

The other important, but increasingly uncommon, cause of vomiting is *Reye syndrome* (see Chapter 93). Although it may occur at any age, it tends to be seen more commonly in toddlers and school-age children. Typically, these children have had a preceding viral illness within the past 2 weeks (especially varicella and influenza in the United States) from which they have just recovered, or they are recovering at the time of presentation. Generally, about 24 hours of severe, recurrent vomiting is followed immediately by progression through the varying stages of encephalopathy. Physical examination at the time of vomiting, before the onset of encephalopathy, may be normal or show only hepatomegaly. Thus, it is crucial to consider Reye or a Reye-like syndrome in the differential diagnosis and to pursue laboratory evidence of hepatic dysfunction in any child with persistent vomiting in association with recent flulike syndromes or varicella or with persistent vomiting with even the mildest change in sensorium toward obtundation and/or delirium. Abnormal laboratory data in Reye syndrome include elevated serum transaminase and ammonia, prolonged prothrombin time, and often, hypoglycemia; bilirubin usually is normal, making other forms of severe liver disease unlikely. Recent research has emphasized that the differential in cases meeting the criteria for Reye syndrome include Reye-like syndromes: inherited metabolic disorders and viral and toxic diseases. After emerging as a discrete diagnosis in the 1960s, there has been a dramatic global decline in cases of Reye syndrome since the late 1970s. Differing experiences internationally, as well as the increased differential diagnostic capabilities in distinguishing Reye-like syndromes, have rekindled the debate over the etiologic association of aspirin in Reye syndrome and the role of its decreased use in the declining frequency of cases. The substitution of acetaminophen for aspirin for antipyresis in childhood viral illnesses will most likely continue in this country until these issues are clarified.

In the discussion regarding older infants, mention was made of occasional inadvertent drug overdose by parents, causing intoxication-related vomiting. In children 1 to 4 years of age, *accidental ingestion* is a common problem. Acute poisonings that cause vomiting as a prominent symptom include aspirin, theophylline, digoxin, and iron sulfate (see Chapter 88). Chronic *lead poisoning* also occurs in this pica-prone age group. Early symptoms of lead intoxication are vomiting, colicky abdominal pain, anorexia, constipation, and irritability. Tragically, many such youngsters have been diagnosed as having nonspecific gastroenteritis syndromes initially, only to return days to weeks later with frank encephalopathy and, ultimately, severe neurologic sequelae. The history of pica and lead paint exposure (peeling paint chips, especially in homes dating back to the 1940s and 1950s) should be sought in every toddler with persistent vomiting. The diagnosis of plumbism can be confirmed with elevated blood levels of lead and erythrocyte protoporphyrins (see Chapter 88).

Finally, the school-age child or adolescent may vomit on a *psychologic* basis. Acutely, brief episodes of vomiting may occur with any emotionally disturbing event. Children with school phobia or other significant psychiatric problems may vomit persistently. Adolescents are at risk for self-induced vomiting in the context of the anorexia nervosa and bulimia syndromes (see Chapter 129 and Chapter 130). Before the physician attributes the vomiting to a psychologic cause, however, a careful history, general examination, and complete neurologic examination are necessary to minimize the likelihood of missing any organic origin. An assessment of disturbed family dynamics, history of emotional disorders, or evidence of depression and/or anxiety during the ED interview may corroborate the suspicion of vomiting on a psychologic basis and may warrant a psychiatric referral.

Suggested Readings

GENERAL

Gale JD. Serotonergic mediation of vomiting. J Pediatr Gastroenterol Nutr 1995;21(Suppl 1):S22–S28.

Grybowski J, Walker WA. Gastrointestinal Problems in the Infant. 2nd ed. Philadelphia: WB Saunders, 1983.

Meadows N. The central control of vomiting. J Pediatr Gastroenterol Nutr 1995;21(Suppl 1):S20–S21.

Moir CR. Abdominal pain in infants and children (Review) (11 refs). Mayo Clin Proc 1996;71(10):984–989, quiz 989.

Roy CC, Silverman A, Cozzetto FJ. Pediatric Clinical Gastroenterology. 3rd ed. St Louis: Mosby, 1983.

MALROTATION

Long FR, Kramer SS, Markowitz RI, et al. Radiographic patterns of intestinal malrotation in children. RadioGraphics 1996;16:547–556.

GERD

Colletti RB, Christie DL, Orenstein SR. Indications for pediatric esophageal pH monitoring. J Pediatr Gastroenterol Nutr 1995;21:253–262.

Faubion WA, Zein NN. Gastroesophageal reflux in infants and children. Mayo Clin Proc 1998;73:166–173.

PYLORIC STENOSIS

Hulka F, Campbell TJ, Campbell JR, et al. Evolution in the recognition of infantile hypertrophic pyloric stenosis. Pediatrics 1997;100(2):E9.

Nagita A, Yamaguchi J, Amemoto K, et al. Management and ultrasonographic appearance of infantile hypertrophic pyloric stenosis with intravenous atropine sulfate. J Pediatr Gastroenterol Nutr 1996;23(2):172–177.

Schechter R, Torfs CP, Bateson TF. The epidemiology of infantile hypertrophic pyloric stenosis. Paediatr Perinatal Epidemiol 1997;11(4):407–427.

INTUSSUSCEPTION

Daneman A, Alton DJ. Intussusception. Issues and controversies related to diagnosis and reduction (Review) (59 refs). Radiol Clin North Am 1996;34(4):743–756.

Ein SH, Alton D, Palder SB, Shandling B, et al. Intussusception in the 1990s: has 25 years made a difference? Pediatr Surg Int 1997;12:374–376.

McAlister WH. Intussusception: even Hippocrates did not standardize his technique of enema reduction (Comment) (Review) (35 refs). Radiology 1998;206(3):595–598.

APPENDICITIS

Hale DA, Molloy M, Pearl RH, et al. Appendectomy: a contemporary appraisal (Review) (23 refs). Ann Surg 1997;225(3):252–261.

Rao PM, Rhea JT, Novelline RA, et al. Effect of computed tomography of the appendix on treatment of patients and use of hospital resources. N Engl J Med 1998;338:141–146.

IMAGING

Gupta H, Dupuy DE. Advances in imaging of the acute abdomen (Review) (47 refs). Surg Clin North Am 1997;77(6):1245–1263.

Weinberger E, Winters WD. Abdominal pain and vomiting in infants and children: imaging evaluation (Review) (21 refs). Compr Ther 1997; 23(10):679–686.

REYE SYNDROME

Casteels-Van Daele M, Van Geet C, Wouters K, et al. The changing clinical pattern of Reye's syndrome 1982–90 (Letter; Comment). Arch Dis Child 1997;76(1):79.

Hardie RM, Newton LH, Bruce JC, et al. The changing clinical pattern of Reye's syndrome 1982–1990 (see Comments). Arch Dis Child 1996;74(5):400–405.

Smith TC. Reye's syndrome and the use of aspirin. Scott Med J 1996; 41(1):4–9.

Stumpf DA. Reye syndrome: an international perspective (Review) (15 refs). Brain Dev 1995;17(Suppl):77–78.

CHAPTER 79

Weakness/Flaccid Paralysis

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A large number of diagnostic considerations exist for the previously well child who presents to the emergency department (ED) complaining of recent onset of weakness or diminished muscle strength (Table 79.1). The initial diagnostic approach may vary considerably, depending on specific clinical and historical features. This chapter limits the discussion to the assessment of the child with acute or subacute onset of weakness and associated flaccid paresis or paralysis on neurologic examination. This chapter does not discuss weakness or inability to move an extremity secondary to pain from trauma (i.e., fracture) or infection (i.e., septic joint) or the child with generalized malaise. Although occasionally children will have weakness on a psychogenic basis, such patients can be distinguished from those with flaccid paralysis on the basis of neurologic examination.

Upper Motor Neuron Disease	Lower Motor Neuron, Neuromuscular Junction, and Muscle Disease
Cerebral Cortex	*Anterior Horn Cell*
Cerebrovascular accident	Poliomyelitis
Hemorrhage	Hopkins' syndrome
Emboli	Werdnig-Hoffman disease
Spinal Cord	*Peripheral Nerve Disease*
Trauma, hemorrhage	Guillain-Barré syndrome
Infection	Heavy metal poisoning
Epidural abscess	Fish toxins
Transverse myelitis	Porphyria
Malignancy	*Neuromuscular Junction*
Disc herniation	Organophosphates
Tethered cord	Tick paralysis
	Botulism
	Myasthenia gravis
	Muscle Disease
	Inflammatory myopathy
	Periodic paralysis
	Rhabdomyolysis
	Muscular dystrophies

Table 79.1. Differential Diagnosis Table

Motor weakness is defined as a decreased ability or inability to move one or several extremities voluntarily against gravity. It may present acutely, subacutely, or intermittently with easy fatigability. It is a reflection of a disease process that may involve the motor neuron unit at any or all of its points. Upper motor disease affects structures extending from the cerebral cortex to, but not including, the anterior horn cell. It is generally characterized by increased deep tendon reflexes and spasticity; however, early in the clinical course there may be flaccid paralysis. Lower motor neuron disease may involve the anterior horn cell, the peripheral axons, the neuromuscular junction (NMJ), or the muscle fibers. In general, it is associated with absent or diminished deep tendon reflexes. Although flaccid paralysis or profound weakness in children is an unusual finding in general, relatively common causes are listed in Table 79.2, and serious life- or limb-threatening causes are listed in Table 79.3.

Infant botulism
Guillain-Barré syndrome
Viral myositis

Table 79.2. Common Causes of Weakness

Cerebrovascular accident	Guillain-Barré syndrome
Spinal cord compression	Infant botulism
Organophosphate or heavy metal poisoning	Rhabdomyolysis
Tick paralysis	

Table 79.3. Life-Threatening Causes of Weakness

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of acute weakness can be categorized by the different parts of the nervous system that are affected. Upper motor neuron disease involves either the cerebral cortex or the spinal cord. The cerebral cortex can be damaged by a cerebrovascular accident (CVA), leading to sudden weakness, usually unilateral. Although CVAs are not as common in children as in adults, various predisposing factors can occur in the pediatric population. Children with sickle cell disease have increased risk for cerebral infarction during a vaso-occlusive crisis. Cerebrovascular disease associated with the rare metabolic disorder homocystinuria may result in CVA. Hypercoagulable states such as found in pregnancy, malignancy, or severe dehydration can lead to venous thrombosis. Embolic causes are to be considered in patients with congenital heart disease, mitral valve prolapse, or a history of rheumatic fever. An intracranial hemorrhage may occur after a relatively minor trauma in a child with hemophilia or spontaneously from a previously undiagnosed tumor or arteriovenous malformation (AVM). Finally, cocaine and amphetamine use have been associated with cerebral infarction in previously healthy patients.

Many processes can damage or compress the spinal cord, leading to weakness or paralysis. Trauma, with or without associated vertebral fracture or dislocation, can lead to spinal cord contusion or transection. The trauma often is severe (i.e., motor vehicle accident, football injury); however, it may be mild, particularly in children with Down syndrome or juvenile rheumatoid arthritis (JRA), in whom there may be an associated cervical vertebral instability. Neck or back trauma can also cause an epidural hematoma, which compresses the spinal cord as it expands. This is of particular concern in patients with hemophilia or other underlying coagulopathy.

Infectious causes of spinal cord compression include epidural abscess and transverse myelitis. An epidural abscess may develop from hematogenous dissemination of staphylococci or arise from direct spread from an adjacent carbuncle or vertebral osteomyelitis. In transverse myelitis (see Chapter 83), areas of inflammation and infarction in the spinal cord result in a clinical picture of flaccid paralysis in both legs, along with rectal and bladder incontinence. Often, cerebrospinal fluid (CSF) protein is elevated and pleocytosis is evident. Although the exact relationship between antecedent viral illnesses and transverse myelitis is not well understood, a history of fever, malaise, and myalgia for several days before the onset of paralysis often occurs.

Other causes of spinal cord compression include hemorrhage from trauma or an AVM, malignancy in the form of a spinal or paraspinal tumor or metastasis, disc herniation, or a tethered cord.

Werdnig-Hoffman disease is a disease of the anterior horn cells that presents with gradual weakness of the proximal muscles. There are three types, with onset of symptoms ranging from birth to 2 years of age. Lower cranial nerve involvement occurs early, leading to feeding difficulties and changes in the quality of the cry. The disease is gradually progressive, with deterioration of muscle strength and eventual respiratory insufficiency.

If peripheral axons are the site of the disease process, motor and sensory functions are both impaired with distal changes usually noted first. In children, one of the more common causes of acute peripheral neuropathy is Guillain-Barré syndrome (GBS) (see Chapter 83). It generally presents as ascending symmetric weakness over days to weeks, usually beginning in the legs and progressing proximally. More than half of the patients will have had an upper respiratory or gastrointestinal infection in the 4 weeks before the onset of the weakness. Many causative agents have been implicated including *Mycoplasma pneumoniae,* influenza virus, adenovirus, varicella, measles, mumps, Epstein-Barr virus, and cytomegalovirus. *Campylobacter jejuni* has been shown to be associated with a more severe form of the disease. A large majority of children report pain in the first week of the illness, with subsequent weakness developing in the painful muscles. Children, if old enough, may also describe paresthesias or numbness, often in a stocking-glove distribution.

The diagnosis of GBS is generally made based on history and clinical presentation. It can be supported, if necessary, by a lumbar puncture showing increased protein concentration in the CSF with no pleocytosis. In fact, the cell count is generally less than 10 lymphocytes/mm^3 and always less than 50 lymphocytes/mm^3. Electrodiagnostic studies can be done as well, if the diagnosis is still in question, and will show a characteristic multifocal demyelination pattern. Factors that make the diagnosis of GBS less likely include fever at the time of presentation, age less than 1 year, sharp sensory level, or persistence of bowel or bladder symptoms. In a young child, when sensory symptoms cannot be described by the patient and sensory examination is limited, GBS is more difficult to diagnose. Other more likely causes in infants with weakness include the anterior horn cell diseases and infantile botulism, discussed in the following.

Heavy metal poisoning from lead, arsenic, or thallium can also cause peripheral nerve damage. Lead poisoning is associated with anemia, abdominal pain, constipation, and often encephalopathy in children (see Chapter 88). Thallium

ingestion may cause ptosis, alopecia, and vomiting in addition to weakness.

Fish toxins can also cause peripheral neuropathy (see Chapter 88). Ciguatera is an illness caused by a toxin from fish that are found generally in the South Pacific, but occasionally near the lower Atlantic states. Nausea and vomiting begin 4 to 36 hours after ingestion of the fish, followed by paresthesias and weakness, and typically reversal of hot/cold sensation. Ingestion of shellfish contaminated during a “red tide” has been associated with paralysis as well. Initial nausea and vomiting are followed rapidly by paresthesias, cranial nerve involvement, and weakness. Supportive care is indicated for both until the paralysis reverses.

Finally, acute intermittent porphyria is a rare autosomal-dominant cause of peripheral neuropathy and paralysis. It is often associated with abdominal pain. Peripheral motor weakness is present and often is more evident in proximal muscles. Central nervous system involvement may be manifested by altered sensorium and hallucinations, and there may be sensory involvement as well. Acute attacks can be precipitated by drugs such as barbiturates, sulfonamides, griseofulvin, estrogens, and alcohol. Detection of increased porphyrin precursors in the urine, as well as familial history, help make the diagnosis.

Diseases involving the NMJ can be recognized by their association with cranial nerve findings and autonomic processes. Importantly, sensation is *not* affected. Organophosphate exposure is a classic example of NMJ dysfunction. Organophosphates, which are used as insecticides, inhibit cholinesterase activity, thereby allowing acetylcholine to remain bound to its postsynaptic receptor. Acetylcholine receptors are found in the brain, at the motor end plate, and in muscarinic receptors in the autonomic nervous system. Thus, in addition to muscle fasciculations, muscle cramps, and weakness, patients exposed to organophosphates may have symptoms of altered mental status and cholinergic stimulation (see Chapter 88).

Tick paralysis is caused by another toxin that is presumed to affect the NMJ, produced by the dog tick *Dermacentor variabilis* and the Rocky Mountain wood tick *Dermacentor andersoni*, as well as several other species of ticks found in the United States. Because exposure to the tick often precedes the paralysis by 5 to 10 days, travel history is important. The paralysis is typically ascending, and progresses rapidly over 12 to 36 hours to the bulbar area. Although 50% of patients may complain of paresthesias, the sensory examination is usually normal. Tick paralysis, although not commonly encountered, should be considered in the differential diagnosis of acute weakness, particularly in spring and summer months of the endemic areas, when ticks are most active in the warm environment.

A third agent that involves the neuromuscular junction is the toxin of *Clostridium botulinum.* It produces paralysis by preventing release of acetylcholine at the myoneural junction. It can be found in improperly home-canned foods or any incompletely reheated or unheated food. It also is found in the soils of some areas of the United States, particularly in eastern Pennsylvania and California. If contaminated food is the source of the toxin, within 24 to 48 hours of exposure, patients will experience nausea and vomiting, followed by diplopia, photophobia, and then dysphagia and dysarthria from sequential involvement of cranial nerves. Skeletal muscle paralysis follows without any sensory involvement. In wound botulism, a wound is contaminated with soil containing the spores of the clostridium. It presents in a similar manner except that no gastrointestinal symptoms are associated.

In infant botulism, the GI tract of the infant is colonized by *C. botulinum.* It has been linked to ingestion of honey, which has a high rate of contamination by the spores. Because the spores are also found in soils, they may gain access to the baby from dust at a nearby construction site, or soil tracked into the home. Although it is generally seen in infants younger than 9 months of age, peak incidence is between 2 and 3 months. It commonly presents with a history of constipation, lethargy, and feeding difficulties. On physical examination, hypotonia, general muscle weakness, pooling of oral secretions, and a decreased gag reflex are present, and dilated pupils respond poorly to light. Sensory examination is normal. Deep tendon reflexes may be normal or diminished early, and are generally diminished or absent late in the disease.

Finally, myasthenia gravis is a disease process involving the transmission of acetylcholine across the synaptic membrane to its receptors. It is recognized by easy fatigability of muscles and worsening weakness over the course of a day. Sensory examination and deep tendon reflexes are normal. Ptosis is often the most easily recognized symptom.

When the muscle itself is the target of the disease, proximal weakness is often noted first, and sensory function is normal. An example of muscle diseases is the group of inflammatory myopathies in which the muscles are often tender. In dermatomyositis or polymyositis, skin and joint manifestations and proximal weakness are usually present. The characteristic rash in dermatomyositis is an erythema of the upper eyelid area, spreading to a malar distribution. Erythema over the extensor surfaces of joints is often present also. Deep tendon reflexes diminish as weakness progresses and muscles atrophy. In viral myositis, a febrile respiratory illness is associated with significant muscle tenderness and weakness. Deep tendon reflexes are generally present, and sensory examination is normal. Serum creatine kinase is elevated in all of the inflammatory myopathies. Another muscle disease involves periodic attacks of paralysis associated with either decreased or increased serum potassium. The hypokalemic form may be associated with an autosomal-dominant familial disorder, hyperthyroidism, hyperaldosteronism, chronic renal disease, ingestion of large amounts of licorice, or chronic vomiting or diarrhea. The familial form often presents in adolescence with sudden paralysis that may involve all the extremities, usually after a large carbohydrate meal or during rest after strenuous exercise. Sensory examination is normal. Autosomal-dominant familial hyperkalemic paralysis usually is less severe. Improvement of paralysis should ensue after intravenous correction of the potassium imbalance.

Rhabdomyolysis from excessive physical exertion, prolonged seizures, stimulant-drug overdose, or genetic disorders may result in acute weakness. Muscles are generally tender, myoglobin is evident in the urine, and serum creatine phosphokinase (CPK) and AST are elevated.

Finally, the muscular dystrophies, although usually thought of as more slowly progressive, can present to an emergency physician as a recently noticed weakness by the patient or their family. The forms that present in childhood involve the

proximal muscles and are primarily X-linked and thus are seen in boys 3 to 15 years of age. They often present first with gait disturbances, including toe walking, waddling, or frequent falling. The children may have trouble getting up from the floor and may demonstrate the Gower sign C, a pushing off the floor with the hands to assist the weak pelvic and thigh muscles. Muscles are generally not tender. Deep tendon reflexes are present early, but disappear later. Sensory examination is normal, and serum creatine kinase is elevated.

EVALUATION AND DECISION

Information most helpful in the critical assessment of the child with sudden paralysis can be discovered in a detailed history, with particular attention to trauma, toxic exposures, prodromal illness, and family history. The physical examination should note the distribution of paralysis—symmetric versus asymmetric, proximal versus distal—as well as associated cranial nerve weaknesses; facial, ocular, or bulbar, deep tendon reflexes, and sensory changes. These clinical features guide the diagnostic approach, as outlined in Figure 79.1.

FIGURE 79.1. Approach to the child with weakness or flaccid paralysis. *SCIWORA,* spinal cord injury without obvious radiographic abnormality; *CT,* computed tomography; *CVA,* cerebrovascular accident; *MRI,* magnetic resonance imaging; *AVM,* arteriovenous malformation; *CPK,* creatine phosphokinase.

The first important historical question is whether the patient had neck or back trauma preceding the weakness. If so, the patient should be immobilized, and spinal radiographs should be taken immediately. Cord transections often result in initially flaccid paralysis and sensory loss below the level of the lesion, along with bladder and rectal incontinence. Deep tendon reflexes may be absent early but will become increased over time. If a patient has these symptoms after trauma, radiographs can confirm a fracture or dislocation. However, a patient with persistent symptoms should remain immobilized until further imaging studies are done because negative radiographs do not completely obviate spinal injury (see Chapter 106).

Once it has been confirmed that the patient has had no trauma, a complete history should be taken. A history of acute weakness associated with abdominal pain or vomiting suggests a possible toxin exposure such as botulism or fish poisoning. Information should be elicited relating to ingestion of potentially contaminated foods and the possibility of other symptomatic exposed individuals. Treatment in either case is supportive, with particular attention to respiratory function. Gastric decontamination and antitoxin may be used if necessary. Gastrointestinal symptoms may also be associated with heavy metal ingestion, either intentional or accidental. Finally, porphyria should be considered as a possible cause of weakness and abdominal pain. A familial history or recent ingestion of a known precipitant and urine studies for porphyrin precursors will help confirm the diagnosis.

A history of constipation for several days before the onset of weakness in a child less than 1 year of age highly suggests infantile botulism. Parents often also report several days of lethargy and feeding difficulties as well. Ingestion of honey or proximity to an endemic area would further support the diagnosis.

Before proceeding with the physical examination, the physician should be sure that the patient has not had an organophosphate exposure. Generally, this is given in the history or easily recognized by the strong garliclike smell on the patient and the readily apparent cholinergic symptoms. Health care personnel caring for such a patient should be careful to protect themselves from exposure by protective clothing and eyewear until the patient is decontaminated.

Upon beginning a thorough physical examination, the patient's respiratory and cardiovascular status should be assessed. Although rare, some diseases cause rapidly progressive weakness and may quickly involve the respiratory muscles or gag reflex. Resuscitation and stabilization should be done before further diagnostic evaluation.

One of the main goals of the physical examination in a patient with acute weakness is to locate the site of the disease process. Acute hemiplegia, with or without facial involvement or mental status changes, should direct the physician to an intracranial process such as a cerebral infarction or hemorrhage. Any child suspected of having a CVA should be stabilized and have a computed tomography (CT) scan of the head done promptly. Rarely, seizures or complicated migraines can be associated with hemiplegia, but usually the possibility of an intracranial event must be eliminated first.

When evaluating a patient with a complaint of weakness, a thorough motor examination will help define and locate the disease process. The physician should assess not only active motor strength but also resistance to gravity. Muscle groups should be inspected for increased or decreased tone, atrophy, and fasciculations. Deep tendon reflexes should be classified as normal, increased, diminished, or absent. A careful sensory examination should be done, both to light touch and to pain, as well as vibratory and position sense. It should be determined whether a spinal level of sensory loss is detectable. The patient's back should be examined for swelling, erythema, loss of normal lordosis, point tenderness,

rigidity, or splinting. Cranial nerves should be carefully assessed, looking specifically for ophthalmoplegia, facial weakness, or bulbar weakness as evidenced by dysphagia, dysarthria, or decreased gag.

Initially, the physician should determine whether the patient has signs and symptoms consistent with an acute or subacute spinal cord compression. A history of back trauma or back pain should be investigated, including eliciting the onset, location, and radiation, if any, of the pain. Other important historical questions pertaining to cord compression include bowel or bladder dysfunction, paresthesias, and abnormal gait. Motor examination would reveal profound weakness or paralysis, with distal more than proximal muscle groups affected. In spinal cord compression, patients may have diminished reflexes early in the course, progressing to increased reflexes later. Often, a spinal sensory level corresponding to the level of loss of motor function is detectable. There may be bladder retention or urinary or fecal incontinence.

Although spinal cord compression may have many etiologies that present similarly, some factors may be helpful in making a diagnosis. Acute onset of symptoms with associated fever and back pain suggests an epidural abscess or transverse myelitis. A patient with an epidural abscess may have point tenderness over the affected area in addition to the paralysis and bowel and bladder dysfunction. In transverse myelitis, prominent sensory loss occurs for pain and temperature, usually with a definable spinal sensory level corresponding to the level of back pain. Acute onset of symptoms without fever may indicate a spontaneous hemorrhage from an AVM. A more subacute presentation may suggest a spinal or paraspinal tumor. Disk herniation is less common in children but may be associated with trauma or symptoms of sciatica. Evidence of spinal dysraphism, such as a hair tuft, dermal sinus, or lipoma, may be a clue to a tethered cord, which also usually presents with a more gradual onset of weakness. Initial evaluation of any patient with signs and symptoms suggesting spinal cord compression should include spinal radiographs, usually followed by magnetic resonance imaging (MRI). Spinal cord compression is an emergency and may require high-dose methylprednisolone to decrease swelling and protect the spinal cord (see Chapter 125).

Assessment of deep tendon reflexes is the next important step in differentiating causes of weakness. Diseases of the NMJ, such as myasthenia gravis, should have normal reflexes. Myasthenia may also be recognized by the involvement of cranial nerves producing dysphagia, facial weakness, and ophthalmoplegia. Rarely, early in their course, other disease processes may present with weakness and normal deep tendon reflexes. However, they can be differentiated from myasthenia gravis by other factors, as explained in the following.

Decreased or absent reflexes, when not caused by early spinal cord compression, are a sign of disease in the anterior horn cell, peripheral nerve, or muscle. The physician can use the sensory examination to further distinguish these. If there is abnormal sensation on examination or subjective reporting of paresthesias, the disease is in the peripheral nerves. The most common cause for an acute or subacute onset of ascending flaccid paralysis, with diminished or absent deep tendon reflexes and associated sensory symptoms, is GBS (see Chapter 83). On physical examination, there may be demonstrable sensory loss, as well as loss of position and vibratory sense. Motor weakness, most often asymmetric and distal, is clinically apparent, although 10 to 20% of patients may have descending paralysis or proximal-greater-than-distal weakness. The combination of lower extremity weakness and sensory changes often cause the child to have an abnormal gait, which is often the presenting symptom. Deep tendon reflexes are diminished or absent in the weak muscles, although they may be preserved in less-affected muscles early in the course of the illness. Associated cranial nerve involvement is seen in 30 to 40% of patients, usually manifested by facial weakness or ocular paresis. Pupillary response is not affected, and rarely are the lower cranial nerves involved. Occasionally, children have autonomic symptoms, including arrhythmias and blood pressure lability. These can become severe and, in some cases, life-threatening. Because up to 20% of children will require assisted ventilation for respiratory paralysis, children with GBS should be admitted to the hospital and observed carefully for progression of weakness. In some cases, plasmapheresis or intravenous immunoglobulin administration may be indicated.

Tick paralysis can present similarly to GBS. Any patient with acute onset of ascending paralysis associated with dysesthesias or cranial nerve palsies should be carefully examined for ticks, particularly in the scalp and hairline. Removal of the tick results in dramatic and complete improvement.

In a patient with decreased or absent deep tendon reflexes but normal sensory examination, the disease process is either in the anterior horn cells or in the muscles. Examples of anterior horn cell disease include spinal muscular atrophy (Werdnig-Hoffman disease), enteroviral infection such as polio, and Hopkins' syndrome. In Werdnig-Hoffman disease, the infants have no associated pupillary or ocular motility dysfunction but are noted to have tongue fasciculations and a decreased gag reflex. Proximal muscles are weak, and deep tendon reflexes are diminished or absent. Sensory examination is normal.

Polio and Hopkins' syndrome both present with a unilateral, asymmetric paralysis. Polio is associated with fever, irritability, and often meningeal signs. There may be an antecedent history of exposure in an unimmunized person or oral vaccine administration in a young infant. Hopkins' syndrome is associated with a preceding asthma exacerbation, but the patient usually has no other symptoms at the time of presentation of the weakness.

Although infant botulism is a disease process of the NMJ and reflexes should be preserved, it may be confused with anterior horn disease because reflexes may in general be difficult to elicit or appreciate in a young infant. It will otherwise present similarly, with progressive weakness and normal sensory examination. Infant botulism may be distinguished by a history of constipation and the presence of dilated pupils poorly reactive to light.

Disease processes involving the muscle present with weakness, decreased reflexes, and a normal sensory examination. These include hypokalemia, hyperkalemia, rhabdomyolysis, inflammatory myopathies, and muscular dystrophies. These can be distinguished from anterior horn cell diseases by history, physical examination, and laboratory evidence of abnormal serum potassium, elevated CPK, or urine myoglobin.

In summary, when presented with a patient with a history of sudden or progressive weakness, the emergency physician

must carefully sort through the history of the weakness for evidence of trauma or toxins, as well as any associated symptoms. The physical examination is important in characterizing the nature, location, and symmetry of the weakness, as well as determining whether there are any accompanying sensory losses. Deep tendon reflexes must be checked carefully, and cranial nerve examination is also important. The history and physical together, as always, can determine the cause of the weakness in most cases. Select diagnostic studies, including imaging modalities, lumbar puncture, and serum and urine laboratory tests, may further clarify the diagnosis. Because of the life-threatening nature of many of the causes of weakness, most patients after ED evaluation and management will require inpatient hospital treatment.

Suggested Readings

Arnon SS. Infant botulism. Ann Rev Med 1980;3:541–560.

David WS, Doye JJ. Acute infantile weakness: a case of vaccine-associated poliomyelitis. Muscle Nerve 1997;20:747–749.

Evans OB, Vedanarayanan V. Guillain-Barré syndrome. Pediatr Rev 1997;18(1):10–16.

Fenichel GM. Clinical Pediatric Neurology: A Signs and Symptoms Approach. 2nd ed. Philadelphia: WB Saunders, 1993.

Ferrari ND, Weisse ME. Botulism. Adv Pediatr Infect Dis 1995;10:81–91.

Jones HR Jr. Childhood Guillain-Barré syndrome. Clinical presentation, diagnosis and therapy. J Child Neurol 1996;11(1):4–12.

Liedholm LJA, Eeg-Olofsson O, Ekenberg BEK, et al. Acute postasthmatic amyotrophy (Hopkins Syndrome). Muscle Nerve 1994;17:769–772.

LoVecchio F, Jacobson S. Approach to generalized weakness and peripheral neuromuscular disease. Emerg Med Clin North Am 1997;15(3):605–23.

Markland LD, Riley HD Jr. The Guillain-Barré syndrome in childhood. Clin Pediatr 1967;6:162–170.

Midura TF. Update: infant botulism. Clin Microbiol Rev 1996;9(2):119–125.

Paine RS, Byers RK. Transverse myelopathy in childhood. Am J Dis Child 1953;85:151–163.

Smith S. Peripheral neuropathies in children. In: Swaiman KF, ed. Pediatric Neurology: Principles and Practice, 2nd ed. St Louis: Mosby, 1994.

Swaiman KF. Anterior horn cell and cranial motor neuron disease. In: Swaiman KF, ed. Pediatric Neurology: Principles and Practice. 2nd ed. St Louis: Mosby, 1994.

CHAPTER 80

Wheezing

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Wheezes are continuous whistling or musical adventitial sounds that are the hallmark of lower airway obstruction. This chapter presents an organized approach to the diagnosis of conditions associated with wheezing in children beyond the newborn period. Emphasis is given to those conditions most likely to be encountered by the emergency department (ED) provider.

PATHOPHYSIOLOGY

Obstruction to air flow is the common denominator in all conditions that produce wheezing. Wheezing usually results from obstruction of the intrathoracic lower airways (bronchi and bronchioles) and only rarely by narrowing of the trachea or major bronchi. Obstruction of the lower airway passages may be anatomic or physiologic and is the result of one or more of several mechanisms: 1) extrinsic airway compression, 2) intrinsic airway narrowing, or 3) intraluminal airway blockage. Table 80.1 provides a simplified pathophysiologic classification of conditions that cause wheezing in children.

Extrinsic Airway Compression
Congenital structural anomalies
Cystic malformations of the lung
Vascular ring
Cardiovascular enlargement
Mediastinal tumors
Teratoma, neuroblastoma, thymoma, ganglioneuroma, pheochromocytoma
Enlarged mediastinal lymph nodes
Tuberculosis, sarcoidosis, leukemia, lymphoma
Intrinsic Airway Narrowing
Structural anomalies
Tracheobronchomalacia/stenosis
Bronchopulmonary dysplasia
α_1-Antitrypsin deficiency
Bronchospasm
Anaphylaxis
Organophosphate poisoning
Inflammation
Asthma
Bronchiolitis
Smoke inhalation
Bronchiectasis
Pulmonary hemosiderosis
Pulmonary aspiration
Gastroesophageal reflux
Swallowing disorders
Tracheoesophageal fistula
Intraluminal Airway Obstruction
Foreign body aspiration
Miscellaneous Causes
Congestive heart failure
Immune deficiency
Cystic fibrosis
Immotile cilia syndrome

Table 80.1. Causes of Wheezing in Childhood

DIFFERENTIAL DIAGNOSIS

Table 80.2 outlines the relative prevalence of those conditions associated with wheezing in children with which the emergency physician should be most familiar, and Table 80.3 lists the life-threatening causes of wheezing.

Disease Prevalence	<1 yr	>1 yr
Common	Bronchiolitis Asthma	Asthma
Less common	Pulmonary aspiration Gastroesophageal reflux Swallowing disorder Foreign body Bronchopulmonary dysplasia Cystic fibrosis	Foreign body aspiration Anaphylaxis
Uncommon	Congenital heart disease Defective host defenses Immune deficiency Immotile cilia syndrome Congenital structural anomalies Tracheobronchomalacia Vascular ring Lobar emphysema Cystic abnormalities Tracheoesophageal fistula	Defective host defenses Mediastinal tumors or enlarged lymph nodes Parasitic infection Pulmonary hemosiderosis α1-Antitrypsin deficiency

Table 80.2. Clinical Classification of Wheezing: Age at Diagnosis and Disease Prevalence

Asthma (severe)	Mediastinal tumor
Bronchiolitis (severe)	Congestive heart failure
Foreign body	

Table 80.3. Life-Threatening Causes of Wheezing

Most Common Conditions

Bronchiolitis is an acute viral infection of the lower respiratory tract caused predominantly by respiratory syncytial virus (RSV). Occurring in epidemics between late fall and early spring, this condition primarily affects infants 2 to 6 months of age but may occur in children as old as 2 to 3 years of age. Rhinorrhea and a low-grade fever typically accompany a prominent staccattolike cough and a variable degree of respiratory distress. The concurrence of respiratory symptoms in other family members is common.

Asthma is a chronic inflammatory disorder of the airways, characterized clinically by *recurrent* episodes of coughing and/or wheezing. Acute exacerbations of asthma are usually triggered by respiratory infections, allergens, and irritants such as cigarette smoke. Patients with asthma have a higher incidence of associated atopic disease, which includes allergic rhinitis and conjunctivitis and atopic dermatitis. Immediate family members are also more likely to be affected by asthma and atopic disease.

Less Common Conditions

Pulmonary aspiration is a less common cause of wheezing that occurs in several fairly characteristic clinical circumstances. Recurrent aspiration of oropharyngeal foodstuffs or gastric contents is usually seen in patients with severe developmental delay and a variety of neuromuscular diseases. Disordered swallowing and esophageal motility, and gastroesophageal reflux typically contribute in varying degrees to the recurrent aspiration that occurs in these patients. Repeated aspiration is also seen in children with structural anomalies of the laryngeal complex or an H-type tracheoesophageal fistula. Patients with chronic recurrent aspiration may develop wheezing and respiratory distress in the absence of a well-defined episode of choking or severe coughing because many such patients have depressed cough reflexes or experience “microaspiration.” Fever often accompanies pulmonary aspiration, reflecting associated chemical inflammation or infection of the tracheobronchial tree.

In otherwise healthy children, the abrupt onset of respiratory distress, associated with an episode of coughing, choking, or gagging, suggests the pulmonary inhalation of a foreign object (see Chapter 29). Foreign body aspiration is typically seen in toddlers, although older infants may aspirate solid food particles or small objects placed within their reach. The aspiration of a small object or food substance may be unwitnessed and go unrecognized for weeks or months until persistent lower respiratory symptoms trigger a search for an underlying cause. In these circumstances, persistent cough, wheezing, and sometimes recurrent fever is associated with an area of consolidation and/or collapse on radiograph; these symptoms fail to resolve despite seemingly appropriate medical therapy for presumed asthma and/or pneumonia.

Wheezing attributable to anaphylaxis is also of sudden onset and usually is accompanied by one or more other clinical findings that include urticaria, angioedema, stridor, and hypotension. When wheezing is the only finding, anaphylaxis may be suspected when the onset of respiratory difficulty is associated closely with Hymenoptera envenomation or food ingestion. Wheezing typically responds promptly to epinephrine administration or to bronchodilator therapy.

Infants and young children with a history of prematurity, oxygen therapy, and ventilatory support for a variety of conditions occurring in the newborn period may have wheezing caused by bronchopulmonary dysplasia (BPD). This condition is the childhood equivalent of chronic obstructive lung disease and represents a pathophysiologic continuum that includes varying degrees of structural damage and airway inflammation. Although gradual improvement in lung

function occurs during infancy and early childhood, bronchial hyperactivity and recurrent episodes of wheezing may persist until later in childhood. Cor pulmonale may exist in patients with severe pulmonary function impairment. Other coexisting problems associated with prematurity such as brain damage and esophageal reflux may complicate the respiratory pathophysiology in patients with BPD.

Uncommon Conditions

Cardiovascular abnormalities are one of many uncommon causes of wheezing in children. Small airway edema in the setting of congestive heart failure or airway impingement by enlarged cardiovascular structures are the usual pathophysiologic mechanisms. Most such cardiac conditions are associated with other abnormal physical findings, including cyanosis, murmurs, abnormal pulses, poor perfusion, or signs consistent with congestive heart failure. A congenital vascular ring may cause wheezing secondary to extrinsic airway compression. Abnormal cardiac physical findings are generally absent in patients with a vascular ring, although concomitant esophageal compression may result in dysphagia. A right-sided aortic arch is associated with this anomaly.

Children with various defects in host defense mechanisms often present with recurrent wheezing and bacterial pulmonary infections. In addition to respiratory tract involvement, patients with cystic fibrosis (see Chapter 96) will often exhibit steatorrhea and growth failure because of pancreatic insufficiency and malabsorption. Children with cell-mediated or humoral immune deficiency syndromes can have opportunistic infections or repeated extrapulmonary infections, including meningitis, otitis media, otitis externa, furunculosis, and mucocutaneous candidiasis. Similarly, patients with the immotile cilia syndrome develop repeated sinusitis and otitis media, often in association with situs inversus viscerum and bronchiectasis (Kartagener's syndrome).

Other uncommon causes of wheezing include extrinsic tracheobronchial compression by an enlarged lymph node or tumor (see Chapter 100). Mediastinal or hilar lymph node enlargement most often is the result of leukemia, lymphoma, sarcoidosis, or a mycobacterial or fungal infection. Mediastinal tumors most likely to produce pulmonary symptomatology include neuroblastoma, pheochromocytoma, ganglioneuroma, thymoma, and teratoma.

Congenital structural anomalies of the respiratory tract, including bronchogenic cysts, intrinsic stenosis, and webs, are among the rarest causes of wheezing in children. Respiratory symptoms typically begin in the neonatal period or early infancy. The predominant clinical features will be determined by the site of abnormality within the tracheobronchial tree. Stridor and a croupy cough are typical of laryngotracheal constriction, whereas wheezing and recurrent pneumonia are more characteristic of bronchial narrowing. Respiratory findings generally worsen with intercurrent respiratory infection and may accentuate with crying and activity.

EVALUATION AND DECISION

Initial Considerations

Table 80.4 presents a capsule summary of the clinical features associated with the more prevalent causes of wheezing in childhood.

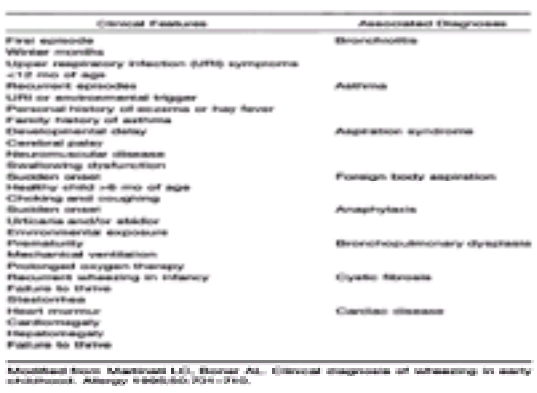

Table 80.4. Clinical Features and Associated Diagnoses

History

Thorough history taking is the key to arriving at an accurate diagnosis in a child with wheezing. In particular, consideration of the age at onset, course and pattern of illness, and associated clinical features, provides a useful framework for approaching a differential diagnosis (Fig. 80.1 and Fig. 80.2).

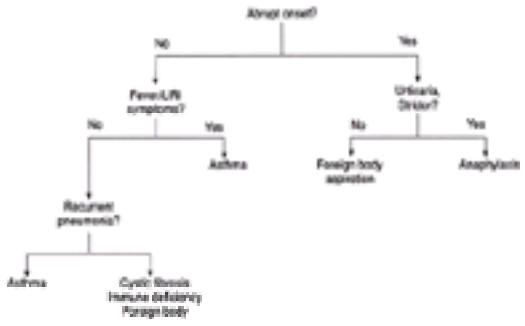

FIGURE 80.1. Approach to wheezing in children under 1 year of age. *URI,* upper respiratory infection.

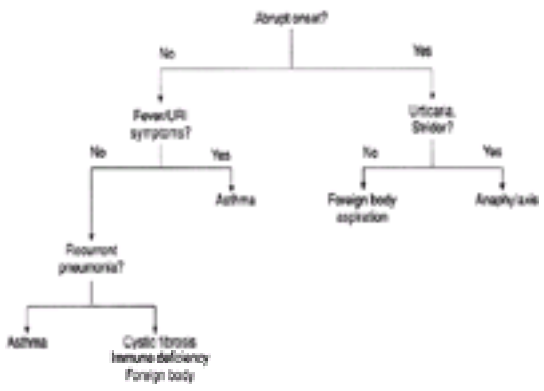

FIGURE 80.2. Approach to wheezing in children 1 year old or older. *URI,* upper respiratory infection.

The onset of wheezing in the neonatal period is associated with congenital structural airway anomalies, although a history of prematurity and mechanical ventilation is more suggestive of BPD. The *first episode* of wheezing in an otherwise healthy infant in association with cold symptoms indicates bronchiolitis. *Recurrent episodes* of wheezing precipitated by colds and a variety of other triggers is the hallmark of asthma. On the other hand, *recurrent wheezing beginning in infancy,* or *"difficult to control asthma,"* at any age should lead to a consideration of cystic fibrosis, gastroesophageal reflux, recurrent pulmonary aspiration or a retained airway foreign body, or immune deficiency. *Persistent wheezing* at any age suggests mechanical airway obstruction from a variety of causes, including congenital airway narrowing, pulmonary foreign body, and compression by a mediastinal tumor. The *sudden onset* of wheezing is characteristic of pulmonary aspiration or anaphylaxis.

As indicated previously, the diagnosis of a chronic wheezing disorder, such as asthma, relies on the identification of recurrent episodes of obstructive lower airway disease. Unfortunately, limitations of parental recall combined with the misdiagnosis of previous bronchospastic episodes as infectious illnesses sometimes make it a challenge to reconstruct an accurate history. In particular, subtle manifestations of asthma are often misinterpreted as episodes of bronchitis, pneumonia, or bronchiolitis. Accordingly, it is often useful to ask if the child has ever had any of these or other "breathing problems" or has ever been treated with a "breathing medicine."

Cough as a salient feature in patients with obstructive lower airway disease cannot be overemphasized. In fact, in many patients with asthma, recurrent cough may be the predominant presenting clinical feature and wheezing may be absent despite careful lung auscultation ("cough variant asthma"). Further inquiry might reveal that a patient usually experiences severe or persistent bouts of coughing in association with colds or that coughing is the cause of recurrent nighttime awakening.

As outlined in Table 80.4, the review of systems may provide additional clues to a variety of disorders. Similarly, the family medical history can contribute significant support for the diagnosis of a number of conditions, particularly asthma and rare inherited disorders such as cystic fibrosis.

Physical Examination

Wheezing must be distinguished from other causes of "noisy breathing" in children, which including the stridor of upper airway obstruction (see Chapter 72), the stertor of nasal congestion, and audible rhonchi. Because of the dynamic flexibility of airway structures, these clinical features of airway obstruction vary in accordance with the respiratory phase. Accordingly, upper airway collapse and stridor are worse on inspiration, whereas lower airway narrowing and wheezing are accentuated on expiration. Moreover, sounds originating in the upper airway passages (e.g., stridor, stertor) are transmitted with uniform quality and intensity across both lung fields. By contrast, wheezes tend to be polyphonic in pitch and distributed somewhat unevenly in intensity and location, despite the fact that most disorders causing lower airway obstruction affect both lungs simultaneously. This auscultatory asymmetry reflects the variation in airway narrowing that typically occurs from one lung segment to another. Conversely, wheezes consistently limited to a single lung field suggest a localized obstructive process, such as a foreign body or an extrinsic mass lesion.

The intensity of wheezes and their pitch and duration are a function of the degree of airway narrowing and the velocity of air flow at the site(s) of obstruction. In patients with minimal airway obstruction, wheezing may be difficult to detect. When such instances are suspected, forced exhalation may reveal low-pitched wheezes limited to the end of expiration. Subtle

wheezes can be accentuated further by combining forced exhalation with simultaneous manual compression applied by the examiner in the anteroposterior dimension of the chest (so-called squeezing the wheeze).

As airway narrowing and minute ventilation increase, wheezes becomes louder and higher pitched. However, as airway obstruction becomes progressively more severe, air flow and wheezes will diminish proportionately. A “quiet chest” in the face of significant respiratory distress may indicate respiratory failure. Conversely, in patients with reversible bronchospasm, air exchange and wheezes are often noted to increase in response to bronchodilator therapy.

As mentioned earlier, the clinical evaluation of a patient with obstructive lower airway disease will invariably reveal a prominent cough. To the experienced clinician or parent, this cough usually will be perceived as having a characteristic whistling or “wheezy” quality that is distinct from the “rhoncorous” cough of bronchitis or the “seal-like” cough of croup. Physical examination of the wheezing child may also reveal inspiratory and expiratory crackles, which are far more often attributable to subsegmental atelectasis than to an associated pneumonia and parenchymal consolidation.

Key “extrapulmonary” physical findings associated with the more prevalent causes of wheezing in childhood are included in Table 80.4.

Diagnostic Tests

Only a limited number of diagnostic modalities are needed to support the ED evaluation of the wheezing child. Many other necessary investigations can be performed as part of a subsequent inpatient or outpatient workup. The chest radiograph is the most important test obtained in the ED. Even so, its value is limited in helping establish a definitive diagnosis in a wheezing child. Rather, the chest radiograph assists in identifying disease complications such as pneumonia, atelectasis, pneumothorax, or pneumomediastinum. Among the few conditions that can be conclusively identified on the basis of plain film findings are heart disease, mediastinal masses, and some foreign bodies of the airway and esophagus.

Varying degrees of hyperaeration, bronchiolar thickening, and subsegmental atelectasis are the most common radiograph findings in patients with bronchiolitis or asthma. Pulmonary infiltrates may reflect the primary viral disease process in bronchiolitis or the complication of airway obstruction in asthma. When either disorder is suspected, a chest radiograph usually can be avoided if the patient is afebrile and little to no respiratory distress is present. The available data do not support the utility of obtaining a chest radiograph for all patients with their first episode of wheezing.

When bronchiolitis is a suspected cause of wheezing, occasionally it is helpful to identify respiratory syncytial virus in nasopharyngeal secretions by performing a rapid immunoassay while the patient is in the ED. The diagnosis of asthma can be supported by demonstrating improvement in peak flow measurements after bronchodilator treatment. Other patients with asthma may benefit from formal pulmonary function evaluation or bronchial challenge testing performed at a later time.

Patients suspected of having aspirated oropharyngeal or gastric contents should have plain radiographs taken of the chest. Nonspecific findings consistent with lower airway obstruction generally proceed the appearance of infiltrates. The location of the latter depends on the position of the patient at the time of the aspiration event. Patients thought to have recurring episodes of pulmonary aspiration subsequently should have further testing to identify swallowing dysfunction, gastroesophageal reflux, or actual tracheobronchial soiling. Such tests might include a barium esophagram, esophageal pH monitoring, esophageal endoscopy and biopsy, or radionuclide scintigraphy (“milk” scan). Fiberoptic bronchoscopy may be required to diagnose patients with a tracheoesophageal fistula.

An immediate and aggressive workup is always justified in patients suspected of having an airway foreign body (see Chapter 29) on the basis of acute and sudden symptomatology. In this setting, chest radiographs are usually normal, although occasionally they can demonstrate a radiopaque object, the faint outlines of a radiolucent foreign body, segmental atelectasis, or a focal area of hyperinflation. Patients with a persistent lower airway foreign body are more likely to show focal collapse and consolidation that is evident on standard chest roentgenograms. Bilateral decubitus views, inspiratory and expiratory radiographs, or airway fluoroscopy may be used to provide additional diagnostic information. Bronchoscopy is the procedure of choice both from a diagnostic and therapeutic perspective when a foreign body is strongly suspected.

The diagnosis of BPD is established on the basis of chronic respiratory symptoms superimposed on a background of neonatal lung disease. Nevertheless, a chest radiograph characteristically shows hyperexpansion and streaky or patchy infiltrates, punctuated by areas of alternating local hyperaeration and atelectasis.

Newborn screening now should identify most children with cystic fibrosis. Nevertheless, infants with recurrent wheezing and those with failure to thrive associated with chronic diarrhea should be referred for sweat chloride testing.

A patient suspected of having congenital or acquired heart disease should have an electrocardiogram (ECG) and a chest radiograph performed in the ED. Definitive diagnosis generally requires echocardiography or cardiac catheterization. A barium swallow is usually sufficient to diagnose the presence of a vascular ring, although angiography is necessary for exact anatomic definition.

Approach

The evaluation of a wheezing child begins with an immediate assessment of the degree of respiratory distress and consideration of the need for general supportive measures. Patients with suspected respiratory failure should be managed aggressively, as outlined in Chapter 5 and Chapter 68. Clinical features suggestive of respiratory failure include severe respiratory distress, agitation or lethargy, dusky mucous membranes, signs of autonomic excess (tachycardia, diaphoresis, peripheral vasoconstriction), poor air movement on lung auscultation, and oxyhemoglobin

saturation of less than 90%. Blood gas analysis will aid in the determination of respiratory failure.

Supplemental oxygen should be offered promptly to any patient with respiratory distress and adjusted to maintain a hemoglobin saturation of 95% or greater. Patients suspected of having reversible bronchospasm should be given a bronchodilator, such as albuterol, by inhalation while further evaluation and management is proceeding according to the priorities established earlier in this chapter.

Expeditious management is essential in patients with poor baseline pulmonary functions because they can develop respiratory failure quickly. Such patients include children with significant BPD and advanced cases of progressive chronic lung disorders such as cystic fibrosis. Moreover, careful titration of inspired oxygen concentration is important in patients with chronic respiratory insufficiency to avoid respiratory drive suppression.

Infant Less Than 1 Year Old

An algorithm for elucidating the cause of wheezing in the child less than 1 year old is presented in Figure 80.1. The abrupt onset of wheezing, often immediately preceded by an episode of choking, gagging, or vomiting, is highly suggestive of pulmonary aspiration; foreign body aspiration should be considered in the previously healthy older infant, whereas gastroesophageal reflux with aspiration of gastric contents is a more likely cause in early infancy. However, most young infants who present with a first episode of wheezing have bronchiolitis. In this illness, fever and upper respiratory symptoms usually accompany wheezing and respiratory distress during the winter months. A similar complex of physical findings in an older infant with a history of bronchiolitis or wheezing and clear improvement after bronchodilator administration is characteristic of asthma.

The remaining disorders often are found in infants who have overt evidence of chronic or severe underlying illness and who typically present with recurrent or persistent episodes of wheezing and respiratory distress. Neurologic disability predisposes a child to gastroesophageal reflux or a swallowing disorder and recurrent pulmonary aspiration. A report of prematurity and/or respiratory difficulty at birth may be a clue to BPD. Recurrent pneumonia, failure to thrive, and steatorrhea are characteristic of infants with cystic fibrosis, whereas pneumonia in association with repeated extrapulmonary infection is suggestive of an immune deficiency. A heart murmur and other clinical findings consistent with congestive heart failure are indicative of congenital heart disease and pulmonary edema. Wheezing accompanied by stridor commonly indicates the coexistence of viral croup but may reflect intrinsic congenital airway narrowing, such as tracheobronchomalacia or extrinsic compression by a mediastinal structure. In the absence of any of the clinical clues listed, the first episode of wheezing in an otherwise healthy child, especially when it occurs during the winter months, is likely to represent bronchiolitis.

Child 1 Year Old or Older

Figure 80.2 outlines an algorithmic approach to the more common causes of wheezing in the child who is 1 year old or older. The sudden onset of respiratory distress and wheezing associated with an episode of choking and coughing is likely to indicate foreign body aspiration, particularly in a toddler who has been eating or playing with a small object. An abrupt onset of wheezing also may accompany stridor, urticaria, and hypotension in the older child with an anaphylactic reaction. However, most first or recurrent episodes of wheezing represent asthma. Typically, such episodes are precipitated by a concurrent upper respiratory infection, and the patient may show responsiveness to bronchodilator administration.

Wheezing and recurrent pneumonia in multiple pulmonary segments are characteristic of patients with defects in host defense mechanisms, such as cystic fibrosis, an immune deficiency syndrome, or the immotile cilia syndrome. Children in this age group who present with these disorders usually have a history of lower respiratory illness that began in infancy, as well as other signs and symptoms suggestive of chronic disease. Repeated pneumonia in the same pulmonary segment in an otherwise healthy child that begins in late infancy or in early childhood is likely to represent a previously unrecognized bronchial foreign body. In the absence of any of the clinical clues previously listed, the first episode of wheezing in an otherwise healthy child is likely to represent asthma.

It is imperative that *all* patients with wheezing receive outpatient follow-up with their primary care provider or, in some instances, with a specialist. With few exceptions, follow-up evaluation should take place within a few days to a week of the ED visit.

Suggested Readings

Chernick V, Kendig EL, eds. Kendig's Disorders of the Respiratory Tract in Children. 5th ed. Philadelphia: WB Saunders, 1990.

Edwards DK. The child who wheezes. In: Hilton SW, Edwards DK, Hilton JW, eds. Practical Pediatric Radiology. 2nd ed. Philadelphia: WB Saunders, 1994.

Felman AH. The Pediatric Chest. Springfield, IL: Charles C Thomas, 1983.

Fireman P. The wheezing infant. Pediatr Rev 1986;7:247–254.

Martinati LC, Boner AL. Clinical diagnosis of wheezing in early childhood. Allergy 1995;50:701–710.

Skoner D, Caliguiri L. The wheezing infant. Pediatr Clin North Am 1988;35:1011–1030.

CHAPTER 81

Weight Loss

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Weight loss occasionally prompts a visit to the emergency department (ED). More commonly, it is an important physical examination finding in a patient presenting with another complaint. Acute weight loss is most commonly caused by a negative fluid balance occurring in the face of illness and can be life-threatening. These problems have special importance to the emergency physician. Chronic weight loss may result from a number of medical and nonmedical causes leading to inadequate nutrition. Any complaint of weight loss, or documented weight loss, is a significant finding that demands careful evaluation and follow-up.

PATHOPHYSIOLOGY

The health of infants and children depends on a balanced intake of fluid and nutrients that serve as building blocks for new tissue. The major determinants of body weight are water and the organic fuels, carbohydrates, protein, and fat. Weight loss occurs when the daily balance of one of these becomes negative (Fig. 81.1). Overall, during childhood, the major cause of weight loss is protein-energy intake inadequate to meet the energy demands of cell metabolism and tissue synthesis. Causes include decreased calorie intake, normal calorie intake with increased metabolic requirement, and normal calorie intake in the face of malabsorption or impaired use. During acute illness, fluid loss in excess of intake, in the presence of protein-calorie malnutrition, is the most common cause. Water losses occur primarily through the gastrointestinal (GI) tract but also through the urine and skin. Fever, infection, trauma, and thermal injury all cause a dramatic increase in metabolism that is rarely balanced with intake. During chronic illness, a cyclic pattern of adequate intake alternating with starvation results in gradual weight loss over time.

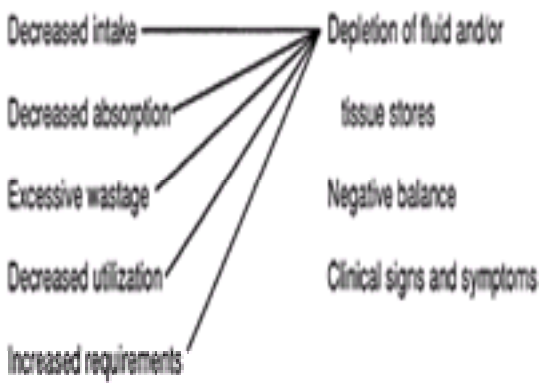

FIGURE 81.1. Pathophysiology.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of weight loss is extensive and can be thought of in major categories, including inadequate fluid and/or caloric intake, decreased absorption, excessive wastage, and increased catabolism or abnormal caloric use (Table 81.1). The single most common cause of acute weight loss in all age groups is dehydration that occurs in conjunction with an acute infectious illness. In the infant and toddler with chronic weight loss or lack of appropriate weight gain, failure to thrive (FTT) or undernutrition is usually identified in the first 3 years of life and is estimated to occur in 10% of children. This is a complex disorder resulting from physical and/or psychosocial problems. In the older patient with chronic weight loss, an underlying medical or psychiatric illness is more likely than in the infant (Table 81.2). In some instances, the exact diagnosis is not made at the time of the ED visit, but a workup may be initiated and an appropriate

referral made.

Table 81.1. Differential Diagnosis of Weight Loss

	Infants	Older Children/Adolescents
Acute	Gastroenteritis	Gastroenteritis
	Acute infectious illness	Acute infectious illness
	Pyloric stenosis	
	Gastroesophageal reflux	
Chronic	Failure to thrive	Inflammatory bowel disease
		Eating disorders
		Affective disorders

Table 81.2. Common Causes of Weight Loss

A few life-threatening diseases associated with weight loss must be separated out from conditions that carry no immediate risk (Table 81.3). Severe dehydration in the presence of gastroenteritis or other acute illness may be catastrophic in any age group (see Chapter 18). In young infants, several disease states need to be considered. The salt-losing form of congenital adrenal hyperplasia (CAH) presents with anorexia, vomiting, dehydration, and progressive weight loss. In female infants, virilization of the external genitalia provides a clue to the diagnosis; however, this is lacking in male infants. The characteristic electrolyte abnormality of hyponatremia and hyperkalemia and the rapidity of onset of the patient's illness support the diagnosis of CAH (see Chapter 97). Inborn errors of metabolism cause a wide variety of symptoms, but poor feeding, anorexia, vomiting, weight loss, and lethargy are always present (see Chapter 98).

Infants	Older Children/Adolescents
Gastroenteritis, secondary dehydration	Gastroenteritis, secondary dehydration
Inborn errors of metabolism	Diabetes mellitus
Congenital adrenal hyperplasia	Addison's disease
Congenital immune deficiencies	
Acquired immunodeficiency	
Congenital heart disease	
Pyloric stenosis	

Table 81.3. Potential Life-Threatening Causes of Weight Loss

Those presenting in the first weeks of life are severe and fatal if the correct diagnosis is not made. These disorders may masquerade as (or be complicated by) sepsis, hypoglycemia, hypocalcemia, or GI obstruction, but an inborn error must always be considered in neonates presenting with weight loss. Clinical deterioration in a previously normal baby, history of previous fetal death, or consanguinity increases suspicion. Infants and young children with congenital immune deficiency syndromes have a significant component of weight loss and wasting, in addition to repeated infections, seborrheic dermatitis, alopecia, chronic diarrhea, and hyperplastic joints. Infants with acquired immunodeficiency often lose weight before diagnosis. Both groups of children are at risk for life-threatening infections. Congenital heart diseases and pulmonary diseases are also associated with poor growth and may require acute intervention.

In older children, a few diseases associated with weight loss are acutely life-threatening and include adrenal crisis, diabetes mellitus with ketoacidosis, and severe dehydration. Older children and adolescents with Addison's disease have gradual onset of fatigue and weakness, anorexia, weight loss, and low blood pressure. If the diagnosis is not made, adrenal crisis may supervene, leading to circulatory failure, which may be rapidly fatal. Evidence of hyperpigmentation (particularly around the genitalia, nipples, axilla, and umbilicus) provides a clue to the diagnosis. Ketoacidosis is the initial manifestation in many children with diabetes; these children often give a history of weight loss in the presence of

polyphagia, polydipsia, and polyuria.

EVALUATION AND DECISION

General Approach

Consideration of the child's age and the severity and duration of the weight loss along with the presence of other systemic symptoms and specific physical examination findings help narrow the extensive differential diagnosis. Many diagnoses are exclusive to specific age groups. For the emergency physician, severity is an important consideration because sudden losses are more suggestive of life-threatening disorders that require prompt recognition and treatment.

Acute weight loss (occurring in less than 2 weeks) is most often caused by anorexia, poor fluid and calorie intake, increased losses, and increased metabolic need in association with an intercurrent illness. Weight loss is a sensitive indicator for dehydration and commonly occurs in the presence of any significant febrile illness. In this setting, the history includes an estimation of intake, losses, and increased need for fluids and calories. The types of losses pinpoint the location of GI pathology. The presence of other symptoms consistent with an acute infectious process substantiates the cause. Circulatory compromise suggests severe depletion (see Chapter 3 and Chapter 18) or adrenal crisis (see Chapter 97).

Chronic weight loss (occurring over more than 2 weeks) results from a combination of factors, including anorexia, poor utilization or malabsorption, and increased requirements, as well as health consequences imposed by the underlying disease state. When considering the cause of chronic weight loss, broad categories exist, including loss 1) secondary to a medical cause (underlying infection, absorptive defect, inflammatory or neoplastic disease); 2) related to a psychosocial or psychiatric cause; or 3) resulting as a consequence of both problems. A complete review of systems, in search of fever, night sweats, arthritis, abdominal pain and/or diarrhea, dermatitis, and other constitutional symptoms, helps the physician reach the diagnosis. A detailed dietary and feeding history (including frequency, types, and amounts of foods ingested) is invaluable. The attitude of the child and family toward food and eating habits should be explored. An estimation of caloric intake is attempted from a record of intake the day preceding the interview. Formula preparation and juice consumption are important historical pieces for babies. Gross overestimation of intake often indicates a psychosocial cause for the poor growth because of an inexperienced parent, poor parental–child interaction, and/or multiple caretakers. A search for a cause of family stress or dysfunction, economic problems, and available resources is essential. The presence or absence of symptoms of depression (poor school performance, disturbed sleep, loss of appetite, and apathy) is also important.

A complete and careful physical examination with attention to vital signs, state of hydration, and findings suggestive of specific disease states (e.g., murmur and/or cyanosis, lymphadenopathy, dermatitis, hyperpigmentation, abdominal mass and/or tenderness, arthritis, neurologic abnormalities) provides useful clues. Nutritional inspection includes an evaluation of body fat, muscle mass, hair, skin, and nails. Physical signs associated with specific vitamin deficiencies are nonspecific and occur late in the course of malnutrition. Dysmorphic features should be noted and a thorough neurologic examination performed. Infants should be observed nursing or being bottle fed, with attention to any gagging, choking, reflux, or respiratory distress.

Measurements of weight, recumbent length in babies younger than 2 years, standing height in children older than 2 years, and head circumferences are necessary components of the physical examination. Normal growth is present when sequential measurements consistently lie within the 5th to 95th percentile on standard growth charts from the National Center for Health Statistics. Values on standard growth curves for children 0 to 36 months are obtained from recumbent length measurements; standing height measurements may be as much as 2 cm shorter. Use of growth charts to evaluate the child's weight and height relative to each other and previous values is important. In infants and toddlers with proportionately low measurements of all parameters, head circumference, weight and height, congenital defects ranging from metabolic/genetic disorders to prenatal or perinatal asphyxia represent the differential diagnosis. This group of children may never achieve normal growth even in the face of nutritional interventions. Infants/toddlers with normal head circumference and mild or proportionate weight reduction to height include those with constitutional growth delay, genetic dwarfism, or endocrine disorders. The final category represents the majority of infants and toddlers. Children in this group have normal head circumference and low weight out of proportion to height. Inadequate nutrition is the cause. Anthropometric measurements may be helpful in sorting out adequacy of growth of children consistently less than 5% after the ED evaluation. Looking at growth parameters over time is extremely helpful, although usually unavailable in the ED; a consistent fall in a downward direction requires a diligent search for an underlying chronic illness.

The severity of malnutrition can be defined further by using the actual weight expressed as a percentage of the ideal weight for the patient's actual height. Mild protein-energy malnutrition exists when the actual weight is 85 to 90% of the ideal body weight for actual height, moderate when this is 75 to 85%, and categorized as severe when this falls below 75%. The degree of malnutrition is important when considering the refeeding regimen and decision making regarding the patient's disposition.

Growth curves have been created for special groups of children who exhibit different growth patterns than the general population. The growth of premature infants can be evaluated based on their corrected age or on special growth graphs created by Babson. The premature infant normally attains “catch-up” growth during the first 2 years of life (after which normal growth curves can be used). Another standard growth curve has been created for the evaluation of growth during the adolescent age that includes the patient's sexual maturity rating. These graphs account for the variability in the timing of the adolescent growth spurt, and deviation from the standard growth curves should be interpreted with caution. No routine screening panel of laboratory tests is indicated in patients with undernutrition. Rather, they should be done as indicated by the history and physical examination. Considerations include complete blood count, erythrocyte protoporphyrin or ferritin, electrolytes, glucose, blood urea nitrogen (BUN), creatinine, serum protein profile, urinalysis, and stool examination (Table 81.4).

Primary	Secondary
Complete blood count	Bone age, chest radiograph
Erythrocyte protoporphirin (or ferritin)	Thyroid function tests
	Immunologic studies
Serum glucose, electrolytes	Chromosomes
BUN, creatinine	Urine for ketones/reducing substances
Serum proteins	Plasma ammonia
Urinalysis, urine culture	Plasma lactate
Stool hemoccult, clinitest	Serum/urine amino acids
Calcium, phosphate	Urine organic acids
Liver function tests	EEG/head imaging
Sedimentation rate	Sweat test
Tuberculin test	Lactose breath test
	Stool for trypsin
	Stool for fat
	B_{12}
	UGI/colonoscopy

EEG, electroencephalogram; UGI, upper gastrointestinal radiographic study.

Table 81.4. Possible Laboratory Test

Infants

Infants should regain their birth weight by 10 to 14 days of age. Average daily weight gain in the first 6 months of life is 20 g and decreases to 15 g in the second half of the first year. During the first 2 months of life, head circumference increases by 0.5 cm per week, and from 2 to 6 months by 0.25 cm per week. When evaluating the infant with weight loss or inadequate weight gain, the physician should include in the history perinatal events and the onset and character of the symptoms. The presence of vomiting and acute weight loss in an otherwise well baby with a good appetite suggests gastroesophageal reflux or pyloric stenosis (Fig. 81.2). Poor sucking or swallowing and delayed development indicates neurologic or neuromuscular disease. Vomiting and anorexia, altered mental status, seizures, and characteristic body fluid odors (see Chapter 98) point to metabolic disease. Renal insufficiency or tubular disease or liver disease also may cause anorexia and vomiting and thus poor growth. Frequent infections, dermatitis, and diarrhea are associated with immune deficiency syndromes. The infant who tires or becomes diaphoretic with feedings may have congenital heart or pulmonary disease. The presence of malodorous, loose stools suggests primary malabsorption or cystic fibrosis. Blood-streaked, water-loss stools may be related to milk protein allergy or infectious (especially bacterial) enteritis.

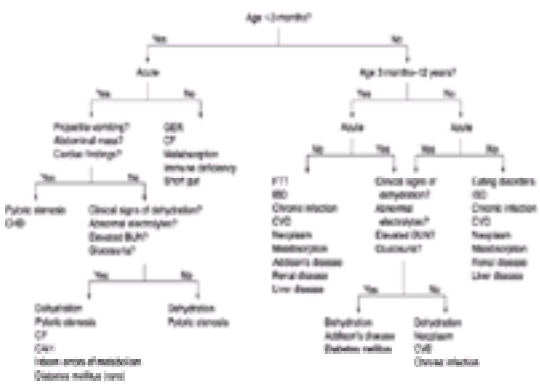

FIGURE 81.2. An approach to the evaluation of weight loss. *BUN,* blood urea nitrogen; *CHD,* congenital heart disease; *CF,* cystic fibrosis; *CAH,* congenital adrenal hyperplasia; *CVD,* collagen vascular disease; *IBD,* inflammatory bowel disease; *FTT,* failure to thrive; *GER,* gastroesophageal reflux.

The physical examination includes measurement of length, weight, and head circumference. When all three parameters are abnormal, a neurologic, genetic, or metabolic cause is suspect. When length and weight are subnormal but proportional, skeletal dysplasias, endocrinopathies, or constitutional short stature are likely. When only weight is below normal, an acute illness, dehydration, or deprivation is probably the culprit; the infant should be evaluated for signs of dehydration, cardiac or pulmonary disease, and neurologic abnormalities. As noted previously, observation of a feeding infant may offer some relevant clues. The evaluation of an infant with suspected failure to thrive as a result of child abuse is discussed in Chapter 128.

Older Children

In approaching children older than 3 to 5 years, the same principles are used (Fig. 81.2). Average weight gain until puberty is 2 kg per year and average growth is 5 cm (2 inches) per year. A careful history, physical examination, and growth assessment identify the child with growth failure that requires further investigation. The most common causes of chronic weight loss in this age group are diabetes mellitus (DM) and inflammatory bowel disease (IBD). Therefore, important historical points include a history of increased appetite, polyuria, and polydipsia. With regard to IBD, colicky abdominal pain, diarrhea, and other symptoms such as arthritis or rash are pertinent. However, weight loss may be the sole manifestation. In this age group, mental health disorders are an important consideration. The incidence of diagnosed depression in children continues to increase because of both improved recognition and a rising incidence in our society.

Adolescents

Growth failure in adolescents may be the harbinger of chronic illness. The history and physical examination again help in uncovering the pathology. Consideration of underlying inflammatory, chronic infectious, or neoplastic disease is

important (Fig. 81.2).

Eating disorders often emerge during adolescence. As in FTT, the diagnosis is made with a careful dietary history and physical examination. These patients exhibit an intense fear of fatness, a relentless pursuit for thinness, a preoccupation with food, and a distorted body image. A limited laboratory evaluation that excludes diseases that may mimic anorexia and bulimia is indicated (see Chapter 130). In cases complicated by severe malnutrition (weight loss over 20% of ideal body weight), metabolic derangements, dehydration, or acute psychosis, patients with eating disorders are cared for initially in the hospital.

Laboratory Tests

The potential list of tests to aid in the evaluation of weight loss is extensive (Table 81.4). Decisions to obtain diagnostic tests should be made individually after a thorough history and examination is completed. A complete blood count serves many purposes and may uncover macrocytosis caused by hypothyroidism or malabsorption of folate or B_{12}; microcytosis caused by iron deficiency, chronic blood loss, or chronic infection; polycythemia related to chronic heart or lung disease; neutropenia indicative of Schwachmann syndrome; elevated neutrophil count secondary to infection; abnormal cell counts or morphologies caused by underlying neoplasm; or thrombocytosis caused by chronic infection or underlying malignancy. Electrolytes may confirm the presence of significant dehydration, and hyponatremia in the presence of hyperkalemia suggests the diagnosis of adrenal insufficiency. Alterations in serum proteins may reflect aberrant absorption, decreased synthesis, or chronic infection. The urinalysis evaluates tubular function and may reveal glucosuria or disaccharide intolerance or galactosemia, as well as infection and renal tubular acidosis. The stool should be checked for the presence of blood, infectious agents, and reducing substances. With the results from this battery of tests, in conjunction with the physical examination and history, a diagnosis or appropriate referral can be made.

SUMMARY

Weight loss is a complaint that requires careful evaluation. With an acute episode of weight loss, many patients seen in the ED will have fluid loss and mild protein energy malnutrition related to an intercurrent illness with anorexia, increased metabolic need, or increased losses. Most of these children will return to baseline spontaneously when their illness resolves. Chronic weight loss or growth failure is indicative of less common diseases, and those for which the differential diagnosis is extensive. In small children, the most common cause is psychosocial growth failure. In older children and adolescents, an organic cause becomes more likely.

Disposition

As always, the general appearance of the infant or child determines the timing and the scope of the evaluation. Hospitalization is indicated in any child who is suspected to have sustained trauma or been abused and may be indicated in those with physical findings consistent with severe malnutrition such as weight 60% less than ideal body weight, hypothermia, bradycardia, or hypotension. Children with mild to moderate protein calorie malnutrition can be referred back to their pediatrician for outpatient management. Nutrition is a central component of well-being in the growing child, and malnutrition carries significant morbidity and mortality. Thus, persons who care for children need to have a sense of normal growth and must develop an approach for the evaluation and treatment of growth failure.

Suggested Readings

Avery M, First L, eds. Pediatric Medicine. Baltimore: Williams & Wilkins, 1989.

Gahagan S, Holmes R. Systemic approach to evaluation of undernutrition and FTT. Pediatr Clin North Am 1998;(45):169–187.

Hubbard V. Clinical assessment of nutritional status. In: Walker WA, Watkins JB, eds. Nutrition in Pediatrics. Boston: Little, Brown, 1985.

Illingworth RS. Common Symptoms of Disease in Children. 7th ed. Oxford: Blackwell Scientific, 1982.

Kien LC. Failure to thrive. In: Walker WA, Watkins JB, eds. Nutrition in Pediatrics. Boston: Little, Brown, 1985.

Kreipe ER. Eating disorders among children and adolescents. Pediatr Clin North Am 1998;45:169–188.

Shils ME, Young VR, eds. Modern Nutrition in Health and Disease. 7th ed. Philadelphia: Lea & Febiger, 1988.

Sills R. Failure to thrive. In: Stockman J, ed. Difficult Diagnosis in Pediatrics. Philadelphia: WB Saunders, 1990.

Tunnessen W. Signs and Symptoms in Pediatrics. 2nd ed. Philadelphia: JB Lippincott, 1988.

Wright CM, Talbot E. Screening for FTT what are we looking for? Child Care Health Dev 1996;22:223–233.

CHAPTER 82

Cardiac Emergencies

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Cardiac problems in infancy and childhood are not rare, often are complex, and always have important implications for the general health of the child. This chapter provides information regarding the evaluation and management of the more common emergencies that arise from the presence of cardiovascular disease in children. For the health care professional not well-versed in childhood cardiac disease, a brief overview is provided as background for the specific problems discussed in detail.

OVERVIEW

As with other pediatric disorders, cardiac diseases in childhood can be congenital or acquired. Major and minor structural diseases of the heart for the most part result from derangements of embryologic development and thus are present in some form at birth. However, the clinical manifestations of such problems may be delayed for days, months, or even years. In contrast, many other disorders of cardiac physiology result from problems superimposed on inherently normal cardiac structures. These problems often are acute in their development when a relatively sudden change from normal cardiac physiology occurs, leading quickly to the development of symptoms. It should be remembered, however, that acquired cardiac diseases also can be associated with prolonged latent periods. For example, the most prevalent form of acquired heart disease in the United States, coronary artery disease, probably has its beginnings in childhood in numerous instances, although its appearance as a clinical entity occurs much later. Examples of forms of congenital and acquired pediatric cardiac problems are reviewed in Table 82.1.

Congenital	**Example**
Disorders of septal or valvar development	Pulmonary, aortic valve stenosis Mitral or tricuspid atresia Ventricular septal defect Atrial septal defect
Disorders of venous or arterial connection	Transposition of the great vessels Anomalies of pulmonary venous return
Disorders of conduction system development	Congenital AV block Persistent bypass tract syndromes (preexcitation)
Acquired	**Example**
Disorders of cardiac muscular function	Cardiomyopathy associated with cancer therapies Acute myocarditis Acute pericarditis
Disorders of valvar function	Acute rheumatic fever Bacterial endocarditis
Disorders of cardiac rhythm	Drug-induced arrhythmias Postsurgical heart block

AV, atrioventricular.

Table 82.1. Examples of Acquired and Congenital Forms of Pediatric Heart Disease

The incidence rate of congenital heart disease has been fairly constant over the past few decades and is estimated to be approximately 8 to 10 cases per 1000 live births (0.8 to 1%). This includes all forms of defects and all ranges of severity but does not include the most common isolated congenital cardiac lesion, bicuspid aortic valve, which occurs in an additional 1 to 2% of people. The prevalence figures for congenital heart disease (CHD) are continually increasing, however, as more and better treatment modalities become available to sustain critically ill patients and because many of these patients are beginning to have offspring of their own. Recently, studies have found the incidence of congenital cardiac problems in children of parents with CHD to range from 5 to 15%. (The disorders seen as part of the spectrum of CHD are extremely variable; comprehensive texts are available for more detailed reviews.)

Age at presentation can vary significantly, depending on the type of congenital cardiac lesion and its severity in terms of effect on cardiac performance. Not all congenital heart defects are clinically apparent at birth. Severe obstruction to pulmonary or systemic flows may be masked in the first few days of life by persistence of a patent ductus arteriosus, whereas the presence of an important ventricular septal defect may not become evident until 4 to 6 weeks of age. This wide disparity in age at presentation is related to the physiologic interactions of the systemic and pulmonary vasculature, as well as to the anatomic specifics of any particular lesion. Table 82.2 presents examples of typical presenting ages for the more common forms of CHD. Although it is not important for the emergency practitioner to be versed specifically in all possible defects, knowledge of the anatomic and physiologic possibilities is important so that the practitioner's awareness of potential problems can lead to appropriate triage and initiation of care. One useful approach is to categorize lesions by the presence or absence of arterial desaturation; thus, there are cyanotic and acyanotic forms of CHD. Table 82.3 reviews some of the congenital lesions that segregate into these two groups.

First 2 Weeks	*6 Weeks to 6 Months*
Left ventricular outflow obstruction	VSD
Coarctation of aorta	AV canal malformations
Severe aortic stenosis	Coronary artery anomalies
Left heart hypoplasia	Truncus arteriosus
Cyanotic lesions	*Over 6 Months*
Transposition of great vessels	VSD
Total anomalous pulmonary venous return	Atrial septal defect
Atrioventricular (AV) canal malformations	Isolated valvar lesions (e.g., pulmonic stenosis, mitral insufficiency)
Truncus arteriosus	Small PDA
First Month	Partial anomalous pulmonary venous return
Coarctation of aorta	Coarctation of aorta
Ventricular septal defect (VSD)	
Patent ductus arteriosus (PDA)	
Truncus arteriosus	
Complex lesions with multiple anomalies (e.g., double-outlet right ventricle)	

[a]There is considerable overlap regarding any particular lesion and the specific clinical setting. This list is representative and illustrative, not all-inclusive.

Table 82.2. Age of Presentation of Congenital Heart Disease[a]

Acyanotic Lesions	*Cyanotic Lesions*
Secundum atrial septal defect	Tetralogy of Fallot
Ventricular septal defect	d-Transposition of the great vessels
Patent ductus arteriosus	Tricuspid atresia variants
Aortic stenosis/regurgitation	Total anomalous pulmonary venous return
Coarctation of the aorta	Truncus arteriosus
Pulmonic stenosis—valvar	Pulmonary atrioventricular fistula
Peripheral pulmonary stenosis	Complete atrioventricular canal defect
Mitral stenosis/regurgitation	Ebstein's anomaly of the tricuspid valve
Partial anomalous pulmonary venous return	Pulmonary atresia with septal defect
Congenitally corrected transposition of the great vessels	Single ventricle states

Table 82.3. Exemplitive Congenital Heart Lesions

Even without recalling specific lesions, however, only a few key principles of cyanosis must be understood (Table 82.4) to be able to consider the possibilities when faced with a particular patient. For example, a patient with anemia may have cyanotic CHD yet not be visibly desaturated; or a patient may have a significant level of pulmonary valve stenosis with significant obstruction to pulmonary blood flow but not appear cyanotic. Conversely, a patient may not have a loud cardiac murmur or any murmur at all and still have a serious cyanotic cardiac lesion (see Chapter 33).

In the presence of normal hemoglobin moieties and normal cardiac output, right-to-left shunting must be present to produce cyanosis; this can be intrapulmonary, intracardiac, or both.
Obstruction to pulmonary blood flow alone does not produce cyanosis.
A critical mass of reduced hemoglobin must be present to allow visual estimation of cyanosis.
Right-to-left shunts are not usually associated with heart murmurs.
The presence of visible cyanosis depends on the interrelationship of pulmonary and systemic blood flows and hemoglobin concentration.

Table 82.4. Useful Rules of Cyanotic Congenital Heart Disease

In diagnosing children with possible CHD, therefore, it is usually not necessary to remember long lists of complex lesions. An appreciation of what is anatomically possible and what is physiologically rational, however, usually leads to the ability to discern what is clinically likely in any particular situation.

The next sections deal with specific examples of the melding of these anatomic and physiologic concerns in emergency

situations that involve cardiovascular diseases in children.

CONGESTIVE HEART FAILURE

Background

Heart failure is best described as a syndrome in which the heart cannot maintain a level of tissue perfusion adequate to meet metabolic needs. During childhood, these needs also include growth and development. This section offers an outline of the primary etiologic and physiologic factors that underlie the clinical presentation of the child in congestive heart failure (CHF) and that influence management decisions.

Etiologic Considerations

Although the primary cause of CHF in infants and children is CHD, a panoply of conditions can be associated with the presentation of CHF in the presence of normal underlying cardiac structure. Table 82.5 lists the more common clinical entities associated with CHF, including primary cardiac disease and conditions in which the heart is affected secondarily. In general, the principal physiologic problems that lead to impaired myocardial performance include 1) excessive pressure loads such as with left-sided heart obstructions; 2) excessive volume loads, such as with large left-to-right shunts, valvar regurgitation, and severe anemia; 3) primary inotropic depression such as with myocarditis, endocrinologic disorders, or coronary perfusion irregularities; and 4) rhythm abnormalities such as supraventricular tachycardia (SVT) or severe forms of heart block.

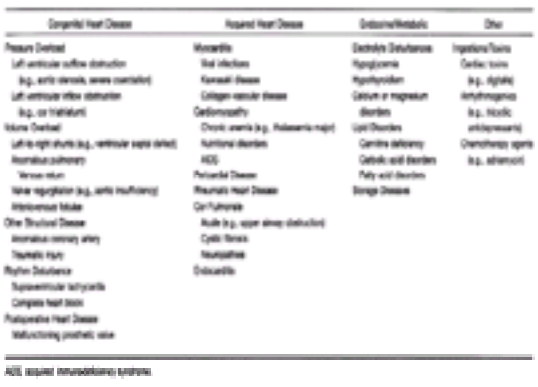


Table 82.5. Etiologic Considerations for Congestive Heart Failure

The history is critical in determining the cause of CHF and should not be glossed over in the rush to treat. Knowledge of preexisting cardiac disease obviously is important. A history of known hematologic disorders such as thalassemia or sickle cell anemia also should be sought. Because heart failure can develop as a consequence of pressure overload of the right side of the heart secondary to pulmonary vasoconstriction and hypoxia, a history of respiratory tract difficulties or breathing pattern irregularities also should be reviewed carefully. A knowledge of other systemic conditions is likewise crucial. For example, several recent studies have indicated that human immunodeficiency virus (HIV) infection can affect cardiac performance seriously on an acute and chronic basis. Late stages of HIV infection often are complicated by cardiomyopathy and CHF. Careful clinical assessment that looks for signs of cardiac involvement should be part of the evaluation of children with HIV who present with hypotension or who have respiratory problems not responsive to direct pulmonary management. As another example, a history for Kawasaki disease should be sought in patients with new-onset CHF that appears to be related to inflammatory myocardial disease, even if other classic signs are not overtly present.

In the presence of appropriate physical findings and historical information, the diagnosis of CHF usually is evident in the older child. The principal problem of diagnosis centers on the infant, in whom differentiation between CHF and primary respiratory tract disease can be difficult. Auscultation of cardiac murmurs is helpful, of course, but such murmurs may not always be audible, particularly in severe failure with low output. Parenchymal lung disease may result in systemic desaturation to the same degree as CHF with associated pulmonary congestion and ventilation-perfusion imbalance. Palpation of the liver edge below the costal margin in an infant may be related to hyperexpansion of the lungs and not to systemic venous congestion. Conversely, respiratory tract signs such as wheezing and retractions may be part of the clinical picture of heart failure in the absence of primary lung disease. The chest radiograph often fails to distinguish clearly between cardiac and pulmonary disease because pulmonary markings often mimic infiltrative patterns. Generally, however, evidence of cardiac enlargement on the radiograph is a useful differential point. Other noninvasive methods, such as echocardiography, also can help establish the diagnosis of cardiac disease.

Pathophysiology

There are four primary determinants of normal cardiac function, each of which may relate to the development of heart failure. The first is preload, the volume at end-diastole that must be ejected by the left ventricle, that closely reflects the intravascular volume status of the child in general, and that directly affects cardiac performance through the Frank-Starling relationship. The second, afterload, is the opposing force to ventricular ejection and relates to the tension that must be developed by the myocardium to eject a given preload. The third determinant, contractility, can be viewed as an intrinsic property of cardiac muscle that permits alterations in cardiac shape necessary for ejection and is determined by fundamental properties of cardiac ultrastructure. The fourth and last determinant, heart rate (HR), is related to intrinsic electrophysiologic capabilities of the specialized cardiac conduction system and to supervening neurologic input. It is

directly related to cardiac output (CO) through the classic relationship:

$$\mathrm{CO} = \mathrm{HR} \times \mathrm{SV}\ (\textbf{stroke volume})$$

Compensatory Responses

To understand the basis for the clinical findings commonly associated with CHF, the physician must direct attention to the physiologic responses to inadequate cardiac function. These include mechanical effects, such as ventricular hypertrophy and ventricular dilation; neurohumoral effects, principally those that involve the adrenergic nervous system; biochemical effects at the cardiac cellular level that alter myocardial energy metabolism and the excitation–contraction coupling process; and hematologic effects that involve oxygen transport. In addition, pulmonary responses, including increased respiratory frequency and altered respiratory patterns, also make up an important part of the clinical picture of CHF.

Clinical Manifestations

The clinical manifestations of CHF are directly related to the compensatory mechanisms already described:

1. Cardiac enlargement usually is the result of ventricular dilation. Although it often may be possible to detect cardiac enlargement by displacement of the cardiac impulse, the chest radiograph remains the most readily available method for assessing ventricular dilation (Fig. 82.1 and Fig. 82.2). Care must be taken to distinguish the normal cardiothymic silhouette from true cardiomegaly in an infant (Fig. 82.2). The other preeminent finding related to mechanical compensatory responses is ventricular hypertrophy, which is easily distinguishable on the electrocardiogram (ECG) (Fig. 82.3). As a compensatory mechanism, hypertrophy occurs before dilation in pressure overload situations, whereas dilation may occur first in volume overload of the heart. As a rule, the cardiac size can be a reliable guide to the overall fluid volume status of the infant or child.

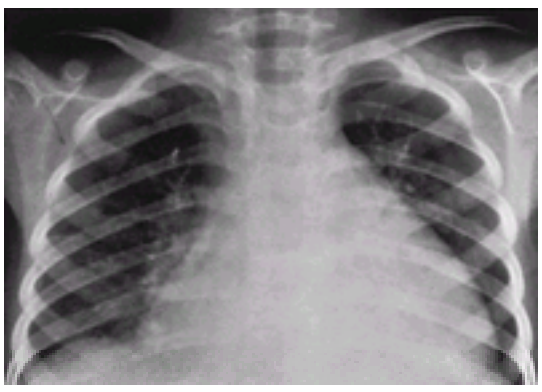


FIGURE 82.1. Chest radiograph of older child with congestive heart failure. Note cardiac enlargement and evidence of pulmonary venous congestion.

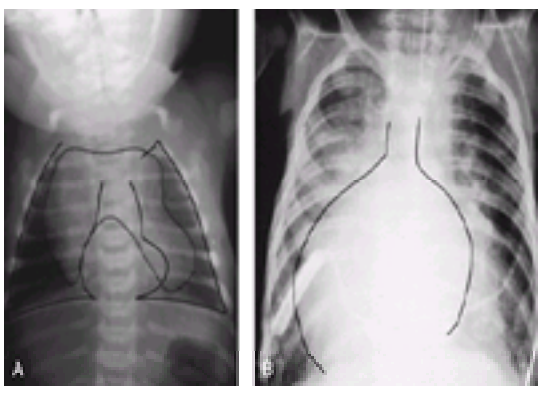


FIGURE 82.2. A. Normal cardiothymic shadow. Thymus is demarcated by speckled line and overlies a portion of the heart shadow. **B.** True cardiomegaly in an infant associated with pulmonary edema. Entire area enclosed in outline is cardiac shadow.

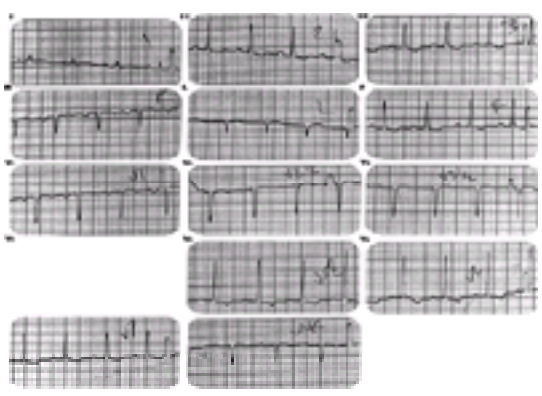


FIGURE 82.3. Electrocardiogram demonstrating left ventricular hypertrophy. Note increased R-wave voltage in left precordial leads and abnormal T-wave changes.

2. Tachycardia is easily detected clinically and, if necessary, confirmed by ECG. A rate of more than 160 beats per minute (bpm) in an infant or 100 bpm in the older child may be a signal for increased adrenergic tone and catecholamine release that is part of the neurohumoral response to diminished cardiac output.
3. Abnormalities of cardiac auscultation also are commonly present. The protodiastolic gallop, or third heart sound (S_3), is a sign of decreased ventricular compliance and increased resistance to filling. Less often, the fourth heart sound, or atrial gallop (S_4), can be heard in children. It should be noted that these auscultatory events can be normal findings in children, and thus, the entire clinical picture must be evaluated before defining their significance in any particular situation.
4. Respiratory responses, notably tachypnea, usually are present as part of the total picture of CHF. Often, rales, rhonchi, and wheezing may be heard and should not be confused as signs of pulmonary parenchymal disease rather than heart failure. In contrast, it is not unusual, particularly in infants, for rales to be absent despite the presence of tachypnea or wheezing because considerable alveolar fluid accumulation is necessary for the development of rales. Thus, the presence of rales usually implies severe failure in an infant, whereas pulmonary interstitial fluid collection, which occurs at an earlier stage, may be represented by tachypnea and wheezing alone. In older children, dyspnea with activity and orthopnea also may be present. A chronic cough also can be a sign of pulmonary congestion associated with CHF and not primary lung disease. Associated with these findings are chest retractions that reflect the large negative intrathoracic pressures needed to ventilate stiff, fluid-filled lungs.
5. Growth failure and undernutrition may be important clinical correlates of chronic CHF. These reflect not only diminished cellular substrate availability as a result of inadequate tissue perfusion but also increased caloric expenditure associated with heightened oxygen consumption and increased work of breathing. Feeding difficulties, which may be associated with the respiratory patterns previously noted, aggravate caloric balance even further.
6. Cool, moist extremities and a generalized pallor also may be present as a result of peripheral vasoconstriction secondary to catecholamine release and the need to maintain blood pressure (BP) in the face of reduced CO.
7. Central and peripheral fluid accumulation with elevated systemic venous pressure also accompanies CHF, reflecting impaired cardiac emptying, as well as impaired sodium and protein balance. Hepatomegaly, jugular venous distension, and peripheral edema represent the clinical manifestations of this aspect of the problem. Peripheral edema, it should be noted, is an unusual finding in young infants. Pulsus alternans, a beat-to-beat variability in the strength of the pulse, also is a clinical sign of cardiac mechanical decompensation.

The child with overt CHF who is seen in the emergency department (ED) may present with nearly all the aforementioned signs and symptoms. If the child is severely ill, pallor will be evident, tachypnea prominent, and intercostal retractions visible. The liver is enlarged and palpable well below the right costal margin; a spleen tip also may be palpable. The pulses are weak and thready, and the skin may be moist and cool to the touch. Auscultation of the chest reveals rales, rhonchi, and sometimes, wheezes. Tachycardia is present, and auscultation of the heart sounds often elicits a gallop rhythm. Murmurs may be strikingly absent unless preexisting heart disease is present. A child with this spectrum of findings deserves immediate attention. Acute heart failure in a child usually implies an unstable situation with possible rapid deterioration.

Laboratory Findings

Usually, a clinical diagnosis of CHF can be made without extensive radiographs or laboratory tests. However, certain objective changes may corroborate the clinical findings:

1. As noted, a chest radiograph shows an increased cardiothoracic ratio, as well as pulmonary congestion. Kerley B lines or platelike atelectasis at the lung bases, which reflect dilated pulmonary lymphatics, may be present. Pleural effusions are common.
2. The ECG is a nonspecific indicator of cardiac decompensation. The precordial voltages decrease in certain conditions associated with CHF, such as myocarditis, but may be normal or increased in other situations despite overt CHF. The ECG is helpful also for establishing the cause of the CHF, such as cardiac arrhythmia or myocardial ischemia.
3. Echocardiography or radionuclide studies can be helpful in evaluating the child with CHF. The differentiation of an enlarged cardiac silhouette secondary to impaired cardiac performance with ventricular chamber enlargement rather than pericardial fluid accumulation can best be made by ultrasound examination. In addition, functional indices can be obtained as an objective measure of cardiac performance and response to therapy.
4. Blood gas abnormalities may be present. Prolonged tissue hypoperfusion can result in metabolic acidosis of a significant degree, and the pulmonary abnormalities already noted may result in hypoxia.
5. Other abnormalities that may be present include electrolyte changes (hyponatremia and hypochloremia) and a reduction in hematocrit, based on dilutional factors. The sedimentation rate (ESR) usually is lowered in active CHF. In addition, in infants with CHF, serum glucose and calcium should be monitored because deficiencies in either may be responsible in large measure for the impaired cardiac function. In situations of suspected perfusion abnormalities or inflammatory myocardial diseases, cardiac enzymes—creatine phosphokinase (CPK) in particular—may be elevated.

Management

For the patient who requires emergency treatment of CHF, initial medical therapy includes several therapeutic measures as outlined in Table 82.6. First, supplemental oxygen should be given through a humidified system. In children, a tight-fitting mask or nasal cannula may not be effective because too much energy is expended fighting such apparatuses. Second, elevation of head and shoulders is helpful in the face of pulmonary edema, with maintenance of the lower extremities in a dependent position to increase peripheral pooling and thus diminish pulmonary blood volume. A “cardiac chair” or appropriate modification of an infant seat establishes this posture in the small baby. Third, morphine sulfate (0.05 to 0.1 mg/kg subcutaneously) can be helpful in the face of agitation and air hunger associated with pulmonary edema. Fourth, positive-pressure respiration by endotracheal intubation is sometimes indicated for severe situations,

particularly if arterial blood gas analysis shows respiratory decompensation (Pa CO_2 50 mm Hg). In infants, the use of controlled mechanical ventilation to improve respiratory status greatly enhances survival. Fifth, bicarbonate therapy sometimes is indicated to correct metabolic acidosis that arises from diminished tissue perfusion. It should be remembered that administration of sodium bicarbonate during respiratory decompensation is hazardous because further PaCO_2 elevation can occur. In addition, bicarbonate given by rapid infusion can promote cerebral edema and rapidly affect serum osmolarity with deleterious effects. Also, an excessive sodium load can result from injudicious use of bicarbonate. Thus, only for severe acidosis (pH less than 7.2) should bicarbonate be considered and, even then, only if respiratory function is satisfactory (see Chapter 95). Last, an intravenous (IV) infusion should be started to aid in the administration of drugs and the strict monitoring and administration of fluids.

Define cause, if possible, through history and physical examination.	Perform arterial blood gas determination.
Elevate head and chest.	Achieve rhythm control.
Ensure adequacy of ventilation.	Treat pulmonary edema (e.g., diuretics, morphine sulfate).
Administer oxygen.	Provide inotropic support (digitalis or catecholamines).
Initiate cardiorespiratory monitoring, including frequent blood pressure measurement.	Consider afterload reduction.
Achieve venous access and obtain laboratory studies (e.g., complete blood count, electrolytes).	

Table 82.6. Emergency Management of Congestive Heart Failure

Blood products in the form of packed cells should be administered if the child is severely anemic. Antibiotics should be reserved for unequivocal evidence of infection or for situations in which circumstantial evidence is strongly suggestive and appropriate cultures have been drawn. The use of corticosteroids may be indicated at times, particularly for heart failure precipitated by rheumatic heart disease. However, decisions of this type should be made with cardiac consultation. Treatment of arrhythmias that result in CHF is discussed subsequently under Cardiac Arrhythmias.

Although newer inotropic agents have become available, the mainstay of the medical management of CHF is still digitalis, or other well-tested inotropic agents, to improve contractility and diuretics to manipulate ventricular preload and intrapulmonary fluid. Recently, pharmacologic adjustments of afterload have become an important part of CHF treatment. Regardless of the specific therapy, frequent reexamination and reevaluation are mandatory.

Digitalis

The drugs of choice for improving the inotropic condition of the heart, unless the CO is severely compromised and the child is acutely ill, are the digitalis glycosides. Several investigations have helped improve our understanding of the mechanisms of action of these agents and how to adjust their administration to specific clinical situations. Although the use of digitalis in infants was reported more than 35 years ago, until recently, dosing regimens have been strictly empiric in their derivations. The use of a radioimmunoassay to determine serum digitalis levels has helped define a dosing format. In infants and children, debate exists about the true therapeutic concentration range. As noted in Table 82.7, the principal clinical value of digitalis pharmacokinetic studies thus far has been to verify that the same unit dose per kilogram of body weight is not necessarily best for children of all ages and that premature infants, in particular, require close adjustment of dose, based on body weight.

I. Usual Doses (IM or oral)		
	Weight (g)	Dose (TDD)*
Premature infants	500–1000	20 µg/kg or 0.02 mg/kg
	1000–1500	20–30 µg/kg or 0.02–0.03 mg/kg
	1500–2000	30 µg/kg or 0.03 mg/kg
	2000–2500	30–40 µg/kg or 0.03–0.04 mg/kg
Term to 12 yr		40–60 µg/kg or 0.04–0.06 mg/kg (no dose greater than 1.5 mg TDD)
II. Alterations in Usual Doses		
Lower if renal function is impaired		
Lower in presence of poor myocardial function (cardiomyopathy, myocarditis)		
Lower in presence of metabolic imbalance (electrolyte abnormalities, hypoxia, acidosis)		
IV dose is 75% of oral or IM dose		

*TDD, Total digitalizing doses. Digitalizing regimen usually given as initial dose = one-half of TDD, second dose = one-fourth of TDD, at 8–12 hr; third dose = final one-fourth TDD at 8–12 hr after second dose. Maintenance is then started as one-eighth TDD every 12 hr. (Note: Parenteral preparation contains 100 µg/mL and oral, 50 µg/mL.)

Table 82.7. Digitalization with Digoxin

The mechanism by which digitalis improves cardiac performance centers on the regulation of the ionic movements that are part of the contractile process. In particular, inhibition of adenosine triphosphatase (ATPase) by digitalis interferes with the sodium pump mechanism, allowing intracellular accumulation of sodium and, consequently, increasing the level of available calcium for contraction. The associated effect of intracellular potassium depletion may be related to the development of toxicity from digitalis preparations.

The result of digitalis administration for CHF is an increase in the force of cardiac contraction and thus an improvement in emptying of the ventricle. Intracardiac filling pressures are reduced, CO rises, cardiac size decreases, and HR slows.

Eventually, use of the compensatory mechanisms (p. 662) to maintain CO is mitigated.

Several important points must be remembered when prescribing digitalis glycosides, regardless of the specific one selected or the route of administration. Diligent care must be used in relaying the prescribing information to avoid errors that may have fatal consequences. Calculations of the total digitalizing dose should be double-checked and clearly recorded. The microgram dosage should be unequivocally clear, and the corresponding volume to be administered also should be written down. If possible, the prescription should be checked by other medical personnel. Decimal errors are inexcusable but all too common.

The route of administration also has a significant bearing on the dosage prescribed. Parenteral digoxin preparations contain 100 µg/mL, and oral preparations contain 50 µg/mL. In the emergency setting, parenteral administration often is the preferred route. If given IV, the calculated oral dose is reduced by 25%, and the child should be monitored for sudden changes in HR or rhythm. Tissue perfusion levels should be assessed as satisfactory before intramuscular (IM) administration is contemplated because poor absorption from an IM dose (which can be painful) may undercut the therapeutic response.

Digoxin is administered at the dosages described in Table 82.7 (maximum 1.5 mg). The total digitalizing dose is given over 24 hours (one-half initially, then one-quarter in 8 to 12 hours, and one-quarter in another 8 to 12 hours). The daily maintenance dosage is one-quarter of the total digitalizing dose divided into twice daily doses.

Poisoning from digitalis ingestion may be the precipitating cause for emergency evaluation and for the development of CHF. Various systemic manifestations may be associated with overdosage, including nausea, vomiting, weakness, and worsening of preexisting heart failure. The ECG manifestations of digitalis excess are reviewed subsequently under Irregular Heart Rates. Care should be taken to clarify the ECG distinctions between “digitalis toxicity” and the more benign “digitalis effect.” In general, it is safest to assume that the appearance of a new major conduction disturbance in a child who takes a digitalis preparation is related to the drug.

Treatment of digitalis toxicity involves cessation of the drug. Potassium supplementation may be helpful, specifically when potassium depletion has occurred because of dietary factors or diuretic therapy. Potassium should not be given to a patient with digitalis-related atrioventricular (AV) block (see Cardiac Arrhythmias). Diphenylhydantoin is particularly useful in disorders of impulse formation related to digitalis therapy (e.g., premature ventricular contractions [PVC]). In instances of severe heart block, pacing may be required. The presence of digitalis is not an absolute contraindication for cardioversion, and in certain instances, cardioversion may be required to convert even a digitalis-induced rhythm disturbance. On occasion, calcium ethylene diamine tetracetic acid (EDTA) (30 mg/kg slow IV over 2 hours) may be used as an adjunct to the previously mentioned forms of therapy. Careful monitoring is required, however, because hypocalcemia may be induced and because EDTA may negate the therapeutic effects of digoxin, resulting in worsening CHF.

The availability of digoxin antibodies is an important adjunct in the treatment of digoxin toxicity. “Digibind” and other such drugs use the Fab fragment of antibody to digoxin to lower serum concentrations rapidly. Although it usually is necessary to combine vigorous antiarrhythmic therapy (see next section) with digoxin antibody treatment, this form of therapy can provide dramatic improvement in as little as 30 minutes. It must be cautioned, however, that Fab treatment in patients receiving chronic digoxin therapy for CHF may exacerbate CO problems by essentially “withdrawing” the drug. Dosage of Fab generally is equimolar to the estimated amount of digoxin in a patient's body. The onset of action is relatively rapid, with some clinical response noted in as little as 30 minutes after completion of infusion. Complete reversal of a toxicity can occur in 3 to 4 hours. However, close supervision of the patient is needed because with the administration of digoxin antibody, rapid occurrence of hypokalemia can occur. In patients with atrial fibrillation, a rapid ventricular response may occur to exacerbate problems, and occasionally a patient may experience hypersensitivity if sheep protein allergy is present. The elimination of digoxin antibody fragments may take up to several days or longer, especially if renal insufficiency is present, and may interfere with subsequent serum digoxin levels done by radioimmunoassay. Thus, other techniques for assessment of digoxin levels may be needed to follow-up patients at risk.

To use digoxin antibodies, it is assumed that the digoxin body burden approximates the nanogram per milliliter digoxin level. Thus, 1.0 ng/mL digoxin level reflects a burden of 1 mg digoxin. Each vial of purified digoxin-specific Fab fragments binds at least 0.4 mg of digoxin. Thus, for a digoxin level of 5 ng/mL, for example, 12.5 vials of Fab fragments will be required. No specific pediatric dose has been derived, and the relationship of digoxin level to body burden may not be as close as these values indicate. Thus, this dosage may not be entirely accurate for infants and small children.

Other Inotropic Agents

In situations of severely compromised CO, isoproterenol or dopamine, both a-receptor agonists, has been used successfully in infants and children. Dobutamine, an analog of dopamine, also has been found to be useful in such circumstances, particularly when impaired myocardial perfusion is part of the underlying problem.

Isoproterenol (Isuprel) has vigorous inotropic effects, as well as significant chronotropic effects. As noted under Cardiac Arrhythmias, for persistent bradycardia, isoproterenol is the drug of choice. Cardiac rhythm effects may limit the use of isoproterenol, however, because induction of tachyarrhythmias is a known consequence of its administration. In addition, hypotension may occur. The starting dose is 0.1 µg/kg per minute by continuous infusion (see Table 82.12).

Dopamine has achieved a wide degree of popularity because of its ability, at low dosages (“dopaminergic effects”), to augment renal blood flow directly, in addition to improving cardiac output. Furthermore, the chronotropic activity of the drug is somewhat lower than isoproterenol, and there is less of a tendency to produce hypotension. Several studies have established the efficacy and safety of dopamine in infants and children. The drug is available in 5-mL ampules that contain 200 mg of dopamine; this must be diluted in 250 to 500 mL of a neutral or acidic solution (usually 5 to 10% dextrose or saline). Dopamine must not be administered through the same IV solution as sodium bicarbonate because

alkali will deactivate the drug. Initial doses in pediatrics range from 2 to 5 µg/kg per minute given by continual infusion. For severe systemic hypotension, 5 to 10 µg/kg per minute may be used as the starting dose. The response should be relatively prompt, with an increase in HR and BP followed by improvement in urine output. Increasing the infusion rate may be necessary, but at higher dosages (15 µg/kg per minute), the beneficial effects of dopamine on renal blood flow are mitigated, and at 20 µg/kg per minute, adrenergic effects predominate and renal blood flow may be reduced. Adverse effects from dopamine include nausea and vomiting and changes in cardiac rhythm, particularly in patients with preexisting arrhythmias and especially at higher infusion ranges (greater than 10 µg/kg per minute). Dopamine also may elevate pulmonary vascular resistance and should be used with caution, if at all, in patients with pulmonary vascular obstructive disease. Monoamine oxidase inhibitors may potentiate the effect of agents such as dopamine.

These agents should be administered under close supervision, optimally with monitoring of arterial pressure, central venous and/or pulmonary wedge pressure, HR, and urinary output.

Dobutamine recently has achieved an increased level of popularity because of its relatively rapid response after initiation of infusion and because of the achievement of favorable hemodynamic effects with less myocardial oxygen debt burden than occurs with dopamine. Fewer chronotropic and arrhythmogenic effects appear to result from dobutamine, and it may have a more direct effect on enhancement of coronary flow. Therefore, dobutamine may have particular efficacy when impaired myocardial perfusion is suspected, as in heart failure from inflammatory myocardial disease or abnormalities that involve the coronary arteries. Dobutamine is administered in similar fashion to dopamine with initial dosages that range from 2.5 to 5.0 µg/kg per minute. Higher dosages may be used, but complicating adrenergic effects, in particular potentiation of rhythm disorders, begin to predominate when high dosages (15 to 20 µg/kg per minute) are used. As with dopamine, dobutamine can be used in concert with other agents, such as afterload reduction drugs.

Concern about the efficacy of dobutamine in the young infant (less than 1 year old) remains. In such infants, dobutamine may improve CO but not result in BP elevation. Thus, in severe hypotension in the young infant associated with septic shock, for instance, dobutamine may be more appropriate as an adjunct, and not as a primary inotrope.

Diuretics

Alterations in renal perfusion and salt and water balance are well known correlates of CHF. Reduced renal blood flow can result in increased circulation volume and increased sodium and water reabsorption (through associated secondary hyperaldosteronism). Thus, diuretics play a critical role in the management of the child with CHF.

The so-called loop diuretics are used most commonly for the acute treatment of CHF. Furosemide (Lasix) is the most popular of these, but ethacrynic acid (Edecrin) also may be used. Through effects on sodium and chloride transport in the loop of Henle, interference with urinary concentrating capability is achieved and diuresis is achieved. An initial dose of 1 mg/kg IV usually results in adequate urine flow within 1 to 2 hours of administration. If 3 to 5 mL/kg per hour urine flow is not achieved, a subsequent dose (at an increment of 1 mg/kg) can be given and repeated at hourly intervals, to a maximum of 3 to 5 mg/kg. Close observation for changes in serum electrolytes, especially potassium, is important because IV digitalis may be given concurrently.

Thiazide diuretics are much less commonly used to treat acute CHF and are now reserved for more chronic situations (as oral agents). Nevertheless, agents such as hydrochlorothiazide or chlorothiazide, working at the tubular level, produce good diuretic effects.

Other classes of diuretics have become available and may be particularly useful in refractory conditions or in cases in which traditional diuretics are already in use. Of these agents, metolazone (Zaroxolyn) has been administered most often. It is for oral use; thus, onset of action is delayed. Particularly intense potassium depletion can result from metolazone, and patients who take this drug should be evaluated for potassium loss whenever they present in the ED.

Spironolactone is considered an adjunctive form of diuretic therapy in most situations and is not a first-line drug because its diuretic effect may not occur for 2 to 3 days. Aldosterone antagonism makes it suitable for use as an additional agent when potassium loss is a problem.

With the proper use of digitalis and diuretic therapy, improvement can be achieved in most children with CHF. Failure to improve an exacerbation of CHF in children already taking these medications requires scrutiny for any of the following: 1) persistent arrhythmia; 2) untreated or unrecognized infection; 3) anemia, especially in the infant with CHF; 4) inadequate or excessive digitalis dose, particularly in the patient with inflammatory myocardial disease; or 5) electrolyte disturbance, such as hypokalemia, which may be worsened with diuretics. If these entities can be ruled out, more intensive treatment is indicated to improve CO.

Other Noncatecholamine Agents

Among recent additions to the pharmacologic tools readily available to treat acute CHF are the bipyridine derivatives amrinone and milrinone. These drugs have the combined effects of inotropic support and peripheral vasodilation. Although the exact mechanism of action is unclear, they are known to be inhibitors of myocardial cyclic adenosine monophosphate (cAMP) phosphodiesterase activity, thereby enhancing intracellular levels of myocardial cAMP. There is also a direct vascular smooth muscle relaxant action. In an ideal sense, amrinone and its class of therapeutic agents could represent a major therapeutic breakthrough because the inotropic effects of these drugs may be additive to other agents, especially to digitalis. However, pediatric experience has been tempered by the profound vasodilatory effects of the drug and the need for careful invasive monitoring when it is used. Adequate filling volumes must be maintained to ensure adequate systemic perfusion. Occasionally, hypersensitivity reactions have occurred with amrinone. Fever and thrombocytopenia are also known side effects, and potassium levels should be followed closely, especially if diuretics are used in conjunction with this drug. Finally, chemical interaction with glucose (dextrose) solutions can occur, and the drug should not be diluted with these fluids. In the patient with CHF in whom standard digoxin and/or catecholamine therapy

does not provide improved peripheral perfusion status, use of amrinone (IV) should be considered. Infusion rates range from 5 to 10 µg/kg per minute with a bolus dosage to start therapy of 0.50 to 2.0 mg/kg given initially over 2 to 5 minutes.

Vasodilators

The management of CHF also includes manipulation of loading conditions, following on the physiologic principles defined earlier in this chapter. Both afterload and preload interventions can be useful. Agents with effects on cardiac loading have been shown to be efficacious in adults with chronic ischemic forms of cardiac dysfunction and are appropriate for infants and children with heart failure unresponsive to more conventional treatment regimens. In heart failure with reduced CO, systemic vascular resistance often is elevated by compensatory mechanisms used to maintain BP. Preload reserve is used through cardiac dilation to increase stroke volume, but this is accomplished at the expense of a decreased ability of the myocardium to shorten as needed to overcome the increase in afterload. Thus, there is a "mismatch" of afterload and preload reserve in heart failure. Vasodilators that work primarily on arteriolar smooth muscle cause afterload reduction while venodilation works on preload by lowering cardiac filling volumes. Some of these agents have mixed effects (Table 82.8). In the vasodilator group, the nitrates, particularly Nitroprusside, have been most actively used in the acute-care setting. Sodium nitroprusside has both arteriolar and venous actions, a prompt onset of action, and usually a short duration of effect. In the patient with impaired renal function, however, caution must be exercised in its use because the byproduct, thiocyanate, can accumulate with neurologic, endocrinologic, and other toxicity. Other drugs used include a-receptor blockers and angiotensin-converting enzyme (ACE) inhibitors, although these are more commonly used in the long-term care setting via the oral route. Calcium channel blockers appear to be most useful for diastolic dysfunction conditions. These situations involve heart failure developing in the face of myocardial hypertrophy and/or normal end systolic volume, such as in hypertrophic cardiomyopathy. The negative inotropic effects of these agents require their use only with careful cardiac monitoring. It is important for the emergency physician to be aware of the chronic use of these drugs in patients who may present with an exacerbation of heart failure because continuation of such therapy in the acute-care setting may be required. Afterload reduction therapy, when used IV, usually requires extensive monitoring because failure to maintain adequate cardiac filling can have serious negative results. Emergency use of afterload reduction should be limited to those well versed in cardiopulmonary physiology, and maintenance of such treatment should be carried out in an intensive care environment.

Agent	Class	Action	Dosage
Nitroprusside	Nitrate	Mixed dilator	1–10 µg/kg/min IV (maximum: 125–10 µg/kg/min)
Nitroglycerin	Nitrate	Venodilator	2–10 µg/kg/min
Hydralazine[a]	Smooth muscle inhibitor	Arteriolar dilator	0.1–0.5 mg/kg/dose q6h or 0.2–1 mg/kg single dose IV
Prazosin	α Blockade	Mixed dilator	0.01–0.05 mg/kg/dose by mouth q8–12hr
Captopril	ACE inhibitor	Mixed	0.1–0.5 mg/kg/dose by mouth q8hr
Enalapril	ACE inhibitor	Mixed	5 or 12 hr (maximum: 5 mg/kg/day)[b]
Diltiazem	Ca^{2+} channel blocker	Arteriolar dilator	0.2–0.5 mg/kg/dose by mouth or sublingual q8hr
Nifedipine	Ca^{2+} channel blocker arteriolar		0.25–1 mg/kg

ACE, angiotensin-converting enzyme; Mixed, both arterial and venous vasoactivity.
[a]Precise mechanism not identified; may inhibit calcium activity.
[b]If Enalapril is available for an every 8-hr regimen (enalaprilat), 0.01–0.05 mg/kg/dose.

Table 82.8. Afterload Reducing Agents Used in Congestive Heart Failure

NEWBORN WITH OBSTRUCTED SYSTEMIC OR PULMONARY BLOOD FLOW

Background

For the baby with cyanotic CHD or with obstruction to systemic blood flow, emergency intervention is crucial, indeed life-sparing, and usually must be delivered before permanent long-term therapy can be undertaken. The emergency physician must be able to recognize when such a life-threatening circumstance is present and must be able to initiate therapy even before a precise diagnosis can be accomplished. Although this is most often a problem for the neonatologist or other physician caring for an infant during the first few days of life, these babies also are brought to EDs after having been at home. Still others may need the help of emergency medical personnel as they are transported to cardiac centers for definitive care. Therapy in these critical infants depends on manipulation of the ductus arteriosus.

Pathophysiology

In fetal life, the ductus arteriosus is the principal conduit allowing the preponderance of right ventricular output to bypass the nonventilating fetal lungs (Fig. 82.4). This pathway thus allows fetal circulatory viability even in the face of extreme disorders of functional cardiac development (Fig. 82.5 and Fig. 82.6). In the case of impaired systemic blood flow, the fetus and later the neonate, survives because the ductus allows right ventricular output to reach the systemic circulation. This right-to-left shunt mitigates the effects of even complete aortic and/or mitral atresia. In situations of obstructed pulmonary blood flow, the ductus serves as a conduit for systemic (left ventricular) output to reach the pulmonary circulation. This left-to-right shunt becomes crucial once the newborn becomes dependent on his or her own pulmonary circulation for oxygenation after delivery. Typical conditions that are "ductal dependent" are listed in Table 82.9.

Ductal-Dependent Pulmonary Blood Flow	*Ductal-Dependent Systemic Blood Flow*
Pulmonary atresia with intact ventricular septum	Coarctation of the aorta
Tricuspid atresia	Aorta arch interruption
Critical pulmonary stenosis	Hypoplastic left heart syndrome (aortic atresia)

Adapted from Gewitz MH. Cardiac disease. In Polin R, Yoder M, Burg F, eds. Workbook in Practical Neonatology. 2nd ed. Philadelphia: Saunders, 1993: Ch. 12.

Table 82.9. Ductal-Dependent Cardiac Lesions

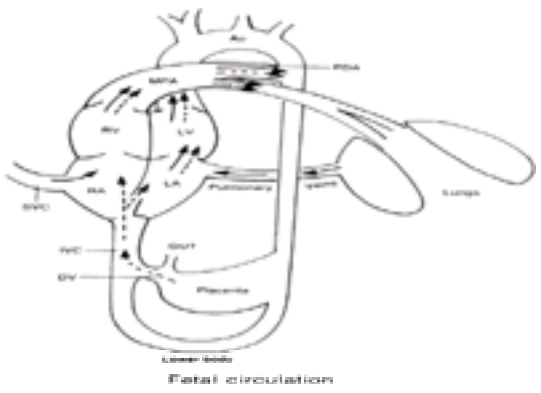


FIGURE 82.4. Normal fetal circulatory pathway including patency of foramen ovale and ductus arteriosus. (Reprinted with permission from Gewitz M. Cardiac disease. In: Polin R, Yoder M, Berg F, eds. Workbook in Practical Neonatology. 2nd ed. Philadelphia: WB Saunders, 1993:Ch. 12.) *Ao,* aorta; *DV,* ductus venosus; *IVC,* inferior vena cava; *LA,* left atrium; *LV,* left ventricle; *PA,* pulmonary artery; *PDA,* patent ductus arteriosus; *RA,* right atrium; *RV,* right ventricle; *SVC,* superior vena cava.

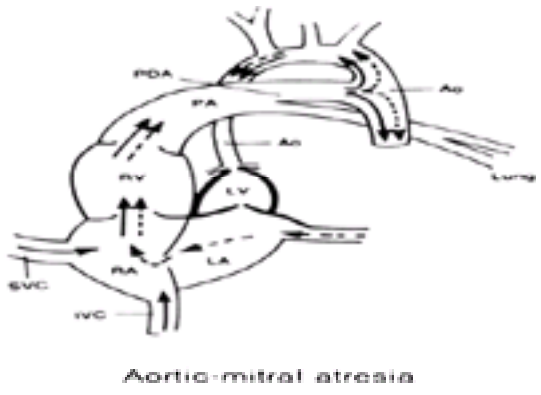


FIGURE 82.5. Aortic and mitral atresia (hypoplastic left heart syndrome in fetus with altered circulatory heart syndrome) in fetus with altered circulatory physiology at ductus and atrial levels. (Reprinted with permission from Gewitz M. Cardiac disease. In: Polin R, Yoder M, Berg F, eds. Workbook in Practical Neonatology. 2nd ed. Philadelphia: WB Saunders, 1993:Ch. 12.) *Ao,* aorta; *DV,* ductus venosus; *IVC,* inferior vena cava; *LA,* left atrium; *LV,* left ventricle; *PA,* pulmonary artery; *PDA,* patent ductus arteriosus; *RA,* right atrium; *RV,* right ventricle; *SVC,* superior vena cava.

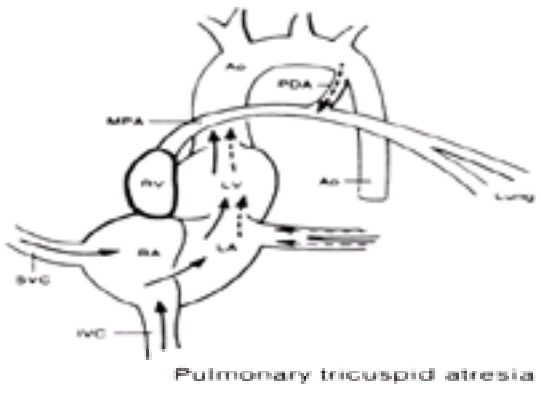


FIGURE 82.6. Pulmonary-tricuspid atresia (right-sided heart hypoplasia) with intact septum in fetus. Altered circulatory physiology at ductus level with enhanced flow at atrial level. (Reprinted with permission from Gewitz M. Cardiac disease. In: Polin R, Yoder M, Berg F, eds. Workbook in Practical Neonatology. 2nd ed. Philadelphia: WB Saunders, 1993:Ch. 12.) *Ao,* aorta; *DV,* ductus venosus; *IVC,* inferior vena cava; *LA,* left atrium; *LV,* left ventricle; *PA,* pulmonary artery; *PDA,* patent ductus arteriosus; *RA,* right atrium; *V,* right ventricle; *SVC,* superior vena cava.

Clinical Manifestations

Any newborn with sudden onset of either collapsed systemic circulation or intense cyanosis should be considered at risk for the presence of a ductal dependent state. In these babies, closure of the ductus unmasks the underlying circulatory insufficiency resulting in the clinical picture of either severe hypoxemia or shock or both. These infants may be as old as 1 or 2 weeks, although usually this catastrophe becomes apparent within the first days of life. However, there are times when the process of ductal closure leading to the presentation can be delayed even longer than 2 weeks.

The mechanisms responsible for ductus closure have been defined over the past several years. Although a prolonged discussion is not relevant in this context, it is important for the emergency physician to understand that these factors center on the balance of dilator and constructor hormones, namely prostaglandins, and that manipulation of this hormonal system can yield prompt and substantial results. Of course, treatment of heart failure and impaired oxygenation involve more than just ductus manipulation as outlined elsewhere in this text.

Management

Based on the previous principles, prostaglandin E_1 (PGE_1) has become the standard medical intervention used in this urgent situation. PGE_1 provides relatively rapid stabilization until more permanent measures can be undertaken. There are few, if any, concerns that would contraindicate the use of PGE_1 when any of the conditions noted above are suspected. Dosage is by infusion at 0.05 to 0.10 µg/kg per minute, after an initial bolus of 0.10 µg/kg. The specific site of infusion is not critical as long as patency of access can be continuously verified. Side effects can be important and, unless prepared for, can be life-threatening. These include bradycardia, apnea, hypotension, and seizures. Rash and hyperthermia also can develop. Therefore, when PGE_1 therapy is initiated, the ability to support respiration and blood pressure should be secured. Intubation may be required and should always be considered if a prolonged transport is planned. It has become evident over the past two decades that manipulation of the ductus by PGE_1 administration has been one of the most important advances in the early treatment of even the most severe forms of CHD.

CARDIAC ARRHYTHMIAS

General Considerations

Background

Disturbances in cardiac rhythm are relatively common in infants, children, and adolescents. An apparent increase in the incidence of cardiac arrhythmias in children can be explained by the extensive use of ECG monitoring equipment in children's hospitals, advances in cardiac surgery that have resulted in the survival of children with complex CHD, new techniques to investigate rhythm disturbances, and an increased awareness on the part of pediatricians and pediatric cardiologists of the manifestations of abnormal cardiac rhythms in children.

Pathophysiology

The electrical impulse that initiates and coordinates the mechanical activity of the heart is propagated in an orderly manner through the normal heart. This electrical activity is initiated in the sinoatrial (sinus) node located at the junction of the superior vena cava and right atrium (Fig. 82.7). Activity then spreads through the atria to the AV node located in the lower part of the right atrium near the coronary sinus and just above the septal leaflet of the tricuspid valve. The impulse continues to the bundle of His, which then divides into the right and left bundle branches in the ventricle. The bundle branches then divide into the Purkinje fibers of the ventricular myocardium, and the entire ventricle is thus depolarized.

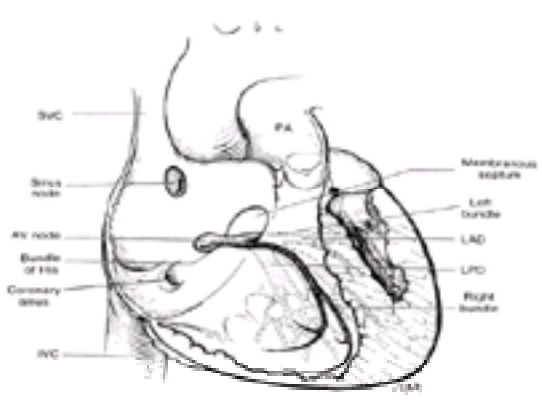


FIGURE 82.7. Schematic representation of the intracardiac conduction system. *IVC,* inferior vena cava; *LAD,* left anterior division; *LPD,* left posterior division; *PA,* pulmonary artery; *SVC,* superior vena cava.

Arrhythmias in children are caused by disturbances in impulse formation or conduction or both. In many children with rhythm disturbances, no cause is recognizable. However, certain children have a propensity to develop arrhythmias. Some types of CHD have a relatively high incidence of associated cardiac arrhythmias. These include corrected transposition of the great vessels, Ebstein's anomaly of the tricuspid valve, congenital mitral stenosis, and the asplenia-polysplenia syndromes. Other children may have congenital complete heart block, Wolff-Parkinson-White (WPW) syndrome, or the long Q-T syndrome. Postsurgical arrhythmias are commonly seen in children after repair of d-transposition of the great vessels, tetralogy of Fallot, endocardial cushion defects, large atrial septal defects, and the

Fontan procedure. Acquired heart diseases with rhythm disturbances may include cardiomyopathies, rheumatic carditis or rheumatic heart disease, Kawasaki disease, viral myocarditis, cardiac tumors, and hemochromatosis.

Systemic diseases or abnormalities associated with cardiac arrhythmias include electrolyte disturbances, neuromuscular disorders (muscular dystrophy, Friedreich's ataxia), endocrine disorders (hyperthyroidism or hypothyroidism), inherited disorders of metabolism (glycogen storage disease, Pompe's disease), mitochondrial disorders (acyl Co-A dehydrogenase deficiency, Kearns-Sayre syndrome), collagen diseases (systemic lupus erythematosus [SLE]), pulmonary diseases (bronchopulmonary dysplasia, cystic fibrosis), hematologic disorders (anemia, thalassemia major), neoplasms, renal diseases (uremia), infectious diseases, and central nervous system (CNS) diseases (increased intracranial pressure [ICP], encephalitides). Drugs and toxic substances (digitalis, general anesthesia, theophylline, sympathomimetic drugs, epinephrine, tricyclic antidepressants) also can lead to abnormalities of cardiac rhythm.

Clinical Manifestations

Many children are not aware of or are unable to express awareness of an abnormal cardiac rhythm. Thus, the physician must suspect the diagnosis from the secondary manifestations. Arrhythmias may surface in the following ways:

1. Symptoms of CHF (see previous section)
2. Symptoms related to decreased cerebral blood flow (syncope, dizziness, irritability, and inappropriate behavior)
3. Symptoms related to decreased coronary blood flow (anginal chest pain)
4. Perception of the rhythm disturbance by the child (palpitations, skipped beats)

Management

Management of the child with a cardiac arrhythmia requires recognition of the manifestations of these disorders, diagnosis of the type of rhythm disturbance, understanding of the mechanism of the abnormality, knowledge of appropriate therapy (pharmacologic or other intervention), judgment about the appropriate timing and urgency of therapy, and understanding of potential side effects of the therapy. Once an abnormality in cardiac rhythm is suspected or found, the precise diagnosis must be made to institute appropriate treatment. This generally depends on evaluation of the ECG. The resting ECG used to evaluate cardiac arrhythmias should include a long rhythm strip in addition to a complete 12- or 15-lead ECG. Lead II often is used for the rhythm strip, but a V_1 lead or another lead in which P waves are prominent may be more helpful. The rhythm on the ECG should be evaluated for rate, regularity, mechanism, and origin of the disturbance.

Cardiac arrhythmias become emergencies when they produce hemodynamic alterations that result in a decreased CO or have the potential to do so. To treat cardiac arrhythmias effectively, one must be able to identify specific arrhythmias, recognize signs and symptoms of cardiac decompensation, and understand which arrhythmias are likely to produce rapid cardiac decompensation. Most infants and children who have symptomatic arrhythmias will require cardiac consultation and admission to the hospital for treatment and observation and continuous ECG monitoring (telemetry or bedside with arrhythmia analysis is preferred). Table 82.10 represents an overview of the emergent management of arrhythmias.

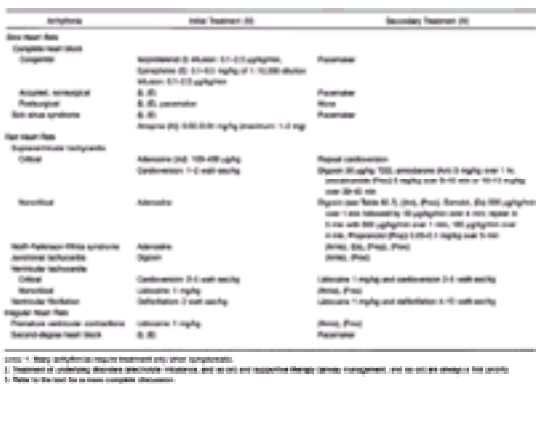


Table 82.10. Emergent Management of Arrhythmias in Children

The cardiac arrhythmias discussed in this section are classified according to their presentation to the physician: slow HRs, rapid HRs, and irregular HRs. Slow HRs that are most commonly seen in the ED include complete (or third-degree) heart block, second-degree heart block, sinus or junctional bradycardia, and the sick sinus syndrome. SVT and ventricular tachycardia (VT) cause a rapid HR. Irregular rhythms usually are caused by premature ventricular or atrial contractions and second-degree heart block.

To determine whether a HR is abnormally fast or slow, one must know the normal range of rates for children of various ages. Results of 24-hour continuous ECG monitoring studies are redefining the normal ranges. Table 82.11 illustrates ranges of rates accepted as normal.

Age	Heart Rate (bpm)
Newborn	80–180
1 wk–3 mo	80–180
3 mo–2 yr	80–160
2 yr–10 yr	65–130
10 yr–adult	55–90

Table 82.11. Normal Heart Rate Ranges

Slow Heart Rates

Complete Heart Block

Complete (third-degree) AV heart block is the most common cause of significant bradycardia in infants and children. Complete heart block, which may be congenital or acquired, results from a complete failure of conduction from atria to ventricles. The atrial rate is usually faster than the ventricular rate, which usually is 40 to 80 bpm. A typical ECG is shown in Figure 82.8.

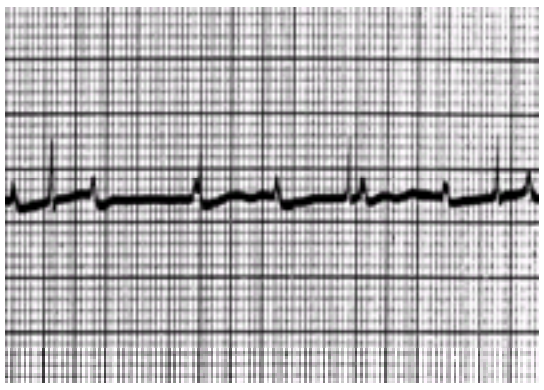


FIGURE 82.8. Example of complete heart block. Note absence of any regular P-R interval. Ventricular rate 62 bpm, atrial rate approximately 95 to 115 bpm.

Congenital Heart Block

Congenital heart block may be idiopathic, associated with specific types of congenital heart defects such as l-corrected transposition of the great arteries or left atrial isomerism/polysplenia syndromes (heterotaxy), or associated with collagen disease in the mother associated with the presence of anti-Ro (SS-A) and anti La (SS-B). Congenital heart block is being diagnosed more often in utero with the use of fetal monitoring and fetal echocardiography, but it may not be recognized for weeks or months after birth.

All infants with congenital heart block have bradycardia. Although some remain asymptomatic, others develop CHF and, occasionally, cardiovascular collapse and sudden death. An ECG will differentiate sinus bradycardia from complete heart block. Sinus bradycardia should respond to the usual resuscitative measures: ventilation and oxygenation, treatment of acidosis, and catecholamine support of HR and BP. The infant with complete heart block who has severe CHF or who is in shock may also require intubation for adequate ventilation, oxygenation, and treatment of acidosis. If no improvement is obtained with these measures, infusion of isoproterenol or epinephrine may increase the HR slightly, allowing time for the placement of a temporary pacemaker (Table 82.12). A hydropic infant may require emergency phlebotomy, as well as a potent diuretic such as furosemide (1 mg/kg IV). Distressed infants with congenital heart block generally have HRs below 50 bpm. If an infant is distressed with a rate greater than 50 bpm, one should suspect significant CHD or some other associated problem such as infection or sepsis in addition to the heart block. The infant in extremis from a slow HR may require immediate temporary pacing in the ED. This may be accomplished by the transcutaneous route or by the transthoracic route. The transcutaneous route, which was first introduced by Zoll in 1952, has recently been reintroduced. This technique involves the use of pacing electrodes placed on the anterior and posterior chest attached to an external current source. Temporary transthoracic pacing is available in most cardioverter defibrillator units. This technique may be used successfully in critical situations to increase the HR but should be replaced as soon as possible with another type of pacemaker because third-degree burns have been noted under the pacing electrodes after short periods in infants. Special wires available for transthoracic pacing can be placed by the subxiphoid route in infants and children. The procedure uses techniques similar to those used for pericardiocentesis. A pacing wire is inserted through a needle that subsequently is removed once the wire is inside the heart. This type of pacing should be replaced by a transvenous pacemaker once the patient is stable. If time allows, placement of a temporary transvenous pacemaker either through the umbilical vein or femoral vein (see Procedures in Section VII) under direct fluoroscopic observation in a cardiac catheterization laboratory is preferred. Temporary transvenous pacing is reserved for infants with signs of CHF, most commonly seen with HRs under 50 bpm or with slightly higher rates in association with a structural congenital heart defect. However, an infant with an HR of 45 bpm should not be paced solely on the basis of HR but should be observed

for signs of CHF such as tachypnea, poor feeding, or hepatomegaly. The width of the QRS on the ECG does not always correlate with the need for a pacemaker, although wider QRS rhythms often are associated with lower escape rates. Only a small percentage of infants with congenital heart block require pacing, and many of these escape the need for pacemakers until they are older.

Drug	Route	Dose
Epinephrine	IV	0.01–0.05 mg/kg (0.1 mL/kg of 1:10,000 solution or 0.01 mL/kg of 1:1,000 solution)
	Infusion	0.1–2.0 μg/kg/min
Isoproterenol	Infusion	0.1–2.0 μg/kg/min

Table 82.12. Treatment of Heart Block

The older child with congenital complete heart block may also present with symptoms associated with CHF. More commonly, dizziness, presyncope, syncope, exercise limitation, or fatigue are the presenting complaints in the older child. At times, the appearance of a ventricular arrhythmia is the presenting sign of difficulty in these patients.

Acquired Nonsurgical Heart Block

Acquired nonsurgical heart block may be idiopathic or associated with congenital heart defects, infectious diseases such as myocarditis (viral or Lyme) or endocarditis, inflammatory processes (lupus, rheumatic fever), Kawasaki disease, muscle diseases, cardiac tumors, or cardiac sclerosis. The emergency treatment for congenital or acquired nonsurgical heart block is similar. Subsequent implantation of a permanent pacemaker is based on the symptoms, CHF, or decreased CO and signs such as low HRs and pauses in rhythm. Resolution of congenital heart block may occur in some inflammatory processes. A single typical Stokes-Adams or syncopal attack not related to neurologic or endocrinologic factors is considered an indication for pacemaker insertion because these attacks may be fatal. The patient with acquired heart block who presents with a syncopal episode requires a temporary transvenous pacemaker. This should be left in place during induction of anesthesia for permanent pacemaker implantation because serious arrhythmias have been noted to occur at this time if adequate CO is not maintained by pacing.

Pharmacologic therapy plays a role if adequate ventilation, oxygenation, and treatment of acidosis does not produce a normalization of the CO as reflected by the BP and peripheral perfusion. The initial drug to be used should be isoproterenol (Table 82.12) because it is most effective in increasing the HR. Adequate intravascular volume should be maintained during isoproterenol infusion because its vasodilatory effect may result in lowered BP. Epinephrine may be tried in place of, or in addition to, isoproterenol if bradycardia persists.

Temporary transvenous pacing may be required during the acute phase of an infectious process, even when permanent pacing is not needed, as in myocarditis. The presence of CHF is an additional indication for pacing.

Postsurgical Complete Heart Block

Postsurgical heart block is less common today than in the early days of surgery for congenital heart defects, with a current incidence of less than 1%. Improved knowledge of the location of the conduction system, as well as the implementation of intraoperative mapping techniques, has helped decrease this serious postsurgical complication. Postsurgical complete heart block may be transient or permanent heart block. Transient complete heart block generally resolves within 8 days. Permanent complete heart block generally presents immediately after surgery but may not occur until many years after surgery. All patients with postsurgical permanent complete heart block should have implantation of permanent pacemakers. Emergency treatment of symptomatic postsurgical complete heart block includes pharmacologic support and temporary pacemaker placement until a permanent pacemaker can be placed.

Sinus Bradycardia

Sinus bradycardia is a HR below the normal range for age (Table 82.11). An ECG is necessary to rule out second-degree or complete heart block; P waves with a normal P-R interval must precede each QRS complex in sinus bradycardia. Sinus bradycardia is commonly associated with sinus arrhythmias. It often occurs in the athletic child or in the adolescent as a normal variant, especially during sleep. Other causes of sinus bradycardia include hypothyroidism, increased ICP, and drugs such as propranolol or digoxin. Therapy of the underlying disorder is indicated, but in symptomatic patients atropine may be useful as a temporizing measure (Table 82.13). Isoproterenol or epinephrine also may be given in this emergency setting (Table 82.12).

Test	Expected Normal Response
Atropine (0.02–0.04 mg/kg)	HR >90 bpm
	>25–50% increase in HR
Isoproterenol (1–3 μg/min)	>25% increase in HR
Exercise	95% of expected normal rate
Electrophysiology study	Normal CSNRT (<550 msec)
	Normal SACT (45–105 msec)
24-hr ambulatory monitor	Normal low rate for age
	Pauses <3 sec

CSNRT, Corrected sinus node recovery time; *HR*, heart rate; *SACT*, sinoatrial conduction time.

Table 82.13. Evaluation of Sick Sinus Syndrome

Sick Sinus Syndrome

Sick sinus syndrome is a condition in which sinus node function is depressed and may present with a sinus bradycardia or a slow junctional rhythm often in association with alternating episodes of tachycardia. Syncopal episodes may occur. This abnormal cardiac rhythm may be seen in children who have undergone atrial surgery for closure of an atrial septal defect, particularly of the sinus venosus type, the Mustard procedure for correction d-transposition of the great arteries, Fontan repair for single ventricle complexes, or in association with a viral myocarditis, or as an idiopathic occurrence. Table 82.13 outlines the evaluation of the child with a suspected sick sinus syndrome. Evaluation will generally be performed by a pediatric cardiologist.

The urgency of the clinical picture determines the treatment of the child with sick sinus syndrome. The asymptomatic patient with a slow HR can be referred for consultation with a cardiologist. The child with CHF or inadequate perfusion from bradycardia or tachycardia requires therapy directed at the specific arrhythmia and admission to the hospital. Isoproterenol or epinephrine infusions (Table 82.12) may increase the HR temporarily in a child with bradycardia but, in this situation, also may precipitate tachyarrhythmias; thus, these drugs should be administered cautiously under continuous monitoring. Symptomatic slow rhythms may require temporary or permanent cardiac pacing.

Pacemakers

Pacemakers are being used more often in infants and children for the treatment of congenital or acquired (usually postsurgical) complete heart block or sick sinus syndrome. In addition, some patients with tachyarrhythmias or with the long Q-T syndrome are being treated with pacemakers. Therefore, it has become important for the emergency physician to recognize the normal and abnormal function of a pacemaker.

Pacemakers have become complex. They may be single-chamber or dual-chamber units. The pacemaker may sense in a chamber, pace in a chamber, be inhibited by a chamber's activity, or perform all functions. Newer pacemakers are rate responsive and sense external factors such as motion or minute ventilation, with subsequent increases in HR.

In pediatrics, the most common pacemaker in the very young is still the pacemaker that senses and paces the ventricle. Motion-sensing, rate-responsive pacers are less useful in young infants than in older children. Rate-responsive and dual-chamber pacemakers that pace or sense in the atrium and the ventricle are being used commonly in children old enough to have two endocardial or epicardial wires placed. Examples of normal function, failure to capture, and failure to sense are shown in Figure 82.9.

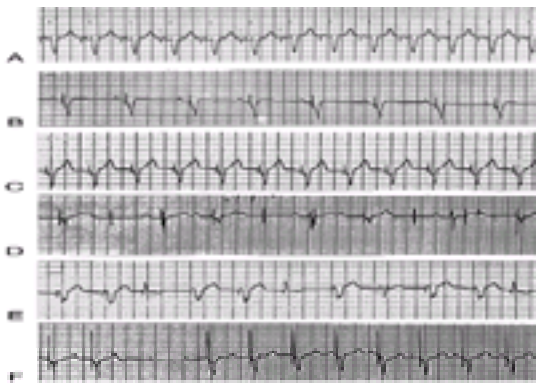


FIGURE 82.9. A. Electrocardiogram showing pacemaker that paces atrium and after 150 msec paces ventricle. Note pacemaker stimulus artifact before the P wave and the QRS complex. **B.** Electrocardiogram showing pacemaker that senses atrium and paces ventricle after a 150-msec delay. Note pacemaker stimulus artifact only precedes the QRS complex. **C.** Electrocardiogram showing ventricular pacing at 100 bpm with normal capture. Note pacemaker stimulus artifact preceding the QRS. **D.** Electrocardiogram showing ventricular pacing at 85 bpm with intermittent failure of capture. Note several pacemaker stimulus artifacts are not followed by a QRS complex, indicating failure of capture. **E.** Electrocardiogram showing ventricular pacing at 90 bpm, showing normal sensing of patient's intrinsic rhythm. **F.** Electrocardiogram showing ventricular pacing at 100 bpm, showing inappropriate sensing and failure to stimulate the heart secondary to wire fracture.

When a pacemaker malfunction is suspected, the specific problem should be identified if possible. A chest radiograph should be obtained to look for wire fractures or lead displacement. Most pacemaker programs now have computerized programmable analysis systems to help identify the problem. An ECG lead that shows the largest possible pacemaker stimulus artifact should be chosen with multiple leads providing more information. A pacemaker stimulus that falls outside the cardiac refractory period and fails to result in a ventricular depolarization indicates a failure of capture. In the currently available pacemakers, the output of the pacemaker may be reprogrammed externally, often resulting in normal capture. As the battery generator is depleted, the rate on most pacemakers decreases to a predetermined end-of-battery life indicator, which reveals impending battery failure.

An abnormally long pause or an earlier-than-expected paced complex indicates a sensing failure, either inappropriate sensing of another electrical signal (e.g., T wave sensing instead of QRS) or failure to sense the QRS. Sensing errors can be identified and external reprogramming accomplished.

Any patient with evidence of pacemaker malfunction should be admitted to the hospital if the problem cannot be resolved in the ED. A consultation with a pediatric cardiologist is generally required to troubleshoot and correct pacemaker problems. The patient with pacemaker malfunction who has symptomatic bradycardia should be managed with the regimens previously discussed Table 82.11.

The procedure for inserting a temporary pacemaker is described in Section VII.

Fast Heart Rates

Supraventricular Tachycardia

Background

Paroxysmal SVT, previously called paroxysmal atrial tachycardia (PAT), is the most common significant arrhythmia seen in pediatric practice. Paroxysmal SVT describes a group of arrhythmias with similar ECG features but different mechanisms. These mechanisms have been clarified by the use of specialized intracardiac pacing and recording techniques known as the electrophysiologic study and have been shown to originate in the sinus node, atrium, and approaches to the AV node or in junctional conduction tissue or to be associated with reentry using atrioventricular accessory pathway.

A typical ECG seen in a pediatric patient with SVT is shown in Figure 82.10. Note the rapid rate and the narrow QRS complex. The P waves are different from the usual sinus P wave but may be obscured by the ST segment and not be visible at all. The rate of tachycardia in infants ranges from 220 to 320 bpm. Older children have tachycardia rates that range from 150 to 250 bpm. Supraventricular tachycardia with aberrancy has an ECG with a wide QRS complex and may resemble VT.

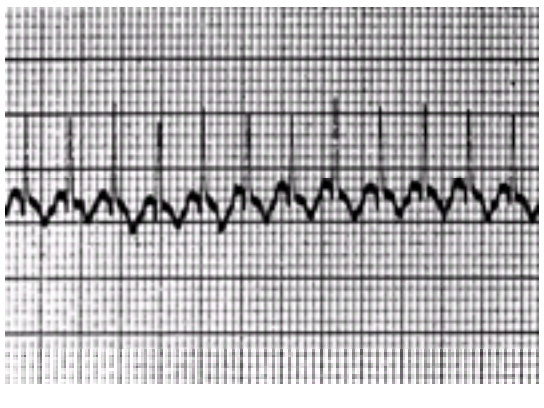


FIGURE 82.10. Supraventricular tachycardia, rate 300 bpm.

The infant with SVT is most commonly less than 4 months of age and is more likely to be male (male:female ratio is 3:2). Although 50% of cases in infants have been classified as idiopathic, 24% have associated conditions such as infection, fever, or drug exposure (most commonly cold medications containing sympathomimetic amines), 23% have CHD (Ebstein's anomaly or corrected transposition are most common), and 22% have WPW syndrome. Those classified as idiopathic generally are considered to have concealed accessory pathways or other reentrant circuits. Older children with SVT are more likely to have WPW, concealed bypass tracts, or CHD.

Pathophysiology

Electrophysiologic catheterization techniques have provided much information about the mechanisms of SVT and have established that SVT actually is several arrhythmias, including 1) those caused by reentry within the sinoatrial node, atrium, AV nodal approaches, and accessory pathways, including the WPW syndrome; and 2) those caused by enhanced automaticity of specialized atrial fibers. The supraventricular arrhythmias most commonly seen in pediatric patients include AV nodal reentrant SVT (usually younger than 5 years of age), WPW with SVT, concealed bypass tracts with SVT, atrial flutter or atrial fibrillation, and ectopic atrial tachycardia.

The concept of AV nodal reentry as a mechanism for SVT was first proposed by Mines in 1913, and the presence of dual

pathways in the AV node was shown subsequently by Moe and colleagues. Recently, programmed stimulation has demonstrated the role of reentry in SVT. By convention, the dual pathways have been labeled a and b. The a pathway is slower conducting but has a shorter refractory period than the faster conducting b pathway. The application of this concept to human SVT is shown in Figure 82.11. During sinus rhythm, the atrial impulse traverses the faster conducting b pathway to produce a single QRS complex. The impulse travels simultaneously down the a (slow) pathway, reaching the His bundle shortly after it has been depolarized and rendered refractory by the impulse that was conducted down the b pathway. In response to an atrial premature depolarization, the impulse is blocked in the b pathway as a result of its longer refractory period and proceeds slowly down the a pathway. If conduction down the a pathway is slow enough to allow the previously refractory b pathway time to recover, a single atrial echo results. An earlier atrial premature depolarization (Fig. 82.11) also blocks in the b pathway, conducts slowly down the a pathway and arrives later to conduct retrograde through the b pathway back to the a pathway with antegrade conduction producing a sustained AV nodal reentrant tachycardia.

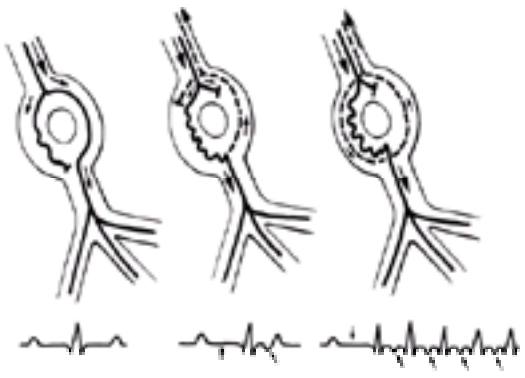


FIGURE 82.11. Schematic representation of conduction pathway and corresponding ECG in the development of AV nodal reentry. See text for full discussion. *LBB,* left bundle branch; *RBB,* right bundle branch.

If conduction delay and refractoriness in both pathways are appropriate, a continuously circulating wavefront of electrical activity ensues, resulting in a reentrant tachycardia. The conditions for reentry may be found in the sinus node, atrium, AV node and accessory pathways, and the Purkinje fibers of the ventricles. The most common form of SVT in pediatrics is caused by sustained reentry within the AV nodal approaches or a bypass tract (accessory pathway).

The rate of SVT appears to reflect the conduction properties of the AV node and bypass tract when involved. Patients whose AV nodes or bypass tracts conduct slowly, either spontaneously or from the effects of drugs like digitalis or propranolol, have slower rates during SVT.

SVT is the usual clinical problem that affects patients with the WPW syndrome. The arrhythmia has all the characteristics of reentry but the reentrant circuit involves an extra AV accessory pathway. WPW complexes, which consist of a short P-R interval and a widened QRS complex with a slurred upstroke (delta wave), generally are not seen during the tachycardia, but only after conversion to normal sinus rhythm as shown in Figure 82.12. The WPW complex represents the presence of a bypass tract connecting atria and ventricles. The short P-R interval and delta wave characteristic of the WPW syndrome are produced by conduction over the accessory pathway, which has different electrophysiologic properties from the normal AV conduction system. The ventricular complex is a fusion beat with a variable contribution from conduction through the accessory pathway and the AV node. The greater the contribution from the accessory pathway, the larger the delta wave and more bizarre the QRS.

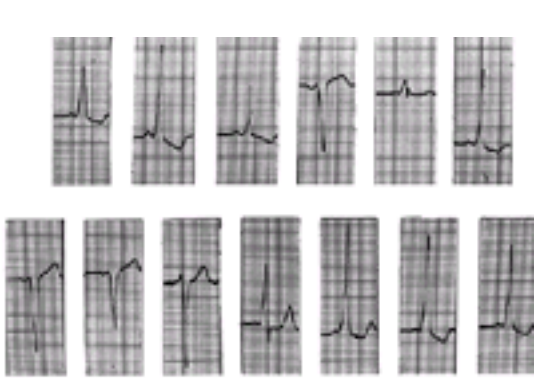


FIGURE 82.12. Electrocardiogram in Wolff-Parkinson-White (WPW) syndrome. Note wide QRS, presence of delta wave (slurred upstroke of R), and short P-R interval.

Episodes of SVT in WPW syndrome usually are initiated by a premature atrial depolarization that blocks antegrade conduction in the accessory pathway and travels to the ventricles over the normal AV conducting system. The impulse, on reaching the ventricular insertion of the bypass tract, can travel retrograde up the bypass tract to the atrium and reenter the AV node to start a “circus movement” or reentrant type of tachycardia.

Therefore, reentrant SVT in WPW syndrome is analogous to AV nodal reentrant SVT with the bypass tract functioning like a b pathway (fast conduction, long refractory period) and the AV nodal-His-Purkinje system functioning like an a

pathway (slow conduction, short refractory period). Because antegrade conduction during SVT occurs over the usual AV conducting system, the QRS complexes will be normal. In rare cases, the route of reentry may be reversed, and the bypass tract forms the antegrade limb. In these cases, the QRS complexes will be wide and bizarre, and the arrhythmia may simulate VT.

A concealed bypass tract indicates that the bypass tract is used only as the retrograde limb of the reentrant circuit during SVT but is not used for antegrade conduction during normal resting rhythm. Thus, the resting ECG appears normal.

Because the ventricle must be depolarized before retrograde conduction up the bypass tract, atrial activation must always follow ventricular activation; therefore, the P wave follows inscription of the QRS complex. The P-R interval is usually less than 50% of the R-R interval. In AV nodal reentrant SVT, the P wave may or may not be visible but, if visible, is generally closely related to the preceding QRS complex.

The identification of the mechanism of the tachycardia in SVT is helpful from a therapeutic point of view so that a medication known to act specifically on the AV node, on the accessory pathway, or on the atrial tissue may be chosen. In addition, if the tachycardia can be initiated reproducibly in the electrophysiology laboratory, the efficacy of a specific drug can be tested in the laboratory rather than empirically chosen.

Clinical Manifestations

The clinical findings in the patient with SVT depend on the duration of the arrhythmia and the presence or absence of an underlying heart defect or myocardial dysfunction. In the patient with no congenital heart defect or myocardial dysfunction, CHF usually appears only after 24 hours of a rapid rate. However, when the patient is first seen in the ED, the precise onset of the SVT is rarely certain. Likewise, the presence of associated heart defects generally is unknown. Therefore, all patients must be treated with some degree of urgency.

The infant with SVT may present with only a fast rate or have varying degrees of CHF (poor feeding, irritability, respiratory distress) or shock. The infant may be acidotic and appear to be septic.

The child older than 5 or 6 years of age will usually complain of some symptom such as chest pain or a rapid heartbeat. The child may also present with signs of CHF but is unlikely to be quite as ill when first seen as the infant or young child, unless a congenital heart defect or primary myocardial disease is present.

Management

Because the mechanisms of SVT are now identifiable, treatment can be chosen for the specific forms of the disease. However, the main principle of treatment in any cardiac arrhythmia is that the type of therapy is determined by the urgency of the situation. Thus, a different mode of treatment is chosen for the patient with tachycardia in shock than for the asymptomatic patient who has only a fast HR (Table 82.14).

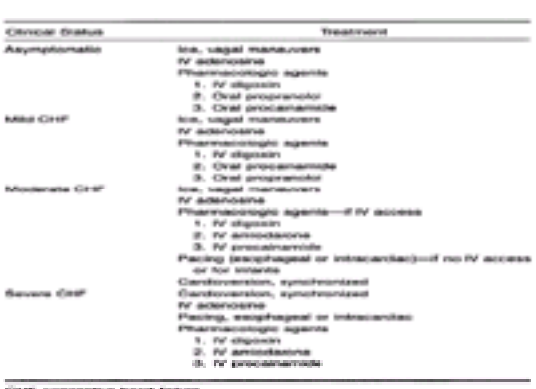
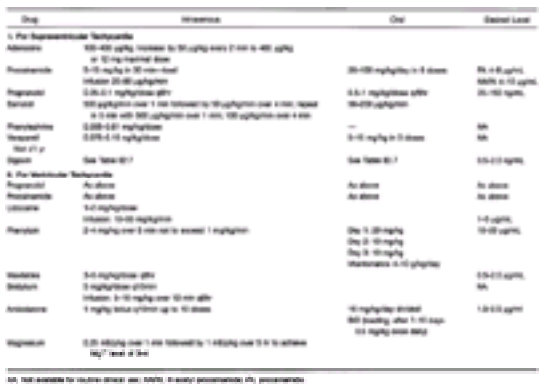


Table 82.14. Treatment of Supraventricular Tachycardia

When the SVT is caused by reentry using the AV node (AV nodal reentry), or AV reentry using a bypass tract, any intervention that interrupts the critical relationship of conduction and refractoriness in the AV node can interrupt the tachycardia. The patient's response to the drugs or maneuvers is not always predictable. The methods used most often are those that slow AV nodal conduction. In adult patients, carotid sinus pressure or the Valsalva maneuver often can terminate the tachycardia by increasing vagal tone, slowing conduction and prolonging refractoriness within the AV node. Although these maneuvers often are ineffective in children, the occasional successful attempt justifies their initial use. Ice water or ice bags applied to the face have been used to recruit the diving reflex and stop the SVT. This technique should be reserved for children who are monitored, with particular caution used in applying these techniques in young infants because significant sinus slowing may occur.

A rapid pharmacologic treatment now available is adenosine. It has a rapid onset of action, usually within 10 seconds, and a short half-life, with side effects lasting less than a minute and rarely being serious. Adenosine may be used in acutely ill patients but should not delay immediate cardioversion in severely compromised patients. Appropriate doses are shown in Table 82.15.



Drug	Intravenous	Oral	Desired Level
I. For Supraventricular Tachycardia			
Adenosine	100–400 µg/kg, increase by 50 µg/kg every 2 min to 400 µg/kg or 12 mg maximal dose		
Procainamide	5–15 mg/kg in 30 min—load Infusion 20–80 µg/kg/min	20–100 mg/kg/day in 4 doses	4–8 µg/mL NAPA in 10 µg/mL
Propranolol	0.25–0.1 mg/kg/dose q6h	0.5–1 mg/kg/dose q6h	25–100 ng/mL
Esmolol	500 µg/kg/min over 1 min followed by 50 µg/kg/min over 4 min; repeat in 5 min with 500 µg/kg/min over 1 min, 100 µg/kg/min over 4 min	50–200 µg/kg/min	
Phenylephrine	0.005–0.01 mg/kg/dose	—	NA
Verapamil Not <1 y	0.075–0.15 mg/kg/dose	3–10 mg/kg in 3 doses	NA
Digoxin	See Table 82.7	See Table 82.7	0.5–2.0 ng/mL
II. For Ventricular Tachycardia			
Propranolol	As above	As above	As above
Procainamide	As above	As above	As above
Lidocaine	1–2 mg/kg/dose Infusion: 10–50 mg/kg/min		1–5 µg/mL
Phenytoin	2–4 mg/kg over 5 min not to exceed 1 mg/kg/min	Day 1: 20 mg/kg Day 2: 10 mg/kg Day 3: 10 mg/kg Maintenance: 4–10 g/kg/day	10–20 µg/mL
Mexiletine	5–6 mg/kg/dose q6h		0.5–2.0 µg/mL
Bretylium	5 mg/kg/dose q12h Infusion: 5–10 mg/kg over 10 min q6h		NA
Amiodarone	5 mg/kg bolus q12h/d up to 10 doses	10 mg/kg/day divided Still loading, after 1–10 days: 5–10 mg/kg once daily	1.0–3.0 µg/ml
Magnesium	0.25–50 mEq/kg over 1 min followed by 1 mEq/kg over 3 h to achieve Mg2+ level of 3 mg		

NA, Not available for routine clinical use; NAPA, N-acetyl procainamide; PA, procainamide.

Table 82.15. Antiarrhythmic Agents

Any patient with SVT who presents with shock, acidosis, or severe hemodynamic compromise should be treated immediately. If adenosine is not effective, synchronized direct current (DC) cardioversion at a dosage of 1 to 2 watt-sec/kg should be used and doubled until effective or until a dosage of 10 watt-sec/kg is reached. If ventricular fibrillation should occur, repeat cardioversion generally converts the patient to normal sinus rhythm. The patient should be given a sedative or short-acting anesthetic, and preparations should be made for airway support and ventilation if needed. The underlying acidosis should be treated and adequate ventilation and oxygenation provided because cardioversion may not be successful in the presence of hypoxia or acid-base imbalance. The presence of digoxin in the patient should not prevent the use of cardioversion when needed. Evidence of digoxin toxicity such as ventricular arrhythmia may be treated with lidocaine. Once the patient's rhythm has been converted, the chosen chronic treatment should be initiated immediately.

Children who have only mild to moderate failure may be treated medically, as described earlier, with adenosine if vagal maneuvers fail because most of these patients will convert rapidly after pharmacologic treatment. Agents, other than digitalis, useful for the treatment of SVT are reviewed in Table 82.15. The preferred medical treatment of AV nodal reentrant SVT in children is digoxin, which works by prolonging AV nodal conduction and refractoriness in both the fast (b) and slow (a) pathways. When refractoriness of the fast pathway is prolonged greater than conduction down the slow pathway, SVT cannot be initiated.

The usual digitalizing dose appropriate for age is used (Table 82.7). The route (IV or IM) depends on the status of the patient. IV digoxin should be used in the presence of CHF when perfusion is decreased. As noted earlier, the IV dose should be calculated to be 75% of the oral or IM dose. The interval of time that precedes the second and third doses of the total digitalizing dose should be determined by the patient's status. These additional doses may be required after only 2 to 4 hours. If the tachycardia persists after three doses, one to two additional doses may be given.

Propranolol, which prolongs AV nodal conduction and refractoriness in both a and b pathways, should be used with caution in the ill child because it may depress cardiac function even more. In instances in which cardiac function is preserved, a slow IV dose may be given. The dosage is 0.1 mg/kg IV. A shorter acting b blocker, esmolol, may be preferred. The doses are shown in Table 82.15. Digoxin seems to be slightly more effective than propranolol in treating SVT. Raising the blood pressure with a-adrenergic agents such as phenylephrine can terminate the SVT by stimulating the vagus through the baroreceptor reflexes (Table 82.15). Termination of AV nodal reentrant SVT by these agents and by vagal maneuvers almost always occurs by gradual slowing and then antegrade block in the slow (a) pathway. In most instances, the tachycardia is terminated by a so-called nonconducted atrial echo.

Procainamide may be used under the guidance of a cardiologist to terminate AV reentrant SVT by blocking retrograde conduction in the fast (b) pathway (Table 82.15). SVT can be converted in the catheterization laboratory by rapid atrial pacing. An electrode catheter is placed in the high right atrium. Rapid atrial pacing (faster than the SVT rate) often interrupts the reentrant cycle and results in normal sinus rhythm. Even when normal sinus rhythm cannot be achieved, a slower rate may be obtained if 2:1 AV nodal block is produced by rapid atrial pacing.

A less invasive method of esophageal overdrive pacing to terminate SVT in infants and children can be used. A small bipolar electrode catheter is passed by the nasogastric route and positioned in the esophagus behind the left atrium. As with intracardiac methods, esophageal rapid atrial pacing at a rate faster than the SVT can capture the atrium and interrupt a reentrant circuit, terminating the SVT.

The treatment of SVT in WPW syndrome or in a concealed bypass tract is similar to the treatment of AV nodal reentrant SVT because the AV node participates in one limb of the reentrant circuit. Propranolol is an exceptionally effective drug because it slows AV nodal conduction and prolongs refractoriness while having no significant effects on the accessory pathway. Digoxin, with similar effects on the AV node, must be used cautiously in patients with WPW syndrome because it can shorten the refractory period of the bypass tract and enhance conduction in the accessory pathway. In a patient with an associated atrial tachyarrhythmia such as atrial flutter or atrial fibrillation, digoxin may predispose to a rapid ventricular response or ventricular fibrillation secondary to rapid conduction of the atrial impulse down the bypass tract. A high percentage of adults with WPW syndrome and SVT also have atrial fibrillation, but the incidence of this association is lower in children. Class IA or IC agents such as procainamide or flecainide are often effective in this type of SVT because they act on the accessory pathway to slow conduction and prolong refractoriness and thus interrupt the retrograde limb of the reentrant circuit.

Although verapamil has been an effective treatment of SVT in adults, serious problems, including hypotension, cardiovascular collapse, and death, have occurred with its use in pediatric patients. This is especially true for patients less than 1 year of age and those in CHF. Therefore, verapamil generally should be avoided in patients less than 3 years

old. Even in older patients, other regimens should be tried first. When used, a dose of 0.075 to 0.15 mg/kg is effective. It is important to give the drug slowly, over at least 2 minutes, while monitoring the child's ECG and BP closely. IV calcium (10% CaCl at a dose of 10 mg/kg) and isoproterenol should be available immediately and should be drawn up in the appropriate dose before verapamil is given. Some recent studies suggest that pretreatment with calcium may prevent the severe hypotension, but this is not the case universally. Verapamil should not be used in patients who have received IV b blockers and should be used with extreme caution in patients who use any other antiarrhythmic agent.

As soon as the patient converts from SVT, an ECG in sinus rhythm should be obtained. This allows diagnosis of the presence of the WPW syndrome or other abnormalities that might direct chronic therapy to be identified. In general, digoxin is not advised in the presence of WPW syndrome unless one knows that the accessory pathway refractory period is relatively long and is unaffected by the digoxin; this drug might shorten the accessory pathway refractory period and promote rapid ventricular conduction of supraventricular impulses to the ventricle. This is particularly problematic in the presence of atrial flutter or fibrillation and WPW syndrome. In the absence of associated atrial arrhythmias and the presence of CHF, digoxin may be used temporarily in these patients until cardiac function improves. The patient may be switched from digoxin to a b blocker at that time.

Infants with SVT should receive maintenance treatment for the first year of life, even if there are no recurrences. This type of therapy should be instituted in the hospital on continuous ECG monitoring to observe for adverse side effects of the antiarrhythmic agent. Studies have shown that only 20 to 30% of patients will have recurrences if the medicines are continued for this period. Treatment is also advised for infants who convert spontaneously from documented episodes of SVT because these children are predisposed to recurrences. Older children or infants, who are difficult to control and have multiple recurrences that often require combinations of digoxin and propranolol or digoxin and procainamide, must be managed individually and should not have their medicines routinely discontinued after 1 year. Children with SVT should not be treated for upper respiratory tract infections with sympathomimetic amines. Instead, if needed, pure antihistamines, such as those listed in Table 82.16 should be used.

Chlorpheniramine Maleate (Chlor-Trimeton—Schering) (Teldin—SKF)	For Coughs: Robitussin
Brompheniramine Maleate (Dimetal—Robbins)	Robitussin DM
Tripolidine HCl (Actidil—Burroughs Wellcome)	Robitussin with codeine—need a prescription
Promethazine HCl (Phenergan—Wyeth)—need a prescription	Phenergan with codeine—need a prescription
Diphenhydramine (Benadryl—Park Davis)	Terpin Hydrate with codeine—need a prescription
	Claritin (Loratadine—Schering)
	Zyrtec (Cetirizine HCl—Pfizer)
	Allegra (Exofenadine—Hoechst)

Table 82.16. Preferable Agents for Treatment of Upper Respiratory Infection in Children with Supraventricular Tachycardia

Atrial Flutter and Atrial Fibrillation

Atrial flutter and fibrillation occur uncommonly in children. Atrial flutter consists of rapid, regular atrial excitation at rates of 280 to 480 bpm (Fig. 82.13). The ventricular response depends on AV nodal conduction that may allow 1:1, 2:1, 3:1, or 4:1 conduction. The typical ECG reveals saw-toothed flutter waves best seen in leads 2 and V_1. Atrial flutter is most commonly seen in children with CHD, especially postoperatively after Mustard repair for d-transposition of the great arteries or the Fontan repair, but it also can occur idiopathically or congenitally.

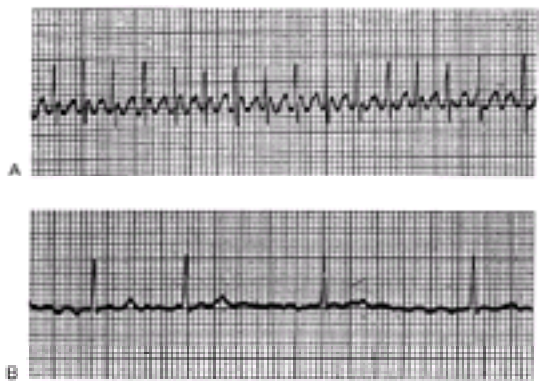


FIGURE 82.13. A. Atrial flutter. Sawtooth baseline is apparent. Regular QRS with ventricular rate of 250. **B.** Atrial fibrillation. Irregularly irregular QRS with course erratic baseline undulations representing fibrillatory waves.

Atrial fibrillation consists of totally disorganized rapid atrial activity (at a rate of 400 to 700 bpm) with a variable ventricular rate secondary to varying AV block. Atrial fibrillation is seen most commonly in adolescents with long-standing rheumatic or congenital mitral disease or in patients with hyperthyroidism.

Children with atrial fibrillation or flutter raise the same therapeutic problems as those with SVT. If cardiac compromise

does not necessitate immediate cardioversion, the initial treatment is digoxin. Adenosine increases the AV block and therefore may help diagnostically, but it usually will not correct the atrial flutter or fibrillation to sinus rhythm. In the child who is stable and has a normal BP and adequate perfusion, the physician should allow 24 hours for a response to digoxin before adding a second drug such as procainamide. Failure to achieve a normal rhythm after an additional 24 to 48 hours calls for cardioversion. Therapeutic drug levels for these agents, which should be obtained in a steady state of drug administration, are listed in Table 82.15.

Automatic Atrial or Junctional Tachycardia

Automatic atrial or junctional tachycardias may be difficult to control and often are associated with inflammatory states such as myocarditis. Digoxin (at relatively low dosages initially to avoid ventricular arrhythmias in a sensitive myocardium), IV procainamide, and IV amiodarone have been effective (Table 82.15). Once the abnormal rhythm has converted, chronic therapy must be initiated, otherwise, the arrhythmia is likely to recur. Chronic drugs for these arrhythmias include digoxin, b blockers, procainamide, flecainide, amiodarone, and sotalol.

Ventricular Tachycardia

VT is defined as three or more consecutive PVCs (Fig. 82.14). The HR usually is 150 to 200 bpm but may be slower or more rapid. These contractions may be hemodynamically inefficient and result in syncope and death. The cause may be electrolyte imbalance, metabolic disturbances, cardiac tumors, drugs, cardiac catheterization or surgery, CHD, cardiomyopathies, acquired heart disease, right ventricular dysplasia, prolonged Q-T syndrome, or idiopathic.

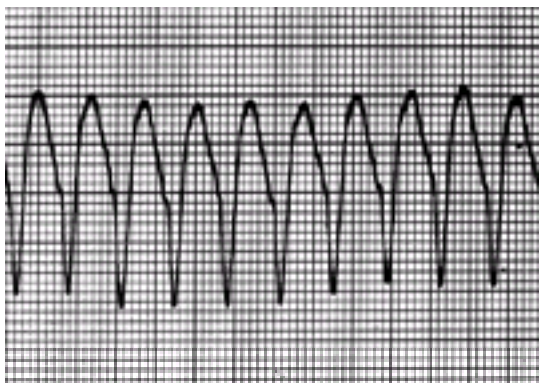


FIGURE 82.14. Ventricular tachycardia. Wide QRS with rate of approximately 250 bpm; sinusoidal pattern.

VT is being seen more commonly in children who can be divided into three groups: 1) patients with identifiable noncardiac causes (electrolyte imbalance, toxins, drug overdoses, or drug toxicity), 2) patients with no known underlying heart disease and no extracardiac disturbances (may have small Purkinje cell tumors or foci in the right ventricular outflow tract or left ventricle septum), and 3) patients with either congenital or acquired heart disease.

In children with identifiable extracardiac abnormalities (group 1), the underlying disturbance is treated. Children with no known cardiac or extracardiac causes for VT (group 2) require treatment if they have a sustained, rapid arrhythmia. As with SVT patients, the urgency of treatment depends on the clinical status. In cases of shock, impending cardiac decompensation, or cardiac failure, synchronized DC cardioversion at 2 to 5 watt-sec/kg up to 10 watt-sec/kg should be used. IV lidocaine at 1 to 2 mg/kg also may be effective (Table 82.15); a continuous infusion of lidocaine at 10 to 50 µg/kg per minute may be required. Other effective drugs include intravenous amiodarone at 5 mg/kg over the first hour, followed by a 5 to 10 µg/kg per minute IV infusion or intravenous procainamide at 10 to 15 mg/kg given over 30 to 45 minutes, followed by a 20 to 80 µg/kg per minute IV infusion. Those with Purkinje cell tumors or ectopic foci may be amenable to catheter radiofrequency ablation or surgical ablation in selected cases.

Rapid ventricular pacing may be used for overdrive suppression for conversion to normal rhythm if pharmacologic therapy fails or is contraindicated. Chronic treatment with nadolol, propranolol, mexiletine, amiodarone, procainamide, sotalol, or propafenone has been effective for long-term control of this arrhythmia.

No data exist that determine whether asymptomatic patients with normal hearts and idiopathic slow VT should be treated with antiarrhythmic agents. Certainly, the patient should not be made more toxic by the therapy than by the arrhythmia. Treatment should be determined by hemodynamic status and potential problems that might occur from the arrhythmia.

Patients with CHD (group 3) and VT raise a different issue. Often, these patients have some degree of hemodynamic compromise associated with ventricular dysfunction and do not tolerate the VT at all. The emergent and chronic treatment is similar to that outlined previously but may be required even more urgently. All patients with CHD and sustained VT over 150 bpm require therapy because sudden death occurs in up to 30% of patients with CHD who have VT. Some patients in this group with slower VT or nonsustained VT may require treatment, especially if hemodynamic abnormalities exist.

The congenital long Q-T syndrome is a special case. Sudden death occurs in 73% of patients who are not treated. The sudden death is secondary to ventricular tachyarrhythmias (torsades de pointes) of the type that often degenerates to ventricular fibrillation. Any patient who presents with VT, especially of the polymorphic or torsades de pointes type, should have corrected Q-T intervals determined in sinus rhythm. A genetic basis for long Q-T syndrome has now been described. A complete family history may reveal the occurrence of sudden death in young relatives, or a family history of syncope associated with exercise or emotional stress. A family history of hearing deficit may be elicited. Emergent

treatment of these patients includes lidocaine and synchronized DC cardioversion when needed. If the VT is polymorphic, nonsynchronized cardioversion or defibrillation is indicated. In addition, temporary atrial or ventricular pacing at a rate 10 to 20% faster than the underlying sinus rate may be needed to control the arrhythmia, especially in patients with underlying bradycardia, a common association. Intravenous propranolol, phenytoin, and magnesium have been used successfully in these patients. The class I agents that prolong the Q-T interval in normal patients should be avoided in patients with these long Q-T intervals. In fact, a number of drugs that may produce this form of VT are shown in Table 82.17. This type of drug effect is believed to be related to QTc prolongation with associated ventricular arrhythmias and bradycardia at times. Temporary pacing and removal of the offending agent are effective therapies. In acquired long Q-T with ventricular arrhythmias, especially if bradycardia is a prominent factor, an isoproterenol infusion may be therapeutic.

Antiarrhythmic Agents	*Psychotropic Drugs*
Quinidine	Tricyclic Antidepressant
Procainamide	Amitriptyline
Flecainide	Phenothiazines
Encainide	Haloperidol
Disopyramide	Risperidone
Amiodarone	Pimozide
Sotalol	Lithium carbonate
Antihistamines	Sertraline hydrochloride
Terfenadine	Nefazodone hydrochloride
Astemizole	Fluvoxamine maleate
Diphenhydramine	*Other*
Antibiotics/Antifungal Agents	Cisapride
Erythromycin	Probucol
Trimethoprim	Anthracyclines
Sulfamethoxazole	Organophosphates
Pentamidine	Epinephrine
Ketoconazole	Diuretics
Fluconazole	Potassium loss
Itraconazole	
Clarithromycin	
Azithromycin	

Table 82.17. Pharmacologic Agents That Prolong Q-T Intervals

Ventricular Fibrillation

Ventricular fibrillation consists of chaotic irregular ventricular contractions with cessation of circulation. Electrical defibrillation with correction of precipitating factors (acidosis, electrolyte imbalance, hypoxia) may result in conversion to normal sinus rhythm. The treatment of cardiac arrest is discussed in Chapter 1.

Irregular Heart Rates

Premature Depolarizations

Background

The primary irregular rhythm that may require attention is the PVC. PVCs are seen as premature, wide, bizarre-shaped QRS complexes. Generally, the T wave is opposite in direction to the main deflection of the QRS. A compensatory pause usually follows the premature beat, and P waves may reveal AV dissociation or retrograde conduction or may be absent. When a rhythmic pattern of PVCs is established, the designation of bigeminy, trigeminy, or quadrigeminy is made, depending on whether that beat followed every second, third, or fourth sinus beat. Nonsustained VT consists of three PVCs or more that last for 10 seconds or less.

Pathophysiology

PVCs often occur without identifiable cause in children and have been considered benign. However, PVCs are seen in children with CHD and CHF, viral myocarditis, Lyme myocarditis, cardiomyopathies, cardiac tumors, hemochromatosis, or electrolyte imbalance. They also are seen in association with various forms of drug administration, including general anesthesia, digoxin, sympathomimetic amines, and phenothiazine tranquilizers. PVCs may be precursors of VT or fibrillation.

Clinical Manifestations

Children who present with PVCs are often asymptomatic and unaware of their arrhythmia, especially if they are younger than 5 years of age. If the PVC is appreciated, the child may complain of a “skipped” or “hard” beat, a fluttering or pounding in the chest, difficulty breathing, or chest pain. If the PVCs are frequent and/or associated with heart disease (congenital or acquired), the child may note dizziness or a rapid heartbeat. Frequent PVCs in the presence of compromised cardiac function may worsen the CO and produce signs and symptoms of CHF.

Management

The only PVCs that require treatment are those that cause or are likely to cause hemodynamic compromise. This generally is seen in the context of frequent PVCs in a patient with abnormal cardiac function as evidenced by cardiomegaly on chest radiograph, abnormal cardiac function or dilated chamber sizes on echocardiography, or abnormal exercise responses on exercise stress testing. Symptoms of dizziness, chest pain, or presyncope may accompany PVCs in patients with abnormal cardiac function. Such patients may be found to have myocarditis, cardiomyopathy or a congenital heart defect (preoperative or postoperative), and abnormal underlying cardiac function. The treatment may include lidocaine, mexiletine, procainamide, propranolol, or amiodarone (Table 82.15) as outlined in the section on VT. Although digoxin is not generally considered for patients with PVCs or VT, patients with poor myocardial function, as evidenced by the clinical findings of CHF and echocardiographic evidence of abnormal

myocardial function, may benefit from digitalization.

Rapid nonsustained VT, usually with rates over 150 bpm, may require treatment even in the presence of a normal heart because of the tendency for symptoms to develop, especially if the abnormal rhythm is present more than 40% of the time.

Isolated multiform or coupled PVCs or nonsustained VT (rate 150 bpm or less) in an asymptomatic patient with a normal heart may not require treatment, but this decision must be individualized after consultation with a cardiologist. Rarely is emergency treatment of this type of patient required, and investigations that use 24-hour continuous ECG monitors, exercise stress testing, echocardiography, and/or the electrophysiologic catheterization studies may be used to determine the appropriate management. It sometimes is helpful to observe the patient's response to activity. If the PVCs abate completely with sinus tachycardia (140 to 150 bpm), they are likely to be benign in terms of clinical significance and need for therapy, although this is not universally true.

Isolated PVCs in an asymptomatic patient in the presence of a structurally and functionally normal heart do not require treatment. Continuous 24-hour ambulatory monitoring should be performed to rule out the presence of undetected VT or complex ventricular arrhythmias. Restriction of caffeine and other stimulants should be recommended in all patients with ventricular arrhythmias.

Premature atrial contractions (PACs) or PVCs generally do not require treatment. Patients with frequent PACs or PVCs should be evaluated appropriately to rule out myocarditis. Continuous 24-hour ambulatory monitoring should be performed to rule out the occurrence of SVT. Elimination of caffeine or other stimulants such as theophylline, pseudoephedrine, and other sympathomimetic amines may decrease the frequency of PACs. Unless the PACs are demonstrated to initiate episodes of SVT, no treatment is indicated. Variations of normal rhythms commonly seen in children and that do not require treatment include sinus arrhythmia and wandering atrial pacemakers as long as rates remain in the normal ranges.

First-Degree and Second-Degree Heart Block

First-degree heart block reflects slowed conduction from the sinus node to the ventricle and is manifested by a prolonged P-R interval. It is seen with digoxin and other antiarrhythmic drugs; certain types of CHD (primum and secundum atrial septal defects); and inflammatory diseases such as rheumatic, viral, or Lyme myocarditis.

Second-degree heart block results in the failure of some impulses to traverse the AV node. The Wenckebach phenomenon, a form of second-degree heart block, is a result of progressive slowing of AV conduction and is seen as progressively prolonged P-R interval and eventual dropped beat. Other forms of second-degree heart block include high grade 2:1, 3:1, and 4:1 block.

Children with first-degree and second-degree heart block rarely are symptomatic unless the associated HR is low enough to decrease the CO. In such instances, signs and symptoms of CHF may be present.

Both first-degree and second-degree heart block may be associated with digitalis toxicity, requiring the digitalis dose be adjusted downward or temporarily held if second-degree AV block is present. Otherwise, first-degree heart block does not need therapy. Second-degree heart block is treated only if it produces a HR sufficiently slow to interfere with cardiac output. In this instance, the management is the same as that outlined for complete heart block (p. 672).

Arrhythmias Associated with Electrolyte Abnormalities

Alterations in electrolyte concentrations may influence cardiac rate, rhythm, and automaticity and may lead to arrhythmias. Potassium and calcium abnormalities are the most common electrolyte alterations that produce arrhythmias, but abnormalities in magnesium and acid-base balance are also important. Commonly, a combination of ionic alterations is responsible for arrhythmias. Any patient with significant arrhythmias should be evaluated for an electrolyte disturbance. The ECG changes may be characteristic and lead to suspicion of a specific electrolyte abnormality. Normal ECG intervals (P-R, QRS, QTc) are listed in Table 82.18.

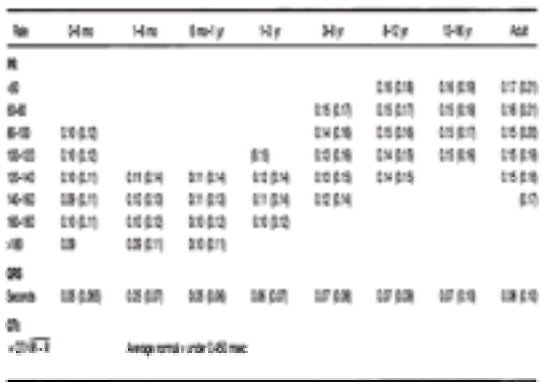


Table 82.18. P-R Interval and QRS Duration Related to Rate and Age (and Upper Limits of Normal)

Hyperkalemia

Hyperkalemia is common in hospitalized children and produces recognizable ECG alterations. Peaked T waves are seen at a serum concentration of 5 to 6 mEq/L, and the QRS widens with a concentration exceeding 6 mEq/L. The Q-T interval increases with the increasing QRS duration. As P wave amplitude decreases, P wave duration increases, and the P-R interval increases above 7 mEq/L. Above 8 to 9 mEq/L, P waves disappear, the ventricular rate becomes irregular, and severe bradycardia with sinus arrest, block, or idioventricular rhythms occur, often with a sinusoidal wave pattern. Ventricular fibrillation or asystole occurs at serum concentrations greater than 12 to 14 mEq/L. Low serum calcium enhances the myocardial toxicity of hyperkalemia. Likewise, acidosis potentiates hyperkalemia by producing potassium ion efflux from cells.

Hypokalemia

Serum potassium concentrations of less than 2.7 mEq/L generally produce typical ECG changes in ventricular repolarization. These changes include a U-wave amplitude greater than 1 mm, seen best in leads V_2 and V_3, and ST-segment depression greater than 0.5 mm. The Q-T interval lengthens and the T wave flattens with progressive hypokalemia. The P-R interval may be prolonged, and intraventricular conduction may be delayed with widening of the QRS complex. With significant hypokalemia, P-wave and QRS amplitude may increase. Other arrhythmias that have been associated with hypokalemia include ectopic atrial and ventricular complexes, ectopic atrial tachycardia with block, AV dissociation, second-degree AV block, ventricular bigeminy, VT, and ventricular fibrillation.

Patients taking digoxin who become hypokalemic are especially susceptible to arrhythmias because of the synergistic effects of digoxin and hypokalemia on automaticity and conduction.

Hypocalcemia

Hypocalcemia produces characteristic ECG changes that consist of Q-T interval prolongation secondary to ST-segment prolongation (Fig. 82.15) and occasionally, reversal of the T wave. The ECG changes correlate with ionized calcium because the degree of Q-T prolongation generally is proportional to the degree of hypocalcemia. Abnormal rhythms, although uncommon, have been reported and include SVT, 2:1 AV block, complete heart block, and torsades de pointes VT. The effects of calcium and potassium on myocardial cells are antagonistic.

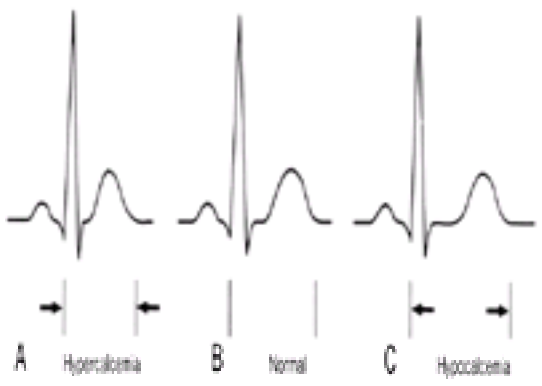


FIGURE 82.15. Electrolyte effects on ECG Q-T interval. Note prolongation with hypocalcemia, shortening with hypercalcemia.

Hypercalcemia

Hypercalcemia, with levels above 12 mg/dL, produces a shortened Q-T interval (Fig. 82.13), a shortened ST segment, and normal or prominent U waves. Severe hypercalcemia causes P-R interval prolongation, QRS prolongation, and occasionally, second-degree and third-degree heart block. Elevated serum calcium decreases the effect of hyperkalemia and potentiates digoxin toxicity. Thus, calcium should be administered cautiously to patients taking digoxin and the HR should be monitored.

Hypomagnesemia

Low magnesium levels often are associated with hypokalemia and hypocalcemia. The ECG abnormalities seen may be those associated with any or all of these aberrations and include prolongation of the corrected Q-T interval. Ectopic beats and T-wave changes are commonly noted. Torsades de pointes VT and ventricular fibrillation have been reported.

Hypermagnesemia

Hypermagnesemia of 3 to 5 mEq/L or higher may be associated with a delay in AV and intraventricular conduction.

Treatment of these electrolyte abnormalities is discussed in Chapter 86.

PERICARDIAL DISEASE

Background

Few medical situations exist in which a simple, quickly performed medical procedure can result in immediate, lifesaving

results. Among these is pericardiocentesis for cardiac tamponade. The technical aspects of pericardiocentesis are discussed elsewhere (see Procedures in Section VII). This section addresses etiologic concerns, clinical findings, and other initial management measures that must be taken to evaluate and treat satisfactorily the child with pericardial disease.

Three forms of illness can affect the pericardium. Pericarditis, usually not a true medical emergency, is a nonspecific term that denotes inflammatory disease. Pericardial effusion, a condition that requires close evaluation but does not necessarily require emergency treatment, implies fluid accumulation within the pericardial space. Cardiac tamponade, a true medical emergency that requires immediate attention, connotes a situation in which impairment of ventricular filling has resulted from pericardial fluid accumulation or from constriction of the heart by an abnormally thickened pericardium, resulting in impairment of CO.

Table 82.19 reviews some of the principal causes of pericarditis in childhood. When considering the cause of pericardial disease and its clinical correlates, it is important to remember that the pericardium is in continuity with the surrounding intrathoracic structures. Thus, conditions that affect the pleura, the mediastinal structures, or the diaphragm may affect the pericardium as well.

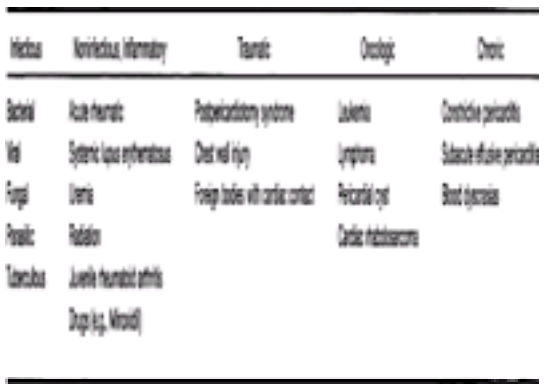


Table 82.19. Causes of Diseases of the Pericardium

Infectious diseases remain the most likely cause of pericarditis in childhood. Although a viral etiology often is presumed to be causative, in only about 20 to 30% of the time is an actual viral pathogen confirmed. Coxsackievirus (group B) and enteric cytopathogenic human orphan (ECHO) viruses are paramount, but other agents, including rubella, Epstein-Barr virus, adenovirus, influenza virus, and mumps virus, have all been associated with pericardial inflammation and pericardial effusion. Rarely do viral diseases result in cardiac tamponade.

Purulent pericarditis is often a medical emergency, however, because of associated cardiac tamponade and because of important sequelae that may be mitigated by early effective treatment. Although it is a disease seen at all pediatric ages, approximately 30% of the cases involve children younger than 6 years of age. *Staphylococcus aureus, Haemophilus influenzae, Neisseria meningitidis, Streptococcus pneumoniae,* and other streptococci are the principal bacterial agents responsible for childhood pyogenic pericarditis, although other pathogens have been recovered occasionally. Other organisms also may cause pericardial infection and chief among these is *Mycobacterium tuberculosis.* In patients from underdeveloped countries, in fact, *M. tuberculosis* may be equally as common as *S. aureus* as a cause of pericardial infection and must not be overlooked. Associated infections, such as respiratory tract disease, osteomyelitis, and pyogenic arthritis, may be present and may be clinically helpful in identifying the specific organism involved. For example, reviews have noted that many cases of staphylococcal pericarditis were associated with infections distant to the pericardium, such as osteomyelitis, whereas most cases of *H. influenzae* pericarditis were associated with respiratory tract infection. Meningococcemia is associated with pericardial involvement in about 5% of cases.

In childhood, noninfectious pericardial disease also can be significant. The postpericardiotomy syndrome is nearly always associated with pericardial inflammation and must be thought of in the postoperative cardiac patient who develops a pericardial effusion, fever, leukocytosis, and a high ESR from after the first week until several weeks after surgery. This syndrome may occur in as many as 15 to 20% of children who undergo open heart surgery. Other important causes of pericardial inflammation include collagen vascular disease and oncologic diseases, especially mediastinal lymphoma.

Pathophysiology

As noted earlier, pericardial inflammation is not usually life-threatening. Of concern for the physician who evaluates a child in an emergency situation are the hemodynamic sequelae of either fluid accumulation in the pericardial space and scarring and thickening of the pericardium, leading to restriction of cardiac filling. Usually, a small amount of intrapericardial fluid (less than 30 to 50 mL) exists in an equilibrium state between secretion into the pericardial space and reabsorption. With a sudden accumulation of fluid or with a more gradual increase of large amounts of fluid within the pericardial sac, interference with ventricular filling occurs, leading to decreased stroke volume and to falling BP. Cardiac filling may be compromised through several interrelated mechanisms, including increased ventricular end diastolic pressure, a decreased gradient for venous return, premature AV valve closure, and shortened diastolic time. The clinical manifestation of these physiologic aberrations, known as cardiac tamponade, is directly related to the severity of these abnormalities and to compensatory mechanisms evoked to overcome them.

Clinical Manifestations

A history of onset of respiratory difficulties after resolution of an upper respiratory illness may indicate pericardial disease in some instances. Chest pain, usually a benign symptom in childhood, is common with pericardial inflammation. This pain varies, depending on position. Occasionally, abdominal pain may be the presenting symptom.

The child with significant pericardial effusion may show clinical signs similar to several of those noted in the preceding section on CHF. Tachypnea secondary to raised pulmonary venous pressures and decreased pulmonary compliance usually is present. This may be associated with intercostal retractions. Reduced CO may result in peripheral vasoconstriction, manifested by cool extremities, pallor, or decreased systemic BP. Elevated systemic venous pressures cause neck vein distension, hepatomegaly, and on occasion in a more chronic picture, protein loss through either the GI tract or the urine. Tachycardia is a universal finding and is representative of an effective compensatory mechanism, but only up to a point. This compensation is limited because diastolic filling times become further shortened by the increased HR.

The cardiac auscultatory findings directly relate to the degree of pericardial fluid accumulation. A friction rub—the scratching, harsh sound commonly heard throughout the cardiac cycle—often is not audible in the presence of significant amounts of intrapericardial fluid and may become apparent only after pericardiocentesis. The heart sounds usually are distant, or muffled, and the apical impulse is weak. In general, the presence of a quiet precordium in the face of these previously noted respiratory and circulatory changes should alert the examiner to the possibility of pericardial disease with effusion.

The sine qua non of cardiac tamponade is pulsus paradoxus. The finding of a paradoxical pulse greater than 20 mm Hg is unequivocal evidence of circulatory compromise. In addition, most investigators assume that as little as 10 mm Hg is suggestive of hemodynamic impairment.

The physiologic mechanisms that underlie pulsus paradoxus can be viewed as exaggerated examples of the integrated functioning of the cardiopulmonary unit. Normally, a small fall (under 10 mm Hg) in systolic BP is noted with inspiration as a result of several factors. As negative intrathoracic pressure is generated by the inspiratory effort, the gradient for systemic venous return increases, favoring right-sided heart filling. At the same time, diaphragmatic descent exerts a traction effect on the heart, limiting filling and ejection. In addition, there may be some decrease in pulmonary venous return because the gradient from pulmonary veins to left atrium probably is reduced. Thus, left-sided heart output and systemic BP are reduced. The pericardium itself is an additional variable factor. In general, because it envelops the heart, the pericardium tends to retard expansion of ventricular volume, normally only to a limited degree. Thus, in normal respiration, the pericardium exerts an additional volume-reducing effect on the left ventricle. In pericardial disease states, as the pericardium itself becomes more rigid or as fluid in the pericardial space increases and intrapericardial pressure rises, the restriction to left ventricular output becomes greater and the consequent decline in systemic BP becomes steeper.

The best method of detecting pulsus paradoxus is to measure BP first in the usual way at expiration and then to inflate the cuff a second time to a few millimeters of mercury above the systolic BP and allow the cuff to deflate slowly. As the pressure falls, the Korotkoff sounds disappear with each inspiration. At the point at which they cease to disappear, becoming equal to that auscultated during expiration, the measured BP is recorded. The difference between the initial maximum systolic BP and the final measurement is the pulsus paradoxus.

It should be noted that pulsus paradoxus is not a finding unique to cardiac tamponade. It is a common finding in respiratory tract disease (asthma) and also may be present in CHF without pericardial effusion.

Laboratory Findings

Laboratory findings vary according to the underlying causes of pericardial disease. Although cardiac tamponade is a clinical diagnosis, certain laboratory tools can be extremely helpful in clarifying the situation.

The ECG shows diminished precordial voltage in most instances of significant intrapericardial fluid accumulation (Fig. 82.16). With pericarditis, an associated current of injury pattern that reflects myocardial involvement, seen as elevations in the ST segments, also may be present. Diffuse T-wave inversions also are common. The heart size is increased on chest radiograph (Fig. 82.17) with pericardial effusion but can be entirely normal if there is no significant amount of intrapericardial fluid. The lung fields may be clear, but one should look for associated bronchopneumonia or pleural effusions that may be helpful for diagnostic considerations. In some situations, as with constrictive pericarditis, the heart size may be relatively small. If the patient has had previous chest radiographs, a sudden increase in heart size always should arouse the suspicion of pericardial effusion.

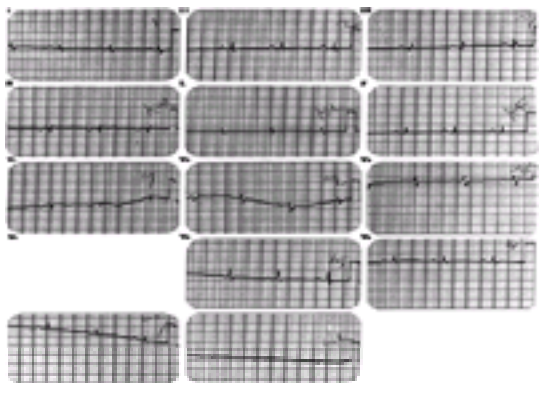


FIGURE 82.16. Electrocardiogram in pericardial effusion. Generalized low voltage present. ST-T wave flattening is

present.

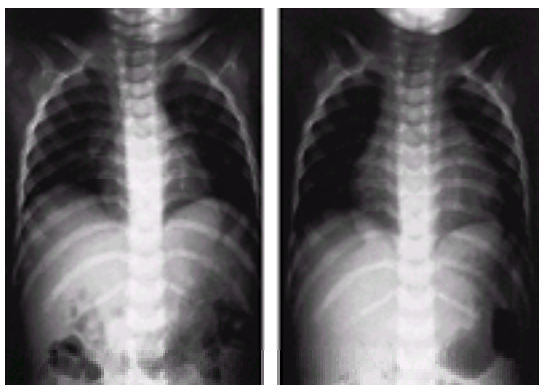


FIGURE 82.17. Radiographs from an infant 4 days before and at the time of the diagnosis of purulent pericarditis. Note the increasing heart size and the “water-bottle” silhouette.

Echocardiography has become the diagnostic procedure of choice for determining the presence and amount of intrapericardial fluid. An echo-free space between the epicardium and the pericardium can be readily identified (Fig. 82.18), with a negligible incidence of false-positive diagnoses in experienced hands. Quantitation is not exact, but evidence of anterior and posterior fluid accumulation suggests a large collection. Either single crystal M-mode echocardiography or two-dimensional real-time echocardiographic studies can be used, although the latter are preferred. In addition, serial evaluation of the pericardial space is easily accomplished by using echocardiography and is helpful for observing the effects of treatment and for evaluating indications for further therapeutic maneuvers after initial drainage measures.

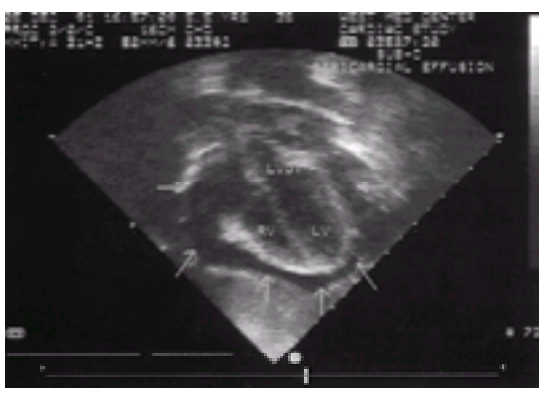


FIGURE 82.18. M-mode echocardiogram in pericardial effusion. Note absence of echoes (clear area) between epicardium and pericardium, representing intrapericardial fluid. *ECG,* electrocardiogram; *ENDO,* endocardium; *EPI,* epicardium; *LV,* left ventricle; *PE,* pericardial effusion; *PERI,* pericardium; *RV,* right ventricle.

When infectious pericardial disease is suspected, as with the evaluation of any other potentially life-threatening infection, a complete bacteriologic evaluation should be initiated before antibiotic therapy is begun.

Management

For pericarditis without evidence of pericardial effusion, emergency invasive treatment usually is not indicated. Symptomatic therapy for pain should be prescribed, and bed rest in the hospital is advisable. The patient should be followed closely for the development of complications such as myocarditis, pericardial effusion, and cardiac tamponade. Diagnostic evaluation to identify the cause should be initiated.

For pericardial effusion, a more definitive approach is needed. Careful evaluation of vital signs and frequent attention to development of pulsus paradoxus are mandatory. Cardiology consultation should be obtained, and the patient should be admitted for evaluation. Diagnostic pericardiocentesis often is required in the de novo presentation, particularly without evidence of other forms of systemic disease; it always is required with the suspicion of a purulent pericardial process. Antibiotic therapy alone is not adequate for treatment of purulent pericarditis. Usually, in the presence of purulent pericarditis, an open drainage procedure is indicated. It is contingent on the emergency physician to ensure cardiovascular stability in the presence of pericardial effusion because tamponade can develop rapidly once maximum pericardial distensibility has been reached (Table 82.20).

1. Ensure adequacy of ventilation and cardiac output
2. Administer oxygen
3. Initiate cardiorespiratory monitoring
4. Obtain laboratory studies (simultaneously with step 5)
Complete blood count, platelet count, electrolytes, blood urea nitrogen, creatinine, glucose, arterial blood gas, blood culture, chest radiograph, electrocardiography, echocardiography
5. Achieve venous access
6. Pericardiocentesis (see Section VII)
Send specimen for laboratory studies: cultures, CIE, viral titers, antinuclear antibody, Gram stain, cytology, cell count and differential, chemical profile
7. Administer antibiotics[a]
Oxacillin (150 mg/kg/day) or nafcillin or methicillin and Chloramphenicol (100 mg/kg/day)
Aminoglycoside (immunocompromised patient)

[a]Select antimicrobials to cover *S. aureus* and *H. influenzae* at least until specific infection is isolated.

Table 82.20. Purulent Pericarditis: Immediate Management

The management of cardiac tamponade requires intense medical vigilance. Although it may be possible in relatively mild or highly selected situations to manage the effusion conservatively, it generally is necessary to remove the fluid. A full discussion of the techniques used for pericardiocentesis in the emergency situation is available in Section VII of this book. This can be a lifesaving technique and, when done successfully, shows clearly the fruitful outcome of appropriate, decisive evaluation and treatment procedures.

INFECTIVE ENDOCARDITIS

Background

One of the persistently complex problems of pediatric cardiovascular medicine has been the evaluation and management of the child with infective endocarditis. Although long-term treatment issues generally are not within the province of emergency medical care, it is critically important for the emergency physician to be aware of the clinical context in which bacterial endocarditis is a consideration. It is also incumbent on the emergency physician to initiate therapy in certain instances, and it is always crucial to avoid unnecessary clouding of the diagnosis.

Etiologic Factors

The clinical picture of infective endocarditis has been evolving steadily over the past 10 to 20 years. Although the most common setting for this problem is the child with preexisting CHD, variability exists in terms of the types of associated lesions (Fig. 82.19), and it is of concern that a substantial proportion of cases develop in children with no history of cardiac abnormality. These children may be among the most ill, presenting with their illness as part of an acute bacterial endocarditis picture.

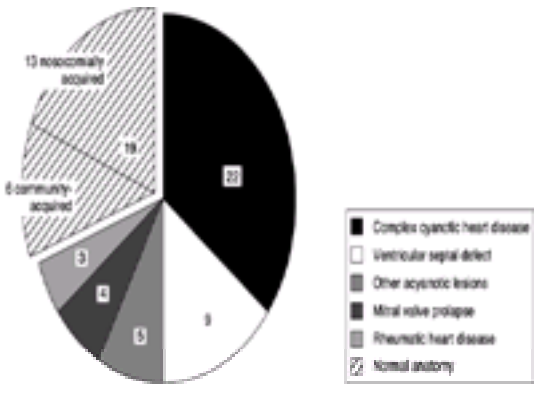


FIGURE 82.19. Distribution of underlying cardiovascular findings in endocarditis in a series of 62 children. (Reprinted with permission from Saiman L, Prince A, Gersony WM. Pediatric endocarditis in the modern era. J Pediatr 1993;122:847–853.)

Certain factors appear to predispose a child to the development of endocarditis. It is widely believed that among these are dental and surgical procedures. Dental procedures even without periodontal disease can yield significant bacteremia. Unfortunately, many ordinary daily events are associated with at least transient bacteremia (Table 82.21). It is small wonder, in fact, that more cases of endocarditis are not evident if bacteremia of oral cavity origin were a singular factor. Conversely, invasive procedures specifically involving the heart, such as cardiac catheterization, in patients with even the highest risk are rarely associated with endocarditis development. Recently, the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the American Heart Association undertook a major review of the guidelines for endocarditis prevention. This review identified the procedures likely to predispose to endocarditis development (Table 82.22) and stratified the cardiac conditions with the highest likelihood of susceptibility regarding their risk potential for endocarditis development (Table 82.23). These findings were based on several emerging trends: 1) children with even the most severe congenital cardiac malformations increasingly survive complicated surgical procedures performed at younger and younger ages, thereby increasing the pool of susceptible children at risk; 2) children who develop endocarditis are more likely to overcome the episode than in the past because of improved clinical and microbiologic technologies and a broadened selection of antimicrobials; 3) the advent of chronic parenteral access catheters for nutritional and pharmacologic therapies of premature neonates and of older children with chronic illnesses, such as oncologic diseases or metabolic and neuromuscular disorders, has created a widening population of susceptible children

with structurally normal hearts; 4) mitral valve prolapse has emerged as a not uncommon finding even in children and reports of endocarditis in this setting have resulted in controversy regarding what distinguishes a truly abnormal valve from the normal variability that enhanced imaging techniques have enabled us to identify as part of the growth and development process.

Procedures	Bacteremia (%)
Tooth extractions (no gingivitis)	34
Tooth extractions (gingivitis)	70–75
Endocontic procedures	4
Chewing mint candy	20
Brushing teeth	40
Oral irrigation device	27–50
Massage of nfected tonsl	23
Urethral surgery	57
Massage of nfected prostate	67
Barium enema	11
Bronchoscopy	15
Sigmoidoscopy	5–10

Modified with permission from Kaye D, ed. Prophylaxis of Endocarditis. Baltimore: University Park Press, 1976:245–255.

Table 82.21. Transient Bacteremia and Various Procedures or Conditions

Table 82.22. Procedures and Endocarditis Prophylaxis

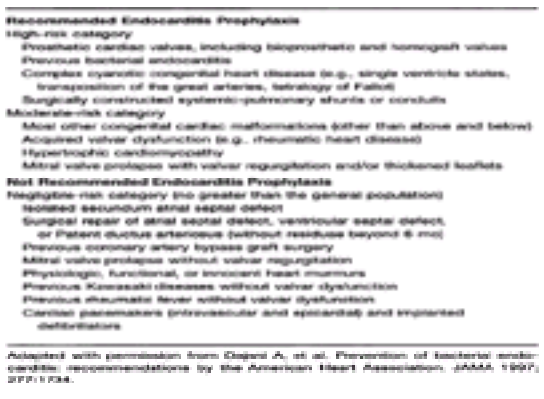


Table 82.23. Cardiac Conditions Associated with Endocarditis

Despite some changes in the specifics of the clinical epidemiology of endocarditis, certain physiologic conditions appear to be consistently most important. Diseases characterized by a highly turbulent stream of blood or a high velocity of flow are particularly prone to this complication. Such lesions include ventricular septal defect, aortic valve stenosis, and mitral valve regurgitation. Children with postoperative systemic-to-pulmonary shunts also are in this category. In contrast, secundum atrial septal defect is a lesion with a negligible risk for endocarditis because the shunt flow is of low velocity. It is presumed that in “high velocity–narrow orifice” conditions, damage to cardiac surfaces occurs, resulting in a nidus for platelet disposition and vegetation formation.

Microbiology

Causative organisms include bacteria and fungi and are often related to the initiating event. Thus, although streptococci in general are the most common causative agents, viridans streptococci are the typical isolates following oral procedures. Staphylococci also are common etiologic agents, especially in children with structurally normal hearts or, ironically, in those with postoperative CHD, such as prosthetic valves. Other bacteria are much less likely to be present in childhood endocarditis. These include Gram-negative organisms, present in immunocompromised children and sick neonates, *Enterococci*, *Pneumococci*, and *Haemophilus* species. These data are reviewed in Table 82.24.

Organism	Frequency (%)
Streptococci	
Viridans	50
Pneumococci	8
β-Hemolytic	5
Enterococci	5
Staphylococci	
S. aureus	25
S. epidermidis	8
Gram-negative bacilli	8
Fungi	8
Culture-negative endocarditis	20

Abstracted with permission from various references and adapted from Friedman RSJ. Infective endocarditis. In: Garson A, ed. The Science and Practice of Pediatric Cardiology. Philadelphia: Lea & Febiger, 1990.

Table 82.24. Agents Associated with Pediatric Infective Endocarditis

Clinical Manifestations

Confirmation of a positive diagnosis depends on the recovery of organisms obtained by blood culture. To arrive at that point, however, a high degree of suspicion must be maintained. Often, early signs and symptoms can be subtle and persist for considerable time before the diagnosis is made. With viridans streptococcal endocarditis, this is a common situation. As a rule, persistence of fever in any child with CHD should prompt the clinician to consider the possibility of endocarditis.

In the clinical context of CHD, certain conditions should prompt a careful evaluation for the presence of endocarditis. These include 1) unexplained fever or a protracted febrile course in a presumed “viral” syndrome, 2) pneumonia, 3) the development of a new neurologic deficit, 4) the onset of hematuria, and 5) signs of systemic or cutaneous embolization.

The classic findings of fever, a change in the cardiac examination, splenomegaly, and evidence of emboli usually are present in severe cases but may require serial examinations. Emboli may be discovered by careful funduscopic examination, by observing for conjunctival lesions, or by meticulous scrutiny of the nailbeds, palms of the hands, soles of the feet, and other skin surfaces. Microscopic hematuria should be recognized as an important sign of endocarditis in the appropriate clinical context. Scrapings of cutaneous emboli may be helpful for rapid identification of infecting organisms.

Complications

The morbidity from infective endocarditis has decreased considerably in recent years. Currently, most series cite a fatality rate of 15 to 20%. Although this is still a high percentage, especially for pediatric illness, it should be remembered that more than 50% mortality was the norm in the 1950s and that the disease was nearly always fatal in the preantiobiotic era.

Other complications occur in as many as 40 to 60% of cases. Systemic or pulmonary emboli, depending on the intracardiac site of the vegetation, are a major source of concern and indicate prompt initiation of treatment. Major neurologic sequelae can arise from focal embolization to the CNS; thus, the presentation of a new neurologic deficit in a child with heart disease can be another clinical clue to the diagnosis of endocarditis. Myocarditis, myocardial abscesses, valvar obstructions associated with large vegetations, and ruptured sinus of Valsalva are other important complications that can be manifested by the appearance of CHF.

Acute bacterial endocarditis, or the development of an acute situation such as new aortic insufficiency, should be considered a true medical emergency. Often, early reparative surgery is required to save the child's life in this situation. These children are critically ill, and CHF is a grave sign in the context of suspected endocarditis. Characteristic heart murmurs may be absent in this setting, and their absence should not be taken as a cause for optimism. Other indications for surgery include the development of a cardiac arrhythmia (heart block), continued embolization, and continued positive blood cultures after initiation of appropriate therapy. Hemodynamic changes can transpire quickly, demanding frequent examinations even while the child awaits hospital admission or transfer from the ED.

Management

Treatment of infective endocarditis should be started as early as possible after appropriate evaluation is completed. Blood cultures must be drawn regardless of the presence or absence of classic clinical findings. To facilitate the diagnosis, the physician, particularly one who evaluates a child with heart disease with unexplained fever, must obtain blood for appropriate cultures at an early stage. In most of cases of endocarditis, the causative organism will be recovered from the initial two blood cultures. Particular emphasis should be placed on avoiding contamination of the sample site. Growth of spurious organisms can be misleading and dangerously time-consuming because bacteria on the skin can be implicated in endocarditis. It is not mandatory to obtain cultures at the time of fever spikes because bacteremia is fairly constant in the untreated patient. Early consultation should be sought from a cardiologist because specialized procedures such as echocardiography may help pinpoint the diagnosis rapidly, even in relatively difficult situations (Fig. 82.20).

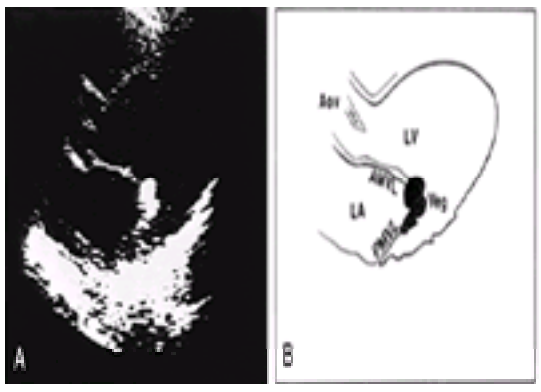


FIGURE 82.20. A. Long axis parasternal two-dimensional echocardiogram in patient with mitral valve pneumococcal endocarditis. **B.** Schematic view of **(A).** *AMVL,* anterior mitral valve leaflet; *Aov,* aortic valve; *LA,* left atrium; *LV,* left ventricle; *PMVL,* posterior mitral valve leaflet; *Veg,* vegetation.

In every instance, the diagnosis of infectious endocarditis implies long-term antibiotic therapy; thus, most management issues arise after the patient has left the emergency area. In general, antibiotic therapy should be instituted as soon as the diagnosis is made. If the patient is critically ill, it may be necessary to initiate therapy even before the results of cultures have returned. Certainly, stabilization of the patient with heart failure, initiation of the diagnostic workup, and mobilization of the relevant medical personnel are the responsibilities of the emergency physician in dealing with the child with suspected endocarditis. If the situation requires the initiation of therapy without definition of the microbial agent, many experts recommend the combination of an aminoglycoside, such as gentamicin (5 to 7.5 mg/kg per day) and a penicillinase-resistant penicillin such as oxacillin (150 mg/kg per day). Others advocate the use of ampicillin (200 mg/kg per day) and gentamicin for the initial therapy in this particular situation. Cephalosporins such as cefuroxime may also play a role in this context.

Much mention has been made of the value of antimicrobial prophylaxis in mitigating the development of infectious endocarditis, although it should be noted that the precise population benefits of prophylaxis never have been fully substantiated. Nevertheless, it remains incumbent on the physician who sees a child with heart disease in the ED to ensure that prophylaxis has been implemented if warranted. Prevention guidelines have been recently modified by the American Heart Association (Table 82.25). As a rule, such measures are practical only in the face of a well-defined, predisposing event. The usual child with heart disease who presents with a routine febrile illness does not require prophylactic antibiotics. Unnecessarily hasty administration of antibiotics when not indicated can be harmful because obfuscation of the ultimate diagnosis may result in damaging delay.

Situation	Agent	Regimen
For Dental, Oral, Respiratory Tract, or Esophageal Procedures		
Standard general prophylaxis	Amoxicillin	Adults: 2.0 g; Children: 50 mg/kg PO; 1 hr preprocedure (maximum: 2.0 g)
Unable to take oral medications	Ampicillin	Adults: 2.0 g IM or IV; Children: 50 mg/kg IV or IM within 30 min of procedure
Allergic to penicillins	Clindamycin	Adults: 600 mg; Children: 20 mg/kg PO; 1 hr preprocedure
	Cephalexin or Cefadroxil	Adults: 2.0 g; Children: 50 mg/kg PO; 1 hr preprocedure
	Azithromycin or Clarithromycin	Adults: 500 mg; Children: 15 mg/kg PO; 1 hr preprocedure
Allergic to penicillin and unable to take oral medication	Clindamycin	Adults: 600 mg; Children: 20 mg/kg IV within 30 min of procedure
	Cefazolin	Adults: 1.0 g; Children: 25 mg/kg IV or IM within 30 min of procedure
For Genitourinary/Gastrointestinal (nonesophageal) Procedures		
High-risk patients	Ampicillin plus Gentamicin	Ampicillin 50 mg/kg IM or IV (max 2.0 g) plus gentamicin 1.5 mg/kg within 30 min of procedure; 6 hr later, ampicillin 25 mg/kg IV or IM or amoxicillin 25 mg/kg PO
High-risk allergic to ampicillin and amoxicillin	Vancomycin plus Gentamicin	Vancomycin 20 mg/kg IV over 1–2 hr plus gentamicin 1.5 mg/kg complete treatment within 30 min of procedure
Moderate-risk patients allergic to Amp/Amox	Vancomycin	Vancomycin 20 mg/kg IV over 1–2 hr—complete treatment within 30 min of procedure

Total children's doses should never exceed adult doses.
Second dose of vancomycin or gentamicin not recommended.
Adapted with permission from Dajani A, et al. Prevention of bacterial endocarditis: recommendations by the American Heart Association. JAMA. 1997;277:1794–1801.

Table 82.25. Endocarditis Prophylaxis Regimens

If systemic antibiotics are contemplated for other infectious indications, in most cases a blood count and a blood culture should be drawn before antibiotic therapy begins. In particular, these measures should be taken for the child with heart disease and a major infection, such as pneumonia or cellulitis, even if no clinical evidence of endocarditis is immediately apparent. It is not mandatory to admit the child with heart disease and an intercurrent febrile illness to the hospital on every occasion, and the previously noted laboratory studies may be helpful in making such a decision. Clinical judgment remains the best immediate guide for hospitalization. Although a high degree of suspicion for the possibility of endocarditis is mandatory, the emergency physician should resist the temptation to administer antibiotics indiscriminately to the child with heart disease.

HYPOXEMIC ATTACKS

Background

Children with cyanotic CHD in which pulmonary blood flow is reduced, such as tetralogy of Fallot, may experience periodic episodes of intense hypoxemia. Emergency attention usually is sought for these episodes at the nearest medical location. Therefore, the emergency physician who cares for children should have a good understanding of the associated physiologic and management principles.

Pathophysiology

The reasons for the acute nature of these episodes have never been defined. Initial thoughts that “cyanotic spells” were caused by spasmodic contraction of the portion of right ventricular outflow tract known as the “infundibulum” cannot provide the entire explanation because children with pulmonary atresia in whom no subpulmonic infundibulum has developed also can experience hypercyanotic attacks. Additional theoretic concerns have focused on 1) sudden changes in systemic vascular resistance and in venous return to the heart, which consequently affect the intracardiac right-to-left shunt; 2) alterations in sensitivity of the respiratory center; 3) significant changes in HR; or 4) some combination of all these factors. A schematic cycle of postulated mechanisms is noted in Figure 82.21.

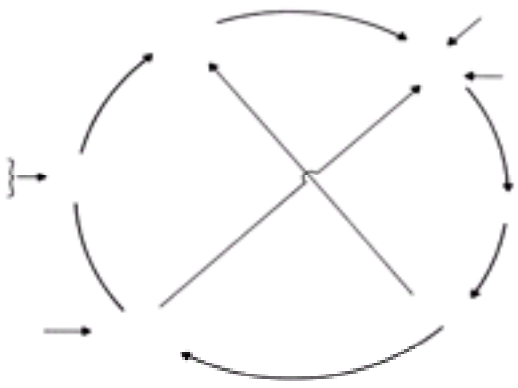


FIGURE 82.21. Schema of interrelated events in the genesis of hypoxemic spells. See text for discussion. (Reprinted with permission from Anthony CL, et al. Pediatric Cardiology, Medical Outline Series. Garden City, NY: Medical Examination Publishing, 1979:193.)

Any number of precipitant events related to these physiologic factors can be associated with the development of cyanotic spells. Often, they are morning events, noted shortly after awakening. This may be related to the sudden changes in CO that occur after arousal from a long sleep. Other likely times for the appearance of hypercyanosis include periods of dehydration, during invasive medical procedures, or other significant stresses.

Clinical Findings

The diagnosis of a hypoxemic spell usually is self-evident. Aside from the obvious cyanosis and the history of heart disease, there also may be a preceding history of squatting with exertion or of other positional vagaries that parents may recall. It is not necessary for the child to have been overtly cyanotic before the onset of a spell because such episodes can occur in children with little or no preexisting cyanosis. During a spell, the child may be irritable and crying or may be lethargic and even unconscious. Hyperpnea is a feature of the syndrome and should be distinguished from tachypnea or other abnormal respiratory patterns that may signal other medical problems associated with cyanosis. During a spell, there may be a notable absence or lessening of a previously heard heart murmur because pulmonary blood flow through the stenotic right ventricular outflow tract is reduced considerably. Laboratory investigations, such as arterial blood gas analysis, ordinarily should be avoided in the initial evaluation. If the attack is prolonged and associated with deepening sensorium changes, assessment of acid-base balance and ventilatory status may be indicated. Monitoring with peripheral oxygen saturation meters (transcutaneous) may be helpful to chart responses to therapy.

Management

The child with hypoxemic spells requires immediate attention. Appropriate positioning, oxygen, and administration of morphine are the standard initial therapeutic measures, and these usually result in prompt abatement of the attack (Table 82.26).

Knee-chest position	IV fluids
Oxygen administration	Vasoconstrictors
Evaluate and treat cardiac arrhythmia	Phenylephrine 0.10 mg/kg bolus, IM or subcutaneous; 2–10 µg/kg/min infusion IV
Morphine sulfate (subcutaneous) (0.1–2 mg/kg)	Methoxamine 0.10 mg/kg IV
Propranolol IV (0.2 mg/kg over 5 min)	Bicarbonate (2–3 mEq/kg, IV; ensure adequate ventilation)

Table 82.26. Acute Management of Hypoxemic Spells

Traditionally, subcutaneous morphine has been used in the treatment of cyanotic spells. Relatively large doses are given (0.1 to 0.2 mg/kg), although the precise mechanism of action is unknown. Morphine probably does not act to inhibit catecholamine action at the cardiac level but may abort the cycle of hyperpnea and vasomotor changes by depressing the respiratory center. A theoretic negative effect of morphine is its tendency to lower systemic vascular resistance.

Oxygen should be administered because Pa_{O_2} levels may be low and some benefit in terms of oxygen saturation may be obtained from even relatively small increments in dissolved oxygen. In the face of significant reduction of pulmonary blood flow, however, as occurs with a “spell,” oxygen may not have a dramatic effect.

The child should be placed in a knee–chest position and calmed, if possible. If the attack persists, additional therapeutic steps are needed. Sodium bicarbonate may be indicated; the dosage depends on arterial pH. Propranolol also has been recognized as efficacious in this situation, and an IV dose of 0.2 mg/kg over 4 to 5 minutes may yield relatively prompt improvement. Whether propranolol primarily affects the infundibular contraction, the hyperpneic ventilatory response, systemic vasomotor tone, or all of these is unclear. It should be remembered that propranolol may exacerbate bronchospasm if the patient has a coincident history of asthma.

IV fluids should be administered during the severe spell in maintenance doses at least because pulmonary blood flow and right ventricular output depend on volume. Functionally, right ventricular outflow obstruction may be heightened in the face of depleted intravascular volume.

Vasopressors have been advocated as alternates or adjuncts in treating hypoxemic spells. Phenylephrine can be given as a dilute IV solution of 10 mg/100 mL and infused at 2 to 10 µg/kg per minute. HR should be monitored and frequent BP assessment carried out if this type of agent is used. Methoxamine (10 mg/ 100 mL) or metaraminol (50 mg/100 mL) also may be used. By increasing systemic vascular resistance, these drugs reduce intracardiac right-to-left shunting favorably and thus improve systemic oxygenation. Digitalis, epinephrine, or norepinephrine should not be used, however, in this setting.

If any underlying condition exists, such as a cardiac rhythm disturbance, prompt correction according to the principles noted under Cardiac Arrhythmias may alleviate this situation quickly. Cardiac consultation is advisable as soon as feasible to make extended management decisions, even if the spell has abated with the measures already mentioned. In most situations, if the spell has required more than oxygen and positional adjustment to abate, hospitalization is indicated. Usually, although relieved with therapy, spells indicate appropriate surgery for the cardiac defect.

ACUTE RHEUMATIC FEVER

Background

Although the large numbers of patients with rheumatic fever seen in the past in the United States have dissipated because of improved diagnosis and treatment of streptococcal infections, the disease still occurs and, recently, has had resurgence. Also, rheumatic fever remains one of the most common causes of cardiovascular morbidity in children from other countries. The most common age of attack in the United States is 5 to 15 years, and winter and spring seasonal peaks are still typical.

Pathophysiology

It has been established clearly that streptococcal infection is a necessary precedent for the development of rheumatic fever. In particular, a history of infection by this organism of the upper respiratory tract should be sought in any suspected case. The precise mechanistic relationship between antecedent streptococcal infection and rheumatic fever, however, remains ill defined. Many serologic types of group A streptococci can be associated with acute rheumatic fever, so the antigenic factors involved are common to various strains of the organism. The particular host factors that determine who succumbs to acute rheumatic fever and who does not, despite identical infections, are also poorly defined. A clear-cut familial pattern has never been identified, although familial susceptibility appears to be a factor. The more common theoretic considerations that relate streptococcal infection and acute rheumatic fever are 1) an immunologic (autoimmune) response that involves host reaction to infection with a target organ being the heart, specifically, endocardial tissue; 2) a persistence of organism despite therapy, with localization to cardiac tissue; and 3) a direct reaction to the organism such as cardiotoxicity from streptolysin O produced by the organism. Thus far, no evidence of direct cardiac infection has developed, making experimental evaluation difficult.

Clinical Manifestations

The diagnosis of rheumatic fever requires a high index of suspicion. The time-honored Jones criteria (Table 82.27), if unequivocally present, usually establish the diagnosis, but the situation may not be always so clear-cut.

Major	Elevated acute phase reactants
Carditis	Erythrocyte sedimentation rate
Arthritis	C-reactive protein
Subcutaneous nodules	Prolonged P-R interval
Erythema marginatum	Supporting evidence of antecedent
Chorea	group A streptococcal infections
Minor	Positive throat culture of rapid
Clinical findings	streptococcal antibody titer
Arthralgia	Elevated or rising streptococcal
Fever	antibody titer
Laboratory findings	

Adapted from Jones TD. The diagnosis of rheumatic fever. JAMA. 1944; 126: 481., as modified in Guidelines for the diagnosis of rheumatic fever. JAMA 1992; 268:2069.

Table 82.27. Rheumatic Fever Manifestations

A complete, careful physical examination is the mandatory first procedure. Special attention should be given to eliciting joint pathology and cutaneous findings whose presence may facilitate the diagnosis in difficult cases. All the major Jones criteria are derived through clinical examination that usually needs to be repeated at frequent intervals. Among the major criteria, carditis can be overdiagnosed easily. Misinterpretation of normal (“innocent”) murmurs, whose auscultation is heightened in the presence of fever or other causes of increased cardiac output, can lead to overdiagnosis. The presence of an apical systolic murmur (characteristic of mitral insufficiency) or of a basal diastolic decrescendo murmur (typical of aortic insufficiency) is an important sign of carditis. The presence of a pericardial effusion, CHF, or pericarditis also strongly suggests the carditis component of acute rheumatic fever, even in the absence of valvar murmurs. Care must be taken to exclude other causes for cardiac findings, such as deteriorating CHD, which may result in cardiac decompensation not related to a rheumatic process. If possible, the examining physician should attempt to document a change in previous clinical findings in children with preexisting heart disease (or previous rheumatic fever episodes). Although regurgitant lesions, such as aortic or mitral insufficiency, are common components of the acute manifestations of rheumatic fever, stenotic lesions, such as aortic or mitral stenosis, usually are not seen with a first attack of acute rheumatic fever.

Polyarthritis is the most commonly found major criterion. It should be remembered that this is true joint inflammation, not arthralgia. Tenderness, motion restriction, heat, redness, and swelling are the typical signs. In contrast to other forms of collagen disease, joint involvement in rheumatic fever usually is migratory and multiple and tends to localize to the larger joints of the extremities. It may be necessary to avoid rapid use of anti-inflammatory agents in patients with suspected acute rheumatic fever to clarify the diagnosis of migratory polyarthritis.

The cutaneous criteria are erythema marginatum and subcutaneous nodules. These findings are not as common as arthritis and carditis and are rarely present as the only major criteria. Nodules usually occur in situations of recurrent rheumatic fever or chronicity. They are found over extensor surfaces of joints, such as elbows or knees; are firm and decidedly nontender; and are movable on palpation. Erythema marginatum characteristically appears on the trunk and proximal extremities and is an extremely evanescent finding. The application of heat may accentuate its appearance. This rash is notable for its fine, lacy appearance with central blanching and a serpiginous pattern. It is not pruritic and is usually distinguished easily from drug rashes or other viral exanthems.

Chorea is the fifth of the major criteria defined by Jones. It is a relatively rare finding limited to children older than 3 years of age and most often occurs some time after the initial streptococcal infection, making accurate diagnosis difficult. Chorea is typified by involuntary purposeless movement of the extremities and facial grimacing. Notable emotional lability is also a part of the picture. The ED diagnosis of acute rheumatic fever rarely depends on chorea as the principal manifestation. The physician should be aware, however, of the possibility of the diagnosis in a child who presents with this finding and should arrange for appropriate further evaluation of the cause of the chorea.

The “minor criteria” defined by Jones are nonspecific indices of inflammatory disease and, often, are sources of overdiagnosis of acute rheumatic fever. The fever associated with acute rheumatic fever is notable for its lack of associated chills or rigor. It typically is low grade, and fevers of greater than 40°C (104°F) or a history of a febrile seizure should point to other illnesses. The wildly fluctuating fever of juvenile rheumatoid arthritis (“quotidian” pattern) usually is not a part of the rheumatic fever picture. Elevation of the ESR or C-reactive protein should be present in acute rheumatic fever, but severe CHF may lower the ESR. A prolonged P-R interval is common in acute rheumatic fever but also is an extremely nonspecific finding. It does not necessarily correlate with the presence of organic murmurs and can be found in other inflammatory cardiac diseases or as a result of certain drugs. Overemphasis of the significance of P-R prolongation is a common cause of improper diagnosis.

It must be emphasized that the modified Jones criteria include evidence of recent streptococcal infection in the history. Culture documentation is helpful, but serologic evidence may be the most rewarding and diagnostic data. The widespread use of the multiple antibody test (Streptozyme) has made serologic confirmation of recent streptococcal infection much easier. The antistreptolysin O test (ASO) is still a commonly used single serologic test and is well standardized. Levels above 250 Todd units in older children and above 333 in younger children are present in active rheumatic fever. As many as 20% of otherwise normal children can have elevated ASO titers, and depending on the time course of the illness, other antibody determinations may be required.

The differential diagnosis of acute rheumatic fever includes many diseases that fall under the classification of “collagen vascular” as well as other types of diseases. The Jones criteria themselves can include a spectrum of illnesses such as juvenile rheumatoid arthritis, serum sickness, systemic lupus, and even bacterial endocarditis or septic arthritis. Viral processes, such as myocarditis or pericarditis, also must be excluded, as well as intracardiac lesions such as left atrial myxoma.

Careful application of the Jones criteria plus documentation of a streptococcal infection of recent onset should enable the physician to diagnose acute rheumatic fever most of the time. Caution must be exercised in arriving at the diagnosis because initiation of therapy may suppress findings critical to the diagnosis. Thus, decisions to treat must be tempered with the understanding that it is vital to collect as much definition of the disease process as possible.

As noted earlier, acute-phase reactants such as the ESR and C-reactive protein are elevated in acute rheumatic fever. A complete blood count should be drawn to screen for anemia or an elevated white blood cell count. Leukocytosis not only is a manifestation of infection but may be considered an acute-phase reactant as well. Throat cultures (at least two) should be obtained before penicillin therapy is started. In addition, the streptococcal screen previously described should be obtained. Blood cultures often are drawn, with appropriate concern, to rule out subacute bacterial endocarditis, a problem that can present in an identical fashion to acute rheumatic fever.

A chest radiograph to assess heart size can be helpful for gauging the severity of carditis, as well as for objectively

verifying its presence. An ECG should be taken to ensure that a rapid pulse rate is the result of sinus tachycardia and to enable measurement of P-R interval. If pericardial disease or intracardiac myxoma needs to be ruled out, an echocardiogram can provide highly sensitive information. These latter procedures usually are completed, of course, after cardiac consultation has been requested. The appropriate laboratory procedures to help rule out other forms of collagen-vascular disease are described in Chapter 101.

Management

Acute rheumatic fever requires admission to the hospital and long-term management. That is, a prolonged treatment course is indicated once the diagnosis is made. Most considerations in caring for a child with acute rheumatic fever are not made in the ED. It should be restated that a rush to treat with anti-inflammatory drugs (aspirin or steroids) in a poorly documented case may obscure the ultimate diagnosis and may delay further therapy, thereby compromising more than helping the patient.

Principles of management include 1) treatment of the active streptococcal infection, 2) rest, 3) anti-inflammatory agents, and 4) treatment of chorea. All patients with acute rheumatic fever should receive a course of penicillin to eradicate any streptococci. Intramuscular benzathine penicillin in appropriate dosage for age and weight (see Chapter 84) is preferable. Bed rest may be helpful for as long as evidence of active inflammation is present. It should be initiated at diagnosis and is best done during the initial period in a hospital setting. Anti-inflammatory drugs (salicylates or steroids) may be indicated, but the tendency to begin such therapy before confirmation of the diagnosis, as outlined already, should be resisted. If arthritis without carditis is present, aspirin usually is sufficient (see Chapter 96). Treatment of carditis may include steroids in selected cases, but that decision should be undertaken only after the child is hospitalized and a cardiologist has been consulted. Treatment of chorea is also a long-term management issue, with agents such as diazepam or haloperidol currently favored (see Chapter 129). Recent evidence suggests a role for steroids in the treatment of chorea and for the presence of carditis.

Occasionally, the child with acute rheumatic fever may present with significant cardiac compromise that involves CHF associated with a large degree of valvar regurgitation or pericardial effusion that results in cardiac tamponade. Initially, the heart failure or tamponade should be managed as outlined in the previous sections, and then consideration should be given to the rheumatic process.

A most important aspect of management of the patient with rheumatic fever is prevention of recurrent attacks. It has been documented clearly that penicillin can be effective in this setting, with minimal patient risk. The most reliable prophylaxis is the IM route, with injections of 1.2 million units of benzathine penicillin G every 28 days being the preferred treatment. Oral penicillin (200,000 units twice daily) is an alternative prophylactic regimen. Sulfonamides may be used, as well as erythromycin, in patients sensitive to penicillin. Recommended dosages for the sulfonamides are 0.5 to 1.0 g/day, depending on weight, and 250 mg twice daily for erythromycin. Although current recommendations about the duration of prophylaxis are under scrutiny, most centers continue to use antibiotics for a minimum of 10 to 15 years after the initial diagnosis, and some use them for life. Increasing age may lessen susceptibility to streptococcal disease, but reliable evidence is lacking to substantiate this impression conclusively. The physician who evaluates a child with known rheumatic heart disease for any reason should review the prophylaxis status of the child at every occasion.

Suggested Readings

General

Emmanouilides GC, Allen HD, Riemenschneider TA, et al, eds. Moss and Adams Heart Disease in Infants, Children, and Adolescents. 5th ed. Baltimore: Williams & Wilkins, 1995.

Dickinson DF, Arnold R, Wilkinson JL. Congenital heart disease among 160, 480 liveborn children in Liverpool, 1960 to 1969: implications for surgical treatment. Br Heart J 1981;46:55–62.

Garson A, Bricker JT, McNamara DG, eds. The Science and Practice of Pediatric Cardiology. Philadelphia: Lea & Febiger, 1990.

Garson AF Jr. The Electrocardiogram in Infants and Children. Philadelphia: Lea & Febiger, 1983.

Gewitz MH, ed. Primary Pediatric Cardiology. Futura Medical Publishing, Armonk, New York, 1995.

Congestive Heart Failure

Chaterjee K, Parmley WW. The role of vasodilator therapy in heart failure. Prog Cardiovasc Dis 1977;19:301.

Driscoll DJ, Gillette PC, McNamara DG. The use of dopamine in children. J Pediatr 1978;92:309.

Friedman WF. The intrinsic physiologic properties of the developing heart. In: Friedman WF, Lesch M, Sonnenblock EH, eds. Neonatal Heart Disease. New York: Grune & Stratton, 1973:21.

Friedman WF, George BL. Treatment of congestive heart failure by altering loading conditions of the heart. J Pediatr 1985;106:697.

Kaplan S. New aspects of management of congestive heart failure. In: Engle MA, ed. Cardio-Vascular Clinics 11/2. Pediatric Cardiovascular Disease. Philadelphia: FA Davis, 1981.

Neill CA. The use of digitalis in infants and children. Prog Cardiovasc Dis 1965;7:399.

Ross J Jr. Afterload mismatch and preload reserve: a conceptual framework for the analysis of ventricular function. Prog Cardiovasc Dis 1976;18:225.

Rudolph AM. Cardiac failure in children: a hemodynamic overview. In: Braunwald E, ed. The Myocardium—Failure and Infarction. New York: HP Publishing, 1974;102.

Smith TW, Butler V, Haber E, et al. Treatment of life-threatening digitalis intoxication with digoxin specific Fab antibody fragments. N Engl J Med

1982;307:1357.

Talner NS. Heart failure. In: Emmanouilides GC, Allen HD, Riemmenschneider TA, et al, eds. Moss and Adams Heart Disease in Infants, Children, and Adolescents. 5th ed. Baltimore: Williams & Wilkins, 1995.

Cardiac Arrhythmias

Benson DW Jr, Sanford M, Dunnigan A, et al. Transesophageal pacing threshold: role of interelectrode spacing, pulse width, and catheter insertion depth. Am J Cardiol 1984;53:63.

Chauder JS, Wolff GS, Garson A Jr, et al. Ventricular arrhythmias in postoperative tetralogy of Fallot. Am J Cardiol 1990;65:655.

Crosson JE, Etheridge SP, Milsrein S, et al. Therapeutic and diagnostic utility of adenosine during tachycardia evaluation in children. Am J Cardiol 1994;74:155.

Fish F, Benson DW Jr. Disorders of cardiac rhythm and conduction. In: Emmanouilliides GC, Allen HD, Riemenschneider TA, et al, eds. Moss and Adams Heart Disease in Infants, Children, and Adolescents. Baltimore: Williams & Wilkins, 1995:1555.

Garson A Jr, Gillette PC, eds. Pediatric Arrhythmias: Electrophysiology and Pacing. Philadelphia: WB Saunders, 1990.

Garson A, Gillette PC, McNamara DG. Supraventricular tachycardia in children: clinical features, response to treatment, and long-term follow-up in 217 patients. J Pediatr 1981;98:875.

Gilette PC, Shannon C, Blair H, et al. Transvenous pacing in pediatric patients. Am Heart J 1983;105:843.

Ludomirsky A, Garson A Jr. Supraventricular tachycardiac. In: Garson A Jr, Gillette PC, eds. Pediatric Arrhythmias: Electrophysiology and Pacing. Philadelphia: WB Saunders, 1990:380.

Park MK, Guntheroth WG. How to Read Pediatric ECGs. Chicago: Year Book, 1981.

Perry JC, Fenrich AL, Hulse JE, et al. Pediatric use of intravenous amiodarone: efficacy and safety in critically ill patients from a multicenter protocol. J Am Coll Cardiol 1996;27:1246.

Pongidglione G, Strasberger JF, Deal BJ, et al. Use of amiodarone for short-term and adjuvant therapy in young patients. Am J Cardiol 1991;68:603.

Roberts ND, Gelband H, eds. Cardiac Arrhythmias in the Neonate, Infant, and Child. New York: Appleton-Century-Crofts, 1977.

Roden DM, Lazzara R, Rosen M, et al. Multiple mechanisms in the long-QT syndrome: current knowledge, gaps, and future directions. Circulation 1996;94:1996.

Smith RT Jr. Pacemakers for children. In: Gillette PC, Garson A, Jr, eds. Pediatric Arrhythmias: Electrophysiology and Pacing. Philadelphia: WB Saunders, 1990:532.

Vetter VL. Ventricular arrhythmias in pediatric patients with and without congenital heart disease. In: Horowitz LN, ed. Current Management of Arrhythmias. Philadelphia: BC Decker, 1990:208.

Vetter VL. What every pediatrician needs to know about arrhythmias in children who have had cardiac surgery. Pediatr Ann 1991;20:378.

Vetter VL, Josephson ME, Horowitz LN. Idiopathic recurrent sustained ventricular tachycardia in children and adolescents. Am J Cardiol 1981;47:315.

Vincent GM, Timothy KW, Leppert M, et al. The spectrum of symptoms and QT intervals in carriers of the gene for the long QT syndrome. N Engl J Med 1992;327:846.

Pericardial Diseases

Engle MA, Zabriskie FB, Senterfit LB, et al. Post-pericardiotomy syndrome: a new look at an old condition: modern concepts. Cardiovasc Dis 1975; 44:59.

Escheverria P, Smith EWP, Ingram D, et al. *Haemophilus influenzae* pericarditis in children. Pediatrics 1975;56:808.

Gersony WM, Hordof AJ. Infective endocarditis and diseases of the pericardium. Pediatr Clin North Am 1978;25:831.

Hirschmann JV. Pericardial constriction. Am Heart J 1978;96:110.

Shabetai R, Fowler ND, Guntheroth WG. The hemodynamics of cardiac tamponade and constrictive pericarditis. Am J Cardiol 1970;26:481.

Van Regen D, Strauss A, Hernandez A, et al. Infectious pericarditis in children. J Pediatr 1974;85:165.

Infectious Endocarditis

Dajani AS, Taubert KA, Wilson W, et al. Prevention of bacterial endocarditis: recommendations by the American Heart Association. JAMA 1997; 277:1734.

Dodo H, Child JS. Infective endocarditis in congenital heart disease. Cardiol Clin 1996;14(3):383.

Durack DT. Prevention of infective endocarditis. N Engl J Med 1996;332:38.

Gewitz MH. Prevention of bacterial endocarditis. Curr Opin Pediatr 1997; 5:518.

Martin JM, Neches WH, Wald ER. Infective endocarditis: 35 years of experience at a children's hospital. Clin Infect Dis 1997;24:669.

Saiman L, Prince A, Gersony WM. Pediatric endocarditis in the modern era. J Pediatr 1993;122:847.

Hypoxemic Attacks

Honey M, Chamberlain DA, Howard J. The effective beta-sympathetic blockage on arterial saturation in Fallot's tetralogy. Circulation 1978; 30:501.

Morgan BC, Guntheroth WG, Bloom RS, et al. A clinical profile of paroxysmal hyperpnea in cyanotic congenital heart disease. Circulation

1965;31:66.

Nudel DB, Berman MA, Talner NS. Effects of acutely increasing systemic vascular resistance on oxygen tension in tetralogy of Fallot. Pediatrics 1976;58:248.

Pinsky WW, Arciniegas E. Tetralogy of Fallot. Pediatr Clin North Am 1990;37:179.

Rheumatic Fever

Albert DA, Harel L, Karrison T. The treatment of rheumatic carditis: a review and meta-analysis. Medicine (Baltimore) 1995;74:1.

Dajani AS, Ayoub E, Bierman FZ, et al. Guidelines for the diagnosis of rheumatic fever. Jones Criteria, updated 1992. JAMA 1992;268:2069.

Dajani A, Taubert K, Ferrieri P, et al. Treatment of acute streptococcal pharyngitis and prevention of rheumatic fever: a statement for health professionals. Pediatrics 1995;96:758.

Kaplan EL. Global assessment of rheumatic fever and rheumatic heart disease at the close of the century. Circulation 1993;88:1964.

Stollerman GH. Rheumatic fever. Lancet 1997;349:935.

Veasey LG, Wiedmeir SE, Garth SO. Resurgence of acute rheumatic fever in the intermountain area of the United States. N Engl J Med 1987;316:421.

CHAPTER 83

Neurologic Emergencies

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Signs and symptoms of neurologic dysfunction in children either are produced by primary nervous system disorders or are secondary to systemic disease. The differential diagnosis of many such neurologic findings can be found in the first section of this book. This chapter focuses on the management of conditions primarily involving the various parts of the nervous system, including the brain, spinal cord, and peripheral nerves. The illnesses are classified by their most prominent clinical manifestations: seizures, altered mental status, headache, weakness, disorders of balance, abnormal movements, and cranial nerve dysfunction.

SEIZURES (SEE ALSO CHAPTER 70)

Seizures are among the more common neurologic symptoms that lead to an emergency department (ED) visit. Epidemiologic studies indicate that from 3 to 6% of children will have at least one seizure in the first 16 years of life; most of these are simple febrile seizures, discussed in the following section. Fortunately, recurrent seizures or other signs of neurologic dysfunction occur in only a small number of these children. However, the first seizure is always frightening and produces anxiety.

A *seizure* is defined as a transient, involuntary alteration of consciousness, behavior, motor activity, sensation, and/or autonomic function caused by an excessive rate and hypersynchrony of discharges from a group of cerebral neurons. The term *convulsion* is often used to describe a seizure with prominent motor manifestations. Epilepsy, or seizure disorder, is a condition of susceptibility to recurrent seizures.

Most seizures are brief, lasting less than 10 to 15 minutes. *Status epilepticus* refers to seizures that are continuous for 30 minutes or longer or to repetitive seizures between which the patient does not regain consciousness.

Pathophysiology

The basic pathophysiologic abnormality common to all seizures and convulsions is the hypersynchrony of neuronal discharges. Many precipitating factors, including metabolic, anatomic, and infectious abnormalities (see Chapter 70), may produce seizures. Seizures that result from an identified precipitant are called symptomatic, or provoked, seizures, whereas those with no precipitating factor are called idiopathic or cryptogenic. Febrile seizures (seizures occurring in association with a febrile illness, without evidence of intracranial infection or other identified cause) are a particular type of provoked seizure seen in children between the ages of 6 months and 6 years. The exact cause of febrile seizures remains elusive. Elevated body temperature lowers the seizure threshold, and the immature brain appears to have a particular susceptibility to seizures in response to fever. It is unclear whether height of fever or rate of temperature rise is more important in inducing febrile seizures, but individual predisposition plays an important role.

During a seizure, cerebral blood flow, oxygen and glucose consumption, and carbon dioxide and lactic acid production increase. If the patient remains well-ventilated, the increase in cerebral blood flow is sufficient to meet the increased

metabolic requirements of the brain. Brief seizures rarely produce lasting deleterious effects on the brain; however, prolonged and serial seizures, especially status epilepticus, may be associated with permanent neuronal destruction.

Clinical Manifestations

When the physician is faced with a child with an acute paroxysmal event, the first step is to distinguish seizures from other nonepileptic phenomena. If the event is indeed a seizure, it may be classified according to type. Finally, a specific causative factor should be sought. The extent of the emergency evaluation is determined by the clinical scenario; some of the diagnostic assessment may be deferred. Of course, when a child is actively seizing, the first priority is to provide necessary resuscitation measures and control the seizures (see Chapter 70 and the following).

Nonepileptic Paroxysmal Events

Paroxysmal events other than seizures that involve changes in consciousness or motor activity are common during childhood and may mimic epilepsy (Table 83.1). Breath-holding spells occur in children 6 months to 4 years of age. Breath-holding spells take two forms: cyanotic and pallid. In the cyanotic form, the infant begins crying vigorously, often in response to an inciting event, then holds his or her breath and becomes cyanotic. After approximately 30 to 60 seconds, the child becomes rigid. As the spell ends, the child becomes limp and may have a transient loss of consciousness and twitching or jerking of the extremities, but quickly returns to full alertness. A pallid breath-holding spell may follow a seemingly insignificant trauma. The child may start to cry, but then turns pale and collapses. There is a brief period of apnea and limpness, followed by rapid recovery. In both types of breath-holding spells, the typical history and lack of postictal drowsiness help determine the diagnosis. Breath-holding spells may be recurrent but disappear spontaneously before school age.

Breath-holding spells	Acute dystonia
Syncope	Gastroesophageal reflux
Migraine	Night terrors
Jitteriness	Sleep paralysis
Benign myoclonus	Narcolepsy
Shuddering attacks	Pseudoseizures
Tics	

Table 83.1. Nonepileptic Events That May Mimic Seizures

Syncope is a brief, sudden loss of consciousness and muscle tone. There are numerous causes of syncope, many of which can be detected on the basis of historical information, physical examination, and simple laboratory tests (see Chapter 73). A syncopal episode can usually be distinguished from a seizure based on the description. The child is typically upright before the event and often senses a feeling of light-headedness or nausea. The child then becomes pale and slumps to the ground. The loss of consciousness is brief, and recovery is rapid. On awakening, the child is noted to have signs of increased vagal tone, such as pallor, clammy skin, dilated pupils, and relative bradycardia. Patients with narcolepsy also experience sudden alterations in alertness, with sleep occurring suddenly and uncontrollably during the daytime. In about half of the patients, narcolepsy is associated with cataplexy, a sudden loss of muscle tone brought on by a sudden emotional outburst. Narcolepsy is far less common than syncope; both occur more often in adolescents than in younger children.

Single episodes of staring, involuntary movements, or eye deviation have been found to occur commonly in the first months of life, although they rarely lead to the parent seeking medical attention. In some children, however, these episodes occur frequently. Children with benign shuddering attacks have episodes of staring and rapid tremors involving primarily the arms and head, sometimes associated with tonic posturing. The episode lasts only a few seconds, and afterward the child resumes normal activity. Acute dystonia, usually seen as a side effect of certain medications, can mimic a tonic seizure. The child having a dystonic reaction, however, does not lose consciousness and has no postictal drowsiness.

Several paroxysmal events are associated with sleep. Night terrors (see Chapter 131) usually begin in the preschool years. The sleeping child wakes suddenly, is confused and disoriented, and appears frightened, often screaming and showing signs of increased autonomic activity (tachycardia, tachypnea, sweating, dilated pupils). Such episodes typically last only a few minutes, and the child does not usually recall the event. Benign myoclonus is characterized by self-limited episodes of sudden jerking of the extremities, usually upon falling asleep. There is no alteration of consciousness. In sleep paralysis, there is a transient inability to move during the transition between sleeping and waking, also with no change in level of consciousness.

Pseudoseizures are occasionally seen, often in patients with an underlying seizure disorder or with a relative with epilepsy. Some features suggestive of pseudoseizures are suggestibility, lack of coordination of movements, moaning or talking during the “seizure,” lack of continence, autonomic changes, postictal drowsiness, and poor response to treatment with anticonvulsant agents.

The most important diagnostic test in distinguishing nonepileptic events from seizures is a careful history, including a detailed description of the event from the person who witnessed it. In atypical or unclear cases, referral for

electroencephalogram (EEG) or video EEG monitoring may help in establishing the diagnosis.

Types of Seizures

Clinically, seizures may be divided into partial and generalized seizures (Table 83.2). Generalized tonic-clonic seizures (previously called grand mal seizures) are the type most often seen in acute pediatric care. The onset of generalized tonic-clonic seizures usually is abrupt, although 20 to 30% of children may experience a sensory or motor aura. If sitting or standing, the child falls to the ground. The face becomes pale, the pupils dilate, the eyes deviate upward or to one side, and the muscles contract. As the increased tone of the thoracic and abdominal muscles forces air through the glottis, a grunt or cry may be heard. Incontinence of urine or stool is common. After this brief tonic phase (10 to 30 seconds), clonic movements occur. The child is unresponsive during the seizure and remains so, postictally, for a variable period. After the seizure, there may be weakness or paralysis of one or more areas of the body (Todd's paralysis). In atonic, or akinetic, seizures (drop attacks), there is abrupt loss of muscle tone and consciousness. Myoclonic seizures are characterized by a sudden dropping of the head and flexion of the arms (“jackknifing”); however, extensor posturing also may occur. The episodes occur quickly and frequently, as often as several hundred times daily.

Generalized	Partial (Focal)
Absence (petit mal)	Simple (no impaired consciousness)
Typical	Motor
Atypical	Sensory
Tonic-clonic (grand mal)	Autonomic
Clonic	Psychic
Tonic	Complex (impaired consciousness)
Myoclonic	Partial seizures becoming partially generalized
Akinetic/atonic (drop attacks)	

Table 83.2. Seizure Types

Absence (petit mal) seizures are generalized seizures marked by sudden and brief loss of awareness, usually lasting 5 to 30 seconds. With typical absence seizures, there is no loss of posture or tone and no postictal confusion. There may be a minor motor component such as eyelid blinking.

The child with simple partial (focal) seizures has unimpaired consciousness. Motor signs are most common in children, although sensory, autonomic, and psychic manifestations are possible. The motor activity usually involves the hands or face and spreads in a fixed pattern determined by the anatomic origin of the nerve fibers that innervate the various muscle groups. Focal seizures may become secondarily generalized, in which case there will be alteration of consciousness. Complex partial seizures, also called psychomotor or temporal lobe seizures, exhibit a diverse set of clinical features, including alterations of perception, thought, and sensation. In children, they are usually marked by repetitive and complex movements with impaired consciousness and postictal drowsiness.

Establishing an Underlying Cause

The first steps in the evaluation of seizures are a thorough history and a physical examination, the results of which are helpful in determining the direction of the search for a specific cause. Important historical items to elicit include fever, trauma, underlying illnesses, current medications, and possible toxic ingestions. A complete neurologic assessment to evaluate for signs of increased intracranial pressure (ICP), focal deficits, or signs of meningeal irritation is also essential.

An important distinction is whether the seizure is associated with fever. Simple febrile seizures are those that are single, brief (less than 15 minutes), and generalized. Approximately 20% of febrile seizures are complex, meaning they are focal, are prolonged (more than 15 minutes), or occur multiple times during the same illness. In children older than 12 months of age with a typical simple febrile seizure and no evidence of meningeal signs, no further evaluation of the seizure is generally required. Clinically unsuspected meningitis is exceedingly rare in such children. In one study of 503 children with meningitis, none presented with an isolated febrile seizure; conversely, two other studies of 803 children found meningitis in none of the children with a febrile seizure but no other clinical findings of meningitis. Lumbar puncture (LP) is mandatory if meningitis is suspected on the basis of physical findings. An LP should be strongly considered in children younger than 12 months of age, in whom signs of meningitis may be subtle, or when the febrile seizure is complex. In addition, LP should be considered for children with prolonged fever before the seizure, particularly those who have sought medical care in the previous 48 hours and who have been found to be at higher risk. Other laboratory tests discussed in the next paragraph have been found to have little yield in the child with a typical febrile seizure and are unnecessary. Other diagnostic tests to determine the source of the fever are determined by other features such as the height of fever and child's age because the frequency of specific infections such as occult bacteremia is not increased in children who have experienced a febrile seizure.

For the child who presents with a first-time, nonfebrile seizure, laboratory or radiologic evaluation to search for a specific treatable cause of the seizure may be indicated. There is little utility in extensive, routine workups; rather, ancillary test selection should be guided by the results of the history and physical examination. Because hypoglycemia may be clinically difficult to detect, a rapid glucose test should be performed on all children. In young infants, children with prolonged seizures, and those with a suggestive history or physical examination, determination of serum sodium and calcium is also indicated. Other laboratory tests that may be indicated, depending on the clinical picture, include serum magnesium, hepatic transanimases, ammonia, and serum or urine toxicology tests. LP is rarely emergently necessary in

the afebrile child without meningeal signs, although it should be considered in neonates even without fever.

In children with a known seizure disorder, subtherapeutic anticonvulsant levels are the most common reason for recurrent seizures. The name and dosage of anticonvulsant medications used should be elicited, as well as the time of the last dose given, any missed doses, the last change in dosage, and recent levels if known. Intercurrent illness may also play a role because the metabolism of some medications is affected by systemic illness. Such children should have blood drawn for measurement of anticonvulsant levels. Although many drugs have a therapeutic range (Table 83.3), individual patients may require levels outside that range for adequate seizure control; conversely, dose-dependent toxic effects may be observed in some children even at typically therapeutic levels.

Drug	Seizure Type	Daily Dose (mg/kg)	Oral Dosage Forms	Serum Half-Life (h)	Therapeutic (Blood Levels (µg/mL)
Carbamazepine (Tegretol)	Generalized motor, partial, complex partial	10–30	Tablets: 100, 200 mg Suspension: 100 mg/5 mL	8–24	4–12
Phenytoin (Dilantin)	Generalized motor, partial, complex partial	3–10	Capsule: 100 mg Chewable tab: 50 mg Suspension: 125 mg/5 mL	10–30	10–20
Phenobarbital	Generalized motor, partial, complex partial	3–4	Tablets: 15, 30, 60, 100 mg Elixir: 20 mg/5 mL	24–96	15–40
Primidone (Mysoline)	Generalized motor, partial, complex partial	10–25	Tablets: 50, 250 mg Suspension: 50 mg/5 mL	12	5–12 (plus measure phenobarbital level)
Valproate (Depakote)	Absence, myoclonic, partial complex, generalized motor	20–40	Tablets: 125, 250 mg Sprinkles: 125 mg Syrup: 250 mg/5 mL	8–18	50–100
Ethosuximide (Zarontin)	Absence	20–40	Capsule: 250 mg Syrup: 250 mg/5 mL	30–60	40–100
Lamotrigine (Lamictal)	Partial, atonic, myoclonic, mixed types	10–15	Tablets: 25, 100, 150 mg	24	Not known
Clonazepam (Klonopin)	Atonic, myoclonic, generalized motor	0.03–0.1	Tablets: 0.5, 1, 2 mg	18–50	0.020–0.080 (20–80 ng/mL)

Table 83.3. Commonly Used Anticonvulsant Agents

Computed tomography (CT) and magnetic resonance imaging (MRI) allow detailed visualization of the gross anatomy of intracranial structures by a noninvasive technique. Presently, CT is more available on an emergent basis in most institutions. It also is a shorter procedure, and patient monitoring is usually easier. CT (or MRI, if available) is indicated in the emergency evaluation of prolonged or focal seizures, when focal deficits are present, when there is a history of trauma, when the child has a ventriculoperitoneal shunt, or when there are associated signs of increased ICP. For other children, an imaging study may be useful in identifying structural anomalies and determining prognosis, but such studies may be deferred to a follow-up visit. Cranial imaging is not indicated in the evaluation of simple febrile seizures.

EEG is also helpful in the evaluation of children with nonfebrile seizures. It is rarely beneficial in acute management, but children with nonfebrile seizures should be referred for outpatient testing.

Management

Resuscitation and Supportive Care

The administration of nasal oxygen and maintenance of an adequate airway are vital parts of the initial management of the unconscious, actively convulsing child (see Chapter 70). Trismus often occurs in generalized seizures but is transient. If the teeth are tightly clenched, even the placement of the airway should be deferred until it can be inserted without undue trauma during a phase of relaxation. Seizure-associated hypoventilation and apnea are common with prolonged seizures, often as a side effect of anticonvulsant medications, and providers caring for such children should be prepared to offer assisted ventilation. Intravenous access should be established promptly; however, because of the potential for increased ICP, fluid therapy should be used judiciously until a more thorough evaluation is performed. The child with active convulsions should be protected from trauma. There is no benefit to placing objects in the child's mouth to prevent tongue biting.

Stopping the Seizure

It is unusual for the child with a brief seizure to arrive in the ED actively convulsing because, by definition, such seizures last less than 15 minutes. Therefore, the actively convulsing child usually is already in a prolonged or serial seizure state, and pharmacologic intervention to terminate the seizure is required (Fig. 83.1).

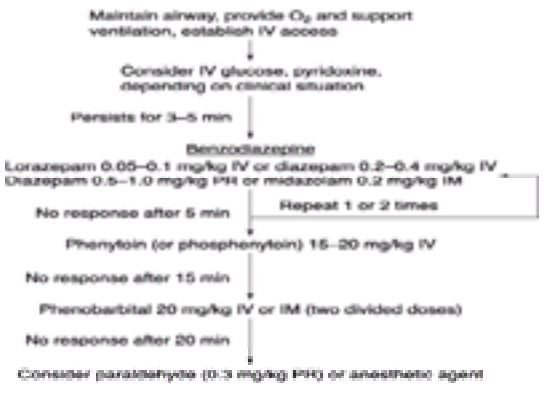

FIGURE 83.1. Treatment of status epilepticus. O_2, oxygen; *IV,* intravenous; *PR,* per rectum; *IM,* intramuscular.

Intravenous access is established, and blood is drawn for diagnostic studies. If hypoglycemia is documented by rapid glucose assay or if rapid determination is unavailable, intravenous glucose is given in a dose of 2 to 4 mL/kg of 25% dextrose in water. In neonates or in children with suspected isoniazid toxicity, pyridoxine 100 mg intravenously (IV) may be administered.

In most situations, benzodiazepines are the first drug of choice for acute seizures because of their rapidity of action. Lorazepam (Ativan) is the preferred agent. Given in a dose of 0.05 to 0.1 mg/kg IV (maximum 4 mg/dose), it has an onset of action of 2 to 5 minutes, and the duration of anticonvulsant effect is 12 to 24 hours. The dose may be repeated after 5 to 10 minutes. An alternative is diazepam (Valium), 0.2 to 0.4 mg/kg IV (maximum 10 mg/dose), which has a similarly rapid onset of action but a much shorter duration of anticonvulsant activity, usually less than 30 minutes. Thus, if diazepam is used, another agent for longer-term control, such as phenytoin, is needed to prevent seizure recurrence. If IV or intraosseous access cannot be established, diazepam may be administered rectally in a dose of 0.5 to 1.0 mg/kg, instilling the IV formulation with a syringe. Intramuscular midazolam (Versed) has also been shown to be effective in a dose of 0.2 mg/kg (maximum 7 mg). Midazolam may also be given intravenously.

All of the benzodiazepines can cause sedation and respiratory depression. Equipment for establishing an airway and supporting respiration must be available, especially if repeated doses are used. Sedation and respiratory depression may persist for hours, particularly with diazepam. Hypotension is uncommon but may be a problem with multiple doses or when barbiturates are administered concomitantly.

If the seizures have not been controlled within 15 minutes with benzodiazepines, phenytoin (Dilantin) should be given. Phenytoin is administered IV, at a loading dose of 15 to 20 mg/kg. In patients known to be taking phenytoin chronically, a smaller dose of 5 to 10 mg/kg should be used initially unless the serum level is known to be very low. Each 1 mg/kg of phenytoin administered raises the serum level by approximately 1 mg/mL. Phenytoin is highly lipid soluble and reaches therapeutic levels in the brain within 10 to 20 minutes, with a duration of action of 12 to 24 hours. Because rapid IV infusion may lead to hypotension and cardiac arrhythmias, the maximum rate of administration is 50 mg/minute, and cardiac monitoring is required. Local reactions such as thrombophlebitis are also common. Fosphenytoin (Cerebyx) is a recently developed prodrug of phenytoin, which is rapidly metabolized to the active form. It offers several advantages over phenytoin, including more rapid administration (150 mg/minute) and fewer local and systemic side effects. Fosphenytoin may also be given intramuscularly, unlike phenytoin. The dose of the two drugs is identical; fosphenytoin doses are expressed as phenytoin equivalents. Although pediatric data are sparse, the pharmacokinetics and safety profile of fosphenytoin appears to be similar to that seen in adult patients. Unlike other anticonvulsant medications, phenytoin does not cause sedation or respiratory depression.

Phenobarbital is the next agent to be added if phenytoin is not effective or contraindicated (e.g., allergy, known therapeutic level). The loading dose of phenobarbital is 20 mg/kg, usually given in two divided doses. The drug is given intravenously over 5 to 10 minutes, or intramuscularly in the absence of IV access. Onset of action is usually within 15 to 20 minutes and lasts more than 24 hours. Phenobarbital, like other barbiturates, may cause significant sedation and hypotension.

Patients with status epilepticus that lasts more than 60 minutes present a special problem. Further management should be done in conjunction with a neurologist and with EEG monitoring when possible. Continuous infusion of benzodiazepines may be used. Paraldehyde is often effective but is difficult to handle because of its foul odor, reactivity with rubber and plastic (necessitating the use of glass syringes), and lack of water solubility. A dose of 0.3 mg/kg diluted 1:1 in oil may be given rectally. Other agents potentially useful in the management of refractory status epilepticus include pentobarbital and general anesthetics such as isoflurane, etomidate, and propofol.

With prolonged seizures, the duration of postictal drowsiness and confusion may also be protracted. However, the child who fails to arouse within 15 to 30 minutes after cessation of seizures should be evaluated carefully to rule out nonconvulsive status epilepticus. Children with status epilepticus, even if successfully treated in the ED, should be admitted to the hospital for monitoring and observation.

Rarely, a child may enter the ED in absence status. In this case, the child may be sitting in a confused or dreamy state. Such attacks may last for hours or even days. The drug of choice in the treatment of absence status is lorazepam or diazepam at the dosages already outlined.

At times, a child may enter the ED with continual focal seizure activity (with or without clouding of consciousness), a condition known as epilepsia partialis continua. The treatment for partial seizures is less urgent than that for generalized seizures, and such seizures are often intractable to anticonvulsant medication. In such cases, phenytoin or fosphenytoin in a dose of 18 to 20 mg/kg can be infused slowly. All such patients should be admitted to the hospital for further observation and evaluation. Other pharmacologic attempts to control these focal seizures should be performed in the hospital.

Initiating Anticonvulsant Medication

Nonfebrile Seizures The decision to initiate long-term prophylactic therapy with anticonvulsant medications is based on a consideration of a number of factors, including the patient's age, type of seizure, risk of recurrence, coexisting medical conditions, and family factors. The consequences of further seizures must be balanced against the potential side effects of the anticonvulsant agents. Treatment is seldom started after a single, uncomplicated nonfebrile seizure because most such patients will not experience a seizure recurrence. On the other hand, a patient who has had two or more such seizures should generally receive anticonvulsant therapy. When possible, it is preferable for long-term treatment decisions to be made in conjunction with the provider who will be responsible for ongoing follow-up of the patient, either a neurologist or the child's primary care physician. Sometimes, it may be necessary to begin prophylactic treatment in the

ED, pending a more complete outpatient evaluation.

A number of drugs are effective in preventing seizures (Table 83.3). Some are better for certain types of seizures, and all have different profiles of adverse effects. The following principles should guide selection of an anticonvulsant medication:

1. Choose a drug that is effective for the particular type of seizure. When more than one agent is available, choose the least toxic one. Initial therapy should be with a single agent.
2. Start at the low end of the dosage range.
3. Arrange for a serum level of the drug to be measured, when appropriate. This is done after a steady state is anticipated, usually five times the half-life of the drug.
4. If a child is already taking an anticonvulsant medication and has an adequate level, consider adding another agent.

Carbamazepine (Tegretol) Carbamazepine is effective against generalized tonic-clonic seizures as well as simple and complex partial seizures. The effective serum concentrations of carbamazepine range between 4 and 12 µg/mL, but with this drug, there is a variable correlation among clinical efficacy, toxicity, and the serum concentration. Recommended maintenance dosages range between 10 and 30 mg/kg per day, divided into three daily doses. The administration of a total maintenance dose to a previously untreated patient often results in drowsiness, blurred vision, and at times, severe lethargy, so this drug should be initiated by gradual increases in dosage (10 mg/kg per day to start, increased by 5 mg/kg per day every 3 to 4 days) until a full maintenance level is reached. Unlike several other agents, it is not available in an IV form. Concomitantly administered medications that may lead to toxic carbamazepine levels include macrolide antibiotics (e.g., erythromycin), isoniazid, cimetidine, verapamil, and diltiazem.

Carbamazepine may cause hepatic and hematologic toxicity but causes little, if any, cognitive dysfunction in most patients. Therefore, it often is the drug of choice for children with generalized seizures.

Phenobarbital Phenobarbital is another broad-spectrum anticonvulsant useful for generalized tonic-clonic and partial (simple and complex) seizures. It remains a commonly used initial drug, primarily because of its low cost and low toxicity. The effective serum concentration ranges between 15 and 40 µg/mL. This serum level usually can be maintained with a dosage of 3 to 6 mg/kg per day in children and 1 to 2 mg/kg per day in adolescents, administered in divided doses twice daily. A loading dosage of approximately twice the maintenance dosage (6 to 10 mg/kg per day in children and 2 to 4 mg/kg per day in adolescents) for 2 to 3 days brings the serum concentration to the therapeutic range within 48 to 72 hours. Such loading dosages usually are associated with considerable transient drowsiness. There is a wide margin between the anticonvulsant and soporific effects of phenobarbital, and drowsiness rarely persists at the recommended dosages. Decreased attention, hyperactivity, and alterations of mood occur in 30 to 50% of children maintained on phenobarbital. These behavioral changes are the most commonly encountered side effects and often are sufficiently undesirable to force the change to another drug. Possible associated long-term cognitive effects also makes phenobarbital problematic, and many clinicians do not consider it a first-line drug.

Primidone (Mysoline) Primidone has a similar effectiveness and side effect profile to phenobarbital, one of its main metabolites. The maintenance dosage is 10 to 25 mg/kg per day, given in two to four divided doses. The therapeutic serum level is 5 to 12 µg/mL; the serum level of phenobarbital should also be monitored when using primidone.

Phenytoin (Dilantin) Phenytoin is another agent effective in the treatment of several seizure types, including generalized motor seizures and both simple and partial complex seizures. The effective serum concentration of phenytoin is between 10 and 20 mg/mL. The usual maintenance dosage is 7 to 10 µg/kg per day in children weighing less than 20 kg, 5 to 7 mg/kg per day in children weighing between 20 and 40 kg, and 5 mg/kg per day in children weighing more than 40 kg, given in once or twice daily doses. However, there is considerable variation in metabolism among patients. Saturation of biotransforming enzyme systems often occurs between serum levels of 10 and 20 µg/mL, so small changes in a dose in this range may lead to relatively large changes in serum levels.

Loading dosages of four times the daily dosage (maximum 20 mg/kg per day) on the first day and two times the daily dosage for the next 2 days will bring serum levels into the therapeutic range within 24 hours; side effects rarely occur with this loading dosage. Gingival hyperplasia is a common side effect and may be seen with phenytoin concentrations in the therapeutic range; this cosmetic side effect and the drug's tendency to cause hirsutism and coarsening of facial features often limit long-term use, especially in girls. Drowsiness, ataxia, nystagmus, and seizures are dose-dependent toxic effects rarely seen with levels in the therapeutic range. Other adverse effects include drug rashes (Stevens-Johnson syndrome) and hematologic and hepatic side effects. Several medications may cause increased phenytoin levels: cimetidine, estrogens, chlorpromazine, chloramphenicol, and isoniazid.

Valproate (Depakote) Valproate is highly effective in the treatment of generalized epilepsy, including especially absence and myoclonic seizures, as well as simple and complex partial seizures. Doses of 20 to 40 mg/kg usually result in therapeutic levels of 50 to 100 µg/mL. However, serum drug levels may not be highly predictive of efficacy or toxicity. The primary side effects include gastrointestinal upset and drowsiness; hepatic, renal, pancreatic, and hematologic dysfunction are also seen. Children younger than 2 years old are at particular risk of idiosyncratic fatal hepatotoxicity. Therefore, valproate is rarely the initial drug of choice for young children with generalized seizures.

Clonazepam (Klonopin) Clonazepam is used to control myoclonic and atonic seizures. The usual dosage is 0.05 to 0.2 mg/kg per day, given in two to four divided doses. The therapeutic range is 0.02 to 0.08 µg/mL (10 to 80 ng/mL). Patients taking clonazepam may experience drowsiness, ataxia, and drooling.

Ethosuximide (Zarontin) Ethosuximide is indicated for the management of absence seizures. It is given at a dosage of 20 to 40 mg/kg per day divided into twice daily doses, with a usual therapeutic level of 40 to 100 µg/mL. Side effects include headache, nausea, and vomiting; erythema multiforme and a lupuslike syndrome have also been reported.

Lamotrigine (Lamictal) Lamotrigine is a new agent available for treatment of partial seizures, atonic and myoclonic

seizures, and intractable mixed seizures (Lennox-Gastaut syndrome). The usual dosage is 10 to 15 mg/kg per day, which is reduced to 5 mg/kg per day when given in conjunction with valproate. Drowsiness, vomiting, and drug rash (including Stevens-Johnson syndrome) are reported side effects.

Other Agents Several new agents are newly available in the United States or currently under investigation. These include vigabatrin (Sabril), gabapentin (Neurontin), and felbamate (Felbatol). Because of the risk of severe hepatotoxicity, felbamate is restricted to use in children with intractable seizures refractory to other treatment.

Febrile Seizures. For children with febrile seizures, the issue of chronic prophylactic medication is more controversial. Presumptive antipyretic therapy for a nonspecific illness does not appear to reduce the risk of seizure recurrence, although it may give the parent a sense of “doing something.” Phenobarbital was widely used in the past for children with recurrent febrile seizures, but this practice is much less common now because of concerns about adverse cognitive and behavioral effects of the medication. To be effective, phenobarbital must be given continuously. Other commonly used anticonvulsant agents such as phenytoin and carbamazepine appear to be ineffective. More recently, some clinicians have used diazepam, administered intermittently during febrile illnesses (0.33 mg/kg every 8 hours), to prevent febrile seizures. One controlled trial showed this treatment to be effective, albeit with a high incidence of side effects; in addition, other studies have failed to confirm the effectiveness of this approach, largely as a result of poor compliance or inadequate recognition of fever. With little evidence that febrile seizures (even febrile status epilepticus) cause permanent neurologic damage or that their control results in a lower incidence of subsequent epilepsy, there is little need to treat most patients. In carefully considered individual cases, long-term continuous therapy with phenobarbital or intermittent therapy with diazepam may be considered. This should usually be done in conjunction with the child's primary care provider.

Disposition Hospital admission is generally required for children who have had a prolonged seizure requiring acute treatment with anticonvulsant medication. Other children, even those with a first-time seizure, can generally be followed as outpatients if they appear well after the seizure, follow-up can be ensured, and the parents are comfortable with home management. Seizure first-aid should be explained to the family before discharge.

After a simple febrile seizure, hospitalization is seldom necessary, and children may be followed by their primary physician. Some useful information can be given to parents after a first febrile seizure. First, they should be informed of the benign nature of the convulsions and the lack of evidence that they cause any type of neurologic injury. Approximately one-third of children with a first febrile seizure will have another one. Of recurrences, 75% occur within 1 year, and they are uncommon beyond 2 years; fewer than 10% of children with febrile seizures have more than three. The recurrence rate is lower if the seizures begin after the first year of life, and the risk is also reduced in children with higher temperature and longer duration of fever before the initial febrile seizure. For example, the recurrence risk is about 35% when the first seizure occurs at a temperature of 38.5°C (101.3°F), compared with a risk of 13% with a temperature of 40°C (104°F). Having a complex first febrile seizure (even febrile status epilepticus) does not increase the risk of recurrence, nor does it increase the chance that a recurrent seizure, if it occurs, will be complex.

Many parents worry that febrile seizures will lead to future epilepsy. A child who has had a febrile seizure but no other risk factors for epilepsy may have a slightly increased risk of future nonfebrile seizures, but the magnitude of this increase is still extremely small: 1 to 2% lifetime risk versus a 0.5 to 1% lifetime risk in the general population. Several risk factors that increase the likelihood of a child experiencing future nonfebrile seizures have been identified. These risk factors include a family history of epilepsy, a complex febrile seizure, and the presence of an underlying neurologic or developmental abnormality. Importantly, even with two or more of these risk factors, the risk of epilepsy is only 10%. Thus, for most children with no risk factors, the parents may be reassured that future epilepsy, although possible, is extremely unlikely. Furthermore, there is no association between febrile seizures and any type of developmental or learning disabilities.

DISORDERS THAT PRESENT WITH ENCEPHALOPATHY (SEE CHAPTER 13)

Encephalopathy is an imprecise term that implies diffuse brain dysfunction with or without alterations in the level of consciousness. The emergency physician often must decide whether the child's degree of irritability, uncooperativeness, and lethargy is proportionate to the degree of systemic illness; whether it is caused by fear; or whether it represents cortical dysfunction. Encephalopathy may be a sign of numerous systemic disorders, or it may result from certain primary disorders of the central nervous system (CNS), discussed next.

Encephalitis

Background

Encephalitis is an inflammation of the brain parenchyma. When there is an associated leptomeningeal involvement (as often occurs), the term *meningoencephalitis* may be applied, whereas *encephalomyelitis* implies involvement of the spinal cord as well. CNS dysfunction is caused by direct invasion of brain by a pathogen, most often a virus, or is secondary to immunologic mechanisms, as in postinfectious encephalomyelitis.

Viral encephalitides are caused by a wide variety of viruses that lead to clinically similar illnesses (Table 83.4). Mumps was the most common cause of meningoencephalitis before the introduction of vaccination, with up to 50% of patients with mumps parotitis having cerebrospinal fluid (CSF) pleocytosis. Classically, the illness occurs several days to 2 weeks after the onset of parotitis but may precede the onset of systemic illness or occur without parotitis and tends to be mild. Measles encephalitis is less common since the advent of widespread live immunization. The onset usually occurs during the prodromal period or after the rash has appeared. Ataxia is the most common neurologic abnormality, and sequelae occur in up to 30% of cases. Varicella encephalitis occurs 2 to 9 days after the onset of the rash; severe infections are uncommon, except in the immunosuppressed host.

Arboviruses	Varicella-zoster
Eastern equine encephalitis	Epstein-Barr
Western equine encephalitis	Cytomegalovirus
St. Louis encephalitis	Mumps
Japanese encephalitis	Measles
California (LaCrosse) encephalitis	Enteroviruses
Herpesviruses	Rabies
Herpes simplex	

Table 83.4. Agents of Viral Encephalitis

The arthropodborne encephalitides--including St. Louis, Western equine, Eastern equine, and California encephalitis--occur in sporadic and epidemic forms, often in late summer or early fall, and tend to cluster in localized geographic areas. Sequelae may be severe and mortality high, especially in eastern equine encephalitis.

Herpes simplex virus (HSV) is a common cause of sporadic encephalitis. Disease in neonates is usually caused by HSV type 2, acquired from perinatal transmission. In previously healthy older children and adults, encephalitis usually results from infection with HSV type 1 and may be a complication of acute primary infection or reactivated latent infection. Recognition of herpes encephalitis is important because specific antiviral therapy reduces the substantial morbidity and mortality of this disease.

Infection with rabies virus, although rare in the United States, is an important cause of encephalitis worldwide. Nonviral pathogens, including *Mycoplasma pneumoniae,* Lyme disease, and rickettsiae, may also cause encephalitis.

Postinfectious encephalitis may follow infection with numerous viruses, including measles, varicella, influenza, and Epstein-Barr virus. The CNS involvement may be confined to a specific area, as in acute cerebellar ataxia after varicella infection, or may be widespread. The latter condition is often designated acute disseminated encephalomyelitis. A particularly virulent form with high mortality is known as acute hemorrhagic leukoencephalitis. A clinical syndrome of encephalopathy after immunization, particularly with whole-cell pertussis vaccine, is also described, although more recent epidemiologic evidence has called into question the association with pertussis immunization.

Pathophysiology

Viral encephalitis usually follows a viremia, although direct spread can occur less commonly via peripheral nerves or the nasal mucosa. Upon reaching the CNS, viral replication in neural cells interferes with cellular function and may lead to cell death. Cerebral edema may result from capillary leakage, with subsequent increased ICP. The degree and extent of neuronal dysfunction depends in part on the pathogen involved and also on host factors, especially immunocompetence. In general, the incidence of overt neurologic findings and sequelae is higher in children younger than 1 year of age.

Postinfectious encephalitis is presumed to be an immune-mediated phenomenon, involving the white matter of the CNS. Demyelination, the pathologic hallmark of the disease, may be focal or widespread.

Clinical Manifestations

The clinical picture of viral encephalitis ranges from a mild febrile illness associated with headache to a severe, fulminant presentation with coma, seizures, and death. The onset of encephalitis may be abrupt or insidious. Typical features consist of fever, headache, vomiting, and signs of meningeal irritation. Altered consciousness, ataxia, and seizures are also seen. Focal neurologic deficits occur in certain types of encephalitis, particularly HSV. Flaccid paralysis may be seen in cases of encephalomyelitis, and rarely, respiratory or cardiac dysfunction results from brainstem involvement. Rash or mucous membrane lesions are often seen with the exanthematous viruses such as measles and varicella; however, cutaneous findings are uncommon with HSV encephalitis.

Laboratory assessment often is nonspecific. The peripheral blood count usually shows a mild polymorphonuclear or mononuclear leukocytosis. With viral encephalitides, CSF pleocytosis is variable and, if present, usually is fewer than 500 cells/mm^3. These cells may be predominantly polymorphonuclear early in the course of the illness; however, a mononuclear predominance is common later. Red blood cells are present in the CSF in approximately 50% of children with herpes encephalitis. Spinal fluid protein and glucose usually are normal with viral encephalitis, but the protein may be greatly elevated in postinfectious encephalomyelitis.

Virus isolation from the CSF may be difficult but should be attempted, as should viral isolation from other body sites, including the nasopharynx, skin lesions, urine, and feces. Serologic evidence for viral infection based on acute and convalescent IgG titers, although useful later, gives little help in making an immediate diagnosis. Infection with arboviruses may be established more rapidly by detecting virus-specific IgM in CSF or serum.

Diagnosis of herpes simplex poses a special problem because early diagnosis is important in instituting effective therapy. When available, polymerase chain reaction (PCR) testing of CSF yields rapid evidence of viral nucleic acid and is highly sensitive and specific. Imaging studies, although less sensitive, may also be useful. Either CT or MRI may demonstrate focal parenchymal involvement or edema of the temporal lobes (Fig. 83.2). MRI is more sensitive than CT, although both may be normal in the early stages of disease. Similarly, EEG may demonstrate focal slowing or epileptiform discharges

localized to the temporal lobes, but absence of such findings does not rule out herpes encephalitis.

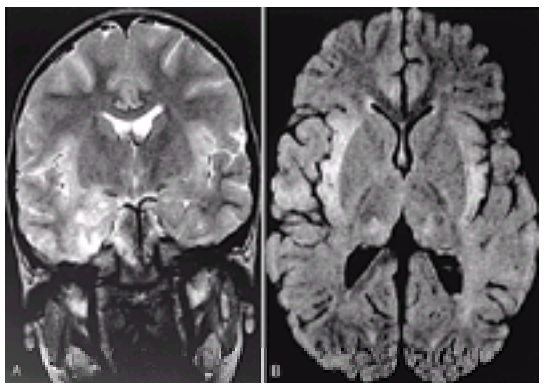

FIGURE 83.2. Coronal **(A)** and axial **(B)** T_2-weighted magnetic resonance images showing multifocal areas of abnormal signal in the medial aspects of both temporal lobes *(large arrow)* and left posterior parietal lobe *(small arrow)* in a patient with herpes simplex encephalitis.

Management

Presently, the treatment of nonherpes viral encephalitis is primarily supportive. Children with very mild manifestations may be followed at home, but those with more significant illness should be hospitalized for observation and monitoring of neurologic status, treatment of increased ICP if present, and fluid restriction and monitoring of urine output and serum sodium because of the risk for inappropriate antidiuretic hormone (ADH) secretion.

Herpes simplex encephalitis causes death or neurologic sequelae in more than 70% of patients. Treatment with acyclovir (30 to 60 mg/kg per day divided three times daily for 14 to 21 days) has resulted in a decrease in mortality and some improvement in morbidity. It must be used early in the disease to achieve maximum benefit. Thus, acyclovir should be considered in all patients suspected of having herpes encephalitis on the basis of clinical or epidemiologic grounds (e.g., history of exposure, oral vesicles, focal neurologic or radiographic findings); because clinical features and laboratory tests are not perfectly sensitive, initial presumptive treatment may be indicated even in the absence of corroborating evidence.

Hemorrhagic Shock and Encephalopathy Syndrome

Hemorrhagic shock and encephalopathy syndrome (HSES), first described in 1983, is a syndrome of catastrophic illness that affects infants less than 1 year of age. It is characterized by the sudden onset of coma, seizures, and shock in a previously healthy infant. Despite the fact that many reported infants with HSES had recent or intercurrent viral illness, no causal link has been established, and the cause and underlying pathophysiology of HSES are unknown. The clinical features are similar to those seen in severe bacterial sepsis, although cultures are by definition negative in children with HSES. The syndrome also resembles heat stroke, leading to speculation that an abnormality of thermal regulation may be responsible.

No standardized diagnostic criteria for HSES exist. Besides the encephalopathy, seizures, and shock, other consistent features of HSES include high fever at the onset, disseminated intravascular coagulation and hemorrhage, and metabolic acidosis. Affected children have a number of characteristic laboratory abnormalities consistent with multiple organ system dysfunction, including elevation of blood urea nitrogen (BUN), creatinine, hepatic transaminases, and creatine phosphokinase (CPK), anemia, thrombocytopenia, and coagulopathy. The laboratory abnormalities peak in the first 48 hours. Brain CT reveals only cerebral edema in the first few days, followed by development of encephalomalacia in severely ill individuals.

Treatment of infants with HSES consists of intensive supportive care. The prognosis is poor, with 50 to 80% mortality and severe neurologic morbidity in most survivors. With increased recognition of the syndrome, however, there have been reports of less severely ill patients with improved outcome.

DISORDERS THAT PRESENT WITH HEADACHE

Headaches of varying character, severity, and origin affect patients of all ages. Much of the CNS, including the brain parenchyma, is devoid of pain sensors. However, headache may result from compression, inflammation, or distortion of a number of pain-sensitive cranial structures, including the proximal portions of the large cerebral arteries, the arteries of the dura and scalp, the intracranial venous sinuses, the dura, the facial sinuses, orbits, teeth, scalp, muscles, and cervical roots of the spinal cord. A full discussion of the differential diagnosis of headache is given in Chapter 56.

Migraine

Background

Migraine—recurrent headaches separated by long, symptom-free intervals—is probably the most common specific cause of episodic headaches in children. In epidemiologic studies, prevalence estimates for migraine in children range from 3 to 10%. A number of forms of migraine are recognized. Migraine is considered “classic” when the headache is well localized and preceded by an aura and considered “common” when it is not. The common form of migraine predominates in

children. Basilar migraine is a migraine variant that involves the posterior cerebral circulation in which brainstem symptoms, possibly including transient loss of consciousness, predominate. Cluster headaches, which are unilateral, occur in runs and are associated with autonomic changes. They represent a rare migraine variant in childhood. Cyclic vomiting, a syndrome of recurrent, discrete attacks of abdominal pain, nausea, vomiting, and pallor, is also believed to be a migraine variant, sometimes called abdominal migraine.

Pathophysiology

The pathogenesis of migraine is not fully delineated, but the headache is thought to be secondary to paroxysmal vascular instability that results in intracranial vasoconstriction followed by vasodilation. These vascular changes often occur sequentially, resulting in premonitory motor, visual, or sensory symptoms (the vasoconstrictive phase), and then headache (the vasodilation phase). The biochemical basis of this instability may be caused by depression of serotonergic brainstem neurons, although many neuronal transmitter abnormalities have been described.

Clinical Manifestations

Prolonged (up to 24 to 48 hours), moderate to severe headache is characteristic of migraine. The headaches may be pulsating and unilateral but assume this pattern less often in children than in adults. Migraine is commonly associated with nausea, vomiting, abdominal pain, and photophobia or phonophobia. Auras occur in less than half of children who experience migraines. During the headaches, analgesics are relatively ineffective, and children seek a quiet, dimly lit area to rest or sleep. Occasionally, the attacks awaken the children from sleep. The physical examination usually shows no focal neurologic deficits, although hemiplegia and ophthalmoplegia may occur in complicated migraine. Unless these episodes have occurred previously, their presence warrants further neurologic evaluation, usually in the form of CT or MRI scanning.

A family history of migraine is helpful in diagnosis, and a disproportionate number of children who experience migraines have episodes of motion sickness, dizziness, vertigo, or frank paroxysmal events. Common trigger factors for migraine in children include emotional stress, lighting changes, and minor head trauma. Particularly in adolescents, it is useful to screen for depression or other psychosocial stressors that may warrant separate treatment. Foods, such as lunch meats, which contain nitrates, and cheeses, which contain tyramine, are less common but important triggers.

The diagnosis of migraine is based almost exclusively on the history and is supported by the absence of abnormalities on examination. There are no diagnostic laboratory tests or imaging studies. Given an accurate history, differentiation from tension headaches, sinusitis, and headaches secondary to intracranial lesions usually is possible; studies such as EEG, CT, and MRI are rarely indicated. Of children who experience migraines, 20 to 90% have been reported to have nonspecific EEG abnormalities, but the EEG usually is not helpful in diagnosis.

Management

A number of agents are available for the treatment of acute migraine (Table 83.5). For many children, mild oral analgesics such as acetaminophen or ibuprofen combined with bed rest may provide sufficient relief and should be considered the first-line agents of choice. Ketorolac (Toradol), a nonsteroidal anti-inflammatory agent for parenteral use, may be used when nausea or vomiting limits oral intake. A short course of a narcotic analgesic such as codeine may occasionally be needed if nonnarcotic agents have failed, especially if the headache prevents sleep.

Drug	Usual Dose
Analgesics	
Acetaminophen	10–15 mg/kg/dose PO or PR q4hr
Ibuprofen	5–10 mg/kg/dose PO q6h
Ketorolac (Toradol)	30 mg initial dose, then 15–30 mg/dose (0.5 mg/kg) IV or IM, or 10 mg/dose PO, q6–6hr
Codeine	0.5–1 mg/kg/dose PO q4–6hr
Antiemetics	
Metoclopramide (Reglan)	0.5–2 mg/kg/dose PO or IV q6–6hr
Prochlorperazine (Compazine)	0.1 mg/kg/dose PO or IM q6hr
Promethazine (Phenergan)	0.25–1.0 mg/kg/dose PO, PR, IV, or IM q4–6hr
Specific Antimigraine Agents	
Dihydroergotamine	0.5–1.0 mg/dose IV or IM, may repeat after 1 hr
Sumatriptan (Imitrex)	6 mg SC or 100 mg PO

(PO, orally; PR, per rectum; IV, intravenously; IM, intramuscularly; SC, subcutaneously.

Table 83.5. Agents for Acute Treatment of Migraine

When nausea and vomiting are severe, antiemetic medications such as metoclopramide (Reglan), prochlorperazine (Compazine), and promethazine (Phenergan) are useful. In addition to their antiemetic effect, these agents often provide some relief of the headache as well and may permit the use of other oral medications. All of these agents have the potential to produce dystonic reactions.

Ergot preparations act primarily as cerebral vasoconstrictors and are specifically indicated for aborting acute migraine attacks. Ergotamine tartrate is administered orally or sublingually, but it must be used early in the headache to be effective, preferably at the outset of the prodrome. Because most young children cannot identify an aura, their use is limited before adolescence. Common side effects of ergot preparations include nausea, vomiting, cramps, and distal paresthesias, all of which may intensify the symptoms of migraine. Chronic ergotism and dependence may occur with repeated use. Dihydroergotamine (DHE) is an injectable ergot derivative with fewer side effects. Ergotamine preparations with additional drugs such as phenobarbital and caffeine are available; however, there is little evidence to show that they

are more efficacious than ergots alone.

For acute migraine, DHE can be given to older children and adolescents in an initial dose of 0.5 mg IM or IV (no milligram-per-kilogram dose has been established). The initial dose of DHE may be repeated in 1 hour if necessary. One study in adults reported that 3 mg administered intranasally is also effective. Antiemetics may be useful to control the nausea and vomiting that often occur after DHE administration.

Sumatriptan succinate (Imitrex) is a serotonergic agent available for either oral or subcutaneous administration. Its effectiveness in relieving symptoms of acute migraine has been demonstrated in adults, but similar evidence is lacking in children. The dose for children 12 years and older is 6 mg subcutaneously or 100 mg orally. Sumatriptan is generally well tolerated; side effects include irritation at the injection site, flushing, tachycardia, disorientation, and chest tightness that lasts for several minutes after parenteral administration. Sumatriptan should not be used concomitantly with ergotamines. A reasonable approach is to use sumatriptan after a trial of analgesics in an older child, although older children or adolescents with recurrent migraine and a history of successful treatment with sumatriptan in the past may benefit from earlier use of this agent.

If migraines are frequent and severe, prophylactic treatment is possible. Many drugs have been used, but because they require close, serial examination and have no effect on the acute attack, they should not be started in the ED. Among the medications used for chronic suppressive therapy are propranolol, tricyclic antidepressants, cyproheptadine (Periactin), valproic acid, and calcium channel blockers.

Idiopathic Intracranial Hypertension (Pseudotumor Cerebri)

Background and Pathophysiology

Idiopathic intracranial hypertension (IIH), also called pseudotumor cerebri, is a poorly understood condition of increased ICP. It may occur at any age during childhood but is more common in adolescents, especially in obese individuals. Females are more commonly affected. A number of other conditions have been reported in association with IIH; these include infections (otitis media, mastoiditis), endocrinologic conditions (hyperthyroidism, Addison's disease), medications (steroid withdrawal, tetracycline, hypervitaminosis A), and mild head trauma. However, a causal relationship remains unproved, and in most cases of IIH, no cause is identified. The mechanism of increased ICP in IIH remains unknown, although several hypotheses have been postulated, including vasogenic brain edema and impaired reabsorption of CSF by the arachnoid villi.

Clinical Manifestations

Headache, of variable severity and duration, is the most common presenting symptom. It is typically worse in the morning. Nausea, vomiting, dizziness, and double or blurred vision also occur. If the process is long-standing, decreased visual acuity or visual field deficits can result. Infants often have nonspecific symptoms of lethargy or irritability. Papilledema is seen in virtually all cases. Other neurologic symptoms and signs are often absent; however, cranial nerve palsies, particularly affecting the sixth cranial nerve, may be seen.

Diagnosis should be considered when a child with a prolonged history of headache is found to have evidence of papilledema without other neurologic findings. Pseudotumor cerebri is a diagnosis of exclusion, and mass lesions and infectious processes must be ruled out. Because posterior fossa tumors and obstructive or nonobstructive hydrocephalus may mimic pseudotumor early in the course of disease, CT or MRI should be obtained in all children with this constellation of findings. In cases of IIH, the ventricles will appear normal or small. If no mass lesion is present, an LP should be performed with a manometer to measure opening pressure. The patient must be recumbent with legs extended to ensure an accurate reading of the opening pressure. Children with pseudotumor have elevated opening pressure (greater than 200 mm H_2O), but normal CSF cell count, protein, and glucose. In children with intermittent symptoms, the opening pressure may be normal when the headache is waning, even though papilledema may persist for several weeks.

Management

Removal of sufficient CSF to normalize ICP usually leads to relief of symptoms. Treatment may then be started with acetazolamide (Diamox) to decrease CSF production (60 mg/kg per day divided four times daily). Although recommended by some authorities, corticosteroids have not been proven to be effective in the management of this condition. However, in cases of IIH following withdrawal of steroid therapy, a course of prednisone or dexamethasone may be beneficial. Patients with mild symptoms and good response to LP may be discharged to home with close follow-up arranged. Children with severe or persistent symptoms or those with visual changes may require hospital admission. Intracranial hypertension may be recurrent or chronic, and long-term monitoring, particularly of visual function, is important.

DISORDERS OF MOTOR FUNCTION (SEE CHAPTER 79)

Every level of the neural axis is involved in the performance of motor tasks. Anatomic localization usually is possible after evaluation of the distribution and character of the deficit (Table 83.6). Paresis refers to partial or complete weakness of a part of the body. Various clinical designations are used to describe some patterns of weakness: paraplegia (or paraparesis), affecting the lower half of the body; quadriplegia, affecting all limbs; and hemiplegia, referring to weakness of one side of the body. Paraplegia most often results from spinal cord involvement, whereas hemiparesis is most often a sign of cortical disease. Some of the common conditions affecting various levels of the neuromotor system that may present with acute motor dysfunction are discussed next.

	Tone	Distribution	Reflexes	Babinski	Other
Upper motor neuron	Increased (may be decreased acutely)	Pattern (e.g., hemiparesis, paraparesis) Distal > proximal	Increased (may be decreased acutely)	Extensor	Cognitive dysfunction possible
Anterior horn cell	Decreased	Variable, asymmetric	Decreased to absent	Flexor	Fasciculations; no sensory involvement
Peripheral nerve	Decreased	Nerve distribution	Decreased to absent	Flexor	Sensory involvement
Neuromuscular junction	Normal	Fluctuating	Usually normal	Flexor	–
Muscle	Decreased	Proximal > distal	Decreased	Flexor	Tenderness; signs of inflammation possible

Table 83.6. Localizing Level of Neuromotor Dysfunction

Stroke/Cerebrovascular Accident

Background

Stroke is defined as a syndrome of acute onset of focal neurologic deficit that persists for more than 24 hours. Stroke is relatively rare in children but may complicate a number of other pediatric medical conditions. For example, among children with sickle cell disease, the incidence of stroke has been reported to be 6 to 9%. Others at risk are those with various forms of cardiac disease, which is one of the most common causes of stroke in children. Table 83.7 lists some of the common causes of stroke, as well as some more uncommon conditions in which stroke is a prominent clinical feature.

Vascular	Drugs
Arteriovenous malformation	Cocaine
Aneurysm	Amphetamines
Moyamoya disease	Oral contraceptives
Fibromuscular dysplasia	Ergot poisoning
Cardiac	Metabolic
Valvular heart disease (including endocarditis)	Homocystinuria
Right-to-left shunts	Fabry's disease
Atrial tumors	Mitochondrial encephalopathies (MELAS syndrome)
Arrhythmia	Organic acidemias
Cardiomyopathy	Hyperlipidemias
Infectious	Hematologic
Meningitis (especially tuberculous)	Sickle cell disease
Mycotic aneurysm	Coagulopathies (e.g., hemophilia)
Mastoiditis, otitis media, or sinusitis leading to venous sinus thrombosis	Anticoagulant deficiency (protein C, protein S, antithrombin III)
Trauma	Polycythemia
Intracranial hemorrhage	Acute myelogenous leukemia
Cervical trauma with vertebral artery injury	Systemic lupus erythematosus
Intracranial trauma with carotid injury	Neurocutaneous Syndromes
Brain tumor	Neurofibromatosis
	Tuberous sclerosis
	Sturge-Weber syndrome

Table 83.7. Causes of Stroke in Children

Pathophysiology

Generally, stroke is classified as either primarily ischemic (including embolic phenomena) or hemorrhagic. In ischemic stroke, there is focal reduction in cerebral blood flow, with hypoxic damage to brain parenchymaleading to neuronal injury and death. Further damage ensues from reperfusion of ischemic areas. More recently, attention has focused on secondary factors that play an important part in determining the extent of damage after acute ischemia; these include excess accumulation of excitatory amino acids and generation of free radicals.

Clinical Manifestations

The presentation of stroke in children is highly variable, influenced by the portion of the cerebral vasculature affected as well as the child's age. Hemiparesis is most often observed, with facial weakness typically ipsilateral to weakness in the rest of the body. Involvement of the anterior cerebral artery leads primarily to lower extremity weakness, whereas compromise of the middle cerebral artery circulation produces hemiplegia with upper limb predominance, hemianopsia, and possibly dysphasia. Less commonly, the posterior circulation is affected, which results in vertigo, ataxia, and nystagmus, as well as hemiparesis and hemianopsia. Older children often have concomitant headache, whereas children less than 4 years of age are more likely to have associated seizures. The child with a stroke may also have a diminished level of consciousness.

Investigations in a child with acute hemiparesis should be directed at confirming the diagnosis of stroke and attempting to identify an underlying cause if none is known. Imaging studies are useful. Cranial CT without contrast is the study of choice for identifying acute hemorrhage. However, CT may be normal in the first 12 to 24 hours after an ischemic stroke; MRI, in contrast, may show changes as early as 6 hours after infarction. A usual approach is to obtain a noncontrast CT, followed by MRI if no hemorrhage is seen.

In a child without a known predisposing condition, ancillary tests may be revealing of the cause of the stroke. Studies worth considering in such patients, depending on the clinical picture, are listed in Table 83.8. In one large series of 129 children with ischemic stroke, no cause was found in 35%.

Brain Imaging	Erythrocyte sedimentation rate
Computed tomography (noncontrast)	Hemoglobin electrophoresis
Magnetic resonance imaging	Protein C and S quantification
Angiography (standard or magnetic resonance)	Antithrombin III level
Cardiac	*Chemistry*
Electrocardiogram	Blood urea nitrogen
Echocardiogram	Cholesterol and triglycerides
Hemetologic	Hepatic transaminases
Complete blood count	Serum amino acids
Prothrombin and partial thromboplastin times	Urine organic acids
Fibrinogen	Toxicology screen
	Lactate
	Lumbar Puncture

Table 83.8. Studies to Consider in the Evaluation of the Child with Acute Stroke

Management

Initial treatment after an acute stroke is focused on stabilization and supportive care, including control of any seizures. Several aspects require special attention. Although evident hypotension should be treated with volume expansion, administration of free water should be restricted because of the potential for edema formation. Hypertension, if present, must be treated cautiously, and the blood pressure lowered gradually. Both hypoglycemia and hyperglycemia can exacerbate ischemic stroke, so careful monitoring of serum glucose is important. Fever, which can occur in children with stroke, may also contribute to ischemic damage and should be controlled with antipyretics.

Further therapy is determined by the type of stoke. With hemorrhagic stroke, neurosurgical intervention may be required to evacuate a hematoma or excise a bleeding arteriovenous malformation (AVM). Catheter-directed embolization may also be possible in cases of AVM. Children with sickle cell disease and stroke should have acute transfusion to decrease the level of hemoglobin S to less than 30%. Thrombolytic and anticoagulant therapies have been shown to be effective in adults with ischemic stroke but remain untested in children. Similarly, novel therapies such as calcium channel blockers and free radical scavengers have not been studied in pediatric patients; their use remains experimental.

Overall, prognosis for children with stroke is better than that in adults. However, regardless of treatment, long-term morbidity of stroke in children is high, with more than 75% of affected children experiencing sequelae such as hemiparesis, seizures, and learning difficulties.

Spinal Cord Dysfunction

Background

Dysfunction of the spinal cord may result from any of a variety of disorders, either intrinsic or extrinsic to the spinal cord, with a great deal of overlap in their clinical presentation. Transverse myelitis is an intramedullary disorder, involving both halves of the cord over a variable length, with involvement of motor and sensory tracts. It occurs in children and adults, although it is rare in the first year of life. Transverse myelitis is believed to be a localized form of acute disseminated encephalomyelitis, discussed previously. Like the latter disorder, transverse myelitis may occur after a number of infections; among those commonly reported are Epstein-Barr virus, cytomegalovirus, measles, mumps, *Campylobacter jejuni,* and *M. pneumoniae.* Transverse myelitis may also result from systemic autoimmune disorders such as lupus erythematosus or scleroderma. In some older children and adolescents, transverse myelitis is a first manifestation of multiple sclerosis.

Acute spinal cord compression in children usually is caused by trauma, infection, or cancer. Spinal trauma may lead to contusion or concussion of the cord with hemorrhage, edema, and local mass effect, or to development of a spinal epidural hematoma. Parenchymal injury usually presents acutely, but an epidural hematoma may develop over several days after the antecedent trauma. Epidural abscess is the most common infectious cause of spinal cord compression. It is usually caused by hematogenous spread of bacteria, with *Staphylococcus aureus* being the most common pathogen. Neoplastic causes include both primary intraspinal tumors (ependymoma and astrocytoma) and extrinsic lesions such as neuroblastoma or lymphoma.

Pathophysiology

Transverse myelitis is believed to be caused by an autoimmune process, with demyelinating lesions found in the spinothalamic and pyramidal tracts and posterior columns of the spinal cord. During the course of the illness, the initial area or areas of spinal cord inflammation may extend rostrally and caudally to involve an extensive portion of the spinal cord.

Mass lesions may cause damage by direct compression of spinal cord tissues or, secondarily, by interference with the tenuous arterial (or, less commonly, venous) blood flow to the spine with resultant spinal infarction.

Clinical Manifestations

Spinal cord dysfunction from any cause is characterized by paraplegia below the level of involvement, hyporeflexia, and sensory symptoms such as bandlike pain at the level of compression, and sensory loss or paraesthesias below the area of damage. If the lower spinal cord is involved (the conus), there usually is early loss of bowel and bladder control. Compression of the cauda equina usually results in asymmetric symptoms, radicular pain, and focal lower extremity

motor and sensory abnormalities.

Transverse myelitis may affect any level of the spinal cord, but thoracic involvement is most common. Initial symptoms include lower extremity paresthesia, local back pain, unilateral or bilateral lower extremity weakness, and urinary retention. A preceding respiratory or gastrointestinal illness is usually reported, and at the time of diagnosis, fever and meningismus are sometimes seen in children. Characteristically, the insidious onset of paresthesia or weakness of the lower extremities progresses over days or, rarely, weeks, and then is replaced by the abrupt occurrence of static paraplegia or quadriplegia and, in the cooperative child, a detectable sensory level. In other children, the course of progression may be less than 12 hours. The sensory loss generally involves all modalities, although a spinothalamic deficit (pain) may occur without posterior column dysfunction (vibration). The weakness usually is symmetric but may be asymmetric. After a variable interval, initial flaccidity may be replaced by spasticity. Sphincter disturbance of the bowel and bladder occurs in most patients, bladder distension being the most common initial sign of damage.

Traumatic and infectious spinal lesions are usually accompanied by relatively acute onset of local back pain, which is exacerbated by direct percussion of the area. Pain may precede other symptoms for days. With tumors, however, there may be weakness in the absence of pain. Patients with epidural abscess often have systemic signs of infections such as fever, headache, vomiting, and perhaps neck stiffness. Bony tenderness in such a patient may indicate vertebral osteomyelitis or discitis, which can also present with weakness, although usually less severe than is seen with actual spinal cord involvement.

Prompt diagnosis of spinal cord lesions requires a high level of expectation. Detailed neurologic examination is essential, with particular attention to quality of deep tendon reflexes, any asymmetry of reflexes or strength, evaluation for a sensory level, and assessment of anal tone and cremasteric reflexes (in males). Note also any point percussion tenderness.

Diagnosis is confirmed by emergency neuroimaging, with precautions to immobilize the patient as much as possible. Plain spine films are useful initially in trauma. MRI of the spine is the procedure of choice to detect compressive mass lesions, but if not immediately available, plain or CT myelography is an alternative. In transverse myelitis, the cord may be widened at the level of involvement. This is easier to detect with MRI, which in some cases may also reveal evidence of focal intramedullary demyelination.

LP alone should not be performed if a diagnosis of spinal cord compression is entertained. If no mass lesion is noted and transverse myelitis is a diagnostic possibility, LP may be useful, showing a normal or slightly elevated opening pressure and a mild pleocytosis in the CSF in nearly 50% of patients at the time of presentation. The CSF protein is often elevated and may demonstrate oligoclonal bands or increased myelin basic protein, but the glucose usually is normal.

Management

Treatment of children with spinal injury from trauma begins with splinting and immobilization of the spine. If trauma is likely, high-dose methylprednisolone, if given within 8 hours of injury at an IV dose of 30 mg/kg followed by infusion at 5.4 mg/kg per hour for 23 hours, improves the quality of neurologic outcome. Neurosurgical consultation should be obtained as soon as possible; however, early surgical attempts to decompress the swollen spine (laminectomy or midline myelotomy) have proven ineffective and, possibly, detrimental.

In cases of possible epidural abscess or tumor-related mass, IV dexamethasone at a loading dose of 2 mg/kg (up to a maximum of 100 mg) should be given, followed by 1 to 2 mg/kg per day IV in four divided doses over the next 24 hours. In patients with a presumed infectious cause and those with cancer of unknown origin, surgical decompression is indicated on an emergent basis to alleviate pressure and pinpoint diagnosis. Further treatment depends on the organism or exact tumor type found.

Treatment of transverse myelitis is supportive, and some degree of recovery occurs in approximately 80% of cases. All children with this syndrome should be hospitalized. Although systemic corticosteroids are often recommended for treatment of transverse myelitis, there is little evidence of their efficacy. Treatment with intraspinal steroids or emergency laminectomy has also not been shown to improve outcome. High-dose dexamethasone as described previously may be begun until cord compression by a mass lesion is ruled out.

Acute Polyneuritis

Background and Pathophysiology

Acute polyneuritis, also called Guillain-Barré syndrome, is characterized by symmetric ascending paralysis. Pathologically, the hallmark of this disease is primary demyelination of motor and sensory nerves, believed to be secondary to autoimmune mechanisms. It occurs in children in all age groups but is uncommon before 3 years of age. An antecedent respiratory or gastrointestinal infection or immunization precedes the onset of illness by 1 to 2 weeks in more than 75% of childhood cases.

Clinical Manifestations

Weakness, commonly with an insidious inset, is the usual presenting complaint. Paresthesias or other sensory abnormalities such as pain or numbness are prominent in up to 50% of cases, particularly in older children. The paresthesias and paralysis usually are symmetric and ascending, although variations may occur. Early in the course of illness, distal weakness is more prominent than proximal weakness. Deep tendon reflexes are depressed or absent at the time of diagnosis. Affected children often have an ataxic gait.

Cranial nerve abnormalities occur during the illness in 30 to 40% of cases and may be the predominant finding,

especially in the Miller-Fisher variant of this syndrome, which is characterized by oculomotor palsies, ataxia, and areflexia without motor weakness of the extremities. The most common cranial nerve deficit is seventh (facial) nerve palsy, followed in decreasing frequency by impairment of cranial nerves IX, X, and XI and oculomotor abnormalities. Autonomic dysfunction occurs commonly and results in blood pressure lability, postural hypotension, and cardiac abnormalities; it is a disproportionate cause of morbidity and mortality. Urinary retention, if it occurs, is usually seen late in the illness. As the paralysis ascends, muscles of breathing may become involved, leading to respiratory embarrassment.

The primary aid in diagnosis is LP, which demonstrates an elevated protein, normal glucose, and fewer than 10 white blood cells/mm^3—the so-called albuminocytologic disassociation. The protein elevation occurs in almost all cases but may be delayed for weeks, usually peaking in the second or third week of illness. Electrophysiologic evidence for Guillain-Barré syndrome is the presence of nerve conduction velocity delay, which usually is not demonstrable until the second or third week of illness. Emergency electromyography (EMG) and nerve conduction velocity testing are not indicated.

Management

Because of the potential for progression to life-threatening respiratory compromise, the child with Guillain-Barré syndrome should be hospitalized and observed closely. Impending respiratory distress must be anticipated, and routine respiratory monitoring should be aided by specific measures of respiratory function, particularly measurement of negative inspiratory force. Because autonomic dysfunction is common, blood pressure must be monitored closely and abnormalities treated vigorously.

Acute polyneuritis is generally self-limiting, with more than 90% of children in most series having complete or near complete recovery. In mild cases, in which children retain the ability to ambulate, only supportive care is required. However, immunomodulatory therapy may be of benefit in more severely affected children. Plasmapheresis and intravenous immunoglobulin have both been used. Although well-controlled, blinded studies of these treatments in children are lacking, the available data suggest both are effective in reducing the duration and severity of illness in those most severely affected, especially when begun early in the course of the disease. Corticosteroids have not been shown to be beneficial in acute Guillain-Barré syndrome.

Myasthenia Gravis

Background and Pathophysiology

In myasthenia gravis, antibodies directed against acetylcholine receptor protein of the postsynaptic neuromuscular junction cause intermittent failure of neuromuscular transmission and fluctuating weakness. Myasthenia manifests by fluctuating weakness of cranial and skeletal musculature, exacerbated by exertion. More commonly a disease seen in adults, myasthenia gravis occurs in children in three major forms: transient neonatal, infantile, and juvenile (most common).

Clinical Manifestations

The juvenile form of myasthenia clinically mimics the adult disease. The mean age of onset is 8 years, with a female predominance of approximately 4:1. The onset of symptoms may be insidious or acute. Most cases affect the cranial nerves, and any cranial nerve can be involved in combination or isolation. Bilateral ptosis is the most common cranial nerve deficit, followed in incidence by oculomotor impairment. Generalized truncal and limb weakness is present at onset in up to half of cases and eventually develops in most children with myasthenia. The diagnosis should be suspected if there is a history of worsening weakness during continual activity or if fatigability of muscle strength is demonstrable. Illnesses confused with myasthenia include the muscular dystrophies, congenital myopathies, inflammatory myopathies, acute and chronic polyneuropathies, and in the infant, botulism.

The Tensilon test is the backbone of diagnosis. In this procedure, the anticholinesterase drug edrophonium (Tensilon), which has a 30-second onset and approximately a 5-minute duration of action, is given slowly by IV at a dosage of 0.2 mg/kg, up to a maximum dose of 10 mg. Atropine should be immediately available to treat potential severe cholinergic reactions (e.g., bradycardia). Initially, one-tenth of the total dose is given, and if no hypersensitivity or severe reactions are noted, the remainder of the dose is administered. Because edrophonium is short-lived, interpretation of the response requires close monitoring of a muscle or muscle group in which improvement can be seen clearly, such as the eyelid elevators. In small children, this often is impossible and longer-acting anticholinesterases such as neostigmine (0.125 mg in an infant and 0.04 mg/kg in an older child) can be used. EMG provides electrophysiologic evidence for myasthenia gravis, with a decremental response to repetitive nerve stimulation, but may be negative when the disease is confined to the cranial nerves.

Management

Although myasthenia gravis is potentially life-threatening, specific management usually can be delayed until after diagnosis is made. If there is evidence of respiratory compromise, ventilatory support is mandatory. Treatment is begun with the use of cholinesterase inhibitors to prolong the availability of acetylcholine at the neuromuscular junction. Presently, the anticholinesterase of choice is pyridostigmine (Mestinon) at a starting dosage of 1 mg/kg by mouth every 4 hours, adjusted according to the clinical response. Other agents, such as corticosteroids or antimetabolites, may be beneficial in selected cases. If there is any concern about respiratory compromise or if severe weakness is present, the child should be hospitalized immediately.

Myasthenia has a fluctuating, unpredictable course that can be exacerbated by intercurrent illness and by certain drugs, particularly the aminoglycoside antibiotics. In a known myasthenic, rapid worsening and respiratory compromise

(myasthenic crises) may be difficult to differentiate from deterioration secondary to overdose of anticholinesterases (cholinergic crises) because the muscarinic side effects of the anticholinesterases, such as nausea, vomiting, cramps, and muscle fasciculations, may be absent. At times, differentiation can be made by giving 1 to 2 mg of IV edrophonium after ensuring respiratory sufficiency. This should result in rapid improvement in the patient with a myasthenic crisis. This procedure may be falsely positive, however, and if the diagnosis is unclear, the patient should be withdrawn from all anticholinesterases and, if necessary, maintained on mechanical respiration for 48 to 72 hours. Cholinergic crises require the immediate withdrawal of all anticholinesterases. Myasthenic crises respond variably to additional anticholinesterases, and plasmapheresis or steroid therapy may be particularly useful in this situation. Both myasthenic and cholinergic crises mandate admission to the hospital.

Botulism

Background and Pathophysiology

Infantile botulism is a cause of acute weakness in previously well infants less than 6 months old. The illness is secondary to intestinal colonization by *Clostridium botulinum,* which produces a neurotoxin that impairs acetylcholine release from the nerve terminal. Spores of *C. botulinum* are of ubiquitous origin, found in soil and agricultural products. Honey has been found to be a particularly significant reservoir. Although infant botulism occurs throughout the United States, the incidence is highest in certain areas; approximately half the cases have been reported from California, Utah, and Pennsylvania. The various host factors that predispose certain infants to intestinal colonization are poorly understood.

Clinical Manifestations

The initial symptom of botulism usually is constipation, followed insidiously by lethargy and feeding difficulties. Physical findings at the time of presentation are hypoactive deep tendon reflexes, decreased suck and gag, poorly reactive pupils, bilateral ptosis, oculomotor palsies, and facial weakness. Differential diagnosis includes all the potential causes of lethargy and poor feeding in infancy, and infants are often misdiagnosed initially. Laboratory studies, including the leukocyte count and LP, are normal. The diagnosis is confirmed by identification of *C. botulinum* toxin (usually type A or B) in the feces or isolation of the organism in stool culture, which is less sensitive. EMG may supply immediate information. Characteristic EMG findings are brief, small-amplitude action potentials; posttetanic facilitation; and normal nerve conduction velocity.

Management

Management of infant botulism is strictly supportive. Affected infants require hospitalization to observe for respiratory compromise. In one large series of 57 patients, 77% required endotracheal intubation because of loss of protective airway reflexes, and 68% received mechanical ventilation for some period. Nasogastric or nasojejunal feedings are usually needed as well. The use of cathartics or other laxatives to reduce the amount of *C. botulinum* present in the intestine has not proved beneficial. Botulinum antitoxin has resulted in anaphylactic reaction in infants and is not recommended. Antibiotics such as penicillin, although widely used, have not been shown to eradicate the organism from the bowel or result in clinical improvement.

Periodic Paralysis

Background and Pathophysiology

Familial periodic paralysis is a rare illness, inherited in an autosomal-dominant fashion that results in episodes of severe weakness associated with an abnormality of circulating potassium during attacks. Two major forms of illness—hyperkalemic and hypokalemic—are recognized. (A third type, normokalemic, has been described but most likely represents a rare variant of the hyperkalemic variety.) Other disorders that can produce weakness and electrolyte abnormalities, such as use of corticosteroids or diuretics, thyrotoxicosis, hyperaldosteronism, and renal insufficiency, may mimic the periodic paralyses. The serum potassium abnormalities in familial periodic paralysis are thought to be epiphenomena of yet undelineated muscle membrane abnormalities.

Clinical Manifestations

Many of the clinical features are common to the various forms of periodic paralysis. Characteristically, a previously well patient develops a flaccid weakness in his or her trunk and upper thighs, and the weakness gradually involves the remainder of the skeletal muscles. Deep tendon reflexes are diminished. The attacks last from hours to days, and between the attacks, muscular strength usually is normal, although a minority of patients have residual muscular weakness.

Hypokalemic periodic paralysis, the most common type, occurs primarily in young adults. Trigger factors include vigorous exercise, heavy carbohydrate meals, alcohol, and the cold. During an attack, potassium levels are usually 2 to 2.5 mEq/L, and electrophysiologic examination demonstrates unstimulatable muscles. The hyperkalemic form usually begins in the first decade of life, and attacks occur predominantly during the period of rest after vigorous exercise or after fasting. The episodes are more common than in hypokalemic paralysis, but often last less than a few hours. Myotonia usually is associated with the illness. During the attack, plasma potassium is moderately elevated, although it is often in the upper normal range. In both forms of periodic paralysis, electrocardiogram (ECG) changes consistent with the serum potassium abnormality may be noted, and cardiac arrhythmias may rarely arise.

Management

Emergency treatment of hypokalemic periodic paralysis includes oral, or rarely IV, potassium. Prophylactically, patients should avoid precipitants such as vigorous exercise or large carbohydrate loads. Recurrences may be prevented with

spironolactone or acetazolamide.

Attacks of hyperkalemic periodic paralysis are often brief enough that acute treatment is unnecessary. In severe attacks, IV calcium gluconate may be helpful. Acetazolamide, thiazide diuretics, and albuterol have been used for prevention of recurrences.

DISORDERS OF BALANCE (SEE CHAPTER 11)

Acute Cerebellar Ataxia

Background and Pathophysiology

Acute cerebellar ataxia, characterized by the acute onset of unsteadiness in a previously well child, is the most common cause of ataxia in young children. It is seen primarily between the ages of 1 and 4 years but can occur at any time during childhood. The exact cause of the illness is unclear; however, it is believed to be a parainfectious or postinfectious demyelinating phenomenon and likely represents a localized form of postinfectious encephalitis. Acute cerebellar ataxia occurs most commonly after primary varicella. Other infections implicated include infectious mononucleosis, enteroviruses, herpes simplex, influenza, *Mycoplasma,* and Q fever. Ataxia is usually seen 5 to 10 days after the onset of illness, although symptoms may be delayed for up to 3 weeks, and there are some reports of cerebellar ataxia preceding the rash of chickenpox.

Clinical Manifestations

The child develops acute truncal unsteadiness with a variable degree of distal motor difficulty, such as tremor and dysmetria. Dysarthria and nystagmus are variably present. Some children have nausea and vomiting, presumably caused by vertigo. Headache is rare.

When acute ataxia follows varicella in a child with no other neurologic findings, the diagnosis may be made on clinical grounds. In atypical cases, CT or MRI may be necessary to rule out a cerebellar mass. LP is not usually necessary in typical cases; if performed, it reveals a mild CSF pleocytosis in approximately half of the cases.

Management

Treatment is supportive. Resolution of symptoms is complete in most children within 2 weeks of onset, but mild residual neurologic deficits have been reported in 10 to 30% of cases. Varicella-associated cases appear to have the most benign prognosis.

Benign Paroxysmal Vertigo

Benign paroxysmal vertigo is an illness that affects children primarily between 1 and 4 years or age, although it can occur any time during the first decade. It manifests with acute episodes of dizziness and imbalance, lasting seconds to minutes. Between episodes, the child is asymptomatic. During the spell, the child characteristically becomes frightened and pale but does not lose consciousness. He or she may have associated nausea, vomiting, or visual disturbance. The physical examination is usually normal except for nystagmus, which may be present. Although the cause of this illness is unknown, it is thought to be a migraine variant. Many children go on to develop more typical migraine headaches later, and there is often a family history of migraine disease. As the name suggests, the course of benign paroxysmal vertigo is self-limiting and benign, and treatment is supportive.

MOVEMENT DISORDERS

Involuntary movements are components of many CNS disorders and tend to be complex. A classification into specific subtypes, based on the character, predominant anatomic localization, rhythmicity, and frequency, is arbitrary but useful in deducing the cause of the disorder (Table 83.9). Movements such as chorea, athetosis, dystonia, ballismus, and certain types of tremors suggest dysfunction of the extrapyramidal nervous system. Involuntary movements also are caused by damage to the cerebellum or its outflow tract, especially static (on maintaining fixed position) and intention tremors. Myoclonus may occur secondary to cerebral cortex, brainstem, or spinal cord disease. Tics, another form of involuntary movement, may be extremely difficult to distinguish from chorea and are best differentiated by their stereotypic character. They probably represent the most common involuntary movement disorder but are not true neurologic emergencies. Many illnesses may present with involuntary movements and are diagnosed by associated neurologic findings.

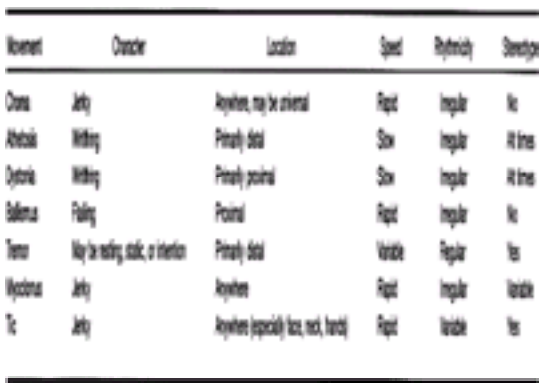

Table 83.9. Categorization of Movement Disorders

Acute Dystonia

Dystonia is marked by involuntary, sustained muscle contractions, typically of the neck and trunk, that cause twisting movements and abnormal postures. In generalized dystonia, the head is usually deviated to the side, and there is grimacing of the face. Acute dystonia in children is nearly always the result of exposure to an antidopaminergic agent such as a neuroleptic, antiemetic, or metoclopramide. Chronic dystonias are rare but may be seen as an isolated disorder or as a manifestation of cerebral palsy. Dystonia must be differentiated from torticollis, an abnormal tilt of the head and neck usually resulting from irritation or spasm of the sternocleidomastoid muscle. Another clinically similar condition is Sandifer's syndrome, which describes intermittent arching of the back and neck observed in infants with gastroesophageal reflux.

Acute dystonia resulting from exposure to antidopaminergic drugs is treated with diphenhydramine (1 mg/kg per dose IV, PO, or IM) or benztropine (Cogentin) (1 to 2 mg/dose IM). Because the half-life of many of the precipitating agents is fairly long, treatment should be continued for 24 to 48 hours.

Sydenham's Chorea

Background

Sydenham's chorea, the most common form of acquired chorea seen in children, occurs primarily between the ages of 3 and 13 years. Marked by involuntary movements, coordination difficulties, and emotional lability, its onset may be abrupt or insidious. Sydenham's chorea is thought to be a poststreptococcal disease and may occur months after the primary bacterial infection. It is one of the major diagnostic criteria for rheumatic fever (see Chapter 82).

Clinical Manifestations

The involuntary movements may be subtle at first and exacerbated by stress. Initially, the movements classically affect the face and distal portion of the upper extremities and consist of rapid, involuntary random jerks. This results in the milkmaid hand, in which the child's hand cannot maintain a uniform strength while grasping the examiner's hand. The involuntary movements disappear during sleep. There usually is associated muscular hypotonia and marked coordination difficulties, and speech is often jerky. Hemichorea, in which the abnormal movements are predominantly unilateral, occurs in some cases. The deep tendon reflexes are normal, although occasionally, the patellar reflex is said to be “hung up.” There is no evidence for upper motor neuron disease.

Serologic evidence for preceding streptococcal infection is absent in up to 25% of cases, and only one-third of patients have associated manifestations of rheumatic fever at the time of diagnosis. In the absence of such confirmatory evidence of a poststreptococcal cause, other disorders that may present with chorea must be considered in the differential diagnosis. These include atypical seizures, drug intoxication, choreoathetoid cerebral palsy, familial choreas, chorea gravidarum, collagen vascular disease, Wilson's disease, and Lyme disease.

Management

Initially, all patients should have a hematologic profile, sedimentation rate, and serologic test for streptococcal infection. An ECG also should be performed to look for evidence of rheumatic carditis (e.g., prolonged P-R interval). If there is a question concerning diagnosis, further tests, such as CT, MRI, LP, and serologic evaluation for collagen vascular disease, might be helpful, but they are not usually necessary on an urgent basis.

The success of any treatment is hard to evaluate because the course is so unpredictable. Haloperidol (0.5 to 1 mg twice daily) has been reported to result in improvement within 2 to 3 days. Because patients with Sydenham's chorea have an increased incidence of rheumatic carditis, prophylactic penicillin should be used, unless another specific cause is determined for the chorea.

DISORDERS OF CRANIAL NERVE FUNCTION

Optic Neuritis

Background

Optic neuritis is an acute inflammation or demyelination of the optic nerve characterized by an impairment of vision, progressing over hours or days and associated with tenderness of the eyeball exacerbated by eye movement. The disease is primarily unilateral, but an increased incidence of bilateral involvement is found in children. Optic neuritis in children is most commonly presumed to be on an autoimmune basis following a viral disease, including the childhood exanthems. At times, a contiguous sinusitis may cause the illness. Of patients with unilateral optic neuritis, 20% will develop multiple sclerosis at a later date, but there is little reason to make this diagnosis before the development of other symptoms of neurologic dysfunction.

Clinical Findings

On examination, decreased visual acuity and decreased color vision are associated with a relative afferent pupillary deficit to light and a central scotoma in the affected eye. The relative afferent pupil defect is demonstrated by the swinging flashlight maneuver, during which the pupil of the affected eye constricts briskly when light is shone into the

contralateral eye (the consensual light reflex) and dilates when light is immediately shone into the affected eye. With bilateral disease, the change in pupillary reflexes may not be apparent. Funduscopic examination discloses a hyperemic, swollen optic disc; in the rare cases of retrobulbar optic neuritis, funduscopic examination is normal.

Optic neuritis must be distinguished from papilledema secondary to increased ICP. Papilledema is almost always bilateral and associated with normal vision and normal pupil reactivity until late in the disease. In cases of bilateral optic neuritis, differentiation may be impossible because funduscopic findings are identical in the two illnesses. If any doubt of increased ICP persists, the patient should undergo evaluation by CT or MRI of the brain and, if normal, CSF analysis. In optic neuritis, the opening pressure is normal, but there may be a mild lymphocytic pleocytosis or elevated CSF protein.

Management

The course of the illness is variable, with most patients recovering to normal or near normal vision over 4 to 5 weeks. Treatment with high-dose systemic corticosteroids, such as prednisone 2 mg/kg per day orally for 7 to 10 days has not been shown to improve the ultimate prognosis but may result in a slightly faster resolution of symptoms.

Facial Nerve Palsy

Background

Weakness in the distribution of the seventh cranial (facial) nerve may be produced by either central (upper motor neuron) or peripheral (lower motor neuron) dysfunction. Peripheral disease is most common in children, particularly when the facial weakness is an isolated finding. Bell's palsy refers to peripheral facial nerve weakness with no identifiable underlying cause. It is believed to be secondary to edema of the facial nerve as it passes through the facial canal within the temporal bone. There is often a history or preceding upper respiratory infection, and in at least a subset of patients, there is evidence of reactivation of infection with Epstein-Barr virus or HSV. Facial palsy may also a manifestation of early disseminated Lyme disease. It usually occurs in isolation, although there may be other signs of CNS disease. Although in general most cases of facial nerve palsy in children are of the idiopathic (or viral reactivation) type, in endemic areas, Lyme disease may be the most common cause.

Clinical Manifestations

Facial weakness may be partial or complete. On the affected side, there is flattening of the nasolabial fold at rest, and the child has difficulty closing the eye or raising the corner of the mouth to smile. In many cases, pain localized to the ear precedes the paralysis. With upper motor neuron involvement, there will be some residual capacity to furrow the brow because of crossed innervation, whereas the entire face is involved with peripheral disease. There may be bilateral involvement in Lyme disease, in contrast to Bell's palsy, which is always unilateral.

In children with facial nerve palsy caused by Lyme disease, other manifestations, such as erythema migrans, are rarely seen (27% in one series). Thus, even in the absence of other findings, serologic evidence for systemic Lyme infection should be sought in all children with isolated seventh nerve paresis in endemic areas. An LP should be performed if there is other evidence of meningoencephalitis such as headache; however, the need for LP in a child at risk for Lyme disease with isolated facial nerve palsy is controversial. Other associated neurologic abnormality, specifically in the other cranial nerves, necessitates further evaluation, including CT or MRI.

Management

Treatment for facial nerve palsy not associated with Lyme disease is somewhat controversial, but steroids may be beneficial when started early in the course of the disease. Some authors have recommended a course of prednisone (2 mg/kg per day in two divided doses) over 7 to 10 days if the patient is seen within the first 24 to 48 hours days of disease. A recent study suggests that acyclovir may be of benefit. Regardless of treatment, complete recovery is seen in 60 to 80% of children, beginning during the second to third week of illness. Those with partial paralysis generally have a better prognosis. During recovery period, special care should be taken to protect the cornea by the instillation of bland ointments (e.g., Lacrilube). The child should be referred for reexamination to ensure a recovery during the expected time period.

Children with clinical or serologic evidence of Lyme-associated facial nerve palsy should be treated with oral antibiotics (amoxicillin, tetracycline, or erythromycin) for 21 to 28 days. The effectiveness of steroids in such patients has not been evaluated. Parenteral antibiotic treatment is recommended for children who also have evidence of meningitis.

Suggested Readings

SEIZURES

Chamberlain JM, Altieri MA, Futterman C, et al. A prospective, randomized study comparing intramuscular midazolam with intravenous diazepam for the treatment of seizures in children. Pediatr Emerg Care 1997;13:92–94.

Dunn DW. Status epilepticus in infancy and childhood. Neurol Clin North Am 1990;8:647–658.

Holmes GL, Russman BS. Shuddering attacks. Am J Dis Child 1986; 140:72–73.

Kenney RD, Taylor JA. Absence of serum chemistry abnormalities in pediatric patients presenting with seizures. Pediatr Emerg Care 1992; 8:65–66.

Maytal J, Shinnar S, Moshe SL, Alvarez LA. Low morbidity and mortality of status epilepticus in children. Pediatrics 1989;83:323–331.

Mosewich RK, So EL. A clinical approach to the classification of seizures and epileptic syndromes. Mayo Clin Proc 1996;71:405–414.

Nypaver MM, Reynolds SL, Tanz RR, Davis AT. Emergency department laboratory evaluation of children with seizures: dogma or dilemma. Pediatr Emerg Care 1992;8:13–16.

Orr RA, Dimand RJ, Venkataraman ST, et al. Diazepam and intubation in emergency treatment of seizures in children. Ann Emerg Med 1991;20:1009–1013.

Reerink JD, Peters ACB, Verloove-Vanhorick SP, et al. Paroxysmal phenomena in the first two years of life. Dev Med Child Neurol 1995;37: 1094–1100.

Runge JW, Allen FH. Emergency treatment of status epilepticus. Neurology 1996;46(Suppl):S20–S23.

Rylance GW. Treatment of epilepsy and febrile convulsions in children. Lancet 1990;336:488–491.

Seigler RS. The administration of rectal diazepam for acute management of seizures. J Emerg Med 1990;8:155–159.

Selbst SM, Clancy R. Pseudoseizures in the emergency department. Pediatr Emerg Care 1996;12:185–188.

Shuper A, Mimouni M. Problems of differentiation between epilepsy and non-epileptic paroxysmal events in the first year of life. Arch Dis Child 1995;73:342–344.

Turnbull TL, Vanden Hoek TL, Howes DS, Eisner RF. Utility of laboratory studies in the emergency department patient with a new-onset seizure. Ann Emerg Med 1990;14:373–377.

Warden CR, Brownstein DR, Del Beccaro MA. Predictors of abnormal findings of computed tomography of the head in pediatric patients presenting with seizures. Ann Emerg Med 1997;29:518–523.

Working Group on Status Epilepticus. Treatment of convulsive status epilepticus. JAMA 1993;270:854–859.

FEBRILE SEIZURES

Annegers JF, Hauser WA, Shirts SB, Kurland LT. Factors prognostic of unprovoked seizures after febrile convulsions. N Engl J Med 1987;316:493–498.

Autret E, Billard C, Bertrand P, et al. Double-blind, randomized trial of diazepam versus placebo for prevention of recurrence of febrile seizures. J Pediatr 1990;117:490–494.

Berg AT, Shinnar S, Hauser WA, et al. A prospective study of recurrent febrile seizures. N Engl J Med 1992;327:1122–1127.

Camfield PR, Camfield CS. Management and treatment of febrile seizures. Curr Probl Pediatr 1997;27:6–13.

Farwell JR, Lee YJ, Hirtz DG, et al. Phenobarbital for febrile seizures: effects on intelligence and on seizure recurrence. N Engl J Med 1990;322: 364–369.

Gerber MA, Berliner BC. The child with a ‘simple' febrile seizure: appropriate diagnostic evaluation. Am J Dis Child 1981;135:431–433.

Green SM, Rothrock SG, Clem KJ, et al. Can seizures be the sole manifestation of meningitis in febrile children? Pediatrics 1993;92:527–534.

Jaffe M, Bar-Joseph G, Tirosh E. Fever and convulsions: indications for laboratory investigations. Pediatrics 1981;67:729–731.

Joffe A, McCormick M, DeAngelis C. Which children with febrile seizures need lumber puncture: a decision analysis approach. Am J Dis Child 19983;137:1153–1156.

Provisional Committee on Quality Improvement, Subcommittee on Febrile Seizures. Practice parameter: the neurodiagnostic evaluation of the child with a first simple febrile seizure. Pediatrics 1996;97:769–775.

Rosman NP, Colton T, Labazzo J, et al. A controlled trial of diazepam administered during febrile illnesses to prevent recurrence of febrile seizures. N Engl J Med 1993;329:79–84.

Rutter N, Smales ORC. Role of routine investigations in children presenting with their first febrile convulsion. Arch Dis Child 1977;52:188–191.

Schnaiderman D, Lahat E, Sheefer T, Aladjem M. Antipyretic effectiveness of acetaminophen in febrile seizures: ongoing prophylaxis versus sporadic usage. Eur J Pediatr 1993;152:747–749.

Verity CM, Butler NR, Golding J. Febrile convulsions in a national cohort followed up from birth. II: Medical history and intellectual ability at 5 years of age. Br Med J 1985;290:1311–1315.

ENCEPHALITIS

Bale JF Jr. Viral encephalitis. Med Clin North Am 1993;77:25–42.

Kesselring J, Miller DH, Robb SA, et al. Acute disseminated encephalomyelitis: MRI findings and the distinction from multiple sclerosis. Brain 1990;113:291–302.

Whitley RJ, Alford CA, Hirsch, et al. Vidarabine versus acyclovir therapy in herpes simplex encephalitis. N Engl J Med 1986;314:144–149.

Whitley RJ. Viral encephalitis. N Engl J Med 1990;323:242–247.

Ziegler DK. Acute disseminated encephalitis: some therapeutic and diagnostic considerations. Arch Neurol 1966;14:476–488.

HEMORRHAGIC SHOCK AND ENCEPHALOPATHY SYNDROME

Jardine DS, Bratton SL. Using characteristic changes in laboratory values to assist in the diagnosis of hemorrhagic shock and encephalopathy syndrome. Pediatrics 1995;96:1126–1131.

Levin M, Kay JDS, Gould JD, et al. Haemorrhagic shock and encephalopathy: a new syndrome with a high mortality in young children. Lancet 1983;ii:64–67.

Levin M, Pincott JR, Hjelm M et al. Hemorrhagic shock and encephalopathy: clinical, pathologic, and biochemical features. J Pediatr 1989;114:

194–203.

MIGRAINE

Burton LJ, Quinn B, Pratt-Chaney JL, Pourani M. Headache etiology in a pediatric emergency department. Pediatr Emerg Care 1997;13:1–4.

Maytal J, Young M, Shechter A, Lipton RB. Pediatric migraine and the International Headache Society (HIS) criteria. Neurology 1997;48:602–607.

Shevell MI. Acephaligc migraines of childhood. Pediatr Neurol 1996;14: 211–215.

Symon DNK, Russell G. The relationship between cyclic vomiting syndrome and abdominal migraine. J Pediatr Gastroenterol Nutr 1995;21(Suppl):S42–S43.

Hamalainen ML, Hoppu K, Valkeila E, Santavuori P. Ibuprofen or acetaminophen for the acute treatment of migraine in children: a double-blind, randomized, placebo-controlled crossover study. Neurology 1997;48: 103–107.

Hamalainen ML, Hoppu K, Santvuori P. Sumatriptan for migraine attacks in children: a randomized placebo-controlled study. Neurology 1997;48:1100–1103.

Gallagher RM, for the Dihydroergotamine Working Group. Acute treatment of migraine with dihydroergotamine nasal spray. Arch Neurol 1996;53:1285–1291.

Welborn CA. Pediatric migraine. Emerg Med Clin North Am 1997;15: 625–636.

PSEUDOTUMOR CEREBRI

Radhakrishnan K, Ahlskog JE, Garrity JA, Kurland LT. Idiopathic intracranial hypertension. Mayo Clin Proc 1994;69:169–180.

STROKE

Dyker AG, Lees KR. The rationale for new therapies in acute ischaemic stroke. J Clin Pharmacol Ther 1996;21:377–391.

Gobel U. Inherited or acquired disorders of blood coagulation in children with neurovascular complications. Neuropediatrics 1994;25:4–7.

Nicolaides P, Appleton RE. Stroke in children. Dev Med Child Neurol 1996;38:172–180.

Ohene-Frempong K. Stroke in sickle cell disease: demographic, clinical, and therapeutic considerations. Semin Hematol 1991;28:213–219.

Pavlakis SG, Gould RJ, Zito JL. Stroke in children. Adv Pediatr 1991;38:151–179.

Riela AR, Roach ES. Etiology of stroke in children. J Child Neurol 1993; 8:201–220.

Trescher WH. Ischemic stroke syndromes in childhood. Pediatr Ann 1992;21:374–383.

SPINAL CORD DYSFUNCTION

Al Deeb SM, Yaqub BA, Bruyn GW, Biary NM. Acute transverse myelitis: a localized form of postinfectious encephalomyelitis. Brain 1997;120:1115–1122.

Bracken, MB, Shepard, MS, Collins WF, et al. A randomized, controlled trial of methylprednisolone or naloxone in the treatment of acute spinal cord injury. N Engl J Med 1990;322:1405–1411.

Lewis DW, Packer RJ, Raney B, et al. Incidence, presentation and outcome of spinal cord disease in children with systemic cancer. Pediatrics 1986;78:438–443.

Menezes AH, Godersky JC, Smoker WRK. Spinal cord injury. In: McLaurin RL, Venes JL, Schut L, Epstein E, eds. Pediatric Neurosurgery. Surgery of the Developing Nervous System. 2nd ed. Philadelphia: WB Saunders, 1989:298–317.

Paine RS, Byers RK. Transverse myelopathy in childhood. Arch Dis Child 1953;85:151–163.

Ropper AH, Proshanzer DC. The prognosis of acute and subacute transverse myelopathy based on early signs and symptoms. Ann Neurol 1978; 4:51–59.

Sebire G, Hollenberg H, Meyer L, et al. High dose methylprednisolone in acute transverse myelopathy. Arch Dis Child 1997;76:167–168.

ACUTE POLYNEURITIS

Gurses N, Uysal S, Cetinkaya F, et al. Intravenous immunoglobulin treatment in children with Guillain-Barré syndrome. Scand J Infect Dis 1995;27:241–243.

Jones HR. Childhood Guillain-Barré syndrome: clinical presentation, diagnosis, and therapy. J Child Neurol 1996;11:4–12.

Korinthenberg R, Monting JS. Natural history and treatment effects in Guillain-Barré syndrome: a multicentre study. Arch Dis Child 1996;74:281–287.

Lichtenfield P. Autonomic dysfunction in the Guillain-Barré syndrome. Am J Med 1971;50:772–780.

Low L, Schneider J, Carter S. Polyneuritis in children. Pediatrics 1958;22:972–990.

Sakakihara Y, Kamoshita S. Age-associated changes in the symptomatology of Guiilain-Barré syndrome in children. Dev Med Child Neurol 1991;33:611–616.

The Guillain-Barré Syndrome Study Group. Plasmapheresis and acute Guillain-Barré syndrome. Neurology 1985;35:1096–1104.

MYASTHENIA GRAVIS

Berrouschot J, Baumann I, Kalischewski P, et al. Therapy of myasthenic crisis. Crit Care Med 1997;25:1228–1235.

Fenichel GM. Myasthenia gravis. Pediatr Ann 1989;18:432–438.

Millichap JG, Dodge PR. Diagnosis and treatment of myasthenia gravis in infancy, childhood, and adolescence. Neurology 1960;10:1007–1014.

Seybold ME. Myasthenia gravis: a clinical and basic science review. JAMA 1983;250:2516–2521.

BOTULISM

Picket L, Berg B, Chaplin E, Brunstetter-Schaefer MA. Syndrome of botulism in infancy: clinical and electrophysiologic study. N Engl J Med 1976;295:770–772.

Schreiner M, Field E, Ruddy R. Infant botulism: a review of 12 years' experience at the Children's Hospital of Philadelphia. Pediatrics 1991;87: 159–165.

Wigginton JM, Thill P. Infant botulism: a review of the literature. Clin Pediatr 1993;32:669–674.

PERIODIC PARALYSIS

Gordon N. Familial periodic paralysis. Dev Med Child Neurol 1973;15: 363–364.

Links TP, Smit AJ, Molenaar W, et al. Familial hypokalemic periodic paralysis: clinical, diagnostic, and therapeutic aspects. J Neurol Sci 1994;122:33–43.

Riggs JE. The periodic paralyses. Neurol Clin 1988;6:485–498.

ACUTE CEREBELLAR ATAXIA

Connolly AM, Dodson WE, Prensky AL, Rust RS. Course and outcome of acute cerebellar ataxia. Ann Neurol 1994;35:673–679.

Gieron-Korthals MA, Westberry KR, Emmanuel PJ. Acute childhood ataxia: 10-year experience. J Child Neurol 1994;9:381–384.

Stumpf DA. Acute ataxia. Pediatr Rev 1987;8:303–306.

BENIGN PAROXYSMAL VERTIGO

Koenigsberger MR, Chutorian AM, Gold AP, Schvey MS. Benign paroxysmal vertigo of childhood. Neurology 1968;20:301–302.

Tusa RJ, Saada AA, Niparko JK. Dizziness in childhood. J Child Neurol 1994;9:261–274.

MOVEMENT DISORDERS

Aron AM, Freeman JM, Carter S. The natural history of Sydenham's chorea. Am J Med 1965;38:83–95.

Klawans HL, Brandabur MM. Chorea in childhood. Pediatr Ann 1993; 22:41–50.

Shenker DM. Grossman HJ, Klawans HL. Treatment of Sydenham's chorea with haloperidol. Dev Med Child Neurol 1973;15:19–24.

Stacy M, Jankovic J. Childhood dystonia. Pediatr Ann 1993;22:53–58.

Swedo SE. Sydenham's chorea: a model for childhood autoimmune neuropsychiatric disorders. JAMA 1994;272:1788–1791.

Vedanarayanan VV. Paroxysmal movement disorders. Pediatr Ann 1997;26:402–408.

OPTIC NEURITIS

Arnason BGW. Optic neuritis and multiple sclerosis. N Engl J Med 1973;289:1140–1141.

Bradley WG, Whitty CW. Acute optic neuritis: prognosis for development of multiple sclerosis. J Neurol Neurosurg Psychiatr 1968;31:10–18.

Williams JR. Optic neuritis in a child. Pediatr Emerg Care 1996;12:210–212.

FACIAL NERVE PALSY

Adour KK. Diagnosis and management of facial paralysis. N Engl J Med 1982;307:348–351.

Adour KK, Ruboyianes JM, Von Doersten PG, et al. Bell's palsy treatment with acyclovir and prednisone compared with prednisone alone: a double-blind, randomized, controlled trial. Ann Otol Rhinol Laryngol 1996;105:371–378.

Albisetti M, Schaer G, Good M, et al. Diagnostic value of cerebrospinal fluid examination in children with peripheral facial palsy and suspected Lyme borreliosis. Neurology 1997;49:817–824.

Bingham PM, Galetta SL, Athreya B, Sladky J. Neurologic manifestations in children with Lyme disease. Pediatrics 1995;96:1053–1056.

Christen H-J, Bartlau N, Hanefeld F, et al. Peripheral facial palsy in childhood: Lyme borreliosis to be suspected unless proven otherwise. Acta Pediatr Scand 1990;79:1219–1224.

Hyden D, Roberg M, Forsberg P, et al. Acute “idiopathic” peripheral facial palsy: clinical, serological, and cerbrospinal fluid findings and effects of corticosteroids. Am J Otolaryngol 1993;14:179–186.

Pachner AR, Steere AC. Neurological manifestations of Lyme disease: meningitis, cranial neuritis and radiculoneuritis. Neurology 1985; 35:47–53.

Smouha EE, Coyle PK, Shukri S. Facial nerve palsy in Lyme disease: evolution of clinical diagnostic criteria. Am J Otology 1997;18:257–261.

CHAPTER 84

Infectious Disease Emergencies

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Although not as dramatic as multiple trauma or cardiac arrhythmias, infection precipitates more emergency department (ED) encounters than either of these other two conditions. Fever is the single most common chief complaint among children seen in the ED at most children's hospitals. Only a small fraction of patients with infections die; however, such diseases, being so much more common than noninfectious conditions with high fatality rates, account for a large percentage of deaths in the ED. Russo et al. reviewed the charts of children dead on arrival and found that infection ranked second only to trauma as a cause of mortality.

The approach to the febrile child is outlined in Chapter 28. In the current chapter, infections are divided anatomically as follows: generalized (bacterial), central nervous system (CNS), upper respiratory, lower respiratory, gastrointestinal (GI), bone and soft tissue, and genitourinary. Systemic nonbacterial illnesses, including the childhood exanthems, and several miscellaneous syndromes are dealt with as a group at the end of this chapter. Infections of the heart are discussed in Chapter 82, and encephalitis is covered as a neurologic emergency (see Chapter 83); human immunodeficiency virus (HIV), a disease of growing importance in children, is discussed in Chapter 85.

For each anatomic area, the relative frequency of disease caused by various pathogens is discussed and an approach is given for establishing a specific cause. A more extensive description is provided for the serious and/or treatable conditions, often clustering similar or less significant pathogens. The recommendations for management are derived from published literature. In certain areas, particularly as regards the indications for admission, scant information exists. Thus, it has been necessary at times to offer, as guidelines, the protocols that the author has found successful in clinical practice in the ED, even though they may not have been subjected to vigorous clinical trials.

INFECTIOUS SYNDROMES

Bacteremia and Sepsis

Bacteremia refers to the presence of bacteria in the blood stream. When bacteremia occurs in a young child and produces relatively few signs or symptoms, other than fever, the patient is considered to have the syndrome of occult bacteremia. The presence or absence of toxicity differentiates occult bacteremia, which is relatively asymptomatic, from sepsis, which is accompanied by findings of serious systemic illness. Because these infections represent a continuum, whereby some children with occult bacteremia proceed to develop the manifestations of sepsis, a separation into distinct diagnostic categories is not always possible. Bacterial infections in the bloodstream may occur in isolation (primary) or in association with focal disease (secondary). This section focuses on primary infections.

Bacteremia

Background

Streptococcus pneumoniae causes 90% of primary occult bacteremias. Less commonly encountered pathogens include *Salmonella*, *Neisseria meningitidis*, group A streptococcus, group B streptococcus, *Haemophilus influenzae* (Hib), and rarely, others. Table 84.1 summarizes the bacteria isolated in prospective studies of bacteremia. Since the introduction of a conjugated vaccine against Hib to be administered to children in the first 6 months of life, the isolation of this organism has declined almost to zero.

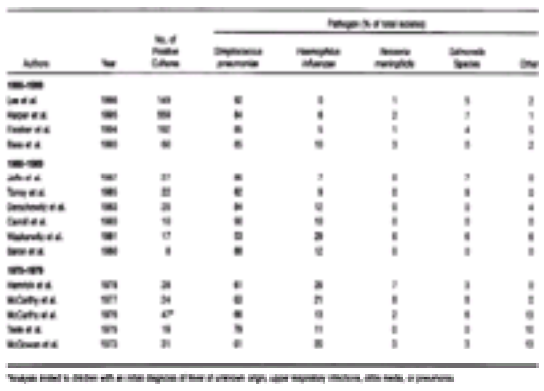

Table 84.1. Organisms Recovered from the Blood of Children with Unsuspected Bacteremia[a]

Occult bacteremia occurs with predictable regularity among febrile children younger than 2 to 3 years of age but perhaps not as often in the first 3 months of life, except in association with *Salmonella* gastroenteritis or urinary tract infections (UTIs). Before the advent of the conjugated vaccine against Hib, McGowan et al. recovered pathogens from 22 of 551 (4%) outpatient children with an elevated temperature. In this series, blood cultures were positive in 1 of 74 (1%) infants 6 months of age or younger, 11 of 116 (9.5%) children 7 to 12 months old, 5 of 131 (3.8%) of those 13 to 24 months old, and 5 of 225 (2.2%) of those more than 24 months of age. In a report of patients from the post–*H. influenzae* type b era, Lee and Harper noted a rate of bacteremia of 1.57% overall and 1.45% *for S. pneumoniae*; the peak incidence occurred in children 6 to 12 months of age. Dershewitz et al. noted similar rates of bacteremia (1.9 to 5.9%) in three groups of children, drawn from varying socioeconomic backgrounds. The introduction of a conjugated vaccine against *S. pneumoniae* will most likely occur within the next decade and would be expected to significantly reduce the incidence of bacteremia.

Pathophysiology

There appears to be a continuum of disease, starting with colonization and progressing through occult bacteremia, that may have three outcomes: 1) spontaneous resolution, 2) sepsis, or 3) focal infection. Clearly, pathogens such as *N. meningitidis* and *H. influenzae* have a greater tendency than *S. pneumoniae* to produce sepsis or invasive disease. Other than the specific organism, factors have not yet been completely defined that determine which children become colonized, which colonized children become bacteremic, and which bacteremic children improve without therapy.

Clearly, exposure to carriers plays an essential role in the process, accounting for the increased incidence of asymptomatic carriage and disease among household contacts of patients with infections caused by *N. meningitidis* or *H. influenzae.* A concurrent viral infection may increase the likelihood of bacteremia in a colonized of a child by disrupting the normal mucosal barrier. Bactericidal antibody in the serum has been shown to protect carriers against meningococcal disease; however, some persons without such antibodies do not progress beyond colonization. Among 492 military recruits studied by Goldschneider et al., 54 lacked bactericidal antibody to meningococcus group C. Of these 54 men, 24 acquired the organism in the nasopharynx, and 11 showed a bactericidal antibody response. Five of those without such a response developed meningococcal meningitis.

Clinical Manifestations

By definition, occult bacteremia causes few symptoms and signs. The complaints are usually those of malaise or an upper respiratory infection (URI). Fever, without evidence of a source, may be the only physical finding, or the patient may have a minor focus of infection, such as otitis media (OM).

McCarthy et al. attempted to define the history and observation variables useful in assessing febrile children. Observation of behavior (playfulness, alertness, and consolability) had the strongest correlation with the overall assessment; however, 9 of 21 children subsequently shown to have serious illnesses were not initially categorized as being moderately or severely ill. A subsequent study by Baker et al. found that this observational scale did not predict serious illness accurately in young infants, and Teach et al. noted a similar lack of success in older children with occult bacteremia. Waskerwitz et al. reported that physician assessment for bacteremia had limited utility with a sensitivity of 47%, a specificity of 83%, and a positive predictive value of 14%.

Children 3 to 24 months of age, who appear well with a fever at or above 39°C (102.2°F), have a 1.5 to 2% incidence of occult bacteremia compared with less than 0.5% for the group with low-grade fever (38° to 38.9°C [100.4° to 102°F]), but further elevations of the temperature above the 39°C (102.2°F) mark only minimally increase the likelihood of bacteremia. Torrey et al. tested whether the initial overall assessment of the response to an antipyretic drug administered in the ED could distinguish febrile children with bacteremia from those with viral infections. Neither the initial assessment nor the magnitude of the decrease in temperature was predictive for the presence or absence of organisms in the bloodstream. Additional studies have confirmed that the response to antipyresis does not correlate with the presence of bacterial infection.

The white blood cell (WBC) count is usually elevated in children with bacteremia, particularly with *S. pneumoniae.* For all pathogens, Jaffe and Fleisher found the sensitivity of the WBC count among children 3 to 36 months of age with a temperature greater than 39°C (102.2°F) to be 92% at 10,000/mm^3 or more, 65% at 15,000/mm^3 or more, and 38% at 20,000/mm^3 or more. Limiting their analysis to pneumococcal bacteremia among a similar group of highly febrile infants and toddlers, Kuppermann et al. reported that 8.2% of highly febrile infants and toddlers with a WBC count of 20,000/mm^3 or more had occult pneumococcal bacteremia. Lee and Harper demonstrated that the risk of bacteremia was greater among the more highly febrile patients with leukocytosis (Table 84.2) and noted an incidence in the range of 10%

among those with significant elevations.

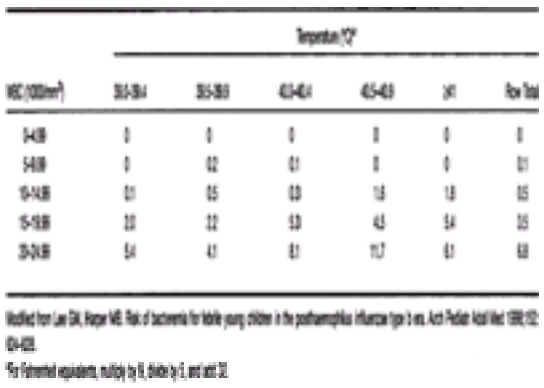

Table 84.2. Proportion of Patients with Bacteremia, Depending on White Blood Cell (WBC) Count and Temperature, 1993–1996

A shift to the left in the differential count and signs of toxicity on the peripheral smear are seen more often in bacteremic children than in those with viral infections, but neither serves to reliably distinguish the two groups. Although both the erythrocyte sedimentation rate (ESR) and the C-reactive protein (CRP) are usually elevated in patients with bacteremia, these tests provide only minimal additional information.

Management

A discussion on the management of bacteremia must address three issues: 1) evaluation for bacteremia in the febrile child with a seemingly trivial febrile illness, 2) treatment of the child with suspected bacteremia but no signs of sepsis or focal disease, and 3) therapy of proven bacteremia. Although controversy plagues all of these areas, the information accumulated over the last decade clearly shows some limitations to clinical judgment and points to the not uncommon occurrence of serious complications from bacteremia.

The likelihood of bacteremia in the child between 3 and 24 months of age with a fever of 39°C (102.2°F) or higher, coupled with the difficulties of clinical assessments in these youngsters, makes obtaining a WBC count, differential, and blood culture useful in many cases but rarely mandatory. With lower fevers in these young children or with highly elevated temperatures in children older than 24 months of age, the physician can rely more firmly on clinical judgment. In addition, children fully immunized against Hib have a reduced risk of bacteremia from this organism, once the most common cause of pediatric meningitis following occult bacteremia. In the patient who is believed to be more irritable or toxic than the usual child with a fever—but not so ill as to require an admission to the hospital—a WBC count and blood culture provides some guidance. In addition, patients who have a lumbar puncture performed in the ED as part of an evaluation for febrile seizures or suspected meningitis should have cultures of their blood. In these situations, the finding of a WBC count of 15,000 to 20,000/mm^3 or more strongly suggests a role for presumptive antibiotic therapy.

The role of presumptive antibiotic therapy for patients at risk for occult bacteremia remains controversial. Several pioneering but uncontrolled studies suggested a decreased incidence of focal bacterial sequelae after treatment with oral antibiotics, such as amoxicillin, but a subsequent randomized and blinded trial by Jaffe et al. did not confirm this observation in a limited sample of patients. Carroll et al. reported that intramuscular penicillin reduced focal complications from bacteremia; however, only 10 children had organisms recovered and a priori penicillin lacks effectiveness against *H. influenzae* and *Salmonella* species. A multicenter comparison of intramuscular ceftriaxone (50 mg/kg as a single dose) and oral amoxicillin (60 mg/kg per day given every 8 hours) described the complete elimination of bacteremia and a significant reduction in definite focal infections with ceftriaxone, a finding confirmed in subsequent, larger retrospective studies and by meta-analysis.

The decision about whether to treat children who are at risk for occult bacteremia hinges in large part on the balance between unnecessary administration of therapy to many children with no organisms in the bloodstream and prevention of serious complications in the few with bacteremia. Because diagnosis based on clinical and laboratory findings at the time of the visit has limited accuracy, physicians must choose between two alternatives, neither of which is completely satisfactory. Presumptive treatment is particularly appropriate for patients 3 to 24 months of age with a fever 39°C (102.2°F) or higher who are at higher-than-average risk (e.g., more irritable or lethargic than the usual child with a fever; WBC count of 15,000 to 20,000/mm^3 or more). For this group, intramuscular ceftriaxone (50 mg/kg) has been demonstrated to be effective. In managing highly febrile, young children not thought to be at higher risk, observation at home and/or oral antibiotic drugs, such as amoxicillin (50 to 75 mg/kg per day for two days) represent the major choices of treatment. If a conjugated vaccine against *S. pneumoniae,* currently under evaluation, proves effective and is introduced for general use, there will most likely be no role for presumptive antibiotic therapy, except in unusual circumstances.

The management of patients with proven bacteremia hinges on the identity of the pathogen. If penicillin-sensitive *S. pneumoniae* is isolated, the clinical findings at repeat examination determine the subsequent treatment. Children without fever or evidence of a serious infection (e.g., meningitis, pneumonia, cellulitis) should receive oral penicillin 50,000 units/kg per day or amoxicillin 50 mg/kg per day for 10 days; those with fever (38.5°C [101.2°F] or higher), clinical toxicity, or a serious focal infection merit initial intravenous (IV) antibiotic therapy. Children returning after a blood culture has grown other pathogens (*H. influenzae, N. meningitidis*, penicillin-resistant *S. pneumoniae*) are managed most prudently with IV antibiotics in the hospital because even those who remain well and afebrile appear to have some potential for persistent bacteremia and/or the continued evolution of focal infections. A few recent studies have

suggested, however, that selected afebrile and well-appearing children with a prior blood culture yielding penicillin-resistant *S. pneumoniae, N. meningitidis,* or *H. influenzae* may be managed with outpatient therapy, such as ceftriaxone (50 mg/kg), pending the results of repeat cultures.

Sepsis

Background

In sepsis, bacteremia exists in association with signs of serious systemic illness. The etiology of sepsis varies with age in the otherwise healthy child.

During the first 2 months of life, group B streptococcus and *Escherichia coli* are the most common isolates with sepsis. The group B streptococci cause more than 10,000 cases of neonatal disease yearly in the United States. At Yale, Gladstone et al. reviewed a decade of neonatal sepsis, from 1979 to 1988, and compared the findings to earlier reports from the same institution. Among 270 infants with sepsis, they isolated group B streptococcus from 64, *E. coli* from 46, *Klebsiella pneumoniae* from 18, and *H. influenzae* from 8. Analysis of the trend showed a steady level of infection for the group B streptococcus, compared with the prior decade, and a slight decrease in incidence for *E. coli.* Although some reports have suggested a downward trend in early onset group B streptococcus sepsis following the introduction of strategies for prophylaxis based on maternal screening, Pena et al. observed a steady incidence in late onset disease in Boston from 1982 to 1996.

N. meningitidis and *S. pneumoniae* infect the newborn only occasionally, but at a later age they emerge as the most common causes of sepsis in children. In a 12-year review, before the advent of the conjugated vaccine against Hib, Jacobs et al. studied 42 cases of “apparent meningococcemia” and found 30 infections with *N. meningitidis* and 12 with *H. influenzae.* Pneumococcal sepsis occurs more commonly in children with an absent or dysfunctional spleen. Group A streptococcus, *Staphylococcus aureus,* and *Salmonella* are recovered from the bloodstream relatively rarely, unless specific focal infections are present or, in the case of *Salmonella,* the patient has typhoid fever.

Sepsis occurs less often than bacteremia; however, large numbers of children with meningococcemia are occasionally seen in epidemics. Approximately 1,500 cases of meningococcal sepsis are reported yearly in the United States, most affecting children. The usual annual incidence has been estimated at 1 per 100,000 population, although in one epidemic, the attack rate was 838 per 100,000 children.

Certain conditions impose an increased susceptibility to sepsis on children. These include neoplasia, immunodeficiency syndromes, immunosuppressive therapy, asplenia, and sickle cell disease. The hemoglobinopathies pose a particularly urgent problem because of their relative frequency and the fact that overwhelming sepsis may occur in the young before the initial clinical manifestation of the underlying hematologic disease. Among 326 consecutive children with sickle cell hemoglobinopathies seen at The Children's Hospital of Philadelphia in a single year, the temperature was 38°C (100.4°F) or higher in 154, and four of those with fever had positive blood cultures. Two of the four were septic and the other two were bacteremic, but in the latter the immediate institution of antibiotic therapy may have prevented the rapid evolution of systemic toxicity.

Pathophysiology

As discussed under occult bacteremia, the first step toward sepsis occurs with colonization of the host by potentially pathogenic bacteria. The site of colonization is usually the pharynx in older children but may be the umbilicus or bowel in the neonate. Among immunosuppressed children, organisms that reside in the GI tract often invade the bloodstream.

After entry into the bloodstream, bacteria increase in number. As some of the organisms are lysed, toxic products, such as endotoxin, are released into the circulation. These products interact with host proteins and bind to receptors on cells of the immune system, as well as endothelia. Activation of the host cells follows, releasing a series of inflammatory mediators into the circulation. The initial cascade includes tumor necrosis factor (TNF), interleukin-1, and interleukin-6. Subsequently, additional interleukins and prostaglandins assume an important role.

Not every child with bacteremia develops the clinical manifestations of sepsis. The intrinsic virulence of the pathogen determines, in part, whether the bacteremia resolves spontaneously. *N. meningitidis* bacteremia results in sepsis or a focal infection in 50 to 75% of children, whereas *Salmonella* almost always remains asymptomatic. Host factors also assume an important role in clearing circulating bacteria. The young child, particularly younger than 2 years old, has a greater tendency to become seriously ill.

Clinical Manifestations

The duration of the history in a child with sepsis varies. Even though some children are febrile for several days during a preceding bacteremia, others develop a sudden dramatic illness. The interval between the initial fever and death may be less than 12 hours in fulminant meningococcemia (Fig. 84.1). The child progresses with continued sepsis from malaise to profound lethargy and, finally, to obtundation. Although fever is the cardinal sign of infection, children younger than 2 to 3 months of age may remain afebrile with sepsis; hypothermia is common in the first month of life.

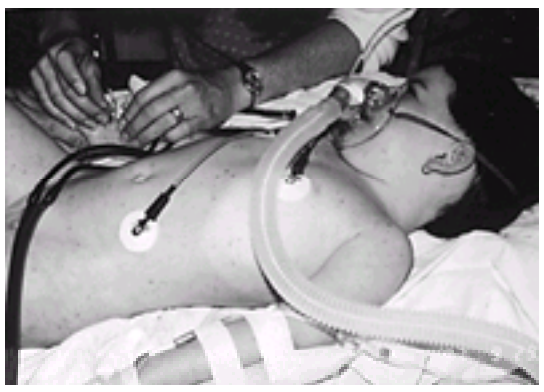

FIGURE 84.1. Sepsis secondary to infection with *N. meningitidis.* Note diffuse purpuric lesions in this critically ill child.

A marked tachycardia occurs early in the course of the disease, at times exceeding 200 beats per minute (bpm) in the first 3 months of life, 175 bpm between 4 months and 2 years of age, and 150 bpm in the older child. Hypotension and tachypnea are present. The skin is cold and poorly perfused; in addition, petechiae and purpura may appear, particularly with *N. meningitidis.*

The hemoglobin and hematocrit are usually normal, falling occasionally from hemolysis as seen with disseminated intravascular coagulation (DIC). Although leukocytosis usually accompanies sepsis, an overwhelming infection occasionally produces neutropenia. The WBC count is rarely normal, and the differential is almost always shifted to the left; metamyelocytes and band forms often make their way into the peripheral blood. As the infection progresses, the platelet count decreases. It is distinctly unusual to have evidence of cutaneous hemorrhage from sepsis without thrombocytopenia. Similarly, the prothrombin time (PT), partial thromboplastin time (PTT), and fibrin degradation products rise with the ongoing consumption of the clotting factors. The electrolytes reflect a metabolic acidosis, and occasionally, mild hyponatremia occurs; the blood urea nitrogen (BUN) is usually normal but may rise in the face of preceding dehydration. A Gram stain of a petechial scraping shows the etiologic agent in one-third of cases. In the infant, hypoglycemia may occur.

Management

Although the initial therapy for sepsis is directed at the preservation of vital functions, every effort must be made to obtain the appropriate diagnostic studies (Table 84.3). Blood should be drawn for culture, complete blood count (CBC), platelet count, PT, PTT, electrolytes, BUN, arterial blood gas (ABG) analysis, serum aspartate aminotransferase (AST, formerly SGOT), and serum alanine aminotransferase (ALT, formerly SGPT) in conjunction with the immediate insertion of an intravenous catheter.

1. Ensure adequate ventilation and cardiac function.
2. Obtain laboratory studies (simultaneously with step 3): CBC, platelet count, PT, PTT, electrolytes, BUN, creatinine, glucose, arterial blood gas, blood culture, fibrin degradation products, AST, ALT.
3. Initiate hemodynamic monitoring and support: peripheral venous access; urinary catheter; central venous and arterial catheters (as indicated); cardiorespiratory monitors; normal saline, starting at 20 mL/kg.
4. Administer drugs and other therapeutic agents:
   Antibiotics: <2 months: ampicillin (50 mg/kg) and gentamicin (2.5 mg/kg)
   >2 months: ceftriaxone (50 mg/kg) or cefotaxime (50 mg/kg) or both ampicillin (50 mg/kg) and chloramphenicol (25 mg/kg)
   Sodium bicarbonate (pH <7.0) 1–2 mEq/kg
   Glucose (serum glucose <50 mg/dL) 0.25–1 g/kg
   Packed red blood cells (Hgb <10 g/dL) 10 mL/kg
   Platelet concentrates (platelet count: <50,000/mm^3) 0.2 unit/kg
   Fresh-frozen plasma (elevated PT/PTT) 10 mL/kg

CBC, complete blood count; PT, prothrombin time; PTT, partial thromboplastin time; BUN, blood urea nitrogen.

Table 84.3. Immediate Management of Sepsis

As initial therapy, normal saline, with or without 5% dextrose, is given at 20 mL/kg per hour or more rapidly, depending on the response. The unstable patient may require central venous, arterial, and urinary catheters. Deterioration often occurs after antibiotic administration because of sudden lysis of organisms, resulting in massive endotoxemia and release of cytokines, particularly TNF. Thus, appropriate venous and arterial access should ideally be in place before or concurrent with the administration of antimicrobial drugs. The initial laboratory studies, the response to the bolus of saline, and the measurements of the intravascular status determine the type and quantity of the subsequent fluids and the need for vasopressors (see Chapter 3).

For children younger than 2 months of age, ampicillin (200 mg/kg per day) and gentamicin (7.5 mg/kg per day) are administered; cefotaxime (150 mg/kg per day) may be used in place of gentamicin for the newborn. Dosages need to be decreased for premature infants in the first month of life and term infants in the first week (Table 84.4A and Table 84.4B). Cefotaxime (200 mg/kg per day) or ceftriaxone (100 mg/kg per day) alone provides effective monotherapy for the child older than 2 months of age. Vancomycin (40 mg/kg per day) may be added for patients who are critically ill or at particular risk of infection with penicillin-resistant *S. pneumoniae,* as in the case, for example, of a patient with sickle cell anemia who is taking daily prophylactic penicillin. In the presence of a focus of infection likely to be staphylococcal, oxacillin (150 mg/kg per day) can be used together with cefotaxime; alternatively, the combination of clavulanic acid and ampicillin (200 mg/kg per day) may be administered. Chloramphenicol (75 to 100 mg/kg per day) or meropenem (60–120 mg/kg per day) should be kept in mind for children with allergies to penicillins and cephalosporins. Corticosteroid therapy

is not recommended for sepsis.

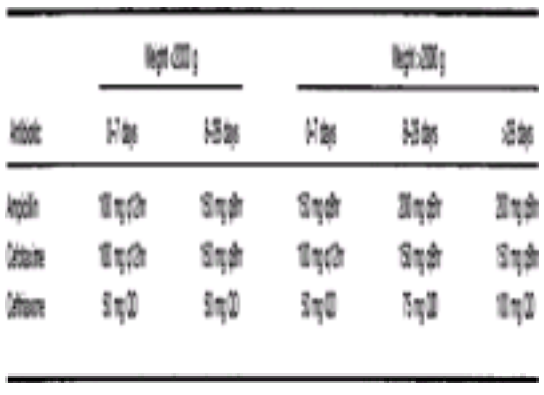

Table 84.4A. Intravenous Antibiotic Dosing for Newborn Infants Based on Age and Weight (Total Daily Dose [mg/kg/day] and Dosing Interval)

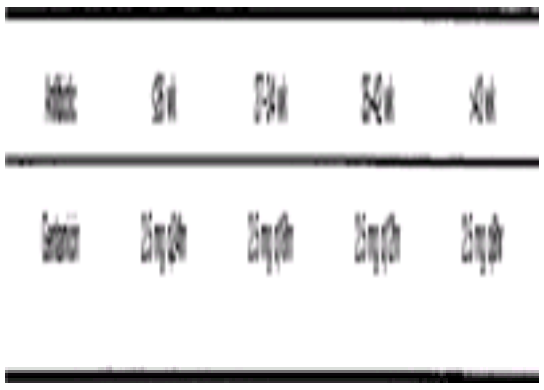

Table 84.4B. Intravenous Antibiotic Dosing for Newborn Infants Based on Gestational Age (mg/kg/dose)

Blood components are given as indicated by the results of the initial hematologic studies. If the hemoglobin is lower than 10 g/dL, packed red cells are administered at 10 mL/kg. Thrombocytopenia (less than 50,000/mm ^{3}) is corrected with platelet concentrates at 0.2 units/kg and decreased clotting factors with fresh-frozen plasma 10 mL/kg. For the child with hypoglycemia (glucose less than 50 mg/dL), glucose should be given at a dose of 0.25 to 1 g/kg in the form of a 25% solution. Heparin plays no role in the initial emergency care of the child with sepsis but may be useful subsequently to treat severe thrombotic episodes. Specific inhibitors of endotoxin and cytokines, such as bacterial polysaccharide inhibiting protein (BPI) are under investigation but have not been demonstrated to be effective as of yet.

Central Nervous System Infections

Three important infectious syndromes involve the CNS: meningitis, encephalitis, and brain abscess. Because encephalitis and brain abscess usually confront the emergency physician as problems in the differential diagnosis of various neurologic manifestations, they are discussed as neurologic emergencies in Chapter 83.

Meningitis, an inflammation of the membranes lining the CNS, results from an infection or irritation on a noninfectious basis. Inflammation of the meninges produces a pleocytosis in the cerebrospinal fluid (CSF), allowing, in most cases, for the diagnosis of meningitis by examination of this readily accessible material. However, organisms may occasionally infect the meninges without eliciting a cellular reaction, either because sufficient time has not elapsed for a leukocyte response or because the pathogen is of low virulence.

In the ED setting, most cases of meningitis result from an infection of the CNS. Table 84.5 lists those organisms that are the more common invaders of the meninges. The most important initial task that confronts the emergency physician is the identification of children with bacterial meningitis, a life-threatening infection amenable to therapy if diagnosed sufficiently early in the course. The sine qua non for the definitive diagnosis of meningitis is examination of the CSF. Routine studies performed on this fluid should include cell count with differential, glucose, protein, Gram stain, and bacterial culture. In selected cases, additional studies, such as latex agglutination; acid fast stain; India ink preparation; serologic testing for Lyme disease and/or syphilis; cryptococcal antigen; polymerase chain reaction (PCR) for herpes; and cultures for anaerobic bacteria, mycoplasma, mycobacteria, and fungi, are indicated. Values of various parameters of the CSF are presented for healthy persons and for those with viral and bacterial meningitis (Table 84.6).

Viruses	*Haemophilus influenzae*
Enteroviruses	*Salmonella* species
Herpes simplex	*Listeria monocytogenes*
Lymphocytic choriomeningitis	*Mycobacterium tuberculosis*
Mumps	Spirochetes—Lyme, syphilis
Other	Fungi
Mycoplasma	*Candida albicans*
Bacteria	*Cryptococcus neoformans*
Streptococcus pneumoniae	Parasites
Neisseria meningitidis	Cysticercosis
Escherichia coli	Amoebae
Group B streptococcus	

Table 84.5. Organisms That Cause Meningitis

	Neonate	Child	Bacterial Meningitis	Viral Meningitis
WBC (per mm^3)	<30	<10	200–20,000	10–1,000
Protein (mg/dL)	<170	<40	>100	40–100
Glucose (mg/dL)	>30	>40	<30	>30

Table 84.6. Usual Ranges for CSF WBC Count, Protein, and Glucose in Normal Infants and Children and in Those with Viral or Bacterial Meningitis

The CSF ordinarily contains no red blood cells. The presence of blood indicates either contamination from a traumatic lumbar puncture or hemorrhage in the CNS. If the density of the red cells is constant from the first to the last tube collected and the cells are crenated, the likelihood of CNS hemorrhage is greater. Certain infectious agents, such as herpes simplex virus, may produce a hemorrhagic meningoencephalitis.

More than 9 WBCs in the CSF from a child and 29 from a neonate indicate inflammation of the meninges. However, a specimen is sometimes obtained early in the course of meningitis before an inflammatory reaction has been invoked. Thus, a child with fewer than 10 cells in the CSF will occasionally later develop the physical and laboratory manifestations of meningitis.

In viral infections of the CNS, the WBC count in the CSF usually ranges from 10 to 1,000/mm^3. Occasionally, a WBC count as high as 2,500/mm^3 may be seen. A predominance of mononuclear cells is usually present, although early in the course of the illness, neutrophils may be in the majority. Bacterial meningitis evokes an intense infiltration of leukocytes, with a marked predominance of neutrophils. The cell count is usually in the range of 1,000 to 20,000/mm^3 but may be even higher.

The CSF glucose is normally one-half to two-thirds of the serum glucose. Equilibration between the serum and CSF glucose levels has been estimated to require at least 30 minutes. Thus, a rapid decrease in the serum glucose may obscure a wide variance from the CSF level, whereas a sudden elevation may lead to a falsely large discrepancy. Because the stress of a lumbar puncture produces hyperglycemia, a serum glucose for comparison with the CSF level should be obtained before this procedure is attempted. In viral meningitis, the CSF glucose, usually in the normal range, may be as low as 30 mg/dL. The glucose in bacterial meningitis often falls below 30 mg/dL. Hypoglycorrhachia accompanying an elevated protein level and a mild mononuclear pleocytosis should arouse a suspicion of tuberculous meningitis.

A normal CSF protein is less than 40 mg/dL in the child and 170 mg/dL in a neonate. Although the protein is minimally elevated in viral meningitis, the level in bacterial meningitis is generally 100 mg/dL or greater.

Additional studies, such as latex agglutination, may be helpful in distinguishing bacterial from viral meningitis.

Bacterial Meningitis

Background

Although almost any bacteria can cause meningitis, more than 90% of the cases in immunocompetent children result from infections with five organisms *: S. pneumoniae, N. meningitidis, E. coli,* group B streptococcus, and *H. influenzae.* The most common organism varies with the age of the child (Fig. 84.2). In the first month of life, *E. coli* and group B streptococcus are usually isolated; *Listeria monocytogenes*, a Gram-positive rod, accounts for 1 to 3% of the cases. Between 30 and 60 days of age, group B streptococcus continues to be recovered frequently, followed by *S. pneumoniae* and *N. meningitidis; H. influenzae* occurs rarely. After the first 2 months of life, *S. pneumoniae* and *N. meningitidis* cause the majority of meningeal infections; *H. influenzae* remains a consideration primarily among children not immunized with

conjugated Hib vaccine or those recently arrived from a third world country. *Salmonella,* an uncommon etiologic agent in the United States, should be suspected in the first few months of life if meningitis occurs in association with gastroenteritis.

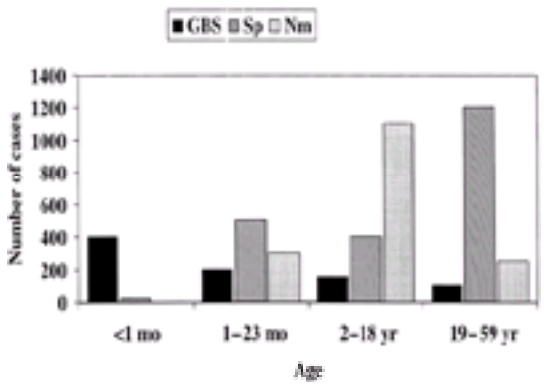

FIGURE 84.2. Projected cases of meningitis by age and organism, excluding *E. coli. GBS,* group B streptococcus; *Nm, Neisseria meningitidis; Sp, Streptococcus pneumoniae.* (Modified from Schuchat, et al. N Engl J Med 1997;337:970–976.)

Meningitis was formerly among the most common life-threatening infections of children, but the incidence of this disease has declined significantly since the introduction of conjugated vaccine against Hib. Schuchat et al. performed an active population-based surveillance of meningitis (excluding disease caused by Gram-negative enteric rods) during 1995 from all the acute-care hospitals in four states with a population of more than 10 million and described the attack rates for the various pathogens, by age (Table 84.7). Based on their findings, they projected the total number of cases of bacterial meningitis during 1995 in the United States, as shown in Figure 84.2, of particular note estimating that only 948 cases occurred in children from 1 to 23 months of age. A conjugated vaccine against *S. pneumoniae* will most likely be introduced within the next decade and would be expected to significantly reduce the incidence of meningitis.

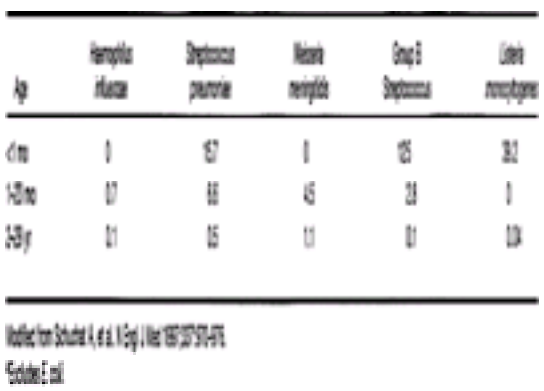

Table 84.7. Age-Specific Incidence (per 100,000) in 1995 of Bacterial Meningitis in the United States during 1995 [a]

Pathophysiology

Microorganisms gain access to the CNS through two potential pathways. Most commonly in children, a preceding bacteremia leads to hematogenous seeding of the meninges. Alternatively, direct extension may occur from a purulent parameningeal focus.

Colonization of the nasopharynx sets the stage for the subsequent development of meningitis in most bacterial CNS infections in the older child. In the infant, who is susceptible to infection with Gram-negative enteric organisms, the bowel is often the source of the pathogen. This is also the case beyond the neonatal period with less common bacteria such as *Salmonella.* A small percentage of the children colonized with a potential pathogen will develop bacteremia, but most will clear the pathogens from their bloodstream spontaneously; more than 80% of cases of bacteremia with *S. pneumoniae* resolve without leading to local infection. Although meningitis is more common after the recovery of *N. meningitidis* from the blood, most such children also escape CNS infection. Splenectomized persons, or those who have infarcted spleens on the basis of inherited hemoglobinopathies, cannot limit the spread of bacteremia as successfully as those who are immunologically intact.

Although a less common predecessor to meningitis, purulent collections contiguous to the CNS may also produce such infections. Sinusitis is the most common offender. Organisms also may invade the meninges on occasion directly from the middle ear. However, meningitis following OM usually results from bacteremia, unless a congenital or posttraumatic fistula in the temporal bone provides access to the CSF.

Clinical Manifestations

The signs and symptoms of meningitis vary with the child's age (Table 84.8). Particularly in the first 3 months of life, the clinician must maintain a high index of suspicion for this disease. In addition, it should be kept in mind that partial

treatment with antibiotics may obscure the typical findings.

		Signs	
Age	Symptom	Early	Late
0–3 mo	Paradoxical irritability	Lethargy	Bulging fontanelle
	Altered sleep pattern	Irritability	Shock
	Vomiting	Fever (±)	
	Lethargy	Hypothermia (<1 mo)	
4–24 mo	Irritability	Fever	Nuchal rigidity
	Altered sleep pattern	Irritability	Coma
	Lethargy		Shock
>24 mo	Headache	Fever	Coma
	Neck pain	Nuchal rigidity	Shock
	Lethargy	Irritability	

Table 84.8. Signs and Symptoms of Meningitis

Before 3 months of age, the history is that of irritability, an altered sleep pattern, vomiting, and decreased oral intake. In particular, paradoxical irritability points to the diagnosis of meningitis. Irritability in the infant without inflammation of the meninges is generally alleviated by maternal fondling; however, in the child with meningitis, any handling, even directed toward soothing the infant, may increase irritability by its effect on the inflamed meninges. The amount of time spent sleeping may either increase because of obtundation or decrease from irritability. Bulging of the fontanel, an almost certain sign of meningitis in the febrile, ill-appearing infant, is a late finding. Vomiting is often a prominent feature of the presentation of infants with meningitis, but when emesis occurs in isolation, particularly in the absence of fever, it more likely points to pyloric stenosis or other disorders of the GI tract.

As the child passes beyond the age of 3 months, the symptoms gradually becomes more specific for involvement of the CNS. A change in the level of activity is almost always noticeable. However, it is only in the child older than 2 years of age that meningitis manifests reliably with complaints of headache and neck stiffness.

The physical examination in the young infant rarely provides specific corroboration, even when the history suggests meningitis. Fever is often absent in these children, despite the presence of bacterial infection. Any child younger than 2 to 3 months of age who is brought to the ED with a documented temperature of 38.0° to 38.5°C (100.4° to 101.2°F) or higher should be considered at particular risk for meningitis. The physical signs are sufficiently elusive that many experts caution that one should not rely exclusively on the examination to rule out meningeal infection. In several studies, 5 to 10% of these young infants had meningitis despite being judged clinically well by experienced pediatric residents.

After 3 months of age, increasing, but not absolute, reliance can be placed on the physical findings; fever is almost inevitably noted. Specific evidence of meningeal irritation often is present, including nuchal rigidity (Fig. 84.3) and, less often, Kernig and Brudzinski signs. When a lumbar puncture fails to confirm the diagnosis of meningitis despite the presence of meningeal signs, other conditions must be pursued that can mimic the findings on physical examination. Conditions capable of producing meningismus (irritation of the meninges without pleocytosis in the CSF) include severe pharyngitis, retropharyngeal abscess, cervical adenitis, arthritis or osteomyelitis of the cervical spine, upper lobe pneumonia, subarachnoid hemorrhage, pyelonephritis, and tetanus.

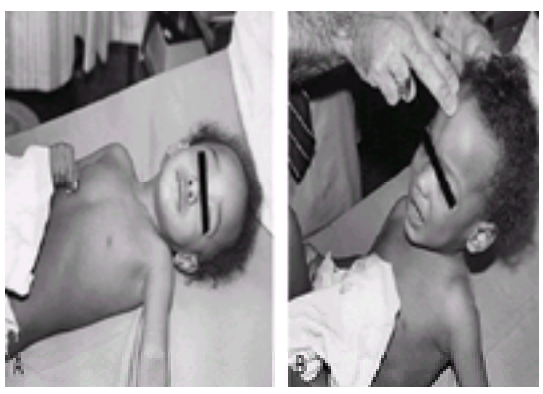

FIGURE 84.3. Child with meningitis who demonstrates ill appearance and nuchal rigidity. **A.** Patient lying supine with neck in neutral position. **B.** Pain grimace and resistance (lifting of shoulders) upon attempted flexion of the neck.

At times, meningitis manifests initially as a convulsion. In the infant less than 6 months of age with a first time seizure, a lumbar puncture is mandatory to discern the presence of CNS infection, unless there are specific contraindications or an alternative diagnosis is readily apparent. The occurrence of a seizure in a febrile child older than 6 months of age presents more of a dilemma for the clinician. Febrile seizures are common, affecting 5% of children, and underlie most of these episodes. However, it may be difficult to distinguish a simple febrile seizure in an ill-appearing child with a high fever from early meningitis because of the vague symptoms and lack of definitive physical findings in the first 2 years of life. In addition, the occurrence of a convulsion may obscure such meningeal signs as nuchal rigidity, which may be masked by the hypotonia of the postictal period. Opinion varies regarding whether a febrile seizure can be distinguished clinically from a seizure secondary to meningitis. In one study reported from the era before widespread administration of the conjugated vaccine against Hib, 20% of children thought to have a first febrile seizure on the basis of the history and

physical examination eventually were determined to have meningitis. However, other investigators have reported more success in making such a clinical differentiation. Because of the difficulty of establishing a clinical diagnosis in the young child, a lumbar puncture should be strongly considered in every child less than 12 months of age with a first febrile seizure. In the older child or in the case of a recurrent febrile seizure, the experienced clinician may choose to be guided by the physical findings and the evolution of the illness over the ensuing 12 to 24 hours.

The child with meningitis often has a complicated course beginning in the ED or even preceding arrival at the hospital (Table 84.9). Shock, seizures, and hyponatremia strike at any age, whereas apnea and hypoglycemia predominantly affect infants less than 3 months of age. Although sterile subdural effusions and, rarely, empyemas usually occur later in the disease, they merit consideration in the infant with signs of herniation and a bulging fontanel.

Early	Late
Apnea	Hyponatremia
Shock	Subdural empyema
Hypoglycemia	Seizures
Hyponatremia	
Seizures	

Table 84.9. Short-term Complications of Meningitis

Management

Bacterial meningitis is a medical emergency that requires the immediate institution of therapy (Table 84.10). If the disease is diagnosed and treated promptly, mortality dwindles to less than 5%. Antibiotics should be given by IV at the completion of the lumbar puncture. Although several studies have shown the average elapsed time between arrival in the ED and delivery of antibiotics averages 2 to 3 hours, every effort should be made to reduce this interval to enhance the theoretic advantages of early therapy. If meningitis is suspected but attempts to obtain CSF are unsuccessful, this failure should not delay the antibiotic administration. It is safer to treat a child presumptively with a sterile pleocytosis than to handle the complications of progressive CNS infection. Similarly, inability to achieve venous access should not retard the initiation of drug therapy. The intramuscular route provides a suitable, temporary alternative.

1. Ensure adequate ventilation and cardiac function.
2. Obtain laboratory studies (simultaneously with step 3):
Cerebrospinal fluid: cell count, glucose, protein, Gram stain, culture, latex agglutination (as indicated)
Blood: complete blood count, platelet count, prothrombin time, partial thromboplastin time, electrolytes, blood urea nitrogen, creatinine, glucose, arterial blood gas, blood culture
3. Initiate hemodynamic monitoring and support.
Achieve venous access; use cardiorespiratory monitors.
4. Administer drugs.
Treat septic shock, if present.
Consider dexamethasone (0.15 mg/kg) before or shortly after antibiotic administration.
Antibiotics: <1 month: ampicillin (50 kg) and cefotaxime (50 mg/kg); >1 month: vancomycin (15/mg) and either ceftriaxone (50 mg/kg) or cefotaxime (75 mg/kg)*
Glucose (if serum glucose <50 mg/dL) 0.25–1 g/kg
Treat acidosis and coagulopathy, if present.

*Chloramphenicol (25 mg/kg) or meropenem (25 mg/kg) may be used in place of cephalosporins for children allergic to those agents.

Table 84.10. Immediate Management of Bacterial Meningitis

The child's age determines the spectrum of microorganisms causing meningitis and the selection of antibiotic therapy (Table 84.10). In the first 30 days of life, the most likely organisms include the Gram-negative enteric rods, such as *E. coli,* and group B streptococcus. The enteric pathogens are almost always sensitive to the aminoglycoside antibiotics; however, recent studies have shown that a third-generation cephalosporin, such as cefotaxime, may be more effective. Penicillin or ampicillin effectively treats the group B streptococcus. Ampicillin and a third-generation cephalosporin (cefotaxime) or an aminoglycoside (gentamicin) thus provide coverage for the most common pathogens in the first month of life. The spectrum of these antibiotics also includes less common organisms in the neonate, such as *S. pneumoniae, N. meningitidis,* and *L. monocytogenes. Salmonella* is a somewhat unusual cause of meningitis in the United States but may be isolated in 1 to 2% of such infections. Increasingly, this organism is resistant to ampicillin. Thus, the isolation or strongly suspected presence of *Salmonella* from the GI tract dictates the inclusion of a cephalosporin (cefotaxime or ceftriaxone). Alternatives for the penicillin-allergic child with *Salmonella* infecting the meninges include chloramphenicol and meropenem.

Between 30 and 60 days of age, group B streptococcus remains the predominant pathogen, but the Gram-negative enteric bacilli decrease in frequency. *S. pneumoniae* and *N. meningitidis* occur sporadically, as does *H. influenzae,* because these children are too young to be immunized against Hib. The usual antibiotic combination is vancomycin to cover penicillin-resistant pneumococci, with either ceftriaxone or cefotaxime (Table 84.10)

After the first 2 months, the predominant pathogens that cause meningitis are *S. pneumoniae* and *N. meningitidis. H.*

influenzae, formerly responsible for most CNS infections in children, has become exceedingly rare. As for the child between 30 and 60 days of age, initial antibiotic therapy includes vancomycin and either ceftriaxone or cefotaxime (Table 84.10). Chloramphenicol is an option for patients with a solid history of a serious reaction to penicillin.

In addition to the antibiotic administration aimed at the eradication of the offending organism, supportive therapy for complications (Table 84.9 and Table 84.10) is an essential ingredient in the care of the child with meningitis. Recommended laboratory studies on every patient include a CBC, electrolytes, BUN, PT and PTT, glucose, and blood culture. A rapid assessment should be made about the adequacy of ventilation. The CNS edema that accompanies inflammation of the meninges may produce obtundation and hypoventilation. Apneic episodes can occur in the infant. Thus, oxygen, intubation, and assisted ventilation all may be required. Bacteremia, which usually accompanies meningitis, may lead to septic shock. This condition demands vigorous fluid resuscitation with normal saline. The response to an initial bolus of 20 mL/kg saline determines the need for further therapy such as the use of cardiotonic agents (see Chapter 3). The urgency to provide adequate perfusion to the vital organs by expanding the intravascular volume takes precedence over concerns about edema in the CNS.

Hyponatremia often accompanies meningitis, resulting from water retention because of inappropriate secretion of antidiuretic hormone (SIADH). Occasionally, the oral administration of hypotonic solutions by the parents during the preceding prodromal illness may produce fluid overload and a low serum sodium (Na). If the child is believed to have seizures on the basis of hyponatremia, the physician may give 3% NaCl (see Chapter 86) at a dosage of 10 to 12 mL/kg over 1 hour.

After the correction of dehydration or shock, the rate of fluid administration to the child with meningitis should be at 75 to 100% of maintenance requirements (see Chapter 86). Generally, D5/0.2% NaCl is used for this purpose. Failure of the serum Na to rise in the hyponatremic child mandates further restriction on hydration.

Hypoglycemia occurs as a reaction to septicemia and stress. It is a more common concomitant of meningitis in the first 3 months of life. If the blood glucose is less than 50 mg/dL, 25% glucose should be given at a dosage of 0.25 to 1 g/kg. This must be followed by an infusion of 5% glucose and monitoring of the response. Occasionally, 10% glucose will be necessary to maintain an acceptable serum level.

Seizures occur in 25% of children with bacterial meningitis and, occasionally, in those with viral infections. One should always be suspicious of derangement of the glucose or sodium as a cause of convulsive activity. However, most seizures are caused by irritation of the brain from the infectious process. They are controlled in the usual fashion with diazepam or lorazepam, phenytoin, and phenobarbital (see Chapter 70 and Chapter 83).

Subdural effusion and, less often, empyema occur in 20 to 40% of children with meningitis but usually appear later in the course and remain asymptomatic. In the rare case of an infant with herniation caused by a subdural collection, percutaneous drainage relieves the pressure on the brain and produces significant improvement.

Some studies have suggested that dexamethasone at a dosage of 0.15 mg/kg per dose given every 6 hours to children older than 2 months of age mitigates the sequelae of bacterial meningitis, particularly sensorineural hearing loss. The mechanism of action has been postulated to involve inhibition of cytokine production in the CSF. Although many clinicians choose to administer dexamethasone to patients beyond the first 2 months of life in whom the diagnosis of bacterial meningitis appears highly likely, contradictory evidence exists in the literature and expert panels have withheld a definitive endorsement. If the decision is made in favor of administration, the drug should be given before antibiotic administration when possible. If dexamethasone is not administered before the initiation of antibiotic therapy, it may be given subsequently but appears to lose its theoretic benefit after an interval or more than 4 hours.

Aseptic Meningitis

Background

The aseptic meningitis syndrome is defined here as an inflammation of the meninges that occurs in the absence of bacterial growth on routine culture media. A child whose initial CSF findings suggest an aseptic meningitis may rarely turn out to have a purulent infection because bacteria do not always elicit a marked polymorphonuclear leukocytosis early in the course of the disease. In addition, bacteria with unusual growth requirements, inhibited by subtherapeutic concentrations of antibiotics, or sequestered in pockets adjacent to but not directly communicating with the CSF, all may produce an aseptic meningitis syndrome.

Both infectious and noninfectious diseases cause the aseptic meningitis syndrome (Table 84.11). By far, the most common cause is viral meningitis; however, the clinician should be alert to unusual pathogens in patients who are immunocompromised or who show atypical clinical features. Despite underreporting, the Centers for Disease Control and Prevention (CDC) notes about 5000 cases annually in the United States.

Infectious	Parasites
Viruses	Trichinosis
Early or partially treated	Toxoplasmosis
bacterial meningitis	Cysticercosis
Parameningeal infection	Malaria
Unusual bacteria	*Naegleria*
Leptospirosis	*Noninfectious*
Syphilis	Neoplasm
Tuberculosis	Hemorrhage
Ehrlichia canis	Hypersensitivity reactions
Borrelia burgdorferi (Lyme)	Heavy metal poisoning
Mycoplasma	Collagen vascular disease
Rickettsia	Sarcoidosis
Fungi	Kawasaki disease (? infectious)
Cryptococcus	
Candida	

Table 84.11. Aseptic Meningitis Syndrome

Aseptic meningitis occurs throughout the year. Because the incidence of enteroviral infections, which are responsible for a large number of the cases, peaks in the summer in temperate regions, outbreaks of aseptic meningitis are more often seen in the warm months.

Pathophysiology

The multiple causes of aseptic meningitis syndrome produce inflammation of the meninges by different mechanisms. Even among the viral infections, the pathogenesis varies considerably. Some viruses lead to an immune reaction in the CNS, whereas others invade the neural tissue directly. Access to the meninges is usually hematogenous but may be achieved by ascension along peripheral nerves.

Clinical Manifestations

The signs and symptoms of aseptic meningitis resemble those of bacterial infections of the CNS but are not usually as severe. The infant shows only lethargy and irritability, whereas the older child complains of a headache and stiff neck. Vomiting may occur and may be persistent. There is often a history of a concomitant upper respiratory or GI viral illness. Fever usually occurs but often hovers around 38.5°C (101.2°F). The infant may appear toxic, but the older child may remain remarkably well. Nuchal rigidity in a patient who is alert and conversant suggests aseptic, rather than bacterial, meningitis. Shining a flashlight in the eyes often elicits photophobia. The fontanel of the infant generally maintains a normal configuration but may bulge rarely. Aside from occasional positive Kernig and Brudzinski signs, the neurologic examination often shows no abnormalities. An altered level of consciousness or focal neurologic deficit points to meningoencephalitis rather than aseptic meningitis (see Chapter 83).

Management

In addition to the routine CSF studies, children with aseptic meningitis usually require a CBC, electrolytes, and a BUN. Most patients need no further tests, but in atypical situations, consideration should always be given to nonviral causes that may mandate additional diagnostic steps or specific therapy. If tuberculosis is suspected based on family contacts, a low CSF glucose, or pulmonary findings, a Mantoux test and chest radiograph are useful for confirmation. In endemic areas, particularly in association with erythema migrans, serologic studies for Lyme disease and antibiotic therapy may be indicated. A computed tomography (CT) scan provides essential information about patients with symptoms or signs of a parameningeal infection or CNS tumors and hemorrhages. Immunosuppressed patients develop infections with a wide variety of unusual bacteria, fungi, and parasites that can be identified in many cases with appropriate examination and culture of the CSF (India ink and acid-fast stains, cryptococcal antigen testing, fungal and mycobacterial cultures). When examining infants, the physician must remain alert to the possibility of a herpetic infection and consider obtaining a PCR for herpes simplex virus.

Therapy for the common viral infections does not currently extend beyond supportive care. Dehydration from prolonged emesis may necessitate IV fluid administration. After any deficit has been corrected, the rate should be set to provide 75 to 100% of the daily maintenance requirement to avoid overhydration and the possible aggravation of cerebral edema in the child who develops an encephalitic component.

Because the CSF findings in aseptic meningitis occasionally overlap those in bacterial infections, hospital admission is usually warranted until the CSF culture results are available. However, the experienced clinician may choose to follow the older child as an outpatient if the family is reliable and nonviral causes (e.g., tuberculosis, cryptococcosis) have been excluded. Generally, to qualify for discharge with aseptic meningitis, a patient must have all CSF parameters pointing away from bacterial infection: less than 500 cells mm^3, less than 50% polymorphonuclear leukocytes, protein less than 100 mg/dL, and glucose greater than 30 mg/dL.

Upper Respiratory Tract Infections

Infections in children involve the upper respiratory tract more often than any other region of the body. Included in this category are nasopharyngitis (common cold), stomatitis, pharyngitis, sinusitis, otitis, peritonsillar abscess, retropharyngeal and lateral pharyngeal abscesses, laryngotracheobronchitis (croup), and epiglottitis. Because the most common causative organism varies between sites, infection in this area demands a specific anatomic diagnosis if the physician is to proceed with the appropriate diagnostic evaluation and therapy.

Nasopharyngitis

Nasopharyngitis (URI), or the common cold, is a viral illness of the upper respiratory tract in children. The most commonly isolated organisms are the rhinoviruses and coronaviruses. Prospective family studies have shown that five or six episodes occur yearly during childhood. The illness is characterized by a fever of less than 39°C (102.2°F) and coryza. There may be a mild conjunctivitis and infection of the pharynx. Although the tympanic membranes may show a slightly dull appearance and decreased mobility, the characteristic features of acute purulent OM (erythema, loss of the landmarks, and bulging) are absent. Therapy is limited to a recommendation for rest, adequate hydration, saline nose drops, and antipyretic agents. Neither antibiotics nor antihistamine decongestant combinations prevent secondary

bacterial infections, such as acute purulent OM.

Stomatitis

Stomatitis, an injection of the mouth, is caused by herpes simplex and the coxsackieviruses at any age and by *Candida albicans* (“thrush”) in the infant (see Chapter 121 and Chapter 124) or in the immunosuppressed child. Viral infections cause vesicular lesions initially (Fig. 84.4) and ulcerations and plaques subsequently. Some coxsackieviruses may involve the hands and feet (Fig. 84.5A) as well as the mouth (coxsackievirus hand–foot–mouth syndrome), and herpetic stomatitis may be complicated by spread of infection to the digits, which is called herpetic whitlow (Fig. 84.5B). For otherwise healthy patients, treatment is limited to systemic antipyretic and analgesic drugs and the local application of topical analgesics, such as 2% viscous Xylocaine or the combination of Kaopectate and diphenhydramine. Oral acyclovir hastens the resolution of herpetic lesions in immunosuppressed patients.

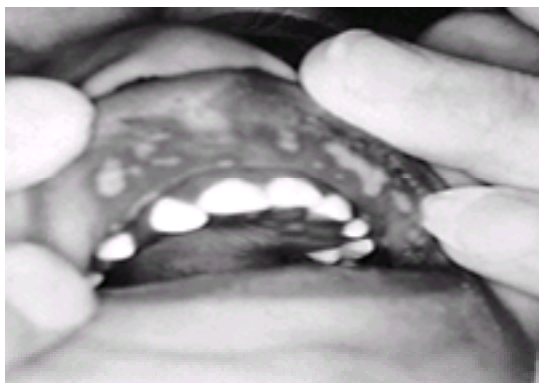

FIGURE 84.4. Lesions of herpetic stomatitis.

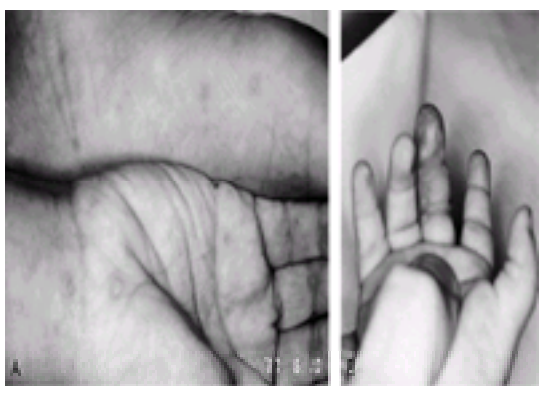

FIGURE 84.5. A. Vesicles in coxsackievirus hand–foot–mouth syndrome. **B.** Herpetic whitlow.

C. albicans produces white plaques on the mucosa that bleed if scraped. Nystatin 200,000 units four times daily leads to a prompt resolution of this condition. Although either oral ketoconazole or fluconazole are effective treatments, neither is indicated routinely for the immunocompetent host.

Pharyngitis

Pharyngitis (see Chapter 71) is an infection of the throat (including the tonsils). In the immunocompetent child, several viruses, perhaps *Mycoplasma pneumoniae* and *Chlamydia trachomatis,* and only a few bacteria cause pharyngitis. Common viral isolates include the adenoviruses, influenza viruses, enteroviruses (including coxsackievirus), parainfluenza viruses, and the Epstein-Barr virus (EBV). Although many bacteria have been reported as possible causes of pharyngitis, only three organisms have well-defined roles: group A streptococcus, *Corynebacterium diphtheriae,* and *N. gonorrhoeae.*

Pharyngitis is a common infection in children. Moffet et al. reported that 128 of 230 visits to an infirmary by youngsters of school age at a children's home were for pharyngeal infections. Group A streptococcus causes almost 50% of such infections between 5 and 15 years of age but is uncommon in the first 3 years of life. In one study of 50 children less than 3 years old with exudative pharyngitis, only 7 had illness from streptococci.

For practical purposes, isolated pharyngitis can be considered as streptococcal (bacterial) or nonstreptococcal (viral). Because the symptoms of the two types overlap, the physician can reliably distinguish the more important streptococcal infections only with the aid of the laboratory. However, certain clinical features favor a bacterial cause. Such infections more often have an abrupt onset with fever and sore throat; cough and coryza are uncommon. Examination of the pharynx shows an erythematous mucosa, often with exudate (Fig. 84.6) and petechiae on the posterior palate. In addition, the cervical lymph nodes often become enlarged and tender.

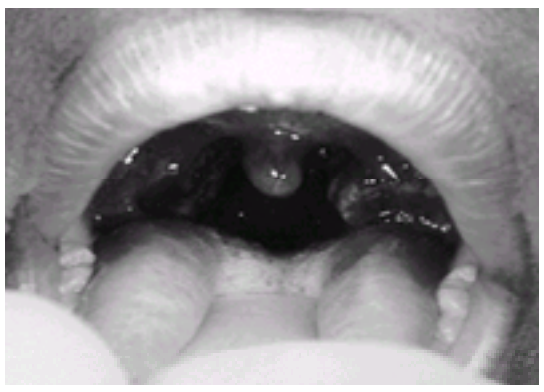

FIGURE 84.6. Examination of the pharynx in a child with streptococcal infection shows enlarged, erythematous tonsils with exudate.

Although unusual, complications may occur with bacterial pharyngitis; both suppurative and nonsuppurative sequelae can result from streptococcal infections. The latter category includes acute rheumatic fever and glomerulonephritis. Viral pharyngitis resolves spontaneously in 2 to 5 days with the exception of EBV, as discussed under Infectious Mononucleosis.

In the ED, a tonsillar swab for antigen detection by latex agglutination or preferably optical immunoassay, if available, should be obtained from children with pharyngeal inflammation and those who complain of sore throat, unless the diagnosis of a generalized viral syndrome can be confidently established on clinical grounds. If the test for antigen is positive, the infection is presumably caused by group A streptococcus and the child is treated with penicillin. Although a single injection of benzathine penicillin (600,000 units if less than 28 kg and 1.2 million units if 28 kg or more) obviates all problems with compliance, oral phenoxymethyl penicillin (250 mg/dose for children and 500 mg/dose for adolescents, given two to three times per day) provides an acceptable alternative, if prescribed for 10 days. Amoxicillin may be used in place of penicillin but offers no advantage. Erythromycin (40 mg/kg per day) is used for penicillin-allergic children; azithromycin for 5 days (10 mg/kg on the first day and 5 mg/kg on subsequent days) represents a more expensive alternative. Shorter courses of therapy, particularly with oral cephalosporins, have been shown to be effective in limited studies but cannot be enthusiastically recommended at present because of the small number of patients treated in various investigations and a lack of data on the prevention of complications. Antipyretic agents, fluids, and adequate rest should be recommended. In children with pharyngitis suggestive of streptococcal disease for which antigen detection is negative or unavailable, a throat culture is indicated. While awaiting the results of cultures, one may choose to treat presumptively children with severe pharyngitis characteristic of streptococcal disease and those unable to reliably return for follow-up. Because antibiotics shorten the course of streptococcal pharyngitis minimally, there is no reason to give these drugs hastily before confirming a bacterial cause. Institution of a liquid diet and acetaminophen provide some symptomatic relief.

Otitis Media

Background

OM refers to inflammation within the middle ear. The disease can be further classified according to the associated clinical symptoms, specific otoscopic findings, duration, and occurrence of complications. Currently, the disease is divided into two broad categories (Table 84.12): acute otitis media (AOM) and otitis media with effusion (OME). AOM (formerly called acute purulent otitis media) is caused by an acute bacterial (or occasionally viral) infection, has a sudden onset, and usually causes symptoms. On the other hand, OME (formerly called serous otitis media) is not primarily bacterial in origin, has a more gradual onset, and often remains asymptomatic. A great deal of overlap exists between these two entities, such that clinical differentiation at a single point in time may not be possible. The bacterial flora of middle ear infections varies somewhat with age. Although Gram-negative enteric bacilli and *S. aureus* cause 15 to 20% of OM during the first month of life, *S. pneumoniae, H. influenzae,* and *Moraxella catarrhalis* predominate at all ages, even in the neonate. *S. pneumoniae* can be recovered from about 40% and *H. influenzae* and *M. catarrhalis* each from 20% of children with OM between 1 month and 10 years of age. Among older children and adolescents, *H. influenzae* decreases in frequency but remains a significant pathogen. More than 90% of the *H. influenzae* that cause OM are nontypeable, and most of the remainder are type b. Most *H. influenzae* and *M. catarrhalis* are resistant to ampicillin. Among pneumococci, the incidence of penicillin resistance varies geographically and continues to evolve; at present, on average, about 25% of these organisms exhibit a high level of resistance (mean inhibitory concentration [MIC] greater than or equal to 2 µg/mL) and another 15 to 20%, an intermediate level.

Type	Duration	Bacteriology	Tympanum	Signs and Symptoms
Acute otitis media (AOM)	Days to weeks	Isolation in 70%	Erythematous or purulent, bulging	Fever (70%), earache, pulse otalgia, irritability
Otitis media with effusion (OME)	Weeks to months	Occasional isolation	Dull, retracted, fluid level	Asymptomatic, decreased hearing, fullness

Table 84.12. Classification of Otitis Media

AOM concerns the emergency physician to a far greater extent than OME. It is the most common bacterial infection in children, affecting an estimated 9 million children annually. Howie and Ploussard found at least one episode of OM in two-thirds of 2-year-old children in their practice, and one in seven children had more than six episodes. Teele et al. reported an average of 0.4 to 1.2 episodes of OM annually among children from birth to age 7 years.

OM is more common in the winter in temperate climates. This is presumably related to the higher incidence of URIs during the colder months.

Pathophysiology

Any discussion of the pathophysiology of OM provokes great controversy among pediatricians and otolaryngologists alike. However, it appears that abnormal function of the eustachian tube contributes to the development of OM in most cases. Possible mechanisms for obstruction of the eustachian tube include hypertrophied nasopharyngeal lymphoid tissue or intrinsic abnormalities of the various components of this structure. Whatever the cause, blockage impairs ventilation of the middle ear, leading to an accumulation of fluid behind the tympanic membrane. This effusion then provides a fertile environment for the proliferation of bacteria from the heavily colonized nasopharynx.

Clinical Manifestations

Studies by Howie, Paradise, Klein, and others have shown the variable spectrum of OM. An infection in the middle ear may produce no symptoms, being detected only on examination, or it may cause obvious localizing pain. In the young child, the initial manifestation is often not otologic but rather fever, irritability, or diarrhea. Children beyond the age of 3 years generally, but not invariably, complain of pain in the ear. Less common symptoms include vertigo and hearing impairment.

Fever, occurring in 25 to 35% of children with AOM, serves only to arouse suspicion of infection in the middle ear. The diagnosis rests in the usual clinical settings on the accurate interpretation of the otoscopic findings, a skill gained only by experience with the pneumatic otoscope (Fig. 84.7). If cerumen obscures the tympanic membrane, a sufficient quantity must be cleared to allow adequate visualization. Either a blunt curette or an apparatus for irrigation adequately removes such material in most cases.

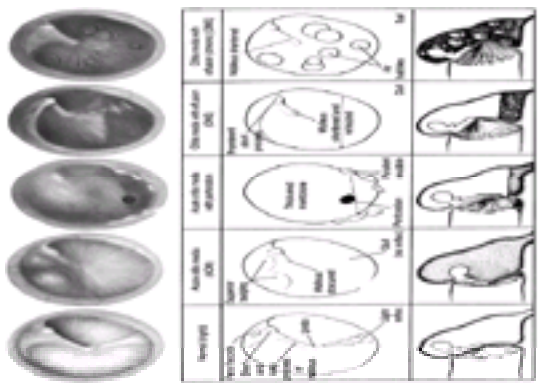

FIGURE 84.7. Appearance of the tympanic membrane in different types of otitis media.

The tympanic membrane in AOM typically bulges out at the examiner as a result of the positive pressure generated by the production of purulent material in the middle ear cavity. Although the drum is sometimes red, it more often appears yellow because of the exudate behind it. A convex contour of the drum secondary to an effusion in the child suspected of having AOM is sufficient to make this diagnosis, regardless of the color of the tympanum. The diffuse injection of the normal tympanum, which is often exaggerated by crying, should not be confused with the intense erythema of infection.

Difficulty arises in differentiating AOM from OME and in diagnosing “early” AOM, particularly in the child with a preexisting middle ear effusion. The tympanic membrane has decreased mobility in both AOM and OME; however, it is usually retracted in the latter condition. During the course of a single examination, the physician may be unable to differentiate with any certainty; in such cases, it is safest to assume a bacterial cause.

The WBC count, if obtained, usually falls within the normal range or shows a mild leukocytosis. Occasionally, a young child may have a count of 20,000 to 30,000/mm^3. Tympanocentesis, when performed as part of a research protocol yields an organism in 60 to 70% of cases. Blood cultures drawn selectively, most often from highly febrile children in the first 2 years of life, show growth of a pathogen in about 3% of cases.

Acute complications have occurred occasionally in AOM subsequent to the advent of effective antibiotics. Local suppuration may involve the mastoids and rarely leads to meningitis or brain abscess. Perforations generally heal spontaneously (see Chapter 121). A child with OM in the first year or two of life may develop dehydration from vomiting

and diarrhea associated with the infection.

Management

Uncomplicated OM in the child older than 1 month should be treated with oral antibiotic therapy on an outpatient basis. Amoxicillin (80 mg/kg per day in three divided doses) is the drug of choice in the United States. The most reasonable alternatives include 1) amoxicillin fortified with clavulanic acid in a 7:1 formulation, 45 mg/kg per day of amoxicillin in two divided doses; 2) cefuroxime axetil, or loracarbef, 30 mg/kg per day in two divided doses; and 3) ceftriaxone, 50 mg/kg intramuscularly as a single dose. Cephalosporins can be used in children with a history of penicillin allergy. Intramuscular ceftriaxone has an advantage in children with persistent vomiting and perhaps in those at high risk of occult bacteremia and also obviates the issue of compliance in high-risk social situations. Whether the course of therapy must be 10 days (except for single-dose ceftriaxone) or can be shortened is controversial. Most authorities believe that antibiotics administered for 5 days are sufficient in children older than 2 years but prefer the longer course in infants. Antihistamines and decongestants have not hastened the resolution of AOM in published studies and are thus not indicated currently.

If AOM persists during therapy with amoxicillin or recurs within 2 days of its discontinuance, ampicillin-resistant organisms emerge as likely causes of the infection. All of the alternatives to amoxicillin represent reasonable agents for a second course and no compelling data favor any particular regimen. Failure of such a second course of antibiotics to eradicate the infection necessitates a third trial of antibiotic therapy and merits consideration of a tympanocentesis for culture.

The management of OM in the first month of life has provoked controversy because of 1) the occurrence of Gram-negative enteric bacilli and *S. aureus* in middle ear infections in these children and 2) the decreased ability of the neonate to resist infection (Fig. 84.8). If a child younger than 1 month of age presents with fever or irritability and is found to have OM, admission for IV antibiotic therapy provides the safest course pending the outcome of cultures of the blood, urine, and CSF. Afebrile infants in the first month of life may be treated as outpatients with the usual oral antibiotics used for older patients and with careful follow-up.

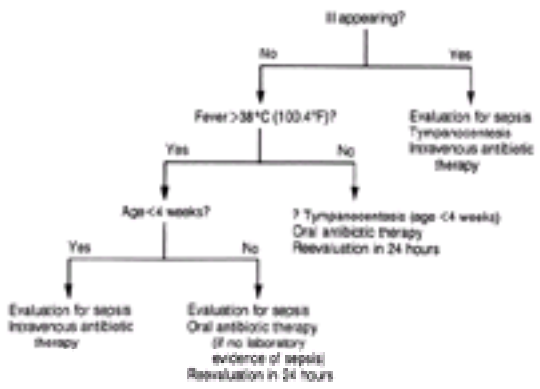

FIGURE 84.8. Diagnostic approach for the management of acute otitis media in the infant during the first month of life.

Infants between 4 and 12 weeks of age with OM can be managed as outpatients because *S. pneumoniae, H. influenzae,* and *M. catarrhalis* are the predominant organisms. However, other sources of infection, including meningitis, must be excluded in the febrile child before attributing the source of a temperature elevation to OM alone.

Otitis Externa

Otitis externa (OE), or swimmer's ear, is an infection of the auditory canal and external surface of the tympanic membrane that spares the middle ear. Multiple organisms, particularly *Pseudomonas aeruginosa,* play a role in this disease. There is usually a history of recent swimming, but occasional cases are seen in children whose only submersion occurs during normal bathing. The first symptom is itching of the ear canal. The child complains subsequently of an earache that may be unilateral or bilateral, and purulent material often drains from the ear. Fever is never present unless cellulitis or another illness is associated. Unlike AOM, pulling on the ear lobe to straighten the canal in preparation for otoscopic examination elicits marked tenderness. A cheesy white or gray–green exudate fills the canal in more than 50% of patients, often obscuring the tympanum.

OE is at times confused with AOM. Although both may cause earache (see Chapter 55), the signs and symptoms usually make an exact diagnosis possible. Occasionally, the physician cannot distinguish OE from AOM with perforation and must treat for both disorders.

Treatment consists of removing the inflammatory debris from the ear canal, eliminating pathogenic bacteria, providing symptomatic relief, and controlling predisposing factors. Usually, dry-mopping of the canal with a cotton-tipped wire applicator cleanses the canal adequately; occasionally, gentle suction is also necessary. The patient should be given commercially available otic drops and a mild analgesic, such as aspirin or acetaminophen. Acetic acid solutions (Otic-Domeboro or Vosol), the combination antibiotic–corticosteroid preparations (Cortisporin, Lidosporin), 4 drops instilled four times daily, or ofloxacin otic, 5 drops instilled twice daily, are effective. In cases with known or suspected perforations, suspensions (but not solutions) are preferred. For the occasional case of a patient with an edematous canal and thick exudate, a wick of cotton or gauze should be inserted 10 to 12 mm into the canal after cleansing to facilitate entry of the medications. All patients should be instructed to avoid swimming until cured.

Children who return without improvement after initial therapy should be examined to be certain that they have OE. If this

diagnosis is confirmed, the canal should be cleansed again and an alternate medication prescribed. The occurrence of a local cellulitis or adenitis requires the addition of an antistaphylococcal antibiotic, such as dicloxacillin (50 mg/kg per day) or cephalexin (100 mg/kg per day). Failure to respond to a second course of therapy or severe local inflammation (necrotizing OE) is an indication for referral to a specialist.

Sinusitis

Background

Sinusitis is an inflammation of the paranasal sinuses: maxillary, ethmoid, frontal, or sphenoid. The ethmoid and maxillary sinuses are present at birth, but the frontal and sphenoid do not become aerated until 4 or 7 years of age. Either an acute or a chronic infection may occur, each characterized by a different but overlapping group of symptoms. Acute sinusitis may occur in children. Among 2613 patients seen in an office practice, Breese et al. made this diagnosis in only 6 (0.23%).

Wald et al. studied the bacteriology of sinusitis in children using cultures of material obtained by antral puncture. They recovered 47 organisms from 30 children: 17, *S. pneumoniae;* 11, nontypeable *H. influenzae;* 9, *M. catarrhalis;* 2, *Streptococcus viridans;* 7, group A streptococcus; and 1, *Moraxella* species. Hamory et al. found a similar spectrum of pathogens in adults with maxillary sinusitis. Although anaerobic organisms and *S. aureus* have been reported occasionally, they do not play a role in most of these infections seen in children. *H. influenzae* type b formerly caused ethmoiditis and periorbital cellulitis but now occurs rarely.

Pathophysiology

Infection of the sinuses arises in a fashion similar to that described for AOM. Organisms ascend from the nasopharynx and cause disease if the mucosal barrier of the sinus or the normal pattern of drainage has been altered.

Clinical Manifestations

The presentation of acute sinusitis varies in some respects with the child's age. Usually, the infection follows a viral URI. Two features that distinguish sinusitis from a viral URI include persistent (longer than 10 days) and/or severe (temperature greater than 39°C [102.2°F] beyond 3 days) symptoms. Cough occurs in 75% of the patients. Unlike adolescents, young children do not often complain of a headache or facial pain. A fever is noted in about half of children with sinusitis. Nasal discharge occurs in almost all of these infections and is often the symptom that prompts a visit. The area of the face that overlies the sinus swells in 10 to 20% of the patients with maxillary disease, and periorbital or orbital edema and cellulitis even more commonly accompany ethmoiditis.

The child with chronic sinusitis complains only of chronic cough and rhinorrhea. Fever, headache, and facial pain are unusual. Often, abnormal findings are not seen on examination.

The WBC count, performed only occasionally, is normal in 60 to 80% of children with sinusitis. In 10 to 30% of these infections, transillumination of the sinuses shows a discrepancy between the two sides. The sinus radiograph is abnormal in almost every child with sinusitis (Fig. 84.9); there may be an air–fluid level, complete opacification, or mucosal thickening (greater than 4 mm). Of 60 sinuses in 30 children evaluated radiographically by Wald et al., 4 were normal, 38 showed complete opacification, 15 showed mucosal thickening, and 3 showed an air–fluid level. Hamory et al. obtained radiographs on 43 patients with 58 episodes of sinusitis. Eighteen had an air-fluid level, 18 had opacification, 12 had mucosal thickening, and 10 had no abnormalities. A CT scan is more sensitive for diagnosis than plain radiograph but is not needed in routine cases.

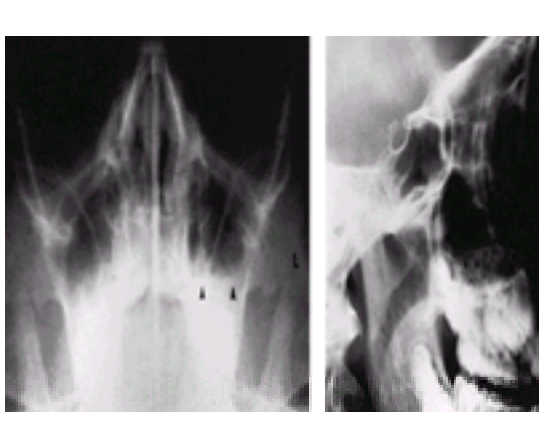

FIGURE 84.9. Anteroposterior **(A)** and lateral **(B)** radiographs show an air–fluid level in the left maxillary sinus *(arrowheads)* and mucoperiosteal thickening on the right side.

Although sinusitis usually responds to oral antibiotic therapy, serious complications occasionally result from the local spread of the suppuration. These include orbital infection, brain abscess, epidural or subdural empyema, and cavernous sinus thrombosis. Proptosis and paralysis of the extraocular muscles point to the accumulation of purulent material within the orbit. After intracranial extension, the child appears toxic and usually has a detectable neurologic deficit.

Management

Children suspected of having acute sinusitis with severe symptoms or an uncertain clinical picture should have a radiograph evaluation of their sinuses. Among this group of patients clinically believed to be at risk for an infection, any abnormality (air–fluid level, opacification, or mucosal thickening) suffices to confirm the diagnosis. Afebrile children with chronic sinusitis diagnosed on the basis of persistent nasal discharge need no laboratory or radiographic evaluation. Possible indications for antral puncture and aspiration of the sinus include 1) associated life-threatening infection, 2) immunocompromise, 3) persistent illness despite therapy, and 4) unusually severe disease. Amoxicillin (40 to 50 mg/kg per day) effectively treats the common pathogens, *S. pneumoniae* and *H. influenzae,* in most cases. Current recommendations call for antibiotic therapy for 10 days, although shorter courses of treatment are under investigation. Alternative drugs for penicillin-allergic children and those with recurrent disease are the same as for OM. Children with acute sinusitis require admission if they appear ill, have facial swelling and tenderness, or develop any complications.

Peritonsillar Abscess

A peritonsillar abscess, or “quinsy,” results from the accumulation of purulent material within the tonsillar fossa. Adolescents develop this condition more often than younger children. Group A streptococcus, various anaerobic organisms, and occasionally *S. aureus* are isolated from these lesions, which are unusual in children, in comparison to uncomplicated tonsillitis.

The complaints of trismus and difficulty in speaking separate a peritonsillar abscess from the far more common pharyngitis. The voice sounds muffled, and the child drools profusely. Both tonsils may swell, but the enlargement of one is more pronounced. Usually, the abscessed tonsil becomes sufficiently large to push the uvula to the opposite side of the pharynx, and the examiner can palpate a fluctuant mass intraorally. The WBC count is often elevated.

All children with a peritonsillar abscess should have the lesion drained in the ED or after admission to the hospital and receive treatment with antibiotics. Penicillin (100,000 units/kg per day) is usually sufficient, but the results of the culture and Gram stain of material from the infected tonsil will determine the final choice of therapy because *S. aureus* is occasionally recovered. In the unusual case of a child with respiratory compromise, aspiration of the abscess can be lifesaving. This is accomplished by using an 18-gauge needle mounted on a 10-mL syringe (see Chapter 121).

Cervical Lymphadenitis

Background

Cervical lymphadenitis is a bacterial infection of the lymph nodes in the neck. This condition must be distinguished from lymphadenopathy, an enlargement of one or more lymph nodes that occurs with viral infections or as a reaction to bacterial disease in structures that drain to the nodes.

S. aureus causes lymphadenitis in most children with an identifiable pathogen. Of 74 children with this condition, Barton and Feigin isolated *S. aureus* from 27 (36%) and group A streptococcus from 19 (26%). Other organisms that may rarely play a role include mycobacteria, *Bartonella henselae* (cat-scratch disease [CSD]), anaerobic bacteria, *Yersinia pestis* (plague), Gram-negative bacilli, *H. influenzae, Francisella tularensis,* Actinomyces, and Nocardia.

Pathophysiology

The causative organisms in cervical adenitis initially colonize the nares or pharynx or less commonly are inoculated transcutaneously. Dental abscesses may also be a source of pathogens. Whether or not they produce a local infection, the bacteria can spread to the lymph nodes in the neck. If not contained by the immune system, they proliferate within the node and evoke an inflammatory response.

Clinical Manifestations

The child with cervical lymphadenitis is usually noted to have swelling in the neck. If sufficiently old, he or she will complain of pain. Fever occurs only occasionally, more often in children less than 1 year old. The infected node may vary in size from 2 cm to more than 10 cm. Initially, it has a firm consistency, but fluctuance (Fig. 84.10) develops in about 25%. The skin overlying the node becomes erythematous, and edema may surround it.

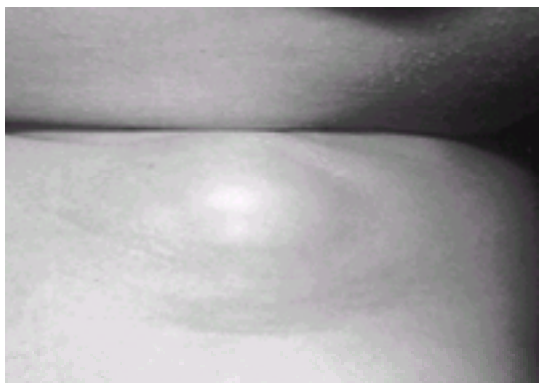

FIGURE 84.10. Lymphadenitis in the inferior cervical chain. The node appears fluctuant.

The WBC count is usually normal but may be elevated in the younger, febrile child. Aspiration of the node often identifies the organism by both Gram stain and culture, even if fluctuance is not appreciated. Children with infections from *M. tuberculosis* usually react to the standard purified protein derivative (PPD-S) skin test and have changes compatible with tuberculosis seen on chest radiograph.

Complications of bacterial adenitis are unusual. Organisms such as *S. aureus* and group A streptococcus can spread locally if unchecked. A sinus tract develops in some children infected with atypical mycobacteria. Recurrence of the infection occurs in conditions such as chronic granulomatous disease.

Management

Figure 84.11 outlines the management of the child with cervical lymphadenitis. Children with cervical adenitis who are otherwise healthy should receive an antibiotic effective against *S. aureus* and the group A streptococcus. Agents such as dicloxacillin (50 mg/kg per day) and cephalexin (50 mg/kg per day) have activity against both organisms. In more severe infections, oxacillin (150 mg/kg per day in four divided doses) can be administered intravenously. If the node is fluctuant, aspiration provides useful etiologic information and speeds the rate of resolution. All children with lymphadenitis should have a PPD skin test and should be followed until the infection subsides.

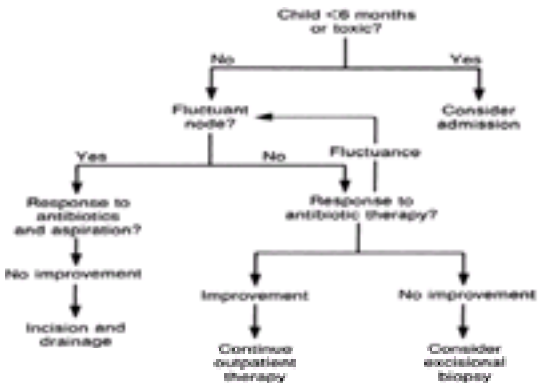

FIGURE 84.11. Diagnostic approach for the management of the child with presumed bacterial lymphadenitis.

Children less than 3 months of age and those who appear toxic or who have developed a draining sinus are best managed in the hospital. A failure to improve with oral antibiotic therapy or a positive skin test for tuberculosis necessitates subsequent hospitalization.

Retropharyngeal and Lateral Pharyngeal Abscess

A retropharyngeal abscess fills the potential space between the anterior border of the cervical vertebrae and the posterior wall of the esophagus. The usual pathogens are group A streptococcus, anaerobic organisms, and occasionally *S. aureus.* These uncommon infections occur most often in children less than 4 years of age. A lateral pharyngeal abscess occurs in the deep soft tissue space of the neck but not in the midline and is less common than a retropharyngeal infection.

The child with a retropharyngeal abscess presents with a clinical picture similar to that seen with epiglottitis, but the onset is less abrupt. Fever and a toxic appearance are common. As purulent material collects, the fluctuant mass obstructs the larynx and esophagus, leading to stridor and drooling. The abscess may cause meningismus; thus, this diagnosis should be considered in the child with nuchal rigidity but no pleocytosis in the CSF.

Although a retropharyngeal infection can rarely be seen as a midline swelling on examination of the pharynx, it is usually difficult to observe this finding in the uncooperative child. If the diagnosis is suspected and the airway is not threatened, a lateral neck radiograph or CT scan should be obtained. The radiograph shows an increase in the width of the soft tissues anterior to the vertebrae and, on occasion, an air–fluid level. Ordinarily, the width of this space is less than half that of the adjacent vertebral body if the examination is done with the neck properly extended.

A lateral pharyngeal abscess causes virtually identical symptoms to an infection in the retropharyngeal area. One important difference is that a lateral pharyngeal abscess, which is not well visualized by radiograph, requires a CT scan for diagnosis.

A retropharyngeal or lateral pharyngeal abscess poses a risk to the patency of the airway. All children with this infection should have careful monitoring in the ED and then be hospitalized in consultation with an otolaryngologist. Unless the airway is in immediate jeopardy, IV access should be secured and treatment given with either clindamycin (30 mg/kg per day in four divided doses) or a combination of penicillin (100,000 units/kg per day) and cefazolin (100 mg/kg per day), both in four divided doses. In the event of respiratory compromise, intubation or, less commonly, tracheotomy becomes necessary. Most patients require drainage, either transcutaneously with ultrasound guidance or at surgery, but a few reports indicate that high-dose antibiotics may suffice, particularly when the CT scan shows cellulitis or only a small collection of pus.

Laryngotracheobronchitis (Croup)

Background

Croup, or laryngotracheobronchitis, is a viral infection that involves the larynx and may extend into the trachea and bronchi. It is a common infection, with thousands of cases of croup to every 1 of epiglottitis among children who present to the ED. Although most children with croup are treated as outpatients, some develop more pronounced respiratory distress and require hospitalization. Hoekelman reported that 3 (1.2%) of 246 healthy term infants in a pediatric practice developed croup during their first year of life. This infection is the most common cause for stridor in the febrile child (see Chapter 72).

Parainfluenza virus can be recovered from about 60% of children with croup. Additional causes of the disease are influenza, adenoviruses, measles, and respiratory syncytial virus (RSV). Bacteria play no role.

Croup occurs more commonly in the winter months. Children between the ages of 6 months and 3 years are most commonly affected. The diagnosis of croup in a child older than 3 years should arouse the suspicion of an underlying anatomic abnormality.

Pathophysiology

The viral pathogens that eventually produce croup invade the epithelium of the pharynx initially. Spread occurs downward to the larynx and occasionally further along the respiratory tract. The infection causes endothelial damage, production of mucus, loss of ciliary function, and edema. Erythema and swelling of the vocal cords and the subglottic larynx are present. A fibrinous exudate partially occludes the lumen of the trachea.

Clinical Manifestations

Croup begins insidiously with the onset of fever and coryza. During the next 1 to 2 days, the infection spreads farther along the airway, producing signs of upper respiratory obstruction. Inspiratory stridor develops at this stage of the illness, and a barking cough is heard. The child may be unable to maintain adequate oral intake.

Although the severity of croup varies, most children appear mildly to moderately ill in contrast to the toxic patients with epiglottitis or retropharyngeal abscess. The fever usually ranges from 38° to 39°C (100.4° to 102.2°F). Tachycardia and tachypnea are evident, but the respirations rarely exceed 40 breaths/minute. Suprasternal and subcostal retractions often accompany croup. On auscultation of the chest, the examiner may hear either stridor alone in mild disease or rhonchi and wheezes with more extensive involvement of the respiratory epithelium. Cyanosis occurs only in the minority of children with severe croup.

Ancillary studies are indicated only occasionally. The WBC count is generally normal; lymphocytosis may occur as with other viral infections. The lateral and anteroposterior neck radiographs show subglottic narrowing (“steeple” sign) from soft-tissue edema in severe disease. However, most of the radiographic studies of the airway are normal or disclose only ballooning of the hypopharynx. Rather than confirm the diagnosis of croup, radiograph examination more often excludes other illness such as epiglottitis or retropharyngeal abscess. The ABG demonstrates neither hypoxia nor hypercarbia unless respiratory fatigue ensues.

Both dehydration and upper airway obstruction may complicate croup. Because of the respiratory distress and the toxicity associated with a febrile illness, the ability to maintain normal hydration will decrease in some children. Dehydration then occurs in the face of increased fluid losses through the pulmonary and cutaneous routes.

Occasionally, a child with croup develops significant upper airway obstruction. Signs suggestive of impending respiratory failure include 1) hypotonicity, 2) noticeable retractions, 3) decreased or absent inspiratory breath sounds, 4) depressed level of consciousness, 5) tachycardia out of proportion to the fever, and 6) cyanosis. Although an ABG is not needed in the evaluation of children with mild croup, this study plays a role in deciding on the therapy in more severe cases. *Respiratory failure* is defined as a partial pressure of arterial carbon dioxide (Pa CO_2) of 60 mm Hg or higher or a partial pressure of arterial oxygen (PaO_2) of less than 50 mm Hg in 100% oxygen. However, significant respiratory compromise is present in croup when the PaCO_2 rises over 45 mm Hg and the PaO_2 falls below 70 mm Hg in room air.

Management

Croup is usually apparent from the history and physical examination. Soft-tissue radiographs of the neck are needed only if the diagnosis is uncertain. Although visualization of the posterior pharynx/epiglottis is not advised routinely when epiglottitis is suspected, this examination may be performed to confirm the absence of tonsillar infection or an obviously enlarged epiglottis in cases in which one is confident that the diagnosis is croup.

Many children with croup are never taken to seek medical attention. Of those who come to the ED, most can be managed as outpatients. Clear indications for admission are dehydration and/or significant respiratory compromise. If any of the signs of respiratory failure are noted, hospitalization becomes necessary. Use of a scoring system may be helpful in deciding on disposition (Table 84.13). Neck radiographs and an ABG may be obtained in cases in which the clinical picture is not decisive. In addition, the physician should consider the social milieu of the family. Hospitalization provides the safest course for the child when the parents are unreliable caretakers or transportation to the ED for a reevaluation presents an obstacle to further treatment.

Score	Degree	Management
4	Mild	Outpatient—mist therapy
5–6	Mild to moderate	Outpatient if child improves in emergency department after mist, is older than 6 months, and has a reliable family
7–8	Moderate	Admitted—racemic epinephrine
≥9	Severe	Admitted—racemic epinephrine, oxygen, intensive care unit

Modified from Taussig LM, et al. Treatment of laryngotracheobronchitis (croup): use of intermittent positive-pressure breathing and racemic epinephrine. Am J Dis Child 1973;125:799.

[a]Any one category with a score of 3 should be considered as a severe disease.

Table 84.13. Scoring System for Assessing Severity of Croup

Mist therapy lessens the severity of croup. Water droplets penetrate to the area of inflammation in the larynx and provide moisture to the mucosa. Because the viral origin of this disease has been well established, antibiotics play no role.

Racemic epinephrine, or more recently in the United States L-epinephrine, is indicated for children with moderate to severe croup who will be hospitalized or for whom admission is being considered. The dose is 0.25 mL of racemic epinephrine, mixed with 3 to 5 mL of saline, delivered by nebulization. If a response is noted and discharge to home is contemplated, the child should be observed in the ED for at least 2 hours to be certain that the respiratory symptoms do not rebound.

Corticosteroids have long been mentioned as potential aids to the treatment of croup, but the early controlled studies on these agents failed to substantiate the early anecdotal successes. Subsequently, Leipzig et al. found dexamethasone effective in a controlled study in 1979, and a meta-analysis of the literature in 1989 supported the use of corticosteroids for hospitalized patents. More recently, controlled trials by Klassen, Schuh, and others have demonstrated that nebulized budesonide decreases the severity of illness in patients with mild to moderate croup. Budesonide has been shown to be slightly less effective than dexamethasone and to provide a slight additive effect. A single report has suggested that the response to budesonide may be equal to that of racemic epinephrine.

Until more data become available, treatment regimens will remain in flux. A reasonable approach for the present is to tailor therapy to the severity of illness. In rare cases with inadequate gas exchange, management of the airway, at times with endotracheal intubation, takes precedence; tracheal edema may make passage of a tube with the usual diameter impossible, and the physician should be prepared with one a size smaller. Patients in the ED who have concerning upper airway obstruction and a high likelihood of hospitalization will benefit from prompt administration of both racemic epinephrine by nebulization and intramuscular dexamethasone at 0.6 mg/kg. For children with moderately severe croup, when hospitalization is being considered, the response to an initial trial of mist and either intramuscular dexamethasone or nebulized budesonide can be assessed. If the response is adequate, the physician could add either a second steroid (e.g., budesonide if dexamethasone was already administered) or racemic epinephrine. Finally, most patients who are mildly ill require only instructions for home care.

Epiglottitis

Background

Epiglottitis is a life-threatening bacterial infection of the epiglottis and the surrounding structures. Recent authors have suggested that supraglottitis would be a more appropriate appellation. Rarely, thermal injury to the epiglottis may cause swelling and clinical findings similar to those seen with infection.

Before the advent of a vaccine against Hib, epiglottitis occurred with regularity in children, accounting for 1 of every 1000 pediatric admissions in the United States. It is now a rare disease in children.

Occasional cases are caused by the group A streptococcus. Although more common in the winter months, epiglottitis may occur throughout the year. The peak incidence during an era of greater prevalence fell between the ages of 3 and 7 years; however, infants and adults with epiglottitis have been well described.

Pathophysiology

The pharynx of normal children is often colonized with potentially pathogenic microorganisms such as *H. influenzae* and *S. pneumoniae.* Occasionally, these bacteria penetrate the mucosal barrier and invade the bloodstream. During the course of bacteremia, focal infection may occur at several sites, including the epiglottis and surrounding structures. Infection causes inflammatory edema, beginning on the lingual surface of the epiglottis, where the submucosa is loosely attached. The swelling progresses rapidly to involve the aryepiglottic folds, the arytenoids, and finally, the entire supraglottic larynx. Tightly bound epithelium on the vocal cords halts the spread at this level. The tremendous reduction in the caliber of the airway results in turbulent air flow on inspiration, appearing clinically as stridor.

Two possible mechanisms may explain the sudden respiratory arrest that can sometimes complicate this disease. The swollen epiglottis may be drawn into the glottis, acting like a plug to obstruct the flow of air, but this seems unlikely because the edematous, inflamed tissues of the supraglottic region become relatively tense. More likely, aspiration of oropharyngeal secretions occludes an already narrowed laryngeal inlet.

Clinical Manifestations

Epiglottitis has an abrupt onset. The duration of illness before presentation is often as short as 6 hours and rarely exceeds 24 hours. Among the 21 children reported by Greenberg and Schisgall, an average of 17 hours elapsed between the first symptom and hospital admission. The parents first note the onset of fever. Shortly thereafter, the child develops stridor and labored respirations. As the disease progresses, the supraglottic edema interferes with the ability to swallow secretions; thus, drooling is a complaint in 60 to 70% of cases. Of the children with epiglottitis, 50% complain of a sore throat. Aphonia, hoarseness, and cough are uncommon. Although both croup and epiglottitis manifest with stridor in a febrile child, the examiner can usually differentiate these two illnesses on the basis of the clinical features (Table 84.14).

	Epiglottitis	Croup
Anatomy	Supraglottic	Subglottic
Etiology	Bacterial: *H. influenzae*	Viral: parainfluenza
Age range	3–7 yr, adults	0.5–3 yr
Onset	6–24 hr	24–72 hr
Toxicity	Marked	Mild to moderate
Drooling	Frequent	Absent
Cough	Unusual	Frequent
Hoarseness	Unusual	Frequent
White blood cell count	Leukocytosis	Normal

Table 84.14. Epiglottitis and Croup: A Comparison

The anxious appearance of most children with epiglottitis strikes the examiner immediately (Fig. 84.12). To maximize air entry, these children assume a sitting position with their jaws thrust forward. Cyanosis may occur in the later stages of the illness. The temperature, almost always elevated, often reaches a level of 40°C (104°F). Tachycardia is a constant feature. Although the patients are universally tachypneic, the respiratory rate rarely exceeds 40 breaths/minute. Stridor can be heard without a stethoscope, but auscultation of the lungs reveals no other adventitious sounds. Marked retractions are seen, predominantly involving the suprasternal and subcostal musculature.

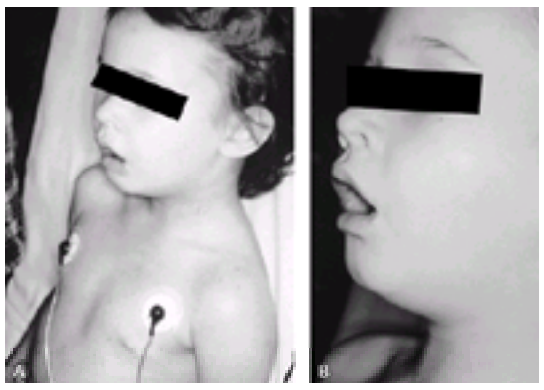

FIGURE 84.12. A 3-year-old girl with epiglottitis has an anxious appearance, assumes the “sniffing” position **(B)**, and prefers to remain sitting **(A).**

As discussed under Management, rigorous attempts to visualize the epiglottis are hazardous and should be avoided in the child with suspected epiglottitis. However, the examiner may view the pharynx without the use of a tongue depressor. The mucosa is seen to be erythematous, and pooled secretions are present in about half the children. Occasionally, a swollen, cherry red epiglottis (Fig. 84.13) protrudes above the base of the tongue and is visible without instrumentation.

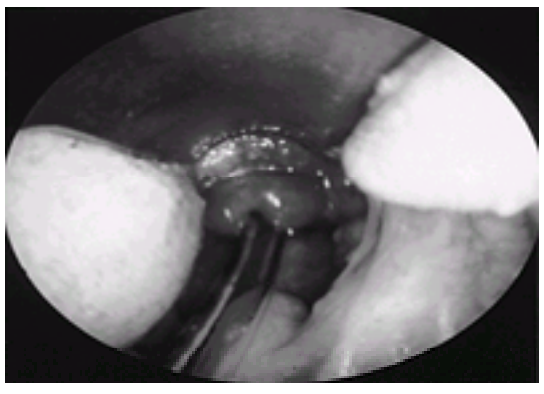

FIGURE 84.13. A swollen, erythematous epiglottis after endotracheal intubation of a child with epiglottitis.

Collection of laboratory specimens is usually delayed until the airway has been secured. The WBC count is elevated in most children with epiglottitis. As in other diseases that result from bacteremia with *H. influenzae,* a leukocytosis in the range of 15,000 to 25,000/mm^3 and a shift to the left occur in response to the infection. Culture of the blood yields a pathogen in 80 to 90% of cases, and of the epiglottis, following the placement of an airway, in about 50% of cases.

A lateral neck radiograph is pathognomonic of epiglottitis. There are three characteristic features: 1) a swollen epiglottis, 2) thickened aryepiglottic folds, and 3) obliteration of the vallecula (Fig. 84.14). The normal epiglottis has a thin, curved silhouette that has been likened to a bent finger, convex on one side and concave on the other. As a result of inflammatory edema from infection, it swells and assumes a configuration that is convex on both sides. This has been called the “thumb sign.” The airway below the level of the vocal cords appears normal on the lateral neck radiograph of a child with epiglottitis.

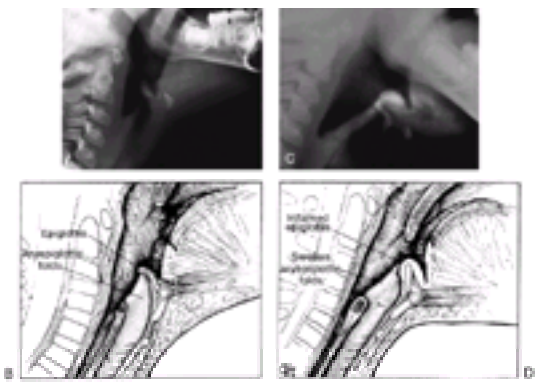

FIGURE 84.14. Appearance of the lateral neck region in a normal child (**A** and **B**) and a child with epiglottitis (**C** and **D**).

The most serious complication of epiglottitis is sudden respiratory obstruction. This may occur unpredictably at any point in the illness, before seeking medical attention, in the ED, or after hospitalization. Although a child with minimal respiratory distress may occasionally have a total obstruction, marked retractions and labored breathing should serve as a warning of an impending airway catastrophe. An additional complication of this illness is extraepiglottic spread of the infection. During the course of the bacteremia, seeding may involve the meninges, lungs, pericardium, synovial membranes, and soft tissues. Thus, the initial examination should attempt to elicit signs of infection at these additional sites.

Management

When a child is suspected of having epiglottitis, the thrust of the management plan is to make a definitive diagnosis and institute therapy before the onset of airway obstruction. The major pitfall in this process is the vigorous examination of the posterior pharynx without having considered the possibility of supraglottic infection. Such manipulation rarely may initiate laryngeal obstruction in a small number of children with epiglottitis.

The initial steps in management are based on the degree of respiratory distress and the likelihood of epiglottitis, as judged from the clinical features (Fig. 84.15). Some children with epiglottitis have total or nearly total airway obstruction as the initial presentation of their disease. In this situation, treatment precedes any diagnostic evaluation and steps to maintain an adequate exchange of air are taken (see Chapter 1 and Chapter 5).

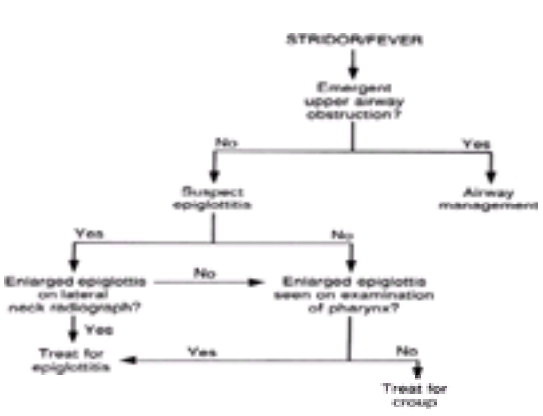

FIGURE 84.15. Diagnostic approach to the child with suspected epiglottitis.

The majority of children, however, manifest lesser degrees of stridor and respiratory compromise with fever. The clinician must decide whether the constellation of historical and physical features points to croup or epiglottitis. In most children with stridor, the history will favor croup, which is the more common of the two diseases. The child will not appear toxic or show signs of air hunger. In such situations, a lateral neck radiograph is not indicated. Rather, the pharynx may be visualized directly with a tongue depressor to confirm the absence of a swollen, inflamed epiglottis.

When the findings weigh in favor of epiglottitis, however, further examination should be postponed and immediate

preparation should be made for the insertion of an artificial airway; this includes collecting the necessary equipment and summoning additional personnel as needed. Anesthesiologists and otorhinolaryngologists alike, if available, should be involved in the care of children with epiglottitis, in addition to the staff in the ED. Following the appropriate preparations, a physician should accompany the child to the radiology department for a lateral neck radiograph, or a portable radiograph may be obtained. An IV infusion using a plastic cannula may be started in the cooperative patient. However, if the child becomes agitated or the procedure lengthy, the radiograph must be obtained quickly, assuming the airway has not been compromised, rather than persisting with attempts to gain IV access. The lateral neck radiograph either confirms or disproves the clinical diagnosis. If epiglottitis is verified radiographically, a skilled physician next performs endotracheal intubation, most often in the operating suite. If intubation is not possible, a surgical approach to the airway is necessary (Table 84.15).

Ensure adequate ventilation.	Endotracheal intubation (or tracheostomy)
Gain peripheral venous access, if tolerated by child.	Defer laboratory studies until airway is secured.

Table 84.15. Immediate Management of Epiglottitis

A review of the mortality statistics in epiglottitis emphasizes the importance of an artificial airway in the management of this illness. Rapkin described a fatal outcome in 20% of the children treated with antibiotics and observation alone. In 1978, Cantrell et al. summarized 749 cases of epiglottitis. The mortality varied with the method of airway management as follows: tracheostomy, 3 deaths among 348 children (0.86%); endotracheal intubation, 2 in 216 (0.92%); no artificial airway, 13 in 214 (6.1%).

Ceftriaxone (100 mg/kg per day in one or two divided doses) and cefotaxime (200 mg/kg per day in four divided doses) serve as single-drug alternatives and are useful particularly for patients allergic to penicillins. Chloramphenicol (100 mg/kg per day in four divided doses) also provides effective therapy. Steroids have not been shown to play a role in epiglottitis.

Bacterial Tracheitis

Infections of the trachea, presumed to be bacterial, were originally described in the period 1920 to 1940. However, the existence of bacterial tracheitis (or membranous tracheitis) was not addressed again until 1979, and some experts still question whether this is a real entity or merely a bacterial colonization of the trachea in a child with viral croup. Thus, bacterial tracheitis is, at best, an unusual infection during childhood. The putative etiologic agents are *S. aureus* and *H. influenzae,* which occur rarely at this point in time. Patients may range from infants to young children.

Clinical Manifestations

Published reports indicate that the signs and symptoms of bacterial tracheitis mimic those of acute epiglottitis but with a somewhat slower onset. The fever is usually greater than 39°C (102.2°F), and the patients are stridorous. Toxicity and respiratory distress occur as a rule. On radiograph, there is tracheal narrowing, and a pseudomembrane may be visible within the tracheal lumen; the supraglottic area is normal.

Management

Children with bacterial tracheitis are often diagnosed initially as having severe viral croup or epiglottitis. Their management is as outlined for these conditions. The first priority is to secure an adequate airway. If bacterial tracheitis is suspected on the basis of a lateral neck radiograph or the findings at laryngoscopy, antibiotic therapy should be initiated with ceftriaxone (100 mg/kg per day in one or two divided doses), ampicillin–sulbactam (200 mg/kg per day of ampicillin in four divided doses), or the combination of oxacillin (150 mg/kg per day) and chloramphenicol (100 mg/kg per day) in four divided doses. Admission to an intensive care unit is essential.

Lower Respiratory Tract Infections

The most common lower respiratory tract infections in children include bronchiolitis and pneumonia, which may be caused by various bacteria or viruses, *C. trachomatis,* or *M. pneumoniae.* Approximately 1 in 50 children in the United States has pneumonia annually. In a 12-year study of approximately 125,000 patients enrolled in the Group Health Cooperative of Puget Sound, the incidence of childhood pneumonia from all causes averaged 19 per 1000 per year. Occasional episodes of pertussis and pulmonary tuberculosis are also seen. Recently described, Hantavirus pneumonia occurs in patients with exposure to rodents and is particularly severe.

Pneumonia is an inflammation of the lung tissue that may follow a noninfectious or an infectious insult. In the ED, the febrile child with an acute onset of pneumonia almost always has an infection. The causative organisms in pneumonia vary according to the age of the child (Table 84.16). Although viral agents account for 60 to 90% of pneumonia, bacteria,

particularly *S. pneumoniae,* play a major role. *M. pneumoniae* increases in frequency after puberty. Unusual causes of pneumonia in the immunocompetent child include *Legionella pneumophilia* (Legionnaires' disease), *M. tuberculosis,* Hantavirus, rickettsia (Q fever), fungi, and protozoa. Children with neoplasms, HIV, and other forms of immunocompromise show susceptibility to a variety of unusual pathogens, including *Pneumocystis carinii* (see Chapter 85 and Chapter 100).

Age	Infecting Organism
2 wk	Bacteria
	Group B streptococcus
	Gram-negative bacilli
	Viruses
2 wk–2 mo	Chlamydia
	Viruses
	Bacteria
	S. pneumoniae
	S. aureus
	H. influenzae
2 mo–3 yr	Viruses
	Bacteria
	S. pneumoniae
	S. aureus
	H. influenzae
3 yr–12 yr	Viruses
	Bacteria
	S. pneumoniae
	M. pneumoniae
13 yr–19 yr	Viruses
	Bacteria
	S. pneumoniae
	M. pneumoniae

Table 84.16. Lower Respiratory Tract Infections

Bacterial Pneumonia

Background

Bacterial pneumonia is an inflammation of the pulmonary parenchyma caused by a bacterial pathogen. In the first weeks of life, the group B streptococcus and Gram-negative bacilli cause most such infections (Table 84.16). Between 2 weeks and 2 months of age, viruses and *Chlamydia* are most common, and viruses remain the most common isolates throughout childhood. Among the bacteria, *S. pneumoniae* predominates at every age beyond the newborn period. *H. influenzae* formerly ranked second to pneumococcus in children 2 months to 3 years of age but now occurs rarely. *S. aureus* causes a severe but uncommon pneumonia in young children; 60% of these infections occur in the first year of life. Group A streptococcus is also uncommon, *N. meningitidis* has been described rarely, and anaerobic bacteria play a role primarily following aspiration.

Definitive studies on the relative frequency of the various pathogens have not been performed in a randomly selected outpatient population of children. Because an organism is not usually recovered from the blood, establishing an etiologic diagnosis requires recovery of the pathogen from either pleural fluid or the pulmonary parenchyma. However, pleural effusion accompanies only a minority of bacterial pneumonias, and a percutaneous or transtracheal aspiration of the lung, although safe, cannot be justified on children who are sufficiently well to be managed as outpatients. Thus, the data collected on hospitalized children or those with more severe infections must be extrapolated to estimate the spectrum of pathogens in uncomplicated bacterial pneumonia. Also, antigen testing may be suggestive on urine or serum specimens.

Pathophysiology

In most pneumonias, the pathophysiology remains unknown. Pathogens reach the lung, either by hematogenous dissemination or by aspiration. In *H. influenzae,* type b, pneumonia, the organism can be recovered from the bloodstream in 90% of children, often 1 to 2 days before the appearance of the infiltrate. This suggests that bacteremia precedes the pulmonary infection. However, bacteremia is found in only 5 to 10% of pulmonary infections with *S. pneumoniae* at the time of diagnosis. Thus, aspiration must play a greater role in the pathogenesis of infections with this organism, or else the preceding bacteremia resolves before the development of pneumonia.

Following invasion of the pulmonary tissue by bacteria, an acute inflammatory reaction ensues. There is an exudation of fluid and polymorphonuclear leukocytes, followed by the deposition of fibrin. Several days later, macrophages appear in the alveoli. The accumulation of fluid in a lobe of the lung leads to the characteristic lobar consolidation seen on the chest radiograph.

Clinical Manifestations

Bacterial pneumonia generally has an abrupt onset with fever, often accompanied by chills. A cough is a common but nonspecific complaint. The young child reacts to bacterial infection in the chest with lethargy and/or a decreased appetite. Occasionally, pleuritic involvement produces pain with respiratory effort.

The observation of the child at rest before the examination often provides the key to the diagnosis of pneumonia. Tachypnea out of proportion to the fever is rarely the only sign, particularly in the first year of life. The infant who breathes at a normal rate, however, seldom has a bacterial infection of the lung. A hasty effort at auscultation that disturbs the quiet infant obscures this finding.

Fever is almost universally present, ranging from 38.5° to 41°C (101.2° to 105.8°F). Grunting respirations in a young child should arouse a strong suspicion of pneumonia. Localized findings, more often seen in the child older than 1 year, include inspiratory rales, decreased breath sounds (sometimes the only abnormality), and less often, dullness to percussion. Gastric dilation may accompany pneumonia; occasionally, the abdominal findings in pulmonary infections mimic appendicitis. With upper lobe pneumonia, the pain may radiate to the neck, causing meningismus; the diagnosis of

pneumonia must therefore be considered in the child with nuchal rigidity and normal CSF.

In the ED, a chest radiograph often assists in the management of a child suspected of having bacterial pneumonia. Although a patient who is dehydrated with pneumonia occasionally may not have an infiltrate, the radiographic evaluation confirms or denies the diagnosis of bacterial pneumonia in most cases. This is important in a clinical setting not conducive to continuity of care. In addition, the radiograph may provide information on the disease process. A lobar consolidation is assumed to be of bacterial origin, needing treatment with antibiotics (Fig. 84.16), whereas a minimal, diffuse interstitial infiltrate in a previously healthy toddler suggests a viral infection that can be managed with symptomatic therapy or, in an adolescent, *M. pneumoniae,* calling for treatment with erythromycin or another macrolide. Bilateral involvement, pleural effusion, and pneumatoceles point to more severe disease.

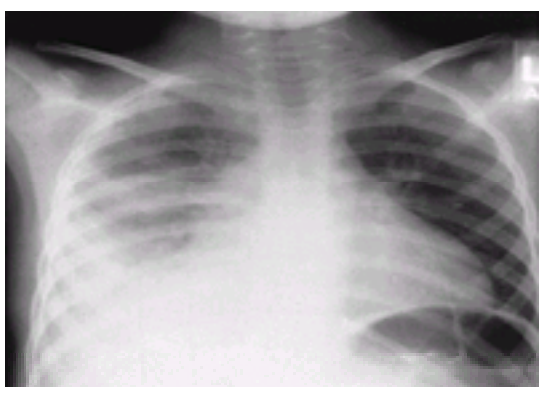

FIGURE 84.16. Radiograph showing lobar consolidation and pleural effusion in a child with bacterial pneumonia.

Further laboratory studies are obtained only on specific indications. A WBC count may be helpful in differentiating viral from bacterial disease or in assessing the likelihood of bacteremia in the young child; the count often exceeds 15,000/mm^3 and occasionally rises above 30,000/mm^3 with bacterial invasion of the pulmonary parenchyma or the bloodstream. Levels of CRP correlate with the bacteremia and lobar infiltrates more closely than the WBC count; however, this test is less readily available than the WBC count.

The most common complication of pneumonia is dehydration, particularly in young children. Electrolytes and a BUN are useful in assessing the degree of fluid loss in a child who appears ill or exhibits dry skin and/or mucosa. Rarely, extensive pulmonary involvement compromises ventilation, leading to respiratory failure. ABGs are indicated for any child with significant respiratory distress or an oxygen saturation below 90%. A pleural effusion accumulates in most infections with *S. aureus* and *H. influenzae,* less often with *S. pneumoniae.* Bacteremia may result in additional foci of infection, including meningitis, pericarditis, epiglottitis, and septic arthritis.

Management

Most healthy children with pneumonia respond to outpatient antibiotic therapy. Because most of the infections are caused by *S. pneumoniae,* amoxicillin (50 mg/kg per day given orally in three divided doses) has been the mainstay of therapy. Ceftriaxone (50 mg/kg) may be administered intramuscularly at the time of diagnosis, especially if there is any concern about oral intake during the first 24 hours. Alternatively, macrolides, including erythromycin (40 mg/kg per day in four divided doses) or azithromycin (10 mg/kg as a single dose on the first day and 5 mg/kg as a single dose on days 2 to 5), may be used in penicillin-allergic children or when mycoplasmal infection is suspected on the basis of age or radiographic findings.

Supportive therapy includes antipyretics and adequate hydration. Antitussives have no place in the treatment of pneumonia. Every child should return within 24 to 48 hours for a second evaluation; patients who do not clinically improve and become afebrile should be evaluated carefully for admission to the hospital.

Any child who appears to be toxic (on the basis of the physician's clinical judgment) or is immunocompromised should be hospitalized. Firmer, but not unarguable, indications for admission are listed in Table 84.17. The child less than 1 year of age does not tolerate bacterial pneumonia as well as those who are older. In addition, potentially serious infections with *S. aureus* and *H. influenzae* occur more often in the first year of life. The radiographic detection of a pleural effusion or pneumatocele also suggests a pathogen other than *S. pneumoniae.* Effusions should be cultured by thoracentesis (see Procedures, Section VII), which requires subsequent observation of the child in the hospital. Although a persistent elevation of the temperature is well described in children with pneumococcal pneumonia that subsequently respond to penicillin, failure of the fever to defervesce within 24 to 48 hours after the administration of antibiotics suggests a greater likelihood of more virulent pathogens.

Age <1 year (lobar infiltrate)	Failure to respond to antibiotic therapy within
Respiratory compromise	24–48 hours
Pleural effusion	Dehydration
Pneumatocele	

Table 84.17. Indications for Admission

Viral Pneumonia

Background

A number of viruses are able to infect the lungs of children and adolescents. Respiratory syncytial and parainfluenza viruses are the most common isolates, particularly in the first year of life. Other viruses, including influenza, adenoviruses, enteroviruses, rhinoviruses, measles, varicella, rubella, herpes simplex, cytomegalovirus, EBV, and hantaviruses, can cause pneumonia. Pulmonary disease complicates upper airway infections with influenza, varicella, and EBV more often in the adolescent. The annual incidence of viral pneumonia peaks in the first 5 years of life at 40 per 1000, and then declines with advancing age.

Pathophysiology

Most viruses that cause pneumonia initially invade the epithelium of the upper respiratory tract and spread locally to the lungs. The infection evokes an inflammatory response that consists primarily of mononuclear cells. After infection, the epithelial cells slough into the airway and obstruct the bronchi, producing the hyperinflation characteristically seen on chest radiograph.

A few viruses may reach the lungs by hematogenous dissemination. These include measles, varicella, rubella, cytomegalovirus, herpes simplex, and EBV.

Clinical Manifestations

Viral pneumonia generally has its onset over a 2- to 4-day period, being more gradual than with bacterial infection. Cough, coryza, and low-grade fever commonly occur. Particularly with RSV infections in the first 3 months of life, an apneic spell may be the first sign to draw attention to the illness.

Fever in viral pneumonia is usually lower than 39°C (102.2°F). As with bacterial infections, tachypnea in the undisturbed child may be the only physical finding. Rales are often audible diffusely throughout the chest, and wheezing may also be present. With more severe disease, the child shows signs of respiratory failure: grunting, cyanosis, and changes in mental status.

The WBC count varies widely in viral pneumonia. Although leukocytosis over 15,000/mm ^{3} may occur in some cases, such elevated counts should arouse suspicion of bacterial disease.

The radiographic examination provides useful clues to the type of pathogen that causes a pneumonia but can never confirm a viral infection or rule out a bacterial cause. Most typically, the radiograph in a child with viral pneumonia shows bilateral air trapping and peribronchial thickening. A diffuse increase in the interstitial markings is also commonly seen. However, the findings can vary from barely detectable increases in volume to segmental infiltrates. Decubitus films occasionally detect small effusions. Because of the limitations in obtaining reliable cultures for bacteria, it is safest to presume a bacterial cause in the child with clinical evidence of pneumonia and a lobar infiltrate, a pleural effusion, a temperature over 39°C (102.2°F), or signs of clinical toxicity. Particularly in a dehydrated child, the chest radiograph may fail to show a lobar consolidation early in the course of a bacterial pneumonia.

Most viral pneumonias resolve without specific therapy. Potential complications include dehydration, apnea, and local progression of the infection. Apnea may occur in the first 3 months of life.

Management

The physician must attempt to make an etiologic diagnosis in pneumonia on the basis of the clinical and radiographic findings without the benefit of definitive laboratory tests. A WBC count should be obtained if there is uncertainty about the likely cause. In such cases, a leukocytosis over 15,000/mm ^{3} would weigh against a viral infection.

If a viral pneumonia is strongly suspected, no specific therapy need be given. An example of such a situation would be a well-hydrated 5-year-old child with a gradual onset of cough, a temperature of 38°C (100.4°F), scattered bilateral rales, WBC count of 8000/mm^3 with predominantly lymphocytes, and the finding of hyperaeration on chest radiograph. Treatment in this case could be limited to antipyresis and hydration with a follow-up visit in 24 hours. Because the infant less than 3 months of age may become apneic during the course of viral pneumonia, these young children may benefit

from observation in the hospital.

Mycoplasmal Pneumonia

Background

M. pneumoniae is one of the most common causes of pneumonia among children older than 5 years. In younger children, infections with this organism are often limited to the upper respiratory tract or, occasionally, to the bronchial tree, although one study has implicated this organism as a common cause of pneumonia in children as young as 18 months. By the end of adolescence, 90% of the population has antibodies to *M. pneumoniae.*

Pathophysiology

The initial infection with M*. pneumoniae* occurs on the surface of the respiratory epithelium. Destruction of these cells causes them to slough into the lumen of the bronchi. The infection evokes an inflammatory response, primarily by mononuclear leukocytes.

Clinical Manifestations

Pneumonia caused by *M. pneumoniae* usually begins insidiously with fever and malaise. After 3 to 5 days, the child develops a nonproductive cough, hoarseness, sore throat, and in one-quarter of cases, chest pain. Fever is almost invariably present and may reach a level of 40°C (104°F). Children seldom develop much respiratory distress, with the exception of those who are less than 5 years old or also have sickle cell anemia or an immunodeficiency. Rales are heard in 75% of these infections, often bilaterally. The pharynx may appear inflamed, and some investigators have noted ear infections, particularly bullous myringitis, in association with pneumonia caused by *M. pneumoniae.* In 10% of patients, a maculopapular or, less often, a vesicular rash occurs; rarely, erythema multiforme, urticaria, or petechiae are seen.

The total WBC count is often normal in infections with this pathogen. A cold agglutinin titer of 32 or higher is found in most patients with lobar infiltrates from an *M. pneumoniae* infection but may also occur, although less often, with viral and bacterial illnesses. The organism may be recovered by culture, and specific diagnosis is possible with measurement of antibody titers in acute and convalescent sera; however, these procedures require 1 to 3 weeks and are not readily available. The radiographic findings show considerable variation. Between 10 and 25% of children will have lobar consolidation. Scattered segmental infiltrates, interstitial disease, and combinations of all these patterns may be seen. Pleural effusions occur in 5% of cases.

Numerous complications are described in association with *M. pneumoniae* infections, but they occur rarely. These include hemolytic anemia, arthritis, encephalitis, meningitis, and neuropathy.

Management

The diagnosis of mycoplasmal pneumonia is presumptively based on the clinical and radiographic findings and, in some cases, on the cold agglutinin titer. An older child or adolescent with the gradual onset of a mild bilateral pneumonia should be treated for this infection. On the other hand, a lobar infiltrate in a 5-year-old child usually is assumed to be of bacterial origin regardless of the level of the cold agglutinins. The results of cultures and specific serologic assays entail too great a delay to be useful to the clinician in the ED. Erythromycin (40 mg/kg per day) provides effective therapy for *M. pneumoniae* infections. The response is more pronounced in the older child with a lobar disease than in the younger child with a diffuse infiltrate.

Chlamydial Pneumonia

Background

Three species of *Chlamydia* cause pneumonia: *Chlamydia psittaci, Chlamydia trachomatis,* and *Chlamydia pneumoniae* (TWAR). Psittacosis, a severe pneumonia caused by *C. psittaci,* is rare but should be suspected in patients with unusual avian exposures. *C. trachomatis* is the most commonly recovered pathogen from children with afebrile pneumonias between 4 and 12 weeks of age. Identified as a new species in 1989, *C. pneumoniae* causes pneumonia primarily in children older than 5 years. In a study from Seattle, the attack rate for this agent was approximately 0.5 to 1 case per 1000 children (5 to 14 years old) per year, compared with 4 to 5 cases per 1000 for *M. pneumoniae*.

Pathophysiology

Among infants born to pregnant women with vaginal colonization by *C. trachomatis,* one-third to one-half acquire the organism. These infants are at risk for the subsequent development of pneumonitis. *C. pneumoniae* spread within families, day-care centers, and schools. Pathologic examination in chlamydial pneumonitis shows a mononuclear consolidation with occasional eosinophils and neutrophils and marked necrotic changes in the bronchioles.

Clinical Manifestations

Infancy. Infants with chlamydial pneumonia usually have a staccato cough that may resemble the paroxysms seen in pertussis but is usually less prolonged. In 50% of cases, conjunctivitis precedes the onset of respiratory symptoms. Pneumonia with this organism only rarely produces a fever. Mild retractions, hyperresonance, and diffuse rales are noted on examination of the chest. Hyperaeration of the lungs depresses the liver, allowing the edge to be palpated 1 to 2 mm

below the right costal margin.

Although the WBC count is usually in the normal range, the eosinophil count rises slightly (400/mm ^{3}, or 5 to 10%) in 75% of these patients. Elevated immunoglobulin levels, although nonspecific, often occur with chlamydial infections, but seldom with viral illnesses. Mild hypoxemia is common. The chest radiograph shows hyperaeration of the lungs and a diffuse increase in the interstitial markings. Lobar consolidations and pleural diffusions are not seen.

Although usually a mild illness, chlamydial pneumonia may be complicated by the occurrence of mucous plugging of the bronchi, apnea, and severe impairment of oxygenation. It is impossible to predict which infants with an initially mild course will have a stormy one.

Childhood. The spectrum of infection ranges from asymptomatic to severe. Adolescents are more likely to have signs of pneumonia than children, who may have clinical findings confined to the upper respiratory tract. Pneumonia is often preceded by sore throat and hoarseness, usually with a brief fever. By the time pneumonia has developed, the fever often resolves. Patients usually have a cough and scattered rales on auscultation. As for the clinical syndrome, the chest radiograph picture resembles that seen with *M. pneumoniae,* consisting of subsegmental lesions rather than lobar consolidation. Leukocytosis is not seen. No specific diagnostic testing is routinely available.

Management

Because of the difficulty in making a definitive etiologic diagnosis and the potential for complications, most young infants with presumed chlamydial pneumonia should be admitted to the hospital. Erythromycin (40 mg/kg per day) may shorten the course and should be given. *C. pneumoniae* infections in older children respond to therapy with macrolide antibiotics, including erythromycin (40 mg/kg per day) or azithromycin (10 mg/kg on day 1 and 5 mg/kg on days 2 through 5).

Bronchiolitis

Background

Bronchiolitis is a pulmonary infection of young children characterized by wheezing. RSV causes most of these illnesses, but other viruses, particularly parainfluenza, are isolated occasionally. In addition, *M. pneumoniae* has been reported as a rare cause of bronchiolitis.

The epidemiology of bronchiolitis primarily follows the pattern of its principal pathogen, RSV. Most of these infections occur in the winter and affect children between 2 and 8 months of age. Although some authorities do not accept the diagnosis of bronchiolitis after the age of 1 year, others believe that the disease occurs until the second birthday.

Pathophysiology

RSV, the most common cause of bronchiolitis, invades the epithelial cells of the nasopharynx and spreads to the mucosa of the lower respiratory tract by cell-to-cell transfer. The infection causes death of the cells that line the bronchi, which then slough into the lumen. The production of mucus increases, and mononuclear cells infiltrate the area. Clumps of necrotic epithelium and mucus initially decrease the diameter of the bronchi, causing turbulent air flow, particularly on expiration when the luminal diameter normally decreases. Eventually, plugging of the bronchi produces hyperinflation and atelectasis.

Clinical Manifestations

Bronchiolitis begins as a URI with cough and coryza. Over 2 to 5 days, signs of respiratory distress appear. The parents can often hear the child wheezing.

Fever occurs in two-thirds of children with bronchiolitis. They often appear ill on overall assessment. The respiratory rate climbs to at least 40 breaths/ minute and may reach 80 to 100 breaths/minute. Nasal flaring and retractions of the intercostal and supraclavicular muscles are noted and increase as the disease progresses. In bronchiolitis and other lower respiratory tract infections, the intercostal retractions are more pronounced than the supraclavicular, the opposite of the findings in croup and epiglottitis. Wheezes and a prolonged expiratory phase are heard in all children with bronchiolitis, at times without a stethoscope; rales are usually minimal. As the ventilatory muscles fatigue, the child will have grunting respirations; only in the most severe cases does cyanosis occur.

The total WBC count in bronchiolitis is normal. Usually, the chest radiograph shows only hyperaerated lungs, but there may occasionally be areas of atelectasis. If respiratory failure supervenes, the Pa O_2 decreases and carbon dioxide is retained.

The complications of bronchiolitis include dehydration, respiratory failure, and rarely, bacterial superinfection. Pneumothorax and pneumomediastinum are rarely seen. The increased respiratory effort in bronchiolitis may prevent an infant from maintaining an adequate oral intake. Careful attention should be paid to the details of fluid balance when taking a history. Of infants with bronchiolitis, 10 to 20% develop significant respiratory compromise. Cyanosis (or an oxygen saturation less than 91%), decreased inspiratory breath sounds, and lethargy on examination point to ventilatory failure. Bacterial superinfection is uncommon in the early stages of the illness, occurring occasionally in hospitalized infants. However, lobar consolidation seen on the chest radiograph suggests a potential bacterial pneumonia, although atelectatic patches may be confused with infiltrates.

Management

In the management of children with suspected bronchiolitis in the ED, a chest radiograph should be considered, both to look for findings compatible with this diagnosis and to help exclude other entities such as lobar pneumonia or a foreign body. Pulse oximetry provides an estimate of the degree of hypoxia. A WBC count, ABG, and/or electrolytes are obtained only if the diagnosis is uncertain or the clinical picture suggests that complications have occurred.

Children with bronchiolitis may benefit from nebulized bronchodilators. Although conflicting reports have been published, at least two studies have described an improvement in clinical status and oxygen saturation following albuterol delivered by nebulization and one group of investigators has found aerosolized epinephrine to be superior. For the child with moderate to severe distress, treatments can be administered every 20 minutes, starting at 0.1 to 0.3 of a 0.5% solution of albuterol or using 3 mL of 1:1000 epinephrine. Patients who show a favorable response to nebulized therapy are candidates to receive further nebulized treatments. Oral albuterol solution at a dosage of 0.1 mg/kg per dose, given every 6 hours, has limited efficacy.

Corticosteroids are not indicated for the treatment of patients with bronchiolitis. In general, it is difficult to differentiate asthma from bronchiolitis during the first 2 years of life; thus, corticosteroids may be given occasionally to some children who may have bronchiolitis or asthma, in accordance with the guidelines for the latter disease.

For the patient who does not respond to nebulized albuterol or has only mild distress, bronchodilators should not be prescribed. Therapy is limited to antipyretics and the encouragement of adequate oral intake, and the infant should be examined again after 24 to 48 hours. Dehydration secondary bacterial infection and significant respiratory distress necessitate admission to the hospital. Although not validated in infants with bronchiolitis, a score of 4 or more on the asthma scale (see Chapter 92) suggests significant respiratory compromise. An oxygen saturation less than 93% or an arterial PaO_2 less than 70 mm Hg in room air also suggests a need for hospitalization. In addition, children with underlying cardiac or pulmonary disease usually require admission. Ribavirin has proved somewhat useful in ameliorating the course of bronchiolitis, when administered by continuous aerosol for 3 to 5 days to severely ill children who are hospitalized. This agent is recommended primarily for patients with underlying cardiac or pulmonary conditions.

Pertussis

Background

Pertussis, or whooping cough, is an infection of the respiratory tract caused by *Bordetella pertussis.* Occasionally, a similar clinical syndrome is caused by *Bordetella parapertussis,* the adenoviruses, or *Chlamydia.* Young children most often contract pertussis, but the incidence in adolescents has increased recently. Although vaccination has contributed to the significant decrease in the frequency of this disease, several thousand cases occur yearly in the United States among unvaccinated children and to a lesser degree among those who have received vaccine, particularly of the whole cell variety.

Pathophysiology

Following inhalation, *B. pertussis* organisms attach to the epithelial cells of the respiratory tract. Multiplication of the bacteria leads to infiltration of the mucosa with polymorphonuclear leukocytes and lymphocytes. Inflammatory debris in the lumen of the bronchi and peribronchial lymphoid hyperplasia obstruct the smaller airways, causing atelectasis.

Clinical Manifestations

Although pertussis can be divided into three stages for discussion, a clinically distinct syndrome does not evolve until the disease has progressed to the second stage. Initially, the symptoms mimic a viral URI. This first stage (catarrhal), characterized by a mild cough, conjunctivitis, and coryza, lasts for 1 to 2 weeks. An increasingly severe cough heralds the onset of the second stage (paroxysmal), which continues for 2 to 4 weeks. After a prolonged spasm of coughing, the sudden inflow of air produces the characteristic whoop. Vomiting often occurs after such an episode. When not coughing, the child has a remarkably normal physical examination, except for an occasional subconjunctival hemorrhage. During the third stage (convalescent), the intensity of the cough wanes.

The WBC count in children usually reaches a level of 20,000 to 50,000/mm^3 with a marked lymphocytosis, but such changes are not often seen in infants less than 3 to 6 months old. Although a chest radiograph occasionally shows the characteristic “shaggy” right heart border, more often the lung fields appear clear. *B. pertussis* can be identified by fluorescent antibody staining of mucus obtained from the nasopharynx or, less commonly, recovered by culture of this material.

The fatality rate for pertussis is approximately 1% for patients in the first month of life and 0.3% for those between age 2 and 12 months. Complications often occur during a bout of pertussis. The most immediately life-threatening complication is complete obstruction of the airway by a mucous plug, leading to respiratory arrest. Although secondary bacterial pneumonia has a more insidious onset, it occurs in 25% of children with pertussis and accounts for 90% of the fatalities from pertussis. Seizures are seen in 3% of patients, and encephalitis in 1%. Sudden increases in intrathoracic pressure can cause intracranial hemorrhages, rupture of the diaphragm, and rectal prolapse.

Management

Except for occasional situations in which fluorescent antibody testing is immediately available, the diagnosis of pertussis rests on clinical grounds. Children with an unmistakable paroxysmal cough followed by a whoop should be assumed to have the disease. When the clinical picture is unclear, a WBC count and chest radiograph may be useful. The radiograph helps eliminate other causes of a severe cough (e.g., foreign body, bacterial pneumonia, cystic fibrosis,

tuberculosis), and the WBC count provides confirmatory evidence if a leukocytosis with marked lymphocytosis is found. Because of the grave risk of complications, all children less than 6 months of age diagnosed firmly as having pertussis should be observed in the hospital. Older children who show signs of respiratory compromise, such as cyanosis during paroxysms of coughing, or who develop complications also require admission. Treatment includes erythromycin (40 mg/kg per day for 14 days), maintenance of adequate hydration, and a level of respiratory support appropriate to the severity of the disease. Clarithromycin and azithromycin are alternative choices. Household and other close contacts require chemoprophylaxis with erythromycin (40 mg/kg per day for 14 days). Children younger than 7 years who are unimmunized or have received fewer than four doses of pertussis vaccine should have their pertussis immunization initiated or continued as soon as possible after exposure. Children who are fully immunized for age but have received only three doses require a fourth dose. Those who have had four doses need a booster unless the last dose has been within 3 years or they are more than 6 years old. DTP_a is preferred.

Tuberculosis

Background

In the United States, tuberculosis is caused almost exclusively by *M. tuberculosis* and occurs in childhood in several clinical forms. Although currently an unusual infection in developed countries, the incidence has increased recently and the disease should be kept in mind as an occasional, treatable cause of morbidity and mortality. At particular risk are children in urban, low-income areas and recent immigrants from underdeveloped countries. In addition, the emergency physician must be concerned about tuberculosis when either the patient or close contacts are infected with HIV (see Chapter 85).

Pathophysiology

Tubercle bacilli enter the body through the respiratory tract, producing an initial focus in the lungs. This lesion usually remains subclinical but may progress locally, resulting in a primary tuberculous pneumonia. During the primary infection, the organisms can disseminate hematogenously. Such spread may remain quiescent or, in a young child, may lead to miliary tuberculosis. Seeding of various organs occurs and may produce focal infections, a particularly serious concern with meningeal involvement.

Usually, the immune system limits the initial infection. However, reactivation of these foci may cause disease years later at any site involved during dissemination. Pulmonary lesions reactivate to produce tuberculous pneumonia in adults and adolescents much more often than in children.

Clinical Findings

Most infections by *M. tuberculosis* in children never cause any significant symptoms. Among the many possible clinical presentations, three stand out as particular concerns to the emergency physician: primary pneumonia, miliary tuberculosis, and meningitis. Pneumonia is by far the most common. Of note, these infections may develop despite prior vaccination against tuberculosis with bacillus Calmette-Guerin vaccine (BCG).

The onset of primary tuberculosis pneumonia resembles that of bacterial infections of the lungs. It begins with fever and tachypnea; rales and an area of dullness are found on examination of the chest. The WBC count may be elevated with a shift to the left, and the chest radiograph shows a lobar consolidation, often accompanied by hilar adenopathy and less often by pleural effusion or cavitation. Although the primary pneumonia often resolves spontaneously, the child occasionally follows a downhill course caused by local progression. In addition to the epidemiologic risks described, clinical findings that should arouse a suspicion of tuberculous pneumonia in the child otherwise thought to have a bacterial infection of the lung include pleural effusion, cavitation, toxicity, and a failure to respond to antibiotic therapy.

Miliary tuberculosis begins with an abrupt rise in temperature but a paucity of other physical findings; it may mimic sepsis. Subsequently, respiratory symptoms and enlargement of the liver, spleen, and superficial lymph nodes occur. The WBC count is usually in the range of 15,000/mm^3. Although the chest radiograph initially shows no lesions, a diffuse mottling of the lung fields appears 1 to 3 weeks after the fever. Miliary tuberculosis is a consideration in a child with a persistent fever and hepatosplenomegaly.

Tuberculous meningitis comes on insidiously with a low-grade fever, apathy, and in 50% of patients, vomiting. After 1 to 2 weeks of nonspecific illness, neurologic signs appear, including drowsiness and nuchal rigidity; if untreated, the child lapses into coma. The CSF shows a mononuclear pleocytosis, an elevated protein concentration, and eventually, a low glucose level.

Management

A child suspected of having pneumonic, meningeal, or miliary tuberculosis should be admitted to the hospital for evaluation and possible chemotherapy. Among inner-city populations, where the risk of tuberculosis is greatest, the routine placement of a tine or Mantoux test in children with lobar pneumonia should be considered. The Mantoux test must be interpreted in accordance with the child's age and the presence of risk factors (Table 84.18) Current treatment for tuberculosis consists of two to four or more drugs (isoniazid, rifampin, pyrazinamide, ethambutol, streptomycin, capreomycin, ciprofloxacin, cycloserine, ethionamide, kanamycin, ofloxacin, para-aminosalicylic acid) for a minimum of 6 months.

Induration ≥5 mm
Children in close contact with known or suspected cases of active tuberculosis, if adequate and timely treatment cannot be verified
Children suspected to have tuberculosis based on a consistent chest radiograph or clinical findings
Children immunosuppressed on the basis of therapy or disease
Induration ≥10 mm
Children <4 years of age
Children with chronic illness including lymphoma, diabetes mellitus, renal failure, and malnutrition
Children born in or traveling to regions of the world with a high prevalence of tuberculosis or exposed to adults likely to be infected
Induration ≥15 mm
Children ≥4 years of age without any risk factors

Modified from Committee on Infectious Diseases, 1997 RedBook. Elk Grove Village, IL: American Academy of Pediatrics, 1997.
[a]Applies regardless of previous BCG vaccination.

Table 84.18. Definition of Positive Criteria for the Standard Mantoux Skin Test (5 Tuberculin Units of PPD) in Children[a]

Hantavirus

The Hantavirus pulmonary syndrome was described in 1994 among 17 adults, of whom 13 died with severe pneumonia and hypotension. In a subsequent series, 8 of 100 patients were 16 years old or younger. Rodents serve as the reservoir for the hantaviruses, of which several varieties infect humans. The syndrome begins with fever, cough, and myalgias, followed shortly thereafter by tachypnea, tachycardia, dyspnea, and finally, hypotension. A marked leukocytosis is common along with thrombocytopenia and elevated clotting studies. The initial chest radiograph shows an interstitial more often than an alveolar infiltrate, with changes starting or becoming bilateral in the majority of cases. Pleural effusions occur in about one-quarter of the patients. The diagnosis should be considered when a severe pneumonia occurs in combination with systemic deterioration and can be confirmed subsequently by specific viral serology. Treatment is supportive.

Gastrointestinal Infections

Gastroenteritis is an inflammation of the alimentary tract that, in its acute form, is overwhelmingly infectious in origin. Viruses are the organisms most commonly found in children with diarrhea in the United States and can be isolated from 30 to 40% of patients. In 10 to 15% of patients, bacteria are recovered, including *Salmonella, Shigella, Campylobacter, Yersinia,* and pathogenic *E. coli; Aeromonas hydrophila* and *Vibrio* species, such as *Plesiomonas shigelloides,* are occasional pathogens. *Clostridium difficile,* which elaborates a toxin, may cause colitis, particularly after the use of antibiotics. Parasitic infestations rarely lead to diarrhea in the developed countries. *Giardia lamblia* and *Cryptosporidium* should be considered, particularly in outbreaks in day-care centers, and *Entamoeba histolytica*, among immigrants or travelers from tropical areas; cryptosporidiosis also affects patients with HIV. Current diagnostic techniques are unable to identify an etiologic agent in most of the remaining episodes.

In the United States, GI infections rank second in frequency to respiratory tract infections during childhood. An estimated 30,000,000 children with gastroenteritis receive treatment at home each year, 3,000,000 visit a physician, and 200,000 require hospitalization. Approximately 12% of all hospitalization of children 1 month through 4 years of age include diarrhea among one of the top three positions on the list of discharge diagnoses. Almost 400 children die annually in the United States from infections of the GI tract.

Viral hepatitis is covered in Chapter 93. Bacterial infections of the liver, almost exclusively abscesses, are rare in otherwise healthy children; more commonly, they complicate an immunosuppressive disease or therapy, or they affect the neonate.

Because calculi in the bile ducts rarely occur during childhood, cholecystitis occurs much less often in children than in adults. Occasionally, episodes are seen in adolescents or children predisposed to stone formation, as in the chronic hemolytic anemias. Less commonly, salmonellosis or leptospirosis produces acalculous cholecystitis.

In childhood, peritonitis almost invariably reflects an intra-abdominal catastrophe that requires surgical intervention. However, the accumulation of ascitic fluid in children with diseases such as nephrosis and cirrhosis allows the development of a primary infection of the peritoneum.

Viral Gastroenteritis

Background

Viral gastroenteritis occurs primarily in two forms caused by different pathogens. The Norwalk virus produces an illness characterized by an explosive onset and vomiting, more severe than the diarrhea that accompanies it. The symptoms are self-limiting, resolving in 2 to 3 days. It occurs in epidemics, most often in the winter, and affects predominantly school-age children. Rotavirus, however, produces a prolonged diarrheal illness of varying severity. It occurs more often in young children, although older family members may be affected. Other viruses, including enteroviruses, coronaviruses, and adenoviruses, may play a role in gastroenteritis.

Viral gastroenteritis is common. Among U.S. families, it trails only the common cold in frequency. Rotaviruses are the most commonly isolated pathogens, particularly among children who develop dehydration. Viral infections of the GI tract cause considerable loss of time from school and occasionally require treatment in the hospital.

Pathophysiology

Rotaviruses invade the intestinal epithelial cells, where they can be visualized by electron microscopy. The histology of the mucosal layer is disturbed during the active infection and for 3 to 8 weeks afterward. Functional abnormalities accompany the morphologic changes, including depressions of disaccharidase levels. Although Norwalk virus may invade the mucosal lining of the intestine, it has not been detected intracellularly. Histologic changes occur and persist for 2 weeks, and disaccharidase levels decline during the infection.

Clinical Findings

Children with viral gastroenteritis are usually brought to the ED with a complaint of diarrhea and/or vomiting. The numbers of stools may vary from 2 or 3 to 15 or 20 daily. Most commonly there are 6 to 8 bowel movements in a 24-hour period; the stools range from semisolid in consistency to watery. Although hematochezia may occasionally occur in viral infections, the presence of blood in the stool should suggest a bacterial gastroenteritis. Vomiting may accompany diarrhea or be the sole manifestation of a viral gastroenteritis. The daily frequency of emesis varies in the same range as for diarrhea. After forceful emesis, streaks of blood may be present in the vomitus. Many children with viral gastroenteritis beyond the age of 2 or 3 years complain of crampy abdominal pain. In more severe illnesses, the parent may relate a history of decreased oral intake and oliguria.

Children with viral gastroenteritis are usually febrile. However, in the child older than 3 years, a temperature over 39°C (102.2°F) may suggest a bacterial enteritis. Tachycardia, hypotension, and lethargy may reflect dehydration in severe episodes. Whereas the respiratory rate is usually normal, tachypnea occurs when acidosis and/or dehydration are present. The abdomen is soft and nondistended in most cases. Although the child may perceive palpation as uncomfortable, this maneuver does not elicit localized or rebound tenderness. Auscultation reveals hyperactive bowel sounds. The skin turgor is decreased and the mucous membranes are dry only in severe gastroenteritis with dehydration (see Chapter 18, Chapter 19, and Chapter 86).

No laboratory studies are indicated in the uncomplicated case of gastroenteritis. The CBC, electrolytes, and BUN usually fall within the normal range. If oral intake fails to keep pace with the efflux of fluids from the alimentary tract, dehydration occurs. The sodium, usually normal, may drop as low as 110 mEq/L or rise to 170 mEq/L, and the bicarbonate is invariably low. With mild dehydration, the serum bicarbonate hovers just below the normal level at 18 to 20 mEq/L; however, values of 10 to 12 mEq/L are usually found in the face of prolonged diarrhea. The BUN reflects the state of hydration and the adequacy at the recent intake of protein. It may climb as high as 100 mg/dL in children who lose more than 10% of their body weight. In a child who has been maintained on clear liquids, however, the BUN will not accurately indicate the degree of dehydration because urea rises as a breakdown product during protein metabolism. Although the hemoglobin and WBC count are usually normal in the child with viral gastroenteritis, hemoconcentration may occur with dehydration.

Management

Uncomplicated viral gastroenteritis usually remits in 2 to 5 days and does not require treatment in the hospital. All children should be weighed, preferably without clothing, to provide a baseline for follow-up. The vomiting will generally respond to a brief cessation of oral intake. After 2 to 4 hours of abstinence, the diet should be resumed gradually. The diarrhea may persist for several days, but hydration can usually be maintained orally after the vomiting has subsided.

Current recommendations for oral therapy emphasize the use of appropriately balanced glucose and electrolyte solutions, as well as the early reintroduction of feedings. Generally, rehydration is initiated, particularly in infants less than 1 year old, with a solution that contains 75 to 90 mEq/L sodium in a ratio with glucose of 1:1 (e.g., Rehydralyte). Older children often tolerate juices and sodas. Some studies have advocated the use of glucose polymers (e.g., Ricelyte) instead of glucose as a means to reduce diarrhea, but significant advantages have yet to be demonstrated for these products. Preparation at home of fluids that contain salt notoriously leads to errors, and this procedure is to be condemned. Similarly, the physician should avoid the use of boiled skim milk, a hypertonic solution that may produce hypernatremia.

Antiemetics and antidiarrheal medications provide minimal, if any, relief to the child. In addition, many of these medications carry significant risks. Kaopectate has been shown to increase stool losses of water and electrolytes. Although the combination of diphenoxylate and atropine (Lomotil) may be successful in adults, toxic reactions in children limit its usefulness. Trimethobenzamide (Tigan) appears to be ineffective as an antiemetic in children. Although the phenothiazine compounds reduce emesis, they occasionally produce adverse side effects, such as extrapyramidal reactions or oculogyric crises that limit their usefulness. Loperamide (0.5 mg/kg per day) has been shown to reduce the severity of diarrhea in conjunction with oral rehydration therapy but is indicated only for unusually severe or prolonged cases of gastroenteritis after excluding a cause that would respond to specific therapy. Although a few studies have suggested a small benefit from bismuth subsalicylate (Pepto-Bismol), this agent is not recommended for routine cases.

Dehydration is the only significant complication of viral gastroenteritis. If the physician suspects that a child has developed more than 5 to 10% dehydration, electrolytes and a BUN should be obtained. These tests establish the degree of acidosis and the presence of hyponatremia or hypernatremia.

Most children with gastroenteritis tolerate oral rehydration. In underdeveloped countries, even patients with severe dehydration are often managed successfully in most cases by using the oral route. However, in the ED, treatment for children with moderate to severe dehydration is usually initiated intravenously. As a rule, all patients with dehydration estimated to be greater than 10%, and many cases falling in the range of 5 to 10%, require IV fluids.

When IV therapy is chosen, a bolus of fluid, such as 10 to 20 mL/kg normal saline, may be administered over 1 hour, or

more rapidly if needed (see Chapter 3). If rehydration is achieved and the child is capable of subsequent oral intake, treatment may be continued at home (as in the milder cases).

Children who are more than 5% dehydrated or have alterations in the serum sodium (less than 130 mEq/L or more than 145 mEq/L) may require hospitalization. IV therapy should be started in the ED, particularly if there is evidence of vascular instability (see Chapter 3, Chapter 18, Chapter 19, and Chapter 86).

Bacterial Gastroenteritis

Background

Five pathogens commonly produce gastroenteritis: *Salmonella, Shigella, Yersinia, Campylobacter,* and pathogenic *E. coli.* Together, these organisms cause 10 to 15% of the diarrheal illnesses seen in children coming to the ED (Fig. 84.17). In underdeveloped countries and occasionally in the United States, *Vibrio* species must also be considered. In addition, *A. hydrophila* has been associated occasionally with diarrheal illnesses in children. *C. difficile* causes a toxin-associated colitis, particularly in patients who receive antibiotics.

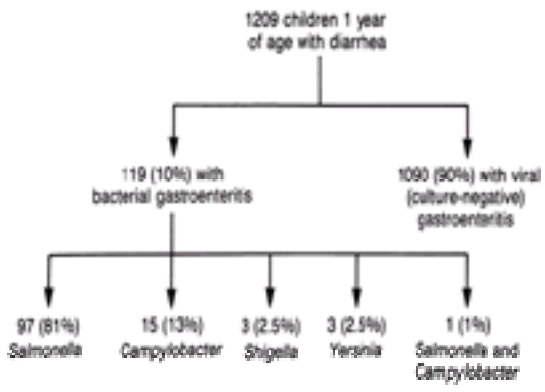

FIGURE 84.17. Etiology of gastroenteritis among consecutively cultured infants less than 1 year of age in an urban emergency department. (Reprinted with permission from Torrey S, Fleisher G, Jaffe D. Incidence of *Salmonella* bacteremia in infants with *Salmonella* gastroenteritis. J Pediatr 1986;108:718.)

Salmonella, Shigella, Yersinia, Gram-negative bacilli in the *Enterobacteriaceae* family, and *Campylobacter* do not normally inhabit the alimentary tract. Thus, recovery of one of these organisms suffices for the diagnosis of gastroenteritis. *E. coli,* however, is part of the normal bowel flora, only occasionally assuming a pathogenic role. Serotyping is useful for detecting *E. coli* O157, a cause of hemolytic uremic syndrome, but identification of other disease-producing strains is not readily available to the clinician.

Pathophysiology

Salmonella species gain access to the small intestine following ingestion. Gastric acid is usually lethal to the organism, but large numbers of bacteria may overcome this defense mechanism. Studies have shown that 10^5 virulent organisms will induce disease in 30% of healthy adults and 10^9 in 95%. Patients with gastrectomies are more susceptible to *Salmonella* infection than those with an intact stomach. *Salmonella* can penetrate the epithelial layer to the level of the lamina propria and evoke a leukocyte response. Generally, the infection extends no further, but bacteremia may occur, especially in young children. Several species, notably *S. choleraesuis* and *S. typhi,* readily enter the circulation through the lymphatics. *Salmonella* produce diarrhea by multiple mechanisms. Several toxins have been identified; in addition, prostaglandins that stimulate the active secretion of fluids and electrolytes may be released.

Certain *Shigella* attach to binding sites on the surface of the intestinal mucosal cells. The organisms penetrate the cells and proliferate within them. Intraepithelial multiplication destroys the cell and produces mucosal ulcerations. Invasion of the epithelium evokes an intense inflammatory response. At the base of the ulcerated lesions, erosion of blood vessels may lead to bleeding. Other *Shigella* elaborate exotoxins that can produce diarrhea. These toxins result in increased secretion of fluid and electrolytes by the intestinal mucosa.

Although the pathophysiology of infection from *Yersinia enterocolitica* has not been completely elucidated, clues are available from animal models and occasional pathologic specimens. The organisms are thought to produce terminal ileitis; inflammatory changes and ulcerations have been visualized with endoscopy. The infection elicits a neutrophilic response, particularly around the Peyer's patches. It then extends to the mesenteric lymph nodes, which are destroyed by microabscess formation and may enlarge considerably. Occasionally, further dissemination occurs with involvement of the liver and spleen. Although an enterotoxin has been identified in cultures of *Y. enterocolitica* maintained at 26° to 30°C (78.8° to 86°F), production in vivo has not been established.

The pathogenesis of *Campylobacter* enteritis remains unknown. Attempts to demonstrate toxin production have not met with success, and the organism has been shown to lack invasive properties. At autopsy, extensive hemorrhagic ulcerations of the bowel have been described.

E. coli may produce diarrhea on the basis of several characteristics. Pathogenic strains have been classified as enteropathogenic, enterotoxic, enteroinvasive, enteroaggregative, enteroadherent, and enterohemorrhagic. The risk of

developing hemolytic uremic syndrome after infection with *E. coli* O157 is estimated to be 10 to 15% in children.

Clinical Manifestations

Signs and Symptoms. A careful epidemiologic history often provides a clue to the diagnosis of *Salmonella* infections. Foodborne outbreaks often occur in the summer. After an incubation period of 8 to 48 hours, the child experiences crampy abdominal pain and nausea. The stools are watery and may contain blood, but this is not the rule. Fever is noted in most children. Unless protracted diarrhea has led to clinically apparent dehydration, the physical examination is unremarkable. Abdominal tenderness and distension are usual findings. The leukocyte count is usually 10,000 to 15,000/mm^3. Methylene blue staining of the stool may show the presence of polymorphonuclear leukocytes, but not in sheets as seen with *Shigella.* A single rectal swab leads to isolation of *Salmonella* from more than 90% of children with this infection.

Shigella may cause an asymptomatic infection, mild gastroenteritis, or bacillary dysentery. Mild illnesses are more common. Children affected in this way complain of frequent watery stools but few constitutional symptoms. The temperature remains normal in many cases, and the physical examination is unremarkable.

Bacillary dysentery begins suddenly with fever and abdominal pain. Diarrhea begins shortly thereafter. The stools, which may average 10 to 12 daily, contain mucus and blood, and tenesmus is common. Children with this form of shigellosis have a fever, often in the range of 39° to 40°C (102.2° to 104°F). Palpation of the abdomen often elicits diffuse tenderness but no evidence of peritoneal irritation.

Occasionally, a *Shigella* infection may produce CNS irritation because of the release of toxin before the onset of diarrhea. Thus, shigellosis must be considered in the differential diagnosis of meningismus in the absence of a pleocytosis in the CSF. A seizure may actually be the first manifestation of the illness.

Certain laboratory abnormalities strongly suggest *Shigella* as the cause of gastroenteritis. The leukocyte count often shows many band forms that exceed the mature neutrophils in number. Poh described this phenomenon in 85% of 123 children between the ages of 2 months and 8 years with shigellosis. The total WBC count may show a leukopenia or a leukocytosis but most commonly hovers in the normal range. Because *Shigella* invades the intestinal mucosa, this infection elicits a profound inflammatory response. The exudation of white cells leads to the finding of sheets of neutrophils in the stool after methylene blue staining. A single rectal swab suffices for the isolation of *Shigella* from most children with this illness.

Children with gastroenteritis caused by *Y. enterocolitica* usually have an abrupt onset of diarrhea. The stools are often watery and may contain blood, but vomiting generally remains inconsequential. Patients with this illness often complain of severe abdominal pain, sometimes before the onset of diarrhea. Delorme et al. noted this symptom in 6 of the 35 children studied, half of whom were 1 to 5 years old. In an epidemic in a school in New York State, 37 of 38 patients had abdominal pain; the potential severity of the abdominal pain in this disease is illustrated by the fact that 16 patients in this outbreak mistakenly underwent an appendectomy.

GI infection with *Y. enterocolitica* usually elicits a febrile response. The mean temperature in the young adolescents reported by Black et al. was 38.7°C (101.6°F), with a range of 37.2° to 40°C (99° to 104°F); it exceeded 37.8°C (100°F) in more than 95% of patients. Younger children appear to develop a fever less often. The abdominal examination is usually benign, but palpation produces marked tenderness in the subset of patients with mesenteric adenitis. Arthritis and skin rashes occur in 5 to 10% of patients with this disease.

The mean WBC count in children with yersiniosis is usually normal, although leukocytosis with a shift to the left occurs occasionally. The electrolytes and BUN are normal except in the face of dehydration. Examination of stool stained with methylene blue reveals polymorphonuclear neutrophils. The organism can be recovered from stool culture but requires enrichment techniques. Although a single specimen is diagnostic in 70 to 80% of illnesses, a second sample should be obtained in the face of a previous negative culture when the clinical suspicion of disease remains strong.

Campylobacter enteritis is characterized by the abrupt onset of fever and abdominal pain, followed shortly by diarrhea. The temperature often remains normal in children less than 3 months old, but ranges up to 40°C (104°F) in the older child. Vomiting occurs uncommonly and resolves rapidly. Two-thirds of children complain of abdominal pain, which may be severe. The number of stools varies from 2 to 20 daily; they are watery and contain blood in at least 50% of cases. Karmali and Fleming found frank blood in the stools of 95% of their patients. The physical examination is generally unremarkable. Although the abdominal pain occasionally simulates appendicitis, palpation of the abdomen elicits minimal tenderness. Signs of dehydration are found only rarely.

The WBC count in *Campylobacter* enteritis usually remains below 12,000/mm^3, the highest being 22,500/mm^3 in one study; on occasion, there may be a shift to the left. The electrolytes and BUN are usually normal. Maki et al. found fecal leukocytes in four of five patients with enteritis caused by *Campylobacter.* The organism is not often isolated from the blood but can be recovered easily from the stool by using appropriate media. When available, phase contrast microscopy can demonstrate the organism in fresh stool specimens.

The clinical picture of diarrhea caused by *E. coli* varies. This organism is suspected most often in the setting of a specific outbreak.

In general, features suggestive of a bacterial rather than a viral gastroenteritis include 1) more than 10 stools per day or diarrhea lasting for more than 4 days, 2) blood in the stool, 3) fever of 39.5°C (103°F) or higher, 4) clinical toxicity, and 5) polymorphonuclear leukocytes in the stool. The presence of these findings enhances the likelihood that a bacterial pathogen is involved, although a viral gastroenteritis is not necessarily ruled out.

Complications. The complications of *Salmonella* gastroenteritis include dehydration and spread of infection beyond the confines of the GI tract. During bacteremia, focal infections, including meningitis, osteomyelitis, and endocarditis, may develop. However, most episodes of bacteremia terminate spontaneously. Dehydration is diagnosed on the basis of the clinical findings: dry mucous membranes, decreased skin turgor, tachycardia, and hypotension. Although the electrolytes are most often normal, both hyponatremia and hypernatremia may occur.

Bacteremia is most common in young children. In a study by Hyams et al., 25% of hospitalized patients with *Salmonella* gastroenteritis had the organism recovered from their blood. However, Torrey et al. noted an incidence of only 6% in an ambulatory population. Although a high fever usually accompanies spread to the circulation, the physical examination is often devoid of any signs of serious illness. In addition, infants in the first 3 months of life often remain afebrile in the face of bacteremia. The WBC count is greater than 15,000/mm^3 in 80%–90% with bacteremia, and culture of the blood leads to recovery of the organism.

Enteric fever also occurs from the dissemination of certain serotypes of *Salmonella;* if *S. typhi* is isolated, the illness is called typhoid fever. The disease is characterized by chills and fever, often rising in a steplike pattern to 40°C (104°F). Diarrhea does not necessarily precede or coexist with the systemic illness. A relative bradycardia in relation to the height of the temperature is a hallmark of enteric fever. Splenomegaly and a macular rash, or rose spots, are detectable in 20 to 30% of patients. Leukopenia characterizes the hematologic picture. Both blood and stool cultures may be negative. The diagnosis may rest on a fourfold rise in the agglutinin titers.

Invasion of the bloodstream may lead to various focal diseases. Meningitis most commonly affects the youngest children. The features are identical to those observed in CNS infections with other purulent organisms. Children with sickle cell hemoglobinopathies have a peculiar predilection for bone and joint involvement. Endocarditis is less commonly seen.

The complications of shigellosis include dehydration, bacteremia, seizures, and colonic perforation. Dehydration often accompanies dysenteric infections and is diagnosed on the basis of the usual clinical findings. Bacteremia and perforation are both rare, occurring in fewer than 1% of GI infections.

Most episodes of gastroenteritis with *Yersinia* are self-limiting, resolving before dehydration develops. Appendicitis occasionally results from obstruction of the appendiceal lumen by swollen lymphoid tissue. The incidence is unknown, but 5 of 38 patients in the New York State epidemic underwent removal of appendices that were suppurative. Bacteremia and focal infection follow gastroenteritis almost exclusively in the compromised host, particularly in association with thalassemia.

Campylobacter infections occasionally lead to dehydration, but less often than is seen with the other bacterial pathogens in the GI tract. Rarely, bacteremic or focal infections occur.

Management

Salmonella gastroenteritis is usually a self-limiting illness. In most cases, the disease is not sufficiently distinct or severe enough to suggest to the clinician the need for a diagnostic evaluation. However, life-threatening complications occur with predictable regularity in infants less than 6 months of age and in children with sickle cell hemoglobinopathies.

The treatment of *Salmonella* gastroenteritis should be directed toward the maintenance of adequate hydration. As with viral infections, limitation of the diet to electrolyte solutions (“clear liquids”) suffices in most children. Antibiotic therapy neither ameliorates the course of the gastroenteritis nor eradicates the organism from the intestinal tract in the immunocompetent host. In fact, several studies have suggested prolonged carriage after the administration of antibiotics.

The indications for admission of a child with diarrhea suspected or proved to be caused by *Salmonella* species are 1) dehydration, 2) focal infection or bacteremia/sepsis, 3) age less than 3 months or temperature over 39°C (102.2°F) in a child under 12 months old (unless blood culture is known to be sterile), and 4) sickle cell anemia. If bacteremia is suspected, IV therapy with cefotaxime (200 mg/kg per day in four divided doses) or ceftriaxone (100 mg/kg per day in two divided doses) should be initiated. Chloramphenicol (75 to 100 mg/kg per day in four divided doses) or, in adolescents, one of the fluoroquinolones (ciprofloxacin, ofloxacin) provides an alternative for cephalosporin-allergic patients. When oral therapy is indicated, TMP-SMZ (8 mg/kg per day of trimethoprim in two divided doses) is the drug of choice.

Shigellosis stands alone as the only form of bacterial gastroenteritis for which antibiotics have proved efficacious. Antimicrobial therapy shortens the course of the illness and the duration of excretion of the organisms in the stool. Treatment alleviates the symptoms and signs of the gastroenteritis and limits transmission of the disease. TMP-SMZ (8 mg of trimethoprim and 40 mg of sulfamethoxazole per kilogram per day) is the initial drug of choice while the results of sensitivity tests are pending. Fluoroquinolones and ceftriaxone are alternatives.

Supportive therapy is an important aspect of the management of shigellosis. The initial oral intake should be limited to solutions with physiologic concentrations of glucose and electrolytes. As the diarrhea begins to abate, solid foods can be added. Dietary manipulation leads to resolution of the disease in some children before the isolation of the organism. Antibiotic therapy may be omitted in such cases, unless there is a particular concern about spread in a closed population.

As with other varieties of infectious gastroenteritis, most medications designed to provide symptomatic relief from diarrhea have no demonstrated efficacy. In particular, paregoric or combinations of diphenoxylate and atropine (Lomotil) are contraindicated. Dupont et al. showed that diarrhea persisted longer in infected volunteers treated with antibiotics and diphenoxylate/atropine than with those who received only antibiotics.

Most episodes of shigellosis can be handled on an outpatient basis. Indications for admission include 1) age 6 months or

younger, 2) dehydration, and 3) bacteremia (rare). Before the definitive diagnosis of shigellosis, particularly with significant bleeding, hospitalization may be required because of a concern about noninfectious entities such as a Meckel's diverticulum.

Most children with yersiniosis can be treated as outpatients. Initially, the diet should be limited to electrolyte solutions (clear liquids). Although *Y. enterocolitica* is usually sensitive in vitro to tetracycline, chloramphenicol, colistin, gentamicin, and kanamycin, current studies have demonstrated no benefit from antibiotic therapy of uncomplicated gastroenteritis. However, persistent diarrhea may respond to antimicrobial treatment. Suspected or proven sepsis merits IV administration of antibiotics such as gentamicin (5 to 7.5 mg/kg per day in three divided doses, beyond the neonatal period). The indications for admission include dehydration, severe abdominal pain suggesting appendicitis, and underlying diseases such as thalassemia.

Campylobacter enteritis is a self-limited but prolonged illness; diarrhea persists for more than 1 week in one-third of children. These organisms exhibit almost universal sensitivity to erythromycin, which can be given orally at a dosage of 40 mg/kg per day; ciprofloxacin is an alternative for adolescents. However, antimicrobial therapy has not proved to decrease the duration of diarrhea.

Antibiotic-Associated Colitis

Background

Children who take antibiotics often develop diarrhea, which varies from mild to severe. For most of the mild cases, no specific diagnosis is established. A small subset of patients, usually with more severe illnesses, have pseudomembranous colitis, caused by *C. difficile.* Although antibiotics are the most important precipitating factor for pseudomembranous colitis, the disease was recognized in the preantibiotic era and still occurs occasionally in the absence of prior antibiotic therapy. Almost every antibiotic has been reported to be associated with pseudomembranous colitis. Clindamycin, lincomycin, and the broad-spectrum b-lactam agents in particular predispose to overgrowth of *C. difficile,* but because these drugs are rarely used for children on an outpatient basis, widely prescribed medications, such as amoxicillin, are implicated more often in the pediatric age group.

Pathophysiology

C. difficile, the etiologic agent in pseudomembranous colitis, is a Gram-positive anaerobic bacillus that may be part of the normal intestinal flora, particularly during the first year of life. Even a short course of antibiotics may lead to overgrowth of this organism. Colitis results from toxin production by *C. difficile* within the intestinal lumen; the two major toxins are known as A and B. These toxins attack the membranes or microfilaments of cells and produce hemorrhage, necrosis, and inflammation.

Clinical Manifestations

Colitis with *C. difficile* varies widely in severity. Typically, profuse watery or mucoid diarrhea begins after several days of antibiotic therapy. Many older children complain of crampy abdominal pain. On examination, the usual findings include fever and diffuse abdominal tenderness. Often, the WBC count rises above 15,000/mm ^{3}. The stool may be guaiac-positive or frankly bloody; leukocytes are found on smears from approximately 50% of patients. An etiologic diagnosis requires the identification of *C. difficile* toxin in the stool; recovery of the organism on culture is suggestive but not sufficient.

If *C. difficile* colitis goes unrecognized and untreated, complications, including toxic megacolon, perforation, and peritonitis, may develop. Case fatality rates as high as 10 to 20% were described before the introduction of specific treatments.

Management

The treatment for children with colitis caused by *C. difficile* depends on the severity of the disease. Mild cases respond to cessation of antibiotics and supportive therapy with fluids and electrolytes. In particular, children seen with a small amount of diarrhea on oral antibiotics for a minor infection, in whom the suspicion of pseudomembranous colitis is low, do not need an extensive diagnostic investigation or institution of specific antimicrobial therapy.

Patients with more severe or persistent antibiotic-associated diarrhea should be evaluated for *C. difficile* with a test for toxin in the stool. Oral metronidazole (30 mg/kg/day in four divided doses) or vancomycin (40 mg/kg per day in four divided doses) are used most commonly. Although used with some success in the past, cholestyramine does not have the same efficacy as oral antibiotics, but it may be used for patients who fail treatment with vancomycin and metronidazole. Antidiarrheal agents should be avoided. When possible, the precipitating antibiotic should be discontinued, but cessation is not essential once specific therapy has been initiated. In general, children who are sufficiently ill to require treatment with vancomycin should be observed in the hospital.

Gastritis

Background

Gastritis is an inflammation of the lining of the stomach. Most cases are noninfectious; however, *Helicobacter pylori* is a Gram-negative rod that is capable of surviving in the acid milieu of the stomach to produce disease. Infection rates are low in young children but increase in adolescence. Chronic infection with this organism is associated with peptic ulcer

and gastric carcinoma.

Clinical Manifestations

Gastritis caused by *H. pylori* manifests in older children and adolescents with persistent epigastric pain, nausea, and vomiting. Often, the stool will test positive for blood. More severe cases are characterized by hematemesis. In younger children and infants unable to verbalize or localize pain reliably, irritability may be the primary manifestation.

H. pylori can be diagnosed by culture of gastric tissue obtained at biopsy, breath testing, and serology. Only serology has applicability in the setting of the ED. Sensitivity of the serologic assay has been reported to range from 70 to 95%, with greater accuracy being observed in older children.

Management

In most cases, the clinician cannot diagnose *H. pylori* infection in the ED with sufficient certainty to warrant the initiation of treatment with antibiotics. Effective two-drug regimens include clarithromycin–omeprazole, amoxicillin–bismuth, and amoxicillin–omeprazole for 2 to 4 weeks. A 1-week course of omeprazole (20 mg twice daily), clarithromycin (250 mg twice daily), and metronidazole (500 mg twice daily) proved effective in a group of 35 children 10 to 19 years of age.

Skin, Soft-Tissue, and Bone Infections

Infections of the skin, soft tissues, and bones include impetigo, cutaneous abscesses, lymphadenitis, cellulitis, fasciitis, pyomyositis, septic arthritis, and osteomyelitis. Chapter 117 deals with cutaneous abscesses. Among the others, impetigo and cellulitis are both common complaints in the ED. Although children with bone and joint infections are seen only occasionally, the differential diagnosis of several common complaints (e.g., fever, limp) often includes these conditions. Thus, the emergency physician who deals with children should be familiar with such infections, particularly because a delay in the institution of therapy can result in appreciable morbidity. Fasciitis has emerged as an important infection in children, particularly as a complication of varicella. In contrast, pyomyositis is rare in the United States, occurring more commonly in tropical regions.

Impetigo

Background

Impetigo is a bacterial infection of the skin confined to the epidermis. A deeper variety of impetigo, ecthyma, involves the dermis as well. Pustules larger than 1 cm in diameter (Fig. 84.18) characterize bullous impetigo. Impetigo is a common infection in children, particularly during the summer months. It occurs in epidemics during the warm weather in confined populations of children.

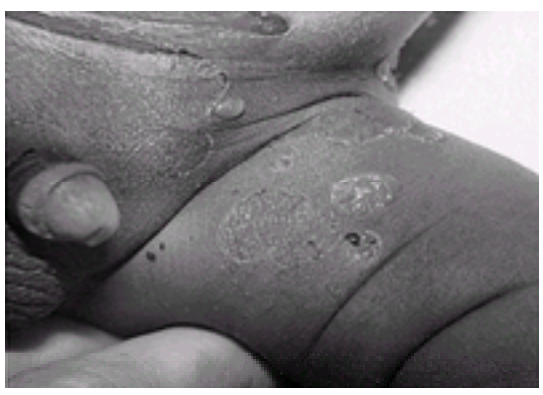

FIGURE 84.18. Bullous impetigo.

Any strain of group A streptococcus, including nephritogenic varieties, can infect the skin and cause impetigo. In recent years, *S. aureus,* the primary agent in bullous impetigo, has become a common cause of nonbullous impetigo as well.

Pathophysiology

The intact epidermis forms a relatively impervious barrier to bacteria. However, a breach in the integument, even if too small to be noticed by the patient or parents, may allow the entry of pathogens and the development of impetigo. In streptococcal infections, toxins, such as streptolysins, elaborated by the organism, promote local spread of the process. Different toxins produced by *S. aureus* lead to the accumulation of purulent material and the evolution of bullae.

Clinical Manifestations

Impetigo is more common in young children, particularly those less than 6 years of age. Typically, a parent will bring a child to the ED complaining of sores on the body. No systemic ailments, such as fever or malaise, are associated. Physical examination shows a healthy child with a normal temperature. The lesions usually ooze serous fluid but may be bullous or crusted as well (Fig. 84.18). Surrounding erythema is minimal, and the regional lymph nodes often do not enlarge noticeably.

Laboratory studies are not routinely obtained in children with impetigo. Cultures, performed only if there is any doubt about the diagnosis, will yield group A streptococci and *S. aureus* in most cases. The WBC count is normal.

The complications of impetigo include spread of the infection locally and remote nonsuppurative disease. Occasionally, impetigo may progress to cellulitis. If the lesions are caused by nephritogenic streptococci, glomerulonephritis may develop 7 to 14 days later. The attack rate for glomerulonephritis has been as high as 1% in certain epidemics, but the incidence is far less in the usual clinical setting.

Management

A single course of antibiotic therapy cures impetigo in 95% of children. Erythromycin (40 mg/kg per day in four divided doses) provides effective oral treatment for the usual pathogens. For nonbullous disease, intramuscular benzathine penicillin (30,000 to 50,000 units/kg) is a reasonable single-dose alternative, if compliance is a concern; patients who fail to improve with this regimen should receive erythromycin for presumed *S. aureus.* Other acceptable oral drugs include dicloxacillin (50 mg/kg per day) or cephalexin (50 mg/kg per day). Mupirocin applied locally is able to eradicate most cases of impetigo, particularly if the disease is limited in distribution. Combination topical and systemic therapy is unnecessary. Vigorous scrubbing does not hasten the resolution, and routine cleanliness is sufficient. Even when systemic antibiotic therapy eliminates the infection, the incidence of glomerulonephritis has not been demonstrated to decrease.

Lymphadenitis

Lymph nodes in any region of the body may become infected. Regardless of the site of involvement, the same considerations apply as discussed under cervical lymphadenitis. *S. aureus* and group A streptococcus are the most common pathogens. The finding of inguinal or axillary adenitis should prompt a meticulous search for a portal of entry for bacteria on the extremities. Locating an impetiginous lesion or other breech in the integument provides reassurance that the lymph node enlargement is caused by infection rather than by neoplasm. History should be requested regarding a cat scratch or bite as a possible etiologic focus. Particularly in the adolescent, inguinal adenitis suggests a need to look for sexually transmitted pathogens. CSD is another important consideration. The child with lymphadenitis should be treated with antibiotic therapy and drainage, if fluctuation occurs. Dicloxacillin (50 mg/kg per day) and cephalexin (50 mg/kg per day) are both effective against the usual pathogens.

Cellulitis

Background

Cellulitis is an infection of the skin and subcutaneous tissues. Any anatomic area may be involved, but the body can be divided, for etiologic considerations, into two regions: 1) the face and 2) the scalp, neck, trunk, and extremities.

Facial cellulitis includes buccal, periorbital, and less often, orbital lesions. Before the introduction of a vaccine against Hib, *H. influenzae* type b caused 50% of these infections. At present, the organisms involved most commonly are *S. aureus,* group A streptococcus, and *S. pneumoniae.* Bacteremia is present in 90% of the cases of disease caused by *S. pneumoniae* and *H. influenzae.*

S. aureus causes most nonfacial cellulitis and has been reported to be recovered from 70% of extremity lesions with an identifiable origin, either as the sole pathogen or in combination with group A streptococcus. Nonfacial cellulitis rarely results from infection *with H. influenzae,* although when this organism is involved, as with facial lesions, it usually invades the bloodstream.

Cellulitis occasionally occurs among immunosuppressed patients. In these cases, unusual organisms, including *P. aeruginosa*, Gram-negative enteric rods, and anaerobic bacteria, must be considered. Even when initial examination suggests minimal inflammation, an extensive infection may exist, because neutropenia often masks the depth of the lesion.

Cellulitis is a common infection that is more often seen in temperate climates when the weather is warm. Precise statistics on the incidence of cellulitis are not available; however, in one study during the summer months, this infection accounted for approximately 1 of every 500 visits to the ED of a children's hospital.

Pathogenesis

Cellulitis follows either hematogenous dissemination of a pathogenic organism or local invasion. Surgical or traumatic wounds may serve as a portal of entry for bacteria. This is the route by which *S. aureus* and group A streptococcus usually gain access to the subcutaneous tissue. Toxins produced by the organisms allow for local spread. Alternatively, invasion of the bloodstream may precede the appearance of cellulitis. The periorbital and facial lesions seen occasionally with *H. influenzae* and *S. pneumoniae* follow a bacteremia, and these organisms often are recovered from the blood. *S. aureus* and group A streptococcus are less often spread by this mechanism.

Clinical Manifestations

The child with cellulitis develops a local inflammatory response (Fig. 84.19) at the site of infection with erythema, edema, warmth, pain, and limitation of motion. There may be a history of a prior wound or insect bite. Facial infections are more common during the first 5 years of life. Fever is unusual, except in bacteremic infections or when the lesions are extensive. Only 10 to 20% of children with cellulitis manifest a fever. The lesion itself is erythematous and tender but not

fluctuant; red streaks may radiate proximally along the course of the lymphatic drainage. The regional lymph nodes usually enlarge in response to the infection.

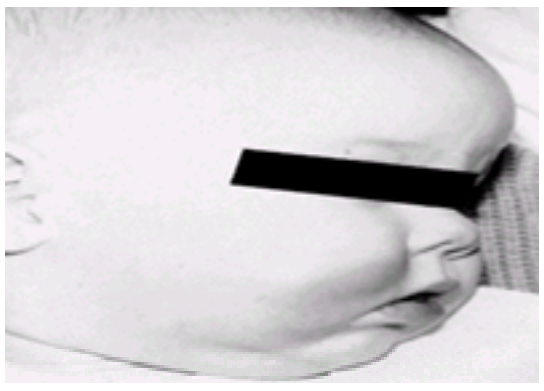

FIGURE 84.19. Infant with buccal cellulitis caused by *H. influenzae*.

With cellulitis caused by *S. aureus* or group A streptococcus, the WBC count is normal in most children. More extensive lesions or bacteremia, seen only occasionally, evoke a leukocytosis. A culture obtained from the central area of the cellulitis will yield a pathogen in 50% of cases, but cultures of the blood usually remain sterile.

Bacteremia accompanies cellulitis caused by *H. influenzae* or *S. pneumoniae;* these organisms are isolated from the blood in 90% of infected patients. The WBC count is greater than 15,000/m ^{3} as a rule, usually with a shift to the left.

The complications of cellulitis, although uncommon, include local and metastatic spread of infection. The organisms may invade deeper tissues, producing septic arthritis or osteomyelitis. During the course of bacteremia with *H. influenzae, S. pneumoniae,* or rarely other organisms, there may be involvement of the meninges, pericardium, epiglottis, or synovial membranes. Multifocal areas of cellulitis should arouse a suspicion of hematogenous dissemination. Occasionally, cellulitis provides a clue to an infection that originates in deeper anatomic structures. As an example, a lesion on the abdominal wall, may be a sign of peritonitis.

Management

Most children with nonfacial cellulitis can receive antibiotic therapy as outpatients, as long as bacteremic disease is unlikely (Fig. 84.20). Because *S. aureus* and group A streptococcus are most commonly isolated, treatment should be directed at these organisms. Acceptable alternatives include a semisynthetic penicillin, such as dicloxacillin (50 mg/kg per day), cephalexin (50 mg/kg per day), or amoxicillin–clavulanic acid (50 mg/kg per day of amoxicillin); *S. aureus* is generally resistant to penicillin and ampicillin. A CBC, blood culture, and aspirate culture are not necessary in afebrile patients.

FIGURE 84.20. Diagnostic approach for the management of the child with soft-tissue swelling and possible cellulitis. *WBC,* white blood cell.

If a child with a nonfacial cellulitis has a high fever (39°C [102.2°F] or higher), the likelihood of a bacteremic infection or lymphangitic spread increases. A WBC count and culture of the blood should be obtained, along with consideration of a culture from the lesion. In cases in which the WBC count is below 15,000/mm ^{3}, antibiotic therapy is given as described for afebrile children, and the patient is asked to return the following day. A leukocytosis in association with a temperature of 39°C (102.2°F) or higher points toward IV treatment, usually on an inpatient basis, with oxacillin (150 mg/kg per day in four divided doses) or cephazolin (100 mg/kg per day in three divided doses). For children not immunized against Hib, consider therapy with cefotaxime (200 mg/kg per day in four divided doses) ceftriaxone (100 mg/kg per day in a single dose), or ampicillin–clavulanic acid (200 mg/kg per day of ampicillin in four divided doses). Children allergic to penicillins and cephalosporins can be given clindamycin (40 mg/kg per day in four divided doses) alone or with chloramphenicol (75 to 100 mg/kg per day in four divided doses) when *H. influenzae* type b is a concern.

Children with facial cellulitis and fever are particularly likely to be bacteremic, in most cases with *S. pneumoniae* or, less commonly, *H. influenzae* type b, and are at risk for local complications. Thus, they should receive IV therapy as listed previously. Those who are afebrile may be managed as outpatients if they do not have risk factors for bacteremic

disease—age less than 3 years, spontaneous cellulitis without a preceding wound, and violaceous discoloration (Fig. 84.20).

Fasciitis

Background

Fasciitis is a deep soft-tissue infection. Unlike cellulitis, it involves the fascial and muscle layers as well as the skin and subcutaneous tissues, but it does not extend per se to the bones or joints. Terms used to refer to this condition include *necrotizing fasciitis, acute streptococcal hemolytic gangrene, Meleney synergistic gangrene,* and *necrotizing erysipelas.* In recent years, the most common cause, by far, has been group A streptococcus; other etiologic agents include *S. aureus* and anaerobic organisms. Although some cases of fasciitis arise spontaneously, most occur as a complication of varicella.

Clinical Manifestations

As occurs with cellulitis, the child with fasciitis develops a local inflammatory response at the site of infection, characterized by erythema, edema, warmth, pain, and limitation of motion.

Fever occurs in almost every case, often exceeding 39°C (102.2°F). In contrast to the usual patient with cellulitis, those with fasciitis almost always appear toxic with a marked tachycardia and, occasionally, hypotension. The local lesion is often described by the family as progressing rapidly and generally exhibits noticeable induration and erythema. Particularly in the presence of varicelliform lesions, the physician should maintain a high index of suspicion for fasciitis, as opposed to cellulitis, in children with extensive local disease, high fever, and any degree of prostration. The WBC count generally reflects a leukocytosis, and blood cultures yield an organism in most cases.

Management

A confirmed case of necrotizing cellulitis should be considered an emergency. The first priorities include supportive therapy for signs of sepsis and initiation of antibiotics, followed promptly by surgical consultation. Appropriate antimicrobial therapy includes penicillin (500,000 units/kg per day in four divided doses) and clindamycin (40 mg/kg per day in four divided doses, intravenously. In a large number of cases, the surgical consultant will elect to incise and/or debride the lesions.

Omphalitis

Background

Omphalitis is an infection of the umbilical cord and surrounding tissues. Although formerly an important cause of neonatal mortality, the disease is now rare in developed countries because of advances in antisepsis and local care of the umbilical cord stump. When infection occurs, the usual pathogens *are S. pyogenes* (group A streptococcus) and *S. aureus*; group B streptococcus, and Gram-negative enteric rods may also be isolated. Children are at risk during the first 2 weeks of life.

Pathophysiology

After ligation at delivery, the umbilical stump undergoes necrosis as a result of interruption of its blood supply. Bacterial colonization follows soon after birth. In rare cases, because of colonization by virulent bacteria or ill-defined host factors, colonizing organisms may invade the umbilical cord stump and surrounding tissues. The initial infection is cellulitis, but peritonitis, liver abscess, and/or sepsis may ensue in short order.

Clinical Findings

Omphalitis is characterized first by drainage and later by erythema around the umbilical cord stump. Late in the course of infection, infants manifest the signs of sepsis, including lethargy, irritability, and hypothermia or hyperthermia. Laboratory studies are normal early in the course.

Because a small amount of drainage and patchy erythema can occur in the absence of infection, the diagnosis of omphalitis may be difficult. There are no definite clinical criteria for early infections, and laboratory tests are not helpful. The findings suggestive of omphalitis are 1) purulent, foul-smelling drainage from the umbilical cord with any erythema of the anterior abdomen, or 2) any drainage with erythema that completely encircles the umbilicus. Induration and erythema of the anterior abdomen wall are definite indicators of infection.

Management

Infants who appear toxic, have induration and erythema of the abdominal wall, or show signs clearly suggestive of omphalitis (purulence and patchy erythema or light drainage plus circumferential erythema) should be presumed to have a significant infection and require IV antibiotic agents. Appropriate therapy is oxacillin (150 mg/kg per day in four divided doses) and gentamicin (7.5 mg/kg per day in three divided doses for term infants). In some cases, minimal drainage or erythema may be present, but the findings are not sufficient for the diagnosis of omphalitis. The parents of these infants should be instructed to swab the cord after each diaper change and to observe the child for any changes in activity or feeding. Reexamination in 24 hours is advisable if the problem has not resolved.

Neonatal Mastitis

Background

Mastitis is an infection of the breast tissue that affects prepubertal children only during the first 2 to 5 weeks of life. In most cases, *S. aureus* is the offending organism, although 5 to 10% of the infections are caused by Gram-negative enteric bacteria. Girls are affected twice as commonly as boys.

Pathophysiology

Maternal hormones cross the placenta during gestation and stimulate hypertrophy of neonatal breast buds in both males and females. Usually, the enlargement subsides within 2 weeks. In occasional cases, bacteria are able to invade the hypertrophied glandular tissue, leading to abscess formation. Manipulation of the breast to excrete “witch's milk” may be a predisposing factor.

Clinical Manifestations

The primary finding in neonatal mastitis is a warm, erythematous, enlarged breast bud (Fig. 84.21). With disease progression, purulent drainage from the nipple may occur and there is tenderness to palpation. Only 25% of infants are febrile or appear ill.

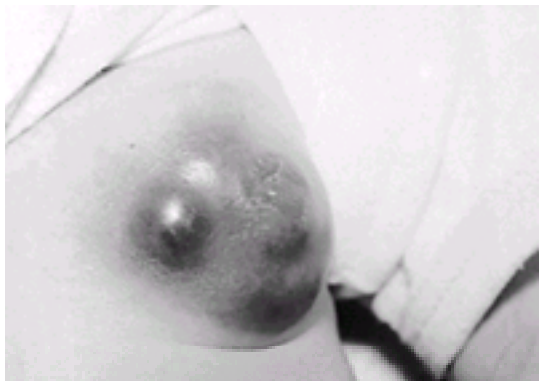

FIGURE 84.21. A 2-week-old infant with mastitis, characterized by erythema and induration.

Mastitis in the infant must be distinguished from physiologic hypertrophy, which resolves spontaneously. The normal breast bud that enlarges in response to stimulation by maternal hormones is neither red nor tender; if any drainage is present, the material is milky white, rather than yellow, and does not contain polymorphonuclear leukocytes or bacteria on Gram stain.

Culture of the purulent drainage yields the pathogen in most cases, but the blood is usually sterile. Because these infections are well localized, the WBC count is usually in the normal range.

Management

Occasionally, an infant will appear septic and require appropriate supportive therapy, as described earlier in this chapter. For the remainder, IV antibiotics should be initiated, pending the results of cultures. Oxacillin (150 mg/kg per day in four divided doses) and gentamicin (7.5 mg/kg per day in three divided doses) provide appropriate coverage for the expected pathogens. Surgical consultation for possible incision and drainage is advisable in the case of local fluctuance.

Septic Arthritis

Background

Septic arthritis is an infection within a joint space. The incidence of this infection is unknown; however, Nelson reported that 20 children were admitted yearly between 1966 and 1970 to two large pediatric services in Dallas. As opposed to adults, children who develop septic arthritis are generally otherwise healthy. Boys are affected twice as often as girls.

The bacterial cause of septic arthritis varies with age. During the first 2 months of life, group B streptococcus and *S. aureus* predominate. Gram-negative enteric bacilli, *Candida* species, and *N. gonorrhoeae* are seen sporadically.

Between 3 months and 3 years of age, *S. aureus* emerges as the single most common pathogen, being isolated from 80 to 90% of children with septic arthritis. The incidence of disease caused by *H. influenzae* has declined significantly since the introduction of the conjugated vaccine, but this organism remains a concern in children in this age range who have not been immunized against this pathogen. Group A streptococcus and *S. pneumoniae* cause occasional cases.

The incidence of gonococcal arthritis in teenagers has varied in different reports, depending on the prevalence of sexual activity in the population studied. In most studies, *N. gonorrhoeae* has been the most common cause of septic arthritis among adolescents, trailed closely by *S. aureus.*

Many other organisms occasionally invade the joint space, some only in special circumstances. *P. aeruginosa* shows a peculiar predilection for septic arthritis of the foot after puncture wounds. In the child with sickle cell anemia, *Salmonella* species often cause septic arthritis. The Gram-negative bacilli are almost never recovered from previously healthy children but are seen in immunosuppressed patients. *M. tuberculosis* is a rare causative agent but may be isolated at any age.

Pathophysiology

Septic arthritis generally results from the hematogenous dissemination of an organism, into either the joint or the bony metaphysis. Rarely, a pathogen gains access to the joint by direct inoculation or spread from a contiguous site of infection. Although many children give a history of recent trauma, the role played by injury remains unknown. In gonococcal arthritis, the initial site of infection may be the genitals, pharynx, or rectum. Dissemination from the cervix follows menstruation when shedding of the organism is highest.

Bacteria in the joint space evoke an inflammatory response with an infiltration of neutrophils. The accumulation of purulent material distends the joint capsule, producing the physical and radiographic findings.

Clinical Features

Infection within a joint produces pain and limitation of motion. Thus, the site of the arthritis determines the specific complaint. Ninety percent of children have a monoarticular arthritis that involves the lower extremity (hip, knee, and ankle). Thus, limp (see Chapter 43) is the most common initial manifestation. If a joint in the arm is involved, mobility of the upper extremity will be decreased (see Chapter 36).

With infections in deeper joints, the pain may radiate to contiguous anatomic structures. Children with a septic hip often complain of an ache at the knee, and sacroiliac arthritis may mimic appendicitis, pelvic neoplasm, or UTIs. Although the duration of symptoms in septic arthritis is less than 3 days in more than 50% of children with these conditions, the delay in diagnosis may reach 3 to 4 weeks with sacroiliac arthritis.

The findings are often vague in the first 6 months of life. Pyoarthritis may cause paradoxical irritability and an increase in crying on being fondled, as seen with meningitis. The infant with a septic hip usually lies quietly, holding the leg abducted and externally rotated.

Of children with septic arthritis, 60 to 70% have a temperature of 38.5°C (101.2°F) or higher. The absence of fever occurs most commonly in the adolescent with a gonococcal infection or in the neonate. Infants with infections caused by *H. influenzae,* rare since the advent of the conjugated vaccine, almost invariably have a high fever. An erythematous swelling may surround a superficial joint that is infected. Although a temperature difference exists between the affected and unaffected sites, it can be difficult to discern in the febrile child. Inflammation within the joint distends the capsule and produces pain with movement. If a child allows the physician to manipulate an extremity through a full range of motion, septic arthritis is unlikely.

The ESR and CRP are the most consistently abnormal laboratory study. Molteni observed an elevated ESR in 32 of 37 (86%) children with septic arthritis; the median value was 50 mm. The peripheral WBC count usually varies from less than 5,000 to more than 20,000/mm^3. Although a leukocytosis with a shift to the left commonly occurs, as many as 20% of children will have a WBC count less than 10,000/mm^3. If septic arthritis is diagnosed early, a radiograph of the joint will not show any pathologic changes. The first radiographic alteration to be noted is edema of the adjacent soft tissues, which is not pathognomonic of inflammation in the joint. Later, distension of the capsule becomes visible, and bony destruction may be seen late in the course of the infection.

The thickness of the tissues that surround the hip joint makes the detection of an effusion difficult by physical examination. A radiograph of the hip should always be obtained if infection in this joint is possible. Early in the course, the tendon of the obturator internus is displaced as the muscle passes over the distended hip capsule. Continued accumulation of an inflammatory exudate forces the femoral head laterally and upward, disrupting the arc formed by the femoral head and the pelvis (Shenton's line). The hip may actually dislocate with intra-articular infection in the young infant, but this is an unusual radiographic finding in older children. Ultrasound examination is useful for the detection of a small effusion not apparent on radiograph.

No constellation of laboratory and radiographic results can rule out the diagnosis of septic arthritis; an analysis of the joint fluid is mandatory if the index of suspicion is high (see Procedures, Section VII). Infection causes an infiltration of polymorphonuclear leukocytes into the joint space. Although intraarticular WBC counts greater than 100,000/mm^3 are traditionally associated with infection, a lesser cellular response is often noted. Nelson found a WBC count in the joint fluid below 25,000/mm^3 in 9 (34%) of 31 children with proven bacterial arthritis. The joint fluid glucose is reduced to less than 40 mg/dL in only 25 to 50% of patients, but the Gram stain of the synovial fluid shows organisms in 75%. Because inflammatory exudates have bacteriostatic properties, cultures of joint fluid yield an organism in only 60% of cases. A pathogen is recovered from the bloodstream in 40% of children with septic arthritis, more commonly if *H. influenzae* or *S. pneumoniae* is the cause of the disease.

The complications of septic arthritis include both local and distant spread of the infection. Osteomyelitis often accompanies joint infections in the first year of life because of the location of the metaphysis within the joint capsule. During the process of hematogenous dissemination, bacteria may invade sites other than the joint. Simultaneous infections may occur in the meninges, pericardium, or the soft tissues; these are particularly common with *H. influenzae.*

Management

Septic arthritis demands prompt management; in particular, infection in the hip joint should be considered an emergency. Pressure generated by purulent material within the joint space can compromise the vascular supply of the femoral head, leading to necrosis and eventual loss of normal ambulation.

The initial treatment is aimed at relieving the pressure within the joint and controlling the infection. At the time of the diagnostic aspiration, as much purulent fluid as possible should be removed. Immediate surgical intervention is needed for hip infections.

All children with septic arthritis require admission to the hospital for IV antibiotic therapy. The initial choice of antimicrobials depends on the child's age and the Gram stain. If no organisms are apparent on examination of the joint fluid, presumptive antibiotic therapy is begun as follows: 1) 2 months of age or younger—oxacillin 150 mg/kg per day in four divided doses and gentamicin 7.5 mg/kg per day in three divided doses; 2) older than 2 months to less than three 3 years of age—cefotaxime 200 mg/kg per day in four divided doses, ampicillin–clavulanic acid 200 mg/kg per day of ampicillin in four divided doses or, for the penicillin-allergic patient, clindamycin 40 mg/kg per day in four divided doses and chloramphenicol 75 to 100 mg/kg per day in four divided doses; 3) 3–12 years of age—oxacillin 150 mg/kg per day up to a maximum of 6 g/day; 4) adolescents—ceftriaxone 100 mg/kg per day. Ceftriaxone (100 mg/kg/day once daily) may be used as a single agent for children older than 2 months; as further experience confirms the virtual disappearance of *H. influenzae,* oxacillin (150 mg/kg per day in four divided doses) will probably prove sufficient as monotherapy during childhood.

Osteomyelitis

Background

Osteomyelitis is an infection of the bone; a variant, discitis, affects the intervertebral disc space. Bremmer and Neligman estimated that 1 in 5000 children less than 13 years old develop osteomyelitis. Approximately 10 children with bone infections were admitted yearly to the pediatric service at Parkland Memorial Hospital in Dallas between 1959 and 1973.

S. aureus causes osteomyelitis in most cases regardless of age. During the neonatal period, group B streptococcus is the second most common isolate; *N. gonorrhoeae* and Gram-negative enteric bacilli are also found. Group A streptococcus causes 5 to 10% of osteomyelitis in children. Other pathogens are recovered rarely, including *S. pneumoniae, H. influenzae, Y. enterocolitica, Brucella* species, anaerobic organisms, *M. tuberculosis,* and *Actinomyces* species.

P. aeruginosa may infect the bones of the foot after a puncture wound. In children with sickle cell hemoglobinopathies, *Salmonella* species account for almost half the cases of osteomyelitis. Unusual pathogens may be recovered from immunocompromised children.

Pathophysiology

In most children with osteomyelitis, bacteria reach the bone through the bloodstream. Occasional infections follow the direct inoculation of pathogens or spread from a contiguous focus. During hematogenous dissemination, organisms lodge in the sinusoidal vessels of the metaphysis at the site of sludging or thrombosis. Bacterial proliferation evokes an inflammatory exudate. Within the confined space of the bone, the pressure generated by the accumulation of purulent material can necrose the cortex and elevate or rupture the periosteum. If the metaphysis is contained within the joint capsule, septic arthritis may ensue.

Clinical Features

Osteomyelitis causes bone pain as the infection progresses. The site of the osteomyelitis determines the presentation of the disease. In 90% of cases, a single bone is involved. The femur and tibia are the most common bones infected, making limp (see Chapter 43) a common presentation. In a study of 100 consecutive children with a limp seen at the ED of The Children's Hospital of Philadelphia, osteomyelitis was diagnosed in 2%.

Osteomyelitis affects the bones of the upper extremity in 25% of cases. These children complain of pain on motion of their upper extremities (see Chapter 36).

The multiplicity of bones that may be involved leads to a wide spectrum of chief complaints. Vertebral osteomyelitis manifests as backache, torticollis, or stiff neck, and involvement of the mandible causes painful mastication. Infection of the pelvis is particularly elusive and may masquerade as appendicitis, neoplasm, or UTI. Infants with osteomyelitis localize the symptoms less well than older children. Initially, irritability may be the only complaint.

Fever exceeds 38.5°C (101.2°F) in 70 to 80% of children with osteomyelitis. The infant with a long bone infection often manifests pseudoparalysis, an unwillingness to move the extremity. Movement may also be decreased in the older child, but to a lesser degree. Point tenderness is seen almost always in osteomyelitis; however, it is found in other conditions such as trauma, may be difficult to discern in the struggling infant, and does not always occur early in the course of the infection. Percussion of a bone at a point remote from the site of an osteomyelitis may elicit pain in the area of infection.

When purulent material ruptures through the cortex, diffuse local erythema and edema appear. This finding occurs often in infants, but late in the course, and is confined primarily to children in the first 3 years of life (before the cortex thickens sufficiently to contain the inflammatory exudate). Weissburg et al. noted swelling of the extremity in 14 of 17 patients with

osteomyelitis less than 1 month old.

The ESR or CRP provides a useful screening test for osteomyelitis because bony infection almost always leads to an elevation. Nelson found an ESR less than 15 mm/hour in only 4 of 88 children with osteomyelitis, and the mean value was 70 mm/hour. Although the WBC count may reach a level of 20,000/mm^3, it falls within the normal range in two-thirds of cases. Cultures from the blood yield an organism in 50% and from the bone in 70% of children with osteomyelitis.

If osteomyelitis is suspected, radiographs of the affected area should always be obtained, even though they are often normal early in the course. The first change, noted after 3 to 4 days, is deep soft-tissue swelling seen as a subtle shift of the lucent deep-muscle plane away from the bone. Within 3 to 10 days, the muscles swell and obliterate the lucent planes that usually separate them radiographically. Visualization of osseous destruction requires the loss of 40% of the bony matrix in an area at least 1 cm in diameter. This amount of demineralization occurs only after 10 to 12 days of infection. At this stage, lytic lesions and periosteal elevation are apparent on the radiograph (Fig. 84.22).

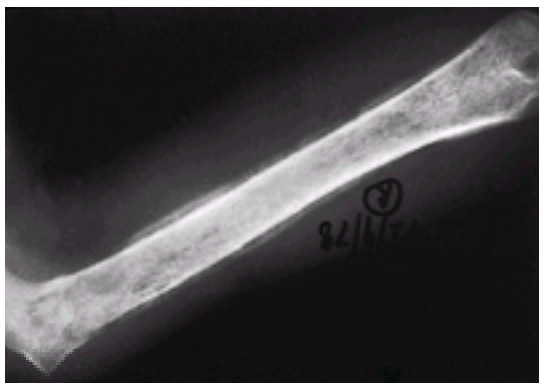

FIGURE 84.22. Radiograph showing lytic lesions and periosteal elevation with osteomyelitis.

Radionuclide scanning provides a useful diagnostic tool for the clinician. Uptake of compounds such as technetium is seen at sites of increased metabolic activity, which occurs in an infection before sufficient bony destruction has occurred to be seen on conventional radiographs. If scintigraphy is available, the patient strongly suspected to have osteomyelitis despite a normal radiograph should have this study. However, the absence of increased uptake does not preclude bony infection. Some patients will have decreased uptake because the accumulation of purulent material lessens the flow of blood to the site; occasionally, in children, the scan may be entirely normal early in the course. When scintigraphy is not diagnostic and clinical suspicion persists, magnetic resonance imaging is useful.

The complications of osteomyelitis include the spread of infection, either locally or to remote sites, chronic infection, and irreparable bony destruction.

Management

All children strongly suspected or known to have osteomyelitis require admission to the hospital for IV antibiotic therapy. Those with a low likelihood of bony infection can be reevaluated in 12 to 24 hours and can have a technetium scan at that time, if the clinical findings are not definitive. The emergency physician should withhold antibiotics until the orthopedic surgeon has been contacted about culturing the bone at the site of infection. Infants should subsequently receive oxacillin, 150 mg/kg per day in 4 divided doses, and gentamicin, 7.5 mg/kg per day in 3 divided doses; older children can be treated with oxacillin alone.

Genitourinary Infections

UTIs in the child are discussed in this section and in Chapter 54. Of the sexually transmitted genital infections, the four most important diseases in the United States are gonorrhea, syphilis, herpes, and *Chlamydia.* When any sexually transmitted disease is diagnosed, concomitant infection with HIV should be considered (see Chapter 85). These diseases are discussed subsequently in this chapter and in Chapter 94.

Urethritis/Cervicitis

Background

Urethritis may have an infectious or noninfectious cause (see Chapter 54). Among the infectious causes, *C. trachomatis* and *N. gonorrhoeae* predominate, with *C. trachomatis* being the single most common cause of urethritis in the United States. In males, approximately 50% of nongonococcal urethritis results from infection with this organism, and 20% of patients with gonococcal urethritis have a concurrent infection with *C. trachomatis.* In addition, most cases of postgonococcal urethritis are caused by this pathogen. In females, *C. trachomatis* causes the dysuria-pyuria syndrome, mucopurulent cervicitis, and pelvic inflammatory disease.

Chlamydial infections are common in adolescents. Although uncommon in prepubertal children, such infections should be considered in the setting of sexual child abuse, particularly in association with rectogenital gonorrhea.

Clinical Manifestations

Urethritis causes dysuria and discharge in the male. Persistent discharge in a young boy or adolescent treated for gonorrhea, in particular, should alert the clinician to infection with *C. trachomatis.*

Mucopurulent cervicitis caused by *C. trachomatis* in the adolescent girl is characterized by an erythematous friable cervix and the accumulation of purulent yellow endocervical secretions. Gram stain of the discharge shows more than 10 polymorphonuclear leukocytes per microscopic field under ×1000 magnification. This organism also causes the "sterile" dysuria-pyuria syndrome; females with this condition complain of dysuria, at times described as originating external to the urethra at the level of the labia, and have pyuria in the face of a negative urine culture. In prepubertal girls, *C. trachomatis* is a cause of vaginitis, usually with scant or no discharge.

Regardless of the clinical findings, chlamydial infection should be suspected with any sexual contact because most infections are asymptomatic.

An etiologic diagnosis in urethritis/cervicitis relies on the clinical syndrome, Gram stain of any discharge, and specific identification of the organism, either by antigen/DNA detection or culture. In general, culture is preferred for prepubertal children.

Management

If an etiologic diagnosis is not possible based on the clinical findings and the results of Gram stain, treatment should be given for both *N. gonorrhoeae* and *C. trachomatis.* Infections with *C. trachomatis* in adolescents and children older than 8 years are treated with doxycycline (100 mg twice daily) or a single dose of azithromycin (1 g). Children less than 8 years of age may be given azithromycin (20 mg/kg as a single dose) or erythromycin (40 mg/kg per day in four divided doses for 10 days) and should have a "test-of-cure" culture after this course of therapy.

Urinary Tract Infection

Background

Infections occur along the urinary tract from the tip of the urethra to the renal parenchyma. Clinical syndromes that may accompany infections include urethritis, cystitis, and pyelonephritis. *Bacteriuria* refers to the presence of bacteria in the urine, arising from any site in the urinary tract, with or without causing symptoms. Significant bacteriuria describes the presence of bacteria in sufficient quantity such that infection is more likely than contamination. Significant bacteriuria may be asymptomatic and the clinical syndromes mentioned above may occur in the absence of infection. Urethritis has been discussed previously. Because cystitis and pyelonephritis may coexist or be difficult to distinguish clinically and share a similar etiology, they are discussed together, using the generic term *urinary tract infection.* Urethritis is covered in a previous section.

The predominant pathogen isolated in UTIs is *E. coli,* which is recovered in 90% of cases. Next in frequency are other members of the *Enterobacteriaceae* family, including *Enterobacter* and *Klebsiella.* Among the Gram-positive organisms, enterococci are seen at all ages, staphylococcal species occur most often in adolescents, and group B streptococci are recovered primarily in infants and during pregnancy. *P. aeruginosa, Candida albicans,* and a number of other bacteria and fungi infect patients with immunocompromise, anatomic obstruction, or indwelling catheters. Cystitis may be caused, in addition, by adenoviruses.

The frequency of infections of the urinary tract varies by age, sex, and race. Overall, infections occur commonly in neonates, decrease in frequency during childhood, and then rise in incidence after puberty in sexually active females. Males are more commonly infected than females in the first 6 months of life, in part because of a higher incidence of congenital urinary tract anomalies, but they rarely acquire infections beyond this period. Females have a rather high incidence of symptomatic infection between 6 months and 2 years of age and of asymptomatic bacteriuria throughout childhood. In several studies, as many as 15% of febrile white girls between 6 months and 2 years of age had positive urine cultures, a rate fivefold greater than seen in African-American females and males of any race. Among males, circumcision decreases the likelihood of UTIs.

Pathophysiology

Bacteria may invade the urinary tract by ascension or hematogenously. In most cases, the organisms colonize the urethral area and ascend to the bladder. The higher incidence of UTI in girls is often attributed to the shorter female urethra. Hematogenous spread to the kidney may occur at times in neonates but rarely thereafter.

Those organisms that cause UTIs have certain distinct properties. In comparison to other Gram-negative rods, the few strains of *E. coli* that are most commonly recovered from the urine share recognized virulence factors, including increased adherence to uroepithelial cells and higher quantities of K antigen.

Children with UTIs have a higher incidence of genitourinary anomalies than the general population, although in most infections, no anatomic or functional abnormalities are identified. Lesions that obstruct the flow of urine and/or predispose to incomplete emptying of the bladder contribute to an increased risk of infection. Additional host factors that play a role include an alkaline urinary pH and glucosuria.

Clinical Manifestations

The manifestations of UTIs vary with age, being particularly nonspecific in infancy. During the neonatal appearance, a septic appearance (see Chapter 69) or fever is often the only finding. UTIs in infants may also cause vomiting, diarrhea,

irritability, and reportedly, meningismus.

Beyond 2 to 3 years of age, symptoms more often point to the urinary tract. For all practical purposes, strict differentiation between upper and lower tract disease is not feasible for the clinician in most cases, and children who are febrile (38.5°C [101.2°F]) should be assumed to have pyelonephritis. On the other hand, some patients will have typical syndromes that localize disease to the upper or lower tract. Typically, children with cystitis appear relatively well and complain of dysuria and suprapubic pain. On examination, they have a lower-grade fever and tenderness on the suprapubic area. In contrast, patients with pyelonephritis may be toxic and usually have additional symptoms, including vomiting and flank pain. The physician is often able to elicit tenderness to percussion in the costovertebral area, either unilaterally or bilaterally.

The mainstays of diagnosis are the urinalysis and culture of the urine, both of which require the clinician to make an interpretation that is influenced by the method of collection and processing, as well as the clinical syndrome exhibited by the patient.

Urine is analyzed directly using both a chemical reagent strip (dipstick) and microscopy, looking most specifically, in regard to infection, for the presence of leukocyte esterase, nitrites, WBCs, and bacteria. Either spun or unspun urine may be studied through the microscope, with or without the aid of a Gram stain. Spinning should be done in accordance with a standardized protocol. When a clean urine specimen is centrifuged at 2000 rpm for 5 minutes and examined under high power (×40), each leukocyte (per high power field [hpf]) represents 5 to 10 cells/mm^3, with 10 to 50 WBC/mm^3 (5 to 10/hpf) being the upper limit of normal. One organism per high power field seen on Gram stain of a spun specimen correlates with a colony count of 10^5 organisms or more.

In interpreting a urine culture in children, the physician must keep in mind that the guidelines for positivity were developed based on data in adults and that the significance of colony counts applies most explicitly to voided specimens. Given these caveats, it is generally accepted that a colony count of greater than 10^5 on a single sample indicates a probability of infection of 80%, increasing to 90% when repeated once and 95% when done a third time. Colony counts between 10^4 and 10^5 of single organisms are somewhat suggestive of infection and merit another culture. On a catheterized specimen, a result of $^3 10^4$ organisms points to infection. The criterion for a suprapubic aspirate is greater than 10^3.

In general, a single negative finding on one parameter of the urinalysis does not exclude a UTI. Taken together, however, negative testing for both leukocyte esterase and nitrates by dipstick alone, or even more so in combination with a microscopic examination that shows the absence of pyuria, makes the diagnosis of a UTI (as opposed to asymptomatic bacteriuria) in a male infant older than 6 months of age or a female older than 2 years, highly unlikely. A schema for the use of urinalysis and urine culture is presented in Figure 84.23. As illustrated for older children who are somewhat older but not yet toilet-trained, in the absence of a high likelihood of UTI a priori, the urinalysis may be collected using a bag. However, if the urinalysis is positive, a specimen for culture should preferably be obtained by catheterization or suprapubic aspiration.

FIGURE 84.23. Diagnostic approach to infants and children with fever, symptoms specifically suggestive of urinary tract infection, and/or non-specific symptoms and signs compatible with urinary tract infection. Use of urinalysis and culture in the diagnosis of UTI **(A)** in children age 2 years or younger and **(B)** in those older than 2 years. [a]UA obtained by catheterization or suprapubic aspiration. [b]UA (only) may be obtained using a urine collection bag.

Bacteremia accompanies UTIs primarily during the first 6 to 12 months of life. In the young infant, bacteremia may be present in the absence of fever and should be suspected in any children during the first year of life with a temperature 39°C (102.2°F) or higher. Indications for a CBC and blood culture with a suspected UTI include 1) signs of clinical toxicity (extreme tachycardia, low blood pressure, shaking chills); 2) age younger than 3 months; and 3) age 3 months to 1 year and temperature greater than 39°C (102.2°F). Only children with dehydration require measurement of electrolytes. When a diagnosis of pyelonephritis, as opposed to cystitis, is being entertained, consideration should be given to ascertaining the BUN and creatinine in the serum.

Management

When the diagnosis of UTI has been established, as a result of earlier urine culture, or is presumed, based on the clinical syndrome and findings on urinalysis, antibiotic therapy is indicated. If available, the results of susceptibility testing should guide the selection of antimicrobial agent. In all other cases, an antibiotic is chosen to cover the most likely pathogens.

Most patients respond to oral antibiotic therapy. Indications for IV administration of antibiotics include 1) clinical toxicity;

2) age younger than 3 to 6 months; 3) vomiting, refusal to drink, or other factors making the delivery of oral medications unreliable; 4) adverse anatomic factors, such as an obstruction to urinary flow; and 5) a known positive culture for a pathogen resistant to oral agents.

For ill-appearing patients, IV ampicillin (200 mg/ kg per day in four divided doses) plus gentamicin (7.5 mg/kg per day in three divided doses, adjusted for gestation age and weight, see Table 84.4) are given. Options for oral therapy include TMP-SMZ (8 mg/kg per day of trimethoprim in two divided doses) or cefixime (8 mg/kg per day as a single dose). If the pathogen is susceptible to ampicillin, amoxicillin provides effective oral coverage.

SPECIFIC INFECTIONS

Sexually Transmitted Diseases

Gonorrhea

Background

The term *gonorrhea* is used in this chapter to describe a genital infection with *N. gonorrhoeae,* but the organism can also produce disease at other sites. The incidence of gonorrhea remains high, and almost 1 million cases are reported yearly in the United States. The highest rate of gonococcal infection is found in adolescents and young adults. When looked for, however, gonorrhea is fairly common in prepubertal youngsters. Paradise et al. studied 38 consecutive prepubertal females with vaginitis from the ED at the Children's Hospital of Philadelphia, recovering *N. gonorrhoeae* from four. The age-specific rates for gonorrhea reported in one study were 6.1 per 100,000 from 0 to 9 years old and 37.4 per 100,000 from 10 to 14 years old. Gonococcal conjunctivitis occurs most often in the first month of life, although cases have been described in older children and adults. Other forms of gonococcal infection are rare in children.

Pathophysiology

A child acquires gonorrhea by direct contact with infected secretions. The gonococci adhere to the surface of columnar epithelial cells and then penetrate through the intracellular spaces to the subepithelial tissues. They evoke an inflammatory response with polymorphonuclear leukocytes. Extension may occur through the lymphatics or the bloodstream.

Clinical Manifestations

The most common form of infection with *N. gonorrhoeae* seen among children is infection of the genitals. Prepubertal girls develop a vaginitis, rather than a cervicitis as seen in adult women, because of differences in the vaginal mucosa. Vaginal irritation, dysuria, and a discharge are the most common complaints. Boys have a urethral discharge and occasionally swelling of the penile shaft (Fig. 84.24) or urinary retention. Fever, systemic signs and symptoms, and spread to the pelvic organs in females occur rarely.

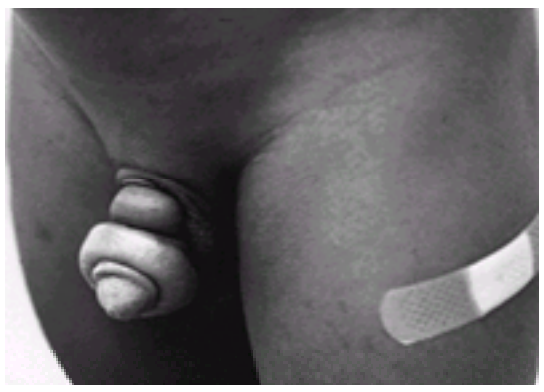

FIGURE 84.24. Penile venereal edema in a 2-year-old boy with gonococcal urethritis.

A Gram stain will show Gram-negative intracellular diplococci in most children with gonorrhea. Cultures allow definitive identification of the organism, a crucial issue from the medicolegal standpoint. Pharyngeal and rectal cultures are positive in 5 to 10% of cases in which the organism is not isolated from the genital tract.

Despite the widespread use of prophylactic solutions in the eyes of newborns, gonococcal conjunctivitis continues to appear sporadically. A thick, purulent discharge quickly replaces the initial mild erythema from chemical irritation. Gram stain of the exudate usually shows the organism, and cultures are almost always positive.

Disseminated gonorrhea (Fig. 84.25) and pelvic inflammatory disease emerge as problems in the sexually active adult. A complete discussion of these entities is found in Chapter 94.

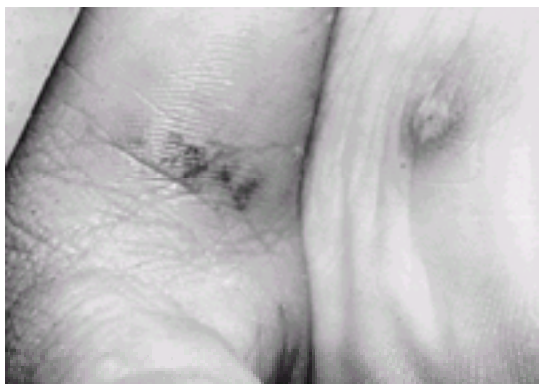

FIGURE 84.25. Disseminated gonococcemia in an adolescent girl. A pustule is seen on the sole of her foot, and a hemorrhagic vesicopustule has formed in the web space between her fingers, which are held adjacent to her foot.

Management

All children with suspected genital gonorrhea should have cultures of the genitals, pharynx, and rectum, as well as serologic tests for syphilis and HIV. Because of the medicolegal considerations, treatment should be delayed in prepubertal children until the diagnosis is confirmed by culture, when possible. If the results of sensitivity testing are not available, ceftriaxone is used for therapy, in a dose of 125 mg intramuscularly. Alternatives for adolescents include single-dose oral therapy with cefixime (400 mg), ciprofloxacin (500 mg), or ofloxacin (400 mg). Spectinomycin is an acceptable alternative for penicillin-allergic patients in a dose of 40 mg/kg (maximum 2.0 g) intramuscularly. Because the usual mode of acquisition often involves sexual contact, a report must be made to the appropriate community department that deals with child abuse. Concomitant therapy should be provided for *C. trachomatis,* with either azithromycin (20 mg/kg, maximum 1 g) or doxycycline (in children older than 8 years).

In infants less than 1 month of age with conjunctivitis, particularly when purulent, consideration should be given to a Gram stain and culture of the ocular exudate. The finding of Gram-negative diplococci on the smear warrants treatment with ceftriaxone (50 mg/kg as a single dose) while awaiting culture results.

Therapy for adolescents with uncomplicated gonorrhea and salpingitis is discussed in Chapter 94.

Syphilis

Syphilis is an infection caused by *Treponema pallidum.* The disease is common among adults in the United States, but it is uncommonly encountered in children. However, scattered congenital infections are seen, and an incidence of 20 per 1 million has been reported in older adolescents.

Congenital syphilis usually presents with the same clinical picture as other intrauterine infections (rubella and cytomegalovirus). Characteristic features include jaundice and hepatosplenomegaly in an ill-appearing newborn. However, some infants have only a few stigmata, and the diagnosis is often overlooked in the nursery. These children may turn up in the first months of life with skin lesions, a persistent nasal discharge, and painful extremities (pseudoparalysis of Parrot). Dark-field examination of cutaneous lesions can identify the spirochetes, and the serologic test for syphilis is positive. In addition, radiographs may show lesions of the long bones. Diagnostic criteria are provided in Table 84.19.

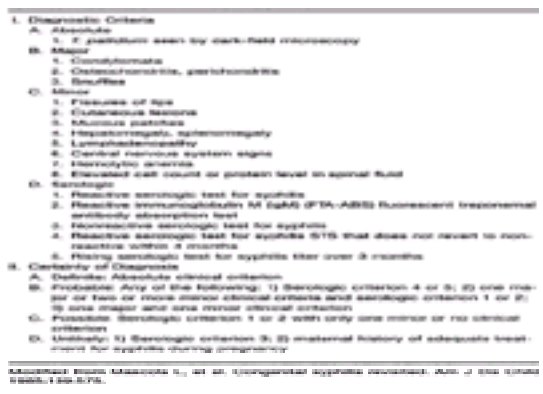

Table 84.19. Criteria for Diagnosis of Neonatal and Early Congenital Syphilis

Acquired syphilis appears in the teenager, as in the adult, in the first stage with a chancre (Fig. 84.26) and in the second stage with cutaneous or mucosal manifestations. The rash of secondary syphilis may resemble pityriasis rosea, and all sexually active patients diagnosed with this disease, particularly with involvement of the palms or soles, should have a serologic test for syphilis. Other lesions include white patches on the mucous membranes and flat-topped warts (condyloma lata) around moist areas. Such lesions shed spirochetes detectable by dark-field microscopy.

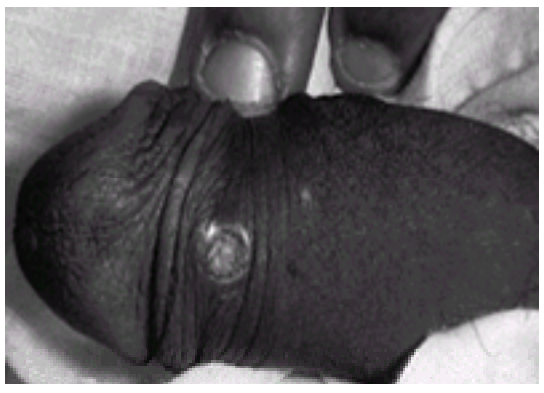

FIGURE 84.26. Chancre in an adolescent with serologically confirmed syphilis.

Congenital syphilis rarely is diagnosed in the ED, and the delay involved in confirmation moves the treatment out of the realm of the emergency physician. All such children require admission to the hospital. In acquired disease, benzathine penicillin (2.4 million units) is given intramuscularly in a single dose for early syphilis and in three doses, each separated by 1 week, for syphilis of more than 1 year's duration.

Herpes Genitalis

Herpes simplex can infect the genitals, as well as other anatomic sites. Although the most common cause of genital ulceration seen among adolescents and adults at venereal disease clinics, this entity is unusual in prepubertal children.

Genital pain is a common complaint with infections caused by herpes simplex and may precede the appearance of the lesions. Characteristically, the virus produces grouped vesicles on an erythematous base (Fig. 84.27); however, erosion of the overlying skin often leaves only painful ulcers at the time of the first visit. Particularly with a primary infection, the inguinal lymph nodes enlarge.

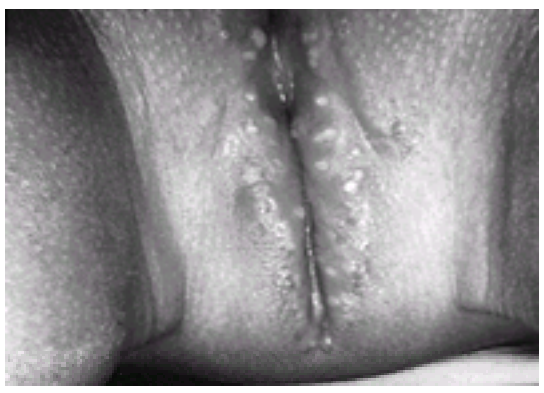

FIGURE 84.27. Genital herpetic lesions in a young girl who was sexually abused (Photo courtesy of Stephen Ludwig, MD)

Visual inspection often suffices for the diagnosis in the adolescent. A Tzanck smear (see Chapter 99) positive for giant cells lends further weight to the clinical impression; either immunofluorescent staining of a scraping from the base of a vesicle or a viral culture can verify the diagnosis. In children, a culture should always be obtained because the disease is rarely seen and needs medicolegal confirmation. Serologic tests for syphilis and HIV and bacterial cultures are appropriate to rule out coexisting sexually transmitted infections. Although it is occasionally spread by nonsexual contact, the physician must explore the possibility of sexual abuse when herpes genitalis occurs before puberty. Oral acyclovir therapy is indicated for primary infections; the dosage is 45–60 mg/kg per day in three divided doses.

Systemic Viral Infections

Viral Syndrome

The term *nonspecific viral syndrome* is used to refer to a generalized illness presumed clinically to be caused by a virus and characterized by malaise and, usually, fever. Numerous agents, including influenza, enteroviruses, and herpesvirus (roseola), have been implicated. Nonspecific viral syndromes and viral URIs account for most of the febrile visits made by children to the ED.

A viral syndrome begins with malaise and usually with fever, as well. The temperature varies from 37°C (98.6°F) to more than 40°C (104°F), greater elevations occurring at times in children less than 2 years old. Particularly with influenza, children who are able to verbalize their discomfort complain of diffuse aching. There may be a cough or occasional bout of emesis. Signs of mild inflammation may be seen in the upper respiratory tract.

The physician arrives at a diagnosis of a nonspecific viral syndrome by excluding other diseases on the basis of the history and physical examination. At times, a WBC count may help the physician determine whether the young child with a high fever is at risk for occult bacteremia. Treatment is limited to antipyresis with acetaminophen (15 mg/kg per dose)

or ibuprofen (10 mg/kg per dose) for patients who do not respond to acetaminophen and the maintenance of an adequate oral intake. Antibiotics will not prevent secondary bacterial infections and are not to be prescribed routinely. The parents must be instructed to seek further care if the fever persists for more than 48 hours.

Erythema Infectiosum (Fifth Disease)

Erythema infectiosum, or fifth disease, is an exanthematous illness of childhood caused by parvovirus B19. It occurs most commonly between 2 and 12 years of age. The appearance of a rash marks the onset of the disease; fever or other prodromal symptoms are uncommon. The rash involves the face initially, conferring on the child a “slapped-cheek” appearance. Maculopapular lesions erupt 24 hours later, initially on the upper portion of the extremities, and they then spread both proximally and distally. Fading of the central portion of the lesions gives a lacelike appearance to the rash. Adolescents in particular may develop arthralgia or arthritis, and patients with chronic hemolytic anemias, such as sickle cell disease, are at risk for aplastic crisis. During pregnancy, infection with parvovirus B19 causes fetal hydrops in approximately 10% of cases. There is no specific therapy in the normal host, but immunocompromised patients may benefit from intravenous gamma globulin.

Infectious Mononucleosis

Background

Infectious mononucleosis (IM) is a disease characterized by malaise, fever, pharyngitis, lymphadenopathy, and splenomegaly. In 1968, Henle and Henle showed that EBV causes this illness.

EBV infections are common, but usually asymptomatic, during the first years of life. In children, sporadic infections occur, and IM is occasionally diagnosed. By late adolescence, 50 to 90% of all people are seropositive, a higher prevalence of antibodies being found in teenagers of lower socioeconomic status. EBV infections are again common between 15 and 25 years of age in more affluent persons; half the seroconversions are accompanied by the clinical manifestations of IM.

Pathophysiology

Several studies have suggested that EBV is transmitted by intimate oral contact. The virus infects B lymphocytes that may spread to the various lymphoid tissues in the body. Sensitized T cells destroy the infected B cells and limit the production of virus; these cells are the atypical lymphocytes that appear in the circulation.

Clinical Manifestations

IM begins insidiously with fever and malaise. Three-fourths of children with this illness complain of a sore throat. Although a child may recover from IM in 7 to 10 days, the symptoms usually last for 2 to 4 weeks. This persistence of symptoms separates the patient with IM from those with pharyngitis caused by group A streptococcus or other viruses. Occasionally, the onset resembles that of infectious hepatitis.

The child with IM is febrile in 90% of cases at presentation. Enlarged lymph nodes are uniformly palpable. Although the lymphadenopathy may be limited to the cervical region, involvement of the axillary and inguinal areas occurs commonly. Pharyngitis caused by any pathogen produces an increase in the size of the anterior cervical nodes, but EBV characteristically affects the posterior cervical and submental glands as well (Fig. 84.28). In 75% of cases, the pharynx is inflamed, often with an exudate. The spleen enlarges in 60% of children and the liver in 25%. Periorbital edema and a diffuse maculopapular rash are seen occasionally.

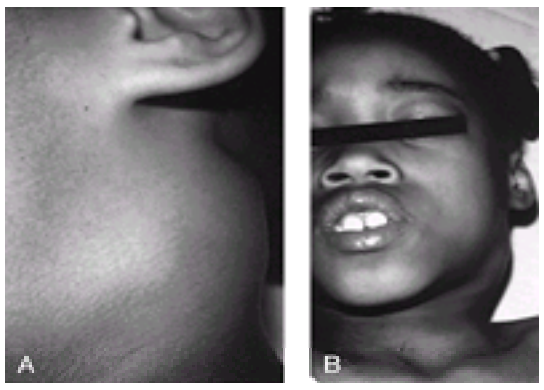

FIGURE 84.28. Posterior cervical adenopathy in a child with infectious mononucleosis. **A.** Close-up of posterior adenopathy. **B.** Anterior and posterior cervical node enlargement.

The hemoglobin and hematocrit are normal in the uncomplicated disease. Although the total WBC count does not often increase much beyond 15,000/mm^3, levels up to 30,000/mm^3 are seen in 10 to 15% of children. A higher leukocyte count casts some doubt on the diagnosis of IM. There is an absolute lymphocytosis with many atypical mononuclear cells; however, 16% of children presenting with IM in one series had fewer than 10% atypical lymphocytes, and 50% had fewer than 20% of such cells. The mainstay for the diagnosis of IM in the adult is the heterophil antibody test, but these antibodies reach levels detectable by routine assays in only 50% of children. Confirmation of a heterophil-negative case of IM requires EBV-specific serologic assays. The AST and ALT levels are elevated in most children.

The most worrisome complications of IM for the emergency physician are splenic rupture and airway obstruction. Even minor trauma can cause a rent in the capsule of the enlarged spleen seen in IM; these children manifest the usual signs of intraperitoneal hemorrhage. Occasionally, massive lymphoid hyperplasia of the tonsils occludes the airway, leading to stridor and retractions. The site of narrowing is easily visualized on examination. Less common complications include encephalitis, pneumonia, myocarditis, hemolytic anemia, and thrombocytopenia.

Management

A WBC count and heterophil antibody titer usually suffice for the confirmation of the clinical diagnosis. EBV-specific antibodies are indicated only for heterophil-negative cases.

Specific therapy is not available. Adequate rest and nutrition should be maintained, and antipyretic agents will increase the child's comfort. The treatment of a child with uncomplicated IM does not require the administration of corticosteroids, but the duration of the illness can be shortened and the patient made more comfortable by judicious use of a short course.

If complications develop, the child should be admitted to the hospital. Corticosteroids almost always shrink dramatically the enlarged tonsils of the child with airway obstruction. Prednisone is given at 2 mg/kg for the first day and tapered over 5 days. Studies with acyclovir have shown only minimal efficacy.

Measles

Background

Measles is a disease caused by a specific myxovirus and is characterized by fever, cough, coryza, conjunctivitis, and a rash. Since the large scale introduction of effective vaccines, the incidence of this disease has decreased significantly. Currently, several thousand cases still are reported each year to the CDC.

Pathophysiology

Measles virus enters the body through the upper respiratory tract, where local replication is believed to occur. A transient viremia ensues, and virus spreads to the reticuloendothelial system. A secondary viremia then follows, producing the clinical disease.

Clinical Findings

Fever and malaise herald the onset of measles. During the course of the illness, the temperature often rises to 40°C (104°F). Within 24 hours, coryza, conjunctivitis, and cough develop. Koplik's spots appear on the buccal mucosa by the third day of fever. These are seen as fine white spots on an erythematous background and have been likened to grains of sand. The rash erupts on the fourth or fifth day. The exanthem is maculopapular in appearance and begins on the face and neck. The lesions are heaviest on the upper portion of the body, often coalescing. As the rash advances down the trunk, the prodromal findings (cough, coryza, conjunctivitis) and the Koplik's spots resolve. The rash involves the extremities on its third day but has already begun to fade on the face.

A leukopenia accompanies uncomplicated measles. Specific antibodies, initially absent from the serum, reach detectable levels 2 weeks after the onset of illness.

Complications, which are unusual, fall into two categories: 1) extension of the viral infection and 2) secondary bacterial infection. The virus itself may produce inflammation of the lower respiratory mucosa, leading to laryngotracheitis, bronchitis, and/or pneumonia. Encephalitis occurs in 1 of every 1000 cases of measles; this is a debilitating illness with a mortality rate of 15%. Thrombocytopenia and corneal ulcerations are seen rarely. Lymphoid hyperplasia in the bowel can occlude the lumen of the appendix, leading to inflammation of this organ; histologic examination of surgically removed tissue confirms the diagnosis of measles on the basis of the characteristic giant cells. Acute purulent OM is the most common bacterial complication of measles, and cervical adenitis occasionally occurs. Pneumonia, although usually viral, may have a bacterial etiology.

Management

The clues gathered from the history and physical examination suffice for the diagnosis of measles by the experienced clinician. However, as the number of cases dwindles, the physician is less likely to be familiar with the disease. Often, serologic studies are required to confirm the cause, particularly among the first few children seen in sporadic outbreaks. Ordinarily, acute and convalescent titers drawn 1 to 2 weeks apart are used for a determination, but some laboratories are able to determine the serodiagnosis on a single specimen by testing for measles-specific IgM antibodies.

Measles runs a self-limited course. Bed rest and antipyretic therapy help keep the child comfortable. Antitussives, antihistamines, and topical ophthalmic preparations have no role. Prophylaxis against secondary bacterial infections with antibiotics is not warranted.

Children with uncomplicated measles or superficial secondary infections such as otitis and cervical adenitis can be treated as outpatients. Hospitalization is required when significantly severe laryngotracheobronchitis is evident, as discussed earlier in this chapter. Lower respiratory tract or CNS involvement necessitates admission to the hospital.

Measles is a preventable disease. Otherwise healthy, susceptible contacts should receive immune serum globulin, 0.25

mL/kg; the dose is increased to 0.5 mL/kg for immunocompromised patients. If within 72 hours of infection, vaccine is indicated in addition, unless the patient is less than six months of age or immunocompromised. (See Appendix D).

Roseola

Roseola infantum, or exanthem subitum, is a common, self-limiting, viral infection of infants caused in most cases by human herpesvirus-6; recently, reports indicate that human herpes virus, which occurs less often, produces a similar syndrome in slightly older children. The child, usually less than 3 years old, presents with a high fever, ranging up to 40.5°C (104.9°F), and a paucity of physical findings. There may be mild irritability but no coryza, pharyngeal infection, or conjunctivitis. After 2 to 4 days of illness, the fever drops precipitously and a rash appears. The lesions are discrete, pink maculopapules, 2 to 3 mm in diameter. They fade with pressure and do not coalesce. The exanthem appears on the trunk initially and spreads outward. Roseola resolves without complications other than an occasional febrile convulsion. The diagnosis of roseola is made on the basis of the clinical course, often in retrospect. If a WBC count is obtained, leukopenia with lymphocytosis will be seen. Treatment is limited to antipyretic agents.

Rubella

Background

Rubella is a childhood infection caused by a specific togavirus. Before the advent of vaccination, epidemics occurred every 6 to 9 years; 488,796 cases were reported to the CDC in the United States in 1964, the year of the last outbreak in this country. Presently, there are an estimated 25,000 to 50,000 cases per year. Rubella traditionally occurs in children 5 to 9 years of age, but the incidence among teenagers is increasing.

The initial site of inoculation is the upper respiratory tract, where local replication occurs. A viremia ensues, disseminating the virus to the skin.

Clinical Manifestations

Only 10% of children experience prodromal symptoms such as fever, malaise, cough, and mild conjunctivitis. However, such complaints are often voiced by the adolescent. The rash begins on the face and spreads downward, reaching the extremities by the end of the second day. The lesions are pink maculopapules that may coalesce. The lymph nodes in the postauricular, suboccipital, and posterior cervical chains enlarge and become somewhat tender. During the first 2 days of illness, the temperature usually rises, but remains under 39°C (102.2°F).

The WBC count often decreases in rubella, and a few atypical lymphocytes may appear. A fourfold rise in specific antibodies occurs after 10 to 14 days.

Complications of rubella are rare in children but include encephalitis, thrombocytopenia, and arthritis or arthralgia. Painful and/or swollen joints often occur in adolescents. Encephalitis has an incidence of 1 in 6000 cases of rubella and usually resolves spontaneously.

Management

Rubella is difficult to diagnose clinically because of its rare occurrence and the plethora of exanthems that have a similar appearance. Situations that require a definite etiologic diagnosis, such as pregnancy in an adolescent, demand serologic confirmation. Children with rubella can be managed as outpatients with antipyretic therapy. Only the rare child with encephalitis or thrombocytopenia requires admission to the hospital.

Varicella/Zoster

Background

Herpesvirus varicellae causes two clinical illnesses, varicella (chickenpox) and zoster (shingles). Varicella occurs during a primary infection and zoster occurs after a reactivation of latent virus.

Varicella is a common infection in children, usually those between 2 and 8 years of age, although the incidence is expected to drop now that an effective vaccine has been introduced. Almost 90% of susceptible household contacts of an index case develop the disease. By adolescence, serologic surveys have shown a seropositive rate of 70 to 80%. Zoster, however, affects adults predominantly. More than 60% of cases occur in persons older than 45 years.

Pathophysiology

The virus enters the body through the oropharynx and replicates locally. Viremia presumably occurs after exposure and before the onset of the exanthem. During an episode of varicella, the virus invades sensory nerve endings and ascends to the dorsal root ganglion where it becomes latent. Zoster follows reactivation of the latent virus.

Clinical Findings

Varicella. A mild prodrome that lasts 1 to 3 days often precedes the exanthem of varicella; however, the first sign of illness may be the rash. Most children develop fever, usually less than 39.5°C (103.1°F) and may complain of malaise. The fever usually has subsided within 24 hours of the appearance of the skin lesions. Recurrence of significant fever should serve as a warning sign for suspicion of complications. Lesions erupt initially on the upper trunk, neck, or face and

spread centripetally. Pruritus is universal.

The abnormal findings on physical examination are limited to the elevated temperature and the skin and mucous membrane lesions. Initially, the exanthem consists of erythematous papules that evolve into vesicles and then pustules over 6 to 8 hours (Fig. 84.29). The early vesicles have a diameter of 2 to 4 mm and a “dewdroplike” appearance. Because new lesions erupt in crops for 2 to 4 days, papules, vesicles, and pustules are usually seen together. An exanthem involves the mucosa of the oropharynx and, occasionally, the vagina. The severity of the cutaneous manifestations varies widely, and there may be from 1 to more than 1000 lesions.

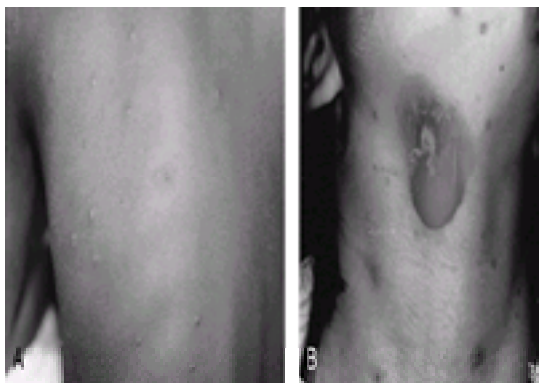

FIGURE 84.29. A. Typical lesions of varicella. **B.** Bullous varicella.

There are few laboratory derangements in varicella. The WBC count occasionally shows a leukocytosis, and the AST and ALT may be mildly elevated. In adolescents, the chest radiograph reveals an interstitial infiltrate in 5 to 10% of patients, even though there may be no respiratory symptoms.

Varicella runs a self-limited course in most cases but is occasionally a more serious illness. Fleisher et al. reviewed 96 children hospitalized with complications of this disease during a 5-year period at The Children's Hospital of Philadelphia. Of the group, 81 were immunocompetent children more than 1 month old; they experienced complications, including encephalitis (20), pneumonia (5), hepatitis (8), bacterial superinfection (22), Reye syndrome (17), unusual cutaneous manifestations (5), medication overdoses (5), exacerbation of an underlying disease (2), and dehydration (1). Simultaneous streptococcal pharyngitis can occur.

Encephalitis takes two forms: 1) a diffuse cerebritis with coma and seizures, and 2) a cerebellitis with ataxia. Both varieties may occur before, during, or after the cutaneous eruption. Because bacterial meningitis can also complicate varicella, an analysis of the CSF should be done even if viral encephalitis is suspected. There will often be a mild pleocytosis (10 to 300 cells) and a slight elevation of the protein (40 to 80 mg/dL). If the encephalopathy is thought to be related to Reye syndrome, a serum ammonia should be obtained.

Starting in approximately 1990, a number of authors reported an increasing incidence of group A streptococcal complications with varicella, including primarily sepsis and necrotizing fasciitis. The diagnosis of streptococcal sepsis should be considered in patients who appear toxic, remain febrile for 5 days or more, or develop a fever after being afebrile for more than 48 hours.

Zoster. Zoster appears suddenly in most children without any warning symptoms (pain or pruritus). The lesions are grouped vesicles on an erythematous base in a dermatomal distribution. In 15 to 20% of cases, extradermatomal cutaneous dissemination is seen. However, spread to the viscera does not occur in the immunocompetent child. If the eruption follows the ophthalmic branch of the trigeminal nerve, the cornea may be involved. The appearance of vesicles on the tip of the nose should evoke a suspicion of ocular involvement that can be best seen after fluorescein staining of the eye.

Management

Visual inspection suffices for the diagnosis of varicella; no laboratory studies are indicated. Acetaminophen is given to control the fever, and antihistaminic drugs provide some relief from the pruritus. Aspirin is contraindicated because of an association with Reye syndrome. Although some investigators have speculated about a relationship between the use of ibuprofen and the development of fasciitis, this remains unproven. Diphenhydramine (5 mg/kg per day), hydroxyzine (2 mg/kg per day), or other antihistamines may be used to decrease pruritus. The child cannot attend school for 1 week after the eruption of the first lesion.

Immunosuppressed children with varicella require hospitalization to receive IV acyclovir, which has been shown to prevent visceral dissemination; recent reports on the use of high-dose oral acyclovir in children with mild immunosuppression await further confirmation before this approach can be routinely recommended. Complications that mandate admission to the hospital for immunocompetent patients include fasciitis, Reye syndrome, pneumonia, and encephalitis, except in the mildest cases. Superficial bacterial infections such as impetigo, cellulitis (if fasciitis is thought to be unlikely), and adenitis can be treated with oral antibiotic therapy such as dicloxacillin 50 mg/kg per day, cephalexin 50 mg/kg per day, or erythromycin 40 mg/kg per day. Children with deeper bacterial infections (i.e., septic arthritis) should receive antibiotics by IV.

For immunocompetent children, oral acyclovir (80 mg/kg per day in four divided doses) given within 24 hours of the onset of the rash reduces the duration of fever and the number and duration of skin lesions. Indications for use have not been

formalized, but consideration is warranted for patients who are at some risk for a particularly severe course: infants less than 6 months old, adolescents (older than 12 years), children receiving long-term aspirin therapy or being treated with oral/inhaled steroid, patients with chronic cutaneous (e.g., atopic dermatitis) or pulmonary (e.g., cystic fibrosis) disorders, and those with fever above 40°C (104°F) and a large number of lesions noted as early as the first day of the eruption (particularly if case follows a household contact). Oral acyclovir should be considered for the pregnant adolescent, but its use remains controversial in this situation.

Zoster usually requires no specific therapy. Although famciclovir and valacyclovir are recommended for adults, they have not been shown to be efficacious in children. Antipruritic and antipyretic agents provide symptomatic relief. Immunocompromised children should be admitted to the hospital. IV acyclovir therapy benefits immunocompetent children with unusually severe disease and reduces the incidence of dissemination in immunocompromised patients. Ocular involvement merits consultation with an ophthalmologist.

Anyone likely to experience a severe episode of varicella should receive prophylaxis after a significant exposure to a patient contagious for varicella-zoster virus (household or close contact for more than 1 hour in a closed environment). Varicella-zoster immunoglobulin (VZIG), 1 vial (125 units) per 10 kg, is indicated for susceptible normal adults, pregnant women, and immunocompromised children. Newborns whose mothers have had onset of varicella within 5 days before or 2 days after delivery should receive 125 units, as soon as possible (see Appendix D).

Miscellaneous Infections

Babesiosis

Babesia species, particularly *B. microti are* protozoa, transmitted by the bite of an *Ixodes* tick, which also serves as a vector in Lyme disease. Cases of this infection have been reported with increasing frequency along both coasts and in the upper Midwest. The clinical picture of babesiosis resembles that of malaria and is characterized by anorexia, malaise, fatigue, and intermittent chills, sweats, and fevers as high as 40°C (104°F). Other than an elevated temperature, physical findings are absent or limited to mild hepatosplenomegaly. Patients with asplenia or immunocompromise are susceptible to severe, or even life-threatening, disease. Laboratory findings include hemolytic anemia with reticulocytosis, a normal or slightly decreased leukocyte count, mild thrombocytopenia, and elevated liver enzymes in half the cases. Microscopic examination of a peripheral smear confirms the presence of intracellular and extracellular ring forms, similar to those of *Plasmodium falciparum;* specific serologic assays are available as well, but the results are often delayed for several weeks and do not distinguish between acute infection and asymptomatic seropositivity. Therapy is reserved for patients with mild to moderate infections or a predisposition to severe disease. The treatment of choice is clindamycin (40 mg/kg per day in four divided doses) and quinine (30 mg/kg per day in three divided doses) intravenously.

Botulism

Background

Botulism is a paralytic illness produced by neurotoxins elaborated by *Clostridium botulinum.* The disease may result from the ingestion of preformed toxin or from the elaboration of toxin by organisms in a wound or in the GI tract. Of particular concern to the physician who cares for children is infantile botulism caused by toxin formed in the intestines. In the United States, approximately 400 cases of this disease were reported to the CDC in 1984.

During growth, *C. botulinum* releases neurotoxins that are the most potent poisons known on a weight basis. They interfere with neurotransmission at peripheral cholinergic synapses by blocking the release of acetylcholine.

Clinical Manifestations

Botulism from the ingestion of toxin causes vomiting in 50% of cases. The patients complain of weakness and a dry mouth; constipation and urinary retention may occur. Paralysis is noted within 3 days, usually affecting the cranial nerves first and then the extremities. The patients are alert and afebrile. Abnormalities of the neurologic examination include ptosis, extraocular palsies, fixed dilated pupils, symmetric weakness, and hyporeflexia. Both the ileus and urinary retention seen in this disease may lead to abdominal distension.

Infantile botulism occurs in children in the first 6 months of life who are otherwise healthy. The duration of symptoms before hospitalization ranges from 1 to 20 days. Breast-fed, Caucasian infants from middle-class families are primarily affected. Constipation is the first symptom of the disease but may not be sufficiently severe to draw attention to any underlying illness. After several days, mild lethargy, weakness, and a decreased appetite are noted. Occasionally, the onset of lethargy and weakness may be so precipitous as to resemble bacterial sepsis or meningitis.

On examination, the infant is quiet, with little discernible movement, and has a weak cry. Fever is not a part of this syndrome. The child sucks on a nipple with difficulty and may be unable to swallow. The absence of a gag reflex, profound hypotonia, and hyporeflexia in infantile botulism helps distinguish this illness from bacterial sepsis.

The WBC count is normal in botulism. Organisms may be recovered by anaerobic culture techniques from the GI tract in infantile or wound botulism, but identification of the toxins requires the specialized facilities of the public health department. Children with botulism have an electromyogram that shows a characteristic pattern of brief duration, small amplitude, overly abundant motor unit action potentials (BSAP).

Respiratory failure is a potential life-threatening complication in botulism of any variety, and ventilatory support is often required. The profound bulbar weakness in infantile botulism often prevents an adequate fluid intake; dehydration occurs

frequently.

Management

Because no test is immediately available to diagnose infantile botulism, the initial evaluation of these infants aims at excluding other causes of lethargy and weakness such as sepsis, poliomyelitis, myasthenia gravis, neuropathy, and drug ingestion. A lumbar puncture is often performed in the ED to rule out meningitis, and electrolytes and a BUN are useful to assess hydration.

The children all require admission to the hospital (Table 84.20). Monitoring of pulse and respiratory rate should start in the ED. An IV line should be started for the administration of fluids to correct dehydration and in anticipation of a possible respiratory arrest. Neither antibiotics nor antitoxin ameliorate the course of infantile botulism. Because they may potentiate the neuromuscular blockade, aminoglycoside antibiotics should be avoided when treating for possible sepsis.

Ensure adequate ventilation.	Achieve venous access and maintain
Administer oxygen, as indicated.	hydration.
Initiate cardiorespiratory monitoring.	Obtain laboratory studies.

Table 84.20. Infantile Botulism: Immediate Management

Children with foodborne and wound botulism also require admission to the hospital. Antitoxin is available from the CDC and should be administered after consultation with the staff at this agency.

Cat-Scratch Disease

CSD is an infection caused by *Bartonella henselae.* Approximately 80% of cases occur in patients less than 20 years of age. Traditionally, this disorder has been thought of primarily as an infection of regional lymph nodes (typical CSD), but the spectrum of the disease has been expanded to include infections of other organ systems by *B. henselae* (atypical CSD). In addition, this same agent causes severe infections (bacillary angiomatosis and peliosis hepatis) in patients with HIV (see Chapter 85).

More than 90% of children with typical CSD have a history of exposure to cats. The most complete form of the illness begins with a the appearance of a pustule at the site contact, 7 to 10 days after exposure. Lymphadenopathy follows within 1 to 6 weeks. The regional nodes enlarge and become mildly to moderately tender on palpation. In one-third of cases, the glands become fluctuant. The epitrochlear, axillary, inguinal, and cervical nodes are commonly affected.

Manifestations of atypical CSD include encephalitis, aseptic meningitis, neuroretinitis (blindness and stellate macular lesions of the retina), Parinaud's syndrome (conjunctivitis and preauricular adenitis), hepatitis, osteolytic lesions, and fever of unknown origin.

In typical cases with a history of exposure, particularly to a kitten, and characteristic lymph node enlargement, no specific diagnostic studies are needed. In atypical disease or with lymphadenopathy that is not characteristic, serum should be sent to a reference laboratory for an indirect fluorescent antibody test.

Various antibiotics have been reported to have some efficacy for CSD, including rifampin, TMP-SMZ, ciprofloxacin, and azithromycin. The only controlled study of patients with typical CSD found that azithromycin (10 mg/kg in a single dose on day 1, followed by 5 mg/kg once daily on days 2 through 5) shortened the duration of adenopathy. Nodes that persist and become fluctuant usually resolve after needle aspiration.

Ehrlichiosis

At least two *Ehrlichia* species cause two similar infectious illnesses: human monocytic ehrlichiosis and human granulocytic ehrlichiosis. *Ehrlichia* species are transmitted by a number of tick vectors throughout the United States. Both forms of the disease resemble Rocky Mountain spotted fever (RMSF) and are characterized by fever, chills, malaise, myalgias, and headache. Unlike RMSF, a rash occurs in only 40% of patients with human monocytic ehrlichiosis and rarely with granulocytic ehrlichiosis. The infection may spread to involve the meninges. Laboratory findings include anemia, thrombocytopenia, hyponatremia, and mildly elevated liver enzymes. Examination of the peripheral smear may show inclusions (morulae) in monocytes or granulocytes (depending on the species), but a definitive diagnosis relies on serology, which is available only from reference laboratories. The treatment of choice is doxycycline (3 mg/kg per day in two divided doses), even for children less than 8 years of age; chloramphenicol (75 to 100 mg/kg per day in four divided doses) offers an alternative.

Malaria

Malaria is a protozoal infection that occurs in humans and is caused by four species: *Plasmodium falciparum, Plasmodium vivax, Plasmodium ovale,* and *Plasmodium malariae.* Although one of the most common infections in the world, malaria is rare in the United States; about 1000 cases occur annually. However, it should be considered in recent immigrants from tropical areas and in travelers; there have been a handful of indigenously acquired cases since 1990, including transmission in the northeastern part of the country. At times, malaria may be fatal, particularly with infections from *P. falciparum.*

Pathophysiology

Plasmodia gain access to the bloodstream when a person is bitten by a female anopheline mosquito or, rarely, by transfusion or transplacentally. The sporozoites invade liver parenchymal cells that subsequently release merozoites. The merozoites enter the erythrocytes and eventually cause their destruction. Subsequent cycles of infection account for the recurrent nature of the illness.

Clinical Manifestations

Fever characterizes malarial infections. Although specific patterns may occur, chaotic elevations of the temperature are more common because of multiple broods of parasites. The typical attack starts with a chill and tachycardia. Within 1 hour, the fever rises to 40°C (104°F) or higher. A profuse diaphoresis follows and lasts for several hours. Hepatosplenomegaly is a common finding. The febrile episodes may be accompanied by hypotension and jaundice. Laboratory findings include anemia, leukocytosis, thrombocytopenia, and hyperbilirubinemia.

Complications of malaria resulting from *P. falciparum* include massive hemolysis (blackwater fever), renal failure, pulmonary edema, and cerebral dysfunction. Neurologic signs of cerebral malaria include decreased level of consciousness, behavioral changes, hallucinations, seizures, and rarely, focal signs. The CSF is usually normal.

Management

Malaria must be suspected in any febrile child recently in an endemic area and in febrile neonates born to women who are recent immigrants from such regions. Thick and thin blood smears should be done and, if positive, will confirm the diagnosis. However, these smears may be negative in some infections and are often not easily interpreted except by experienced personnel. In such circumstances, a CBC is helpful. Thrombocytopenia often occurs in children during febrile episodes; the platelet count usually falls in the range of 50,000 to 100,000/mm ^{3}.

Children with suspected or proven malaria from areas without resistant *P. falciparum* should be given chloroquine phosphate. (Updated information on the geographic prevalence of resistance is available from the CDC or the World Wide Web at www.CDC.gov.) The initial dose is 10 mg/kg of base (maximum 600 mg) followed by 5 mg/kg at 6, 24, and 48 hours. Patients unable to take oral medication require IV quinidine, beginning with a loading dose of 10 mg/kg infused over 1 hour with cardiac monitoring. Uncomplicated infections with chloroquine-resistant *P. falciparum* can be treated with oral quinine sulfate (30 mg/kg per day in three divided doses) plus oral pyrimethamine–sulfadoxine (Fansidar) (less than 1 year old, ¼ tablet; 1 to 3 years, ½ tablet; 4 to 8 years, 1 tablet; 9 to 14 years, 2 tablets; older than 14 years, 3 tablets). Fansidar should not be used in patients with known sensitivity to sulfa drugs. An alternative regimen for chloroquine-resistant *P. falciparum* is mefloquine, 15 to 25 mg/kg (maximum 1250 mg) as a single oral dose, but this drug should not be used in children who weigh less than 15 kg, in pregnant women, or in patients with epilepsy or psychiatric disorders. In life-threatening infections with known or suspected resistant organisms, quinidine should be administered by IV, as already described; this drug carries a significant risk of adverse reactions and is thus reserved for emergencies.

Parasitic Infestations of the Gastrointestinal Tract

A variety of parasites can infest the GI tract or invade the body via this route. These infestations may be asymptomatic, being detected on screening examination of the stool. In the ED, the diagnosis is usually entertained when a parent reports observing a worm (pinworm, *Enterobius vermicularis*) or in acute diarrheal diseases, particularly among patients who are immunosuppressed or recently in underdeveloped countries. Two pathogens deserve particular consideration in the United States among immunocompetent children. *Cryptosporidium parvum,* first reported as a cause of human disease in 1976 and well known as a pathogen in children with HIV, has been reported to be the responsible pathogen in 2 to 5% of cases of nonspecific, watery diarrhea and has been implicated in several large, waterborne outbreaks, one affecting an estimated 400,000 persons in Milwaukee, Wisconsin, in 1993. *Giardia lamblia* is also a waterborne parasite that can survive even in running waters and has been described as a cause of diarrhea among campers and hikers who have ingested water from streams. Both cryptosporidiosis and giardiasis merit consideration in day care settings. Table 84.21 summarizes the clinical symptoms and treatment of gastrointestinal parasites in children.

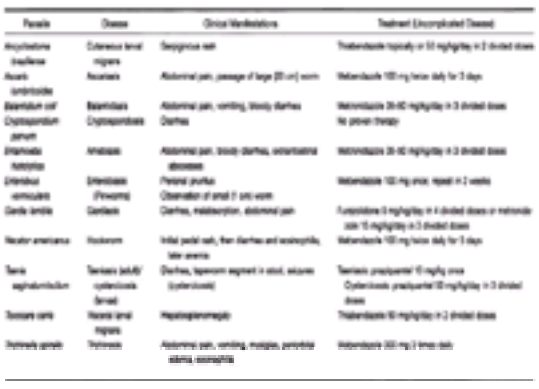

Table 84.21. Parasitic Diseases

Rabies

Rabies is a viral infection of the brain that is almost invariably fatal. Although the actual disease is rare in the United States, potential exposure in the form of animal bites commonly occurs. Dogs bite 1 million to 2 million people per year, and 75% of the victims are children.

The decision whether or not to give prophylaxis for rabies is influenced by the species of animal, the condition of the animal, the ability to study the animal, the type of exposure, and the prevalence of rabies in the region (Fig. 84.30). The incidence of rabies in the area should be available from the local health department. If a sleeping or preverbal child has had close exposure to a bat in an area where rabies is endemic in this species, prophylaxis is indicated even in the absence of a visible bite wound because of the occurrence of several pediatric cases in this circumstance. When the physician determines that prophylaxis is necessary, human rabies immune globulin (HRIG) 20 IU/kg and human diploid cell vaccine (HDCV) are used. After cleaning the wound, half the HRIG is given locally and the remainder at a distant site. Vaccine must be given in the deltoid muscle (not the thigh or buttock) in a different extremity then that used for the HRIG. (See Practical Information, Appendix D.

FIGURE 84.30. Diagnostic approach for the management of the child with a mammalian bite wound.

Rocky Mountain Spotted Fever

Background

RMSF is an infection caused by *Rickettsia rickettsii.* It is the most commonly occurring rickettsial disease in the United States. Ticks harbor the organism and transmit it to humans during blood sucking. Although the disease is named for the area of the country in which the causative agent was discovered, most cases of RMSF occur in the states along the eastern coast of the United States. The incidence of the disease peaks during the warmer months. It affects persons of all ages, but two-thirds of the victims are children and adolescents. Each year, approximately 1000 cases are reported to the CDC.

Pathophysiology

Rickettsiae are inoculated during blood sucking by a tick and replicate locally. In animal models, the organisms disseminate hematogenously and invade the endothelial lining of the small blood vessels. The infection induces an inflammatory reaction in these cells that leads to swelling, necrosis, thrombosis, and finally, occlusion of the vascular lumina. The diffuse vasculitis underlies the widespread clinical manifestations that may involve almost every organ.

Clinical Manifestations

The incubation period of RMSF ranges from 2 to 10 days but usually lasts 1 week. The initial symptoms of headache and malaise are followed by fever. The rash erupts on the third or fourth day of illness. In more than half the cases reviewed by Vianna, the exanthem appeared first on the wrists and ankles and then spread inward toward the trunk. The initial lesions are maculopapular but become hemorrhagic in the ensuing 24 to 48 hours if the disease remains unchecked (Fig. 84.31).

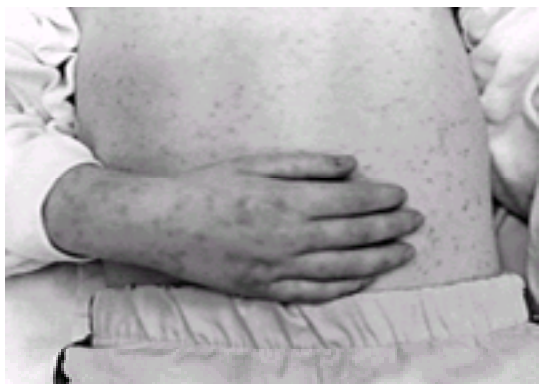

FIGURE 84.31. Rocky Mountain spotted fever.

The findings on examination vary with the duration of the disease. Early in the course of the illness, the child remains alert. Conjunctivitis and a rash may be the only signs. Edema begins in the periorbital regions and involves the extremities as the vasculitis progresses. Mild splenomegaly is found in one-third of cases. Vomiting is common. Although the sensorium is clear initially, obtundation and, finally, coma develop after several days of illness.

The WBC count remains normal or rises slightly with RMSF. Thrombocytopenia occurs in 75% of patients during the first stages of the disease; later, DIC may develop with a prolonged PT and PTT, as well as elevated fibrin split products. Most patients have hyponatremia but no other electrolyte abnormalities. Bradford and Hawkins noted a decrease in the serum sodium among 88% of children. Immunofluorescent staining has been used to identify rickettsiae in the endothelial cells of dermal vessels from skin biopsies but is not routinely available for diagnosis. Even when myocarditis remains clinically silent, the electrocardiogram may show signs of cardiac dysfunction. The earliest changes consist of an elevation of the ST segment; later, the P-R interval may become prolonged and arrhythmias may occur. In some cases, mild increases in the CSF cell count and protein concentration are seen.

Complications of RMSF that demand immediate attention include shock and seizures. Vascular collapse occurs from the combination of endothelial damage and inadequate hydration in the vomiting, obtunded patient. Tachycardia, hypotension, and an impaired peripheral perfusion point to a decrease in the intravascular volume. Convulsions may occur in the comatose child with RMSF. Either hyponatremia or a cerebral vasculitis may underlie the seizure activity. Occasionally, the hemorrhagic diathesis needs immediate treatment in the ED. Myocarditis and nephritis are also seen.

Management

A CBC, platelet count, electrolytes, PT, PTT, and serologic titers should be obtained on the child with suspected RMSF. These studies help pin down the diagnosis and influence the management. Because no routinely available test confirms the diagnosis of RMSF early in its course, treatment must be initiated presumptively. The mildly ill child with a fever, maculopapular exanthem, and a history of a tick bite can be treated as an outpatient. Chloramphenicol (50 mg/kg per day) is the drug of choice for patients less than 8 years of age and tetracycline (50 mg/kg per day) in older youths.

Admission is indicated when there is 1) clinical evidence of toxicity, 2) encephalitis, 3) thrombocytopenia (platelet count less than 150,000/mm^3) or derangements in the clotting studies, and 4) hyponatremia (Na less than 130 mEq/L). In the ED, an IV infusion should be started and sufficient fluids administered to maintain an adequate blood pressure as discussed under Septic Shock (see Chapter 3). Chloramphenicol (50 mg/kg per day) can be given alone if the illness is clearly felt to be RMSF; in practice, however, broader antibacterial coverage (e.g., chloramphenicol plus ampicillin or oxacillin) is often used because bacterial sepsis cannot be excluded.

Tetanus

Clinical tetanus is caused by the toxin produced by *Clostridium tetani.* The disease is rare in the United States (approximately 50 cases annually) because of widespread use of the vaccine. Neonatal tetanus from infections of the umbilicus by the organism continues to be reported occasionally. However, the more common problem for the emergency physician is the use of prophylaxis after traumatic wounds. Both tetanus toxoid (0.5 mL) and human tetanus immunoglobulin (250 units) may be indicated, depending on the wound and the immunization history (Table 84.22). Tetanus-prone wounds include punctures, crush injuries, and injuries contaminated by animal excreta or those left untreated for more than 24 hours. (See Practical Information, Appendix D.)

No. of Primary Immunizations	Years Since Last Booster	Type of Wound	Recommendation
≤2	Irrelevant	Low risk	T
		Tetanus prone	T + TIG
3	10	Low risk	T
		Tetanus prone	T
3	5–10	Low risk	No treatment
		Tetanus prone	T
3	<5	Low risk	No treatment
		Tetanus prone	No treatment

T, tetanus toxoid; TIG, human tetanus immune globulin.

Table 84.22. Guidelines for Tetanus Prophylaxis

Toxic Shock Syndrome

Background

Toxic shock syndrome (TSS) is characterized by severe, prolonged shock and is caused by a toxin produced by *S. aureus.* Todd et al. initially described this syndrome in seven children, aged 8 to 17 years, but most of the subsequently reported episodes have occurred in postpubertal females, often after a menstrual period. About 400 cases of TSS occur annually in the United States.

Colonization by a phage-group-1 toxin–producing staphylococcal strain sets the stage for the development of TSS. The enterotoxins of these organisms are pyrogenic and enhance the susceptibility to shock from endotoxins.

Clinical Manifestations

TSS begins suddenly with high fever, vomiting, and watery diarrhea. Pharyngitis, headache, and myalgias may also occur, and oliguria rapidly develops. Within 48 hours, the disease progresses to hypotensive shock. The patient has a fever, usually 39° to 41°C (102.2° to 105.8°F), a diffuse, erythematous maculopapular rash, and hyperemia of the mucous membranes. Often, marked disorientation evolves.

The WBC count is elevated, with a shift to the left. Thrombocytopenia commonly occurs, being present in more than 75% of children reported by Todd. Most patients develop DIC and have an elevated PT and PTT. Additional abnormalities in the laboratory studies may include an elevated AST, ALT, BUN, creatinine, and creatinine phosphokinase. The serum calcium and phosphate may be decreased.

Management

The initial diagnosis of TSS rests on the constellation of clinical and laboratory findings. The following laboratory tests should be obtained from all children suspected of having this syndrome: CBC, platelet count, PT, PTT, fibrin split products, electrolytes, BUN, creatinine, AST, ALT, and creatinine phosphokinase. Cultures of the blood, urine, stool, throat, and vagina serve to isolate *S. aureus* and to rule out other infectious causes of shock. A lumbar puncture often is required to exclude bacterial meningitis.

The management of TSS is the same as that for shock caused by other organisms (see Chapter 3). The physician should secure venous access with a plastic cannula and administer sufficient fluids to maintain an adequate blood pressure, beginning with 20 mL/kg of normal saline. Monitoring of the intravascular volume and urine output usually requires the placement of central venous and peripheral arterial lines and a urinary catheter.

Suggested Readings

GENERAL

Committee on Infectious Diseases. 1997 Redbook. Elk Grove Village, IL: American Academy of Pediatrics, 1997.

Russo RM, Gururaj VJ, Sheth KA, et al. Pediatric DOA: causes of death. N Y State J Med 1978;78:1045.

BACTEREMIA AND SEPSIS

Alpert G, Hibbert E, Fleisher GR. Case-control study of hyperpyrexia in children. Pediatr Infect Dis J 1990;9:161.

Bas JW, Steele RW, Wittler RR, et al. Antimicrobial treatment of occult bacteremia: a multicenter cooperative study. Pediatr Infect Dis J 1993;12:466–473.

Baskin MN, O'Rourke EJ, Fleisher GR. Outpatient treatment of febrile infants 28–89 days of age with intramuscular administration of ceftriaxone. J Pediatr 1992;120:22.

Bone RG, Fisher CJ, Clemmer TP, et al. A controlled clinical trial of high-dose methylprednisolone in the treatment of severe sepsis and septic shock. N Engl J Med 1987;317:653.

Bratton L, Teele DW, Klein JC. Outcome of unsuspected pneumococcemia in children not initially admitted to the hospital. J Pediatr 1977;90:703.

Fleisher GR, Rosenberg N, Vinci R, et al. Intramuscular vs. oral antibiotic therapy for the prevention of meningitis and other bacterial sequelae in young, febrile children at risk for occult bacteremia. J Pediatr 1994;124:504–512.

Goldschneider T, Gotschlich EC, Artenstein IM. Human immunity to the meningococcus. I. The role of humoral antibodies. J Exp Med 1969; 129:130.

Hanninen P, Terho P, Toivanen A. Septicemia in a pediatric unit. Scand J Infect Dis 1971;3:20.

Harper MB, Bachur R, Fleisher GR. Effect of antibiotic therapy on the outcome of outpatients with unsuspected bacteremia. Pediatr Infect Dis J 1995;14:760–767.

Jacobs RF, Hsi S, Wilson CB, et al. Apparent meningococcemia clinical features of disease due to *H. influenzae* and *N. meningitidis.* Pediatrics 1983;72:469.

Jaffe DM, Fleisher GR. Temperature and total white blood cell count as indicators of bacteremia. Pediatrics 1991;87:670.

Jaffe DM, Tan RR, Davis T, et al. Antibiotic administration to treat possible occult bacteremia in febrile children. N Engl J Med 1987 (in press).

Johnston RB, Sell SH. Septicemia in infants and children. Pediatrics 1964;34:473.

Kuppermann N, Fleisher GR, Jaffe DM. Predictors of occult pneumococcal bacteremia in young febrile children. Ann Emerg Med 1998;31: 679–687.

Lee GM, Harper MB. Risk of bacteremia for febrile young children in the post- *Haemophilus influenzae* type b era. Arch Pediatr Adolesc Med 1998;152:624–628.

Lieu TA, Schwartz S, Jaffe JM, et al. Strategies for diagnosis and treatment of children at risk for occult bacteremia: clinical effectiveness and cost-effectiveness. J Pediatr 1991;118:21.

McCarthy PL, Grundy GW, Speisel SZ, et al. Bacteremia in children: an outpatient clinical review. Pediatrics 1976;57:861.

McCarthy PL, Jekel JF, Dolan TF. Comparison of acute phase reactants in pediatric patients with fever. Pediatrics 1978;62:716.

McCarthy PL, Jekel JF, Staskwick CA, et al. History and observation variables in assessing febrile children. Pediatrics 1980;65:1090.

Pena BMG, Harper MB, Fleisher GR. Occult bacteremia with group B streptococci in an outpatient setting. Pediatrics 1998;102:67–72.

Roberts KB, Borzy MS. Fever in the first eight weeks of life. Johns Hopkins Med J 1977;141:9.

Schutzman SA, Petrycki S, Fleisher GR. Bacteremia with otitis media. Pediatrics 1991;87:48.

Stiehm ER, Damrosch DS. Factors in the prognosis of meningococcal infection. J Pediatr 1966;68:457.

Teele DW, Pelton SI, Myles JAG, et al. Bacteremia in febrile children under 2 years of age: results of culture of blood of 600 consecutive children seen in a “walk-in” clinic. J Pediatr 1975;87:27.

Waskerwitz S, Berkelhammer JE. Outpatient bacteremia: clinical findings in children under two years with initial temperatures of 39.5°C or higher. J Pediatr 1981;99:231.

MENINGITIS

American Academy of Pediatrics, Committee on Infectious Diseases. Dexamethasone therapy for bacterial meningitis in infants and children. Pediatrics 1990;86:130.

Aufricht C, Tenner W, Stanek G. Aseptic meningitis in the decennium of *Borrelia burgdorferi* infection (Lyme disease). Pediatrics 1991; 87:268.

Baker CJ, Barrett FF, Gordon RL, et al. Suppurative meningitis due to streptococci of Lancefield group B: a study of 33 infants. J Pediatr 1973;82:724.

Baraff LJ, Lee SI, Schriger DL. Outcomes of bacterial meningitis in children: a meta-analysis. Pediatr Infect Dis J 1993;12:389–394.

Centers for Disease Control. Progress toward elimination of *Haemophilus influenzae* disease among infants and children—United States, 1987–1995. MMWR 1996;45:901–906.

Feigin RD, Shackelford PG. Value of repeat lumbar puncture in the differential diagnosis of meningitis. N Engl J Med 1973;289:571.

Friedman A, Fleisher G. Meningitis: update of recommendations for the neonate. Clin Pediatr 1980;19:395.

Golden SE. Aseptic meningitis associated with *Ehrlichia canis* infection. Pediatr Infect Dis J 1989;8:335.

Jaffe A, McCormick M, DeAngelis C. Which children with febrile seizures need a lumbar puncture? A decision analysis approach. Am J Dis Child 1983;137:1153.

Kalio MJT, Kilpi T, Antilla M, et al. The effect of a recent previous visit to a physician on outcome after childhood bacterial meningitis. JAMA 1994;272:787–791.

Kornelisse RF, Westerbeek CML, Spoor AB, et al. Pneumococcal meningitis in children: prognostic indicators and outcome. Clin Infect Dis 1995;21:1390–1397.

Laxer RM, Marks MI. Pneumococcal meningitis in children. Am J Dis Child 1977;131:850.

Lepow ML, Carver DH, Wright HT Jr, et al. A clinical, epidemiological, and laboratory investigation of aseptic meningitis during the four year period 1955–1958. Observations concerning etiology and epidemiology. N Engl J Med 1962;266:1181.

McGowan JE, Chesney PJ, Crossley KB. Guidelines for the use of systemic glucocorticoids in the management of selected infections. J Infect Dis 1992;165:1.

Moore CM, Ross M. Acute bacterial meningitis with absent or minimal cerebrospinal fluid abnormalities. Clin Pediatr 1973;12:117.

Odio CM, Faingezicht I, Paris M, et al. The beneficial effects of early dexamethasone administration in infants and children with bacterial meningitis. N Engl J Med 1991;324:1525.

Peltola H, Antilla M, Renkonen OV. Randomized comparison of chloramphenicol, ampicillin, cefotaxime, and ceftriaxone for childhood bacterial meningitis. Lancet 1989;1:1281.

Quaglierello VJ, Scheld WM. Treatment of bacterial meningitis. N Engl J Med 1997;336:708–715.

Rodewald LE, Woodin KA, Szilagyi PG, et al. Relevance of common tests for cerebrospinal fluid screening for bacterial meningitis. J Pediatr 1991;119:363.

Rothrock SG, Green SM, Wren J, et al. Pediatric bacterial meningitis: is prior antibiotic therapy associated with an altered clinical presentation? Ann Emerg Med 1992;21:146.

Schaad UB, Suter S, Gianella-Borradori A, et al. A comparison of ceftriaxone and cefuroxime for the treatment of bacterial meningitis in children. N Engl J Med 1990;322:141.

Schuchat A, Robinson K, Wagner JD, et al. Bacterial meningitis in the United States in 1995. N Engl J Med 1997;337:970–976.

Silver TS, Todd JK. Hypoglycorrhachia in pediatric patients. Pediatrics 1978;58:67.

Syrogiannopoulos GA, Lourida AN, Theodoridous MC, et al. Dexamethasone therapy for bacterial meningitis in children: 2- versus 4-day regimen. J Infect Dis 1994;169:853–858.

Swartz MN, Dodge PR. Bacterial meningitis—a review of selected aspects. N Engl J Med 1965;272:725, 779, 842, 898, 954, 1003.

Wald ER, Kaplan SE, Mason EO, et al. Dexamethasone therapy for children with bacterial meningitis. Pediatrics 1995;95:21–28.

Waler JA, Rathore MH. Outpatient management of pediatric bacterial meningitis. Pediatr Infect Dis J 1995;15:89–92.

Wenger JD, Hightower AW, Facklam RR, et al. Bacterial meningitis in the United States, 1986: report of a multistate surveillance study. J Infect Dis 1990;160:1316.

NASOPHARYNGITIS

Clemens CJ, Taylor JA, Almquist JR, et al. Is an antihistamine-decongestant combination effective in temporarily relieving symptoms of the common cold in preschool children. J Pediatr 1997;130:463–466.

Davis SD, Wedgewood RS. Antibiotic prophylaxis in acute viral respiratory disease. Am J Dis Child 109:544, 1965.

Gwaltney JM, Phillips D, Miller RD, et al. Computed tomographic study of the common cold. N Engl J Med 1994;330:25–30.

Townsend EH Jr, Radebaugh JF. Prevention of complications of respiratory illnesses in pediatrics practice. A double blind study. N Engl J Med 1962;266:683.

PHARYNGITIS

Breese BB, Disney FA. The accuracy of the diagnosis of beta streptococcal infections on clinical grounds. J Pediatr 1954;44:670.

Brien JA, Bass JW. Streptococcal pharyngitis: optimal site for throat culture. J Pediatr 1985;106:781.

El-Daher NT, Hijazi SS, Rawasdeh NM, et al. Immediate vs. delayed treatment of group A beta-hemolytic streptococcal pharyngitis with penicillin V. Pediatr Infect Dis J 1991;10:126.

Honikman LH, Massell BF. Guidelines for the selective use of throat cultures in the diagnosis of streptococcal respiratory infections. Pediatrics 1971;48:573.

Komaroff AL, Aronson MD, Pass TM, et al. Serologic evidence of chlamydial and mycoplasmal pharyngitis in adults. Science 1983;222:907.

Lieu TA, Fleisher GR, Schwartz JS. Rapid streptococcal diagnosis in clinical action. Benefits and limitations. Pediatr Infect Dis J 1986; 5:665.

Lieu TA, Fleisher GR, Schwartz JS. Clinical evaluation of a latex agglutination test for streptococcal pharyngitis: performance and impact on treatment rates. Pediatr Infect Dis J 1988;7:847.

Lieu TA, Fleisher GR, Schwartz JS. Cost-effectiveness of rapid latex agglutination testing and throat culture for streptococcal pharyngitis. Pediatrics 1990;85:246.

Mehra S, Van Moerkerke M, Welck J, et al. Short course therapy with cefuroxime axetil for group A streptococcal tonsillopharyngitis in children. Pediatr Infect Dis J 1998;17:452–457.

Moffet HL, Cramblett HG, Smith A. Group A streptococcal infections in a children's home. II. Clinical and epidemiological patterns of illness. Pediatrics 1964;33:11.

Moffet HL, Siegel AC, Doyle HK. Nonstreptococcal pharyngitis. J Pediatr 1968;73:51.

Pichichero ME, Gooch M, Rodriquez W, et al. Effective short-course treatment of acute group a streptococcal tonsillopharyngitis. Arch Pediatr Adolesc Med 1994;148:1053–1060.

Veasey LG, Weidmeier SE, Ormond GS, et al. Resurgence of acute rheumatic fever in the intermountain area of the United States. N Engl J Med 1987;288:181.

Weisner PJ, Tronca E, Bonin P, et al. Clinical spectrum of pharyngeal gonococcal infection. N Engl J Med 1973;288:181.

OTITIS MEDIA AND EXTERNA

Arola M, Ruuskanen O, Ziegler T, et al. Clinical role of respiratory virus infection in acute otitis media. Pediatrics 1990;86:848.

Barnett ED, Teele DW, Klein JO, et al. Comparison of ceftriaxone and trimethoprim-sulfamethoxazole for acute otitis media. Pediatrics 1997;99:23–28.

Berman S. Otitis media in children. N Engl J Med 1995;332:1560–1565.

Brook I, Frazier EH, Thompson DH. Aerobic and anaerobic microbiology of external otitis. Clin Infect Dis 1992;15;955–958.

Brownlee RC Jr, Deloach WR, Cowan CC Jr, et al. Otitis media in children: incidence, treatment, and prognosis in a pediatric practice. J Pediatr 1969;75:636.

Del Beccaro MA, Mendelman PM, Inglis AF, et al. Bacteriology of acute otitis media: a new perspective. J Pediatr 1992;120:81.

Fries SM. Diagnosis of group A streptococcal pharyngitis in a private clinic: comparative evaluation of an optical immunoassay method and culture. J Pediatr 1995;126:933–936.

Giebink GS, Canafax DM, Kempthorne J. Antimicrobial treatment of acute otitis media. J Pediatr 1991;119:495.

Grundfast KM. Management of otitis media. Pediatr Infect Dis J 1991; 10:269.

Howie VM, Ploussard JG, Slayer J. The “otitis-prone” condition. Am J Dis Child 1975;129:676.

Jenkins BH. Treatment of otitis externa and swimmer's ear. JAMA 1961;175:402.

Jones RN, Milazzo J, Seidlin M. Ofloxacin otic solution for treatment of otitis externa in children and adults. Arch Otolaryngol Head Neck Surg 1997;123:1193–1200.

Kozyrskyj AL, Hildes-Ripstein E, Longstaff SEA, et al. Treatment of acute otitis media with a shortened course of antibiotics. JAMA 1998;279:1736–1742.

Le Ct, Freeman DW, Fireman BH. Evaluation of ventilating tubes and myringotomy in the treatment of recurrent or persistent otitis media. Pediatr Infect Dis J 1991;10:2.

Marchant CD, Shurin PA, Turcyzk VA, et al. Course and outcome of otitis media in early infancy: a prospective study. J Pediatr 1984; 104:826.

Marcy SM. Infections of the external ear. Pediatr Infect Dis J 1985;4:192.

Paradise JL. Otitis media in infants and children. Pediatrics 1980;65:917.

Paradise JL. Short-course antimicrobial treatment for acute otitis media: not best for infants and young children. JAMA 1997;278:1640–1642.

Paradise JL, Smith CG, Bluestone CD. Tympanometric detection of middle ear effusion in infants and young children. Pediatrics 1976;58:198.

Pichichero ME, Cohen R. Shortened course of antibiotic therapy for acute otitis media, sinusitis, and tonsillopharyngitis. Pediatr Infect Dis J 1997;16:680–695.

Shurin PA, Pelton SI, Donner A, et al. Persistence of middle ear effusion after acute otitis media in children. N Engl J Med 1979;300:1121.

Tetzlatt TR, Ashworth C, Nelson JD. Otitis media in children less than twelve weeks of age. Pediatrics 1977;59:827.

Teele DW, Klein JO, Rosner B, et al. Epidemiology of otitis media during the first seven years of life in children in Greater Boston: a prospective cohort study. J Infect Dis 1989;160:83.

SINUSITIS

Brooks I, Friedman EM, Rodriques WJ, et al. Complications of sinusitis in children. Pediatrics 1980;66:568.

Hamory BH, Sande MA, Sydnor A, et al. Etiology and antimicrobial therapy of acute maxillary sinusitis. J Infect Dis 1979;139:197.

Shopfner CE, Rossie JO. Roentgen evaluation of paranasal sinuses in children. Am J Radiol 1973;118:176.

Wald ER. Acute sinusitis in children. Pediatr Infect Dis J 1983;2:61.

Wald ER, Milmoe GH, Bowen A, et al. Acute maxillary sinusitis in children. N Engl J Med 1981;304:749.

Wald ER, Reilly JS, Casselbednt M, et al. Treatment of acute maxillary sinusitis in childhood: a comparative study of amoxicillin and cefaclor. J Pediatr 1984;104:297.

Wald ER. Sinusitis in children. N Engl J Med 1992;326:319.

Wald ER. Chronic sinusitis in children. J Pediatr 1995;127:339–347.

PERITONSILLAR AND RETROPHARYNGEAL AND LATERAL ABSCESSES

Broughton RA. Nonsurgical management of deep neck infections in children. Pediatr Infect Dis J 1992;11:14–18.

Flodstrom A, Hallander HO. Microbiological aspects of peritonsillar abscesses. Scand J Infect Dis 1976;8:157.

McCurdy JA Jr. Peritonsillar abscess. Arch Otolaryngol 1977;103:414.

Nagy M, Pizzuto M, Blackstom J, Brodsky L. Deep neck infections in children: a new approach to diagnosis and treatment. Laryngoscope 1997;107:1627–1634.

Quraishi MS, O'Halpin DR, Blayney AW. Ultrasonography in the evaluation of neck abscesses in children. Clin Otolaryngol 1997;22:30–33.

CERVICAL LYMPHADENITIS

Barton LL, Feigin RD. Childhood cervical lymphadenitis: a reappraisal. J Pediatr 1974;84:846.

Bodenstein L, Altman RP. Cervical lymphadenitis in infants and children. Semin Pediatr Surg 1994;3:134–141.

Dajani AS, Garcia RE, Wolinsky E. Etiology of cervical lymphadenitis in children. N Engl J Med 1963;268:1379.

Fleisher G, Grosflam J, Selbst S, et al. Management of lymphadenitis in childhood. The role of percutaneous needle aspiration. Ann Emerg Med 1984;13:908.

Gorenstein A, Somekh E. Suppurative cervical lymphadenitis; treatment by needle aspirations. Pediatr Infect Dis J 1994;13:669–671.

Marcy SM. Infections of lymph nodes of the head and neck. Pediatr Infect Dis 1983;J 2:397.

Medina M, Goldfarb J, Traquina D, et al. Cervical adenitis and deep neck infection caused by *Streptococcus pneumoniae.* Pediatr Infect Dis J 1997;16:823–824.

LARYNGOTRACHEOBRONCHITIS (CROUP)

Adair JC. Ten year experience with IPPB in the treatment of acute laryngotracheobronchitis. Anesth Analg 1971;50:649.

Eden AN, Larkin VD. Corticosteroid treatment of croup. Pediatrics 1964; 33:768.

Fitzgerald D, Mellis C, Johnson M, et al. Nebulized budesonide is as effective as nebulized adrenaline in moderately severe croup. Pediatrics

97;5:722–725.

Gardner HG, Powell KR, Roden VJ, et al. The evaluation of racemic epinephrine in the treatment of infectious croup. Pediatrics 1973;52:52.

Hal CB, German JM, Breese BS, et al. Parainfluenza viral infections in children: correlation of shedding with clinical manifestations. J Pediatr 1977;91:194.

Hoekelman RA. Infectious illnesses during the first years of life. Pediatrics 1977;59:119.

Johnson DW, Jacobson S, Edney PC et al. A comparison of nebulized budesonide, intramuscular dexamethasone, and placebo for moderately severe croup. N Engl J Med 1998;339:498–503.

Kairys SW, Olmstead EM, O'Connor GT. Steroid treatment of laryngotracheitis: a meta-analysis of the evidence from randomized trials. Pediatrics 1989;83:683.

Klassen TP, Watters LK, Feldman ME, et al. The efficacy of nebulized budesonide in dexamethasone-treated outpatients with croup. Pediatrics 1996;97:463–466.

Klassen TP, Feldman ME, Watters LK. Nebulized budesonide for children with mild-to-moderate croup. N Engl J Med 1994;331:285–289.

Koren G, Frand M, Barzilay Z, et al. Corticosteroid treatment of laryngotracheitis v. spasmodic croup in children. Am J Dis Child 1983; 137:941.

Ledwith CA, Shea LM, Mauro RD. Safety and efficacy of nebulized racemic epinephrine in conjunction with oral dexamethasone and mist in the outpatient treatment of croup. Ann Emerg Med 1995;25:331–337.

Liepzig B, Oski FA, Cummings CW, et al. A prospective randomized study to determine the efficacy of steroids in the treatment of croup. J Pediatr 1979;94:194.

Taussig LM, Castro O, Biandry PA, et al. Treatment of laryngotracheitis (croup): use of intermittent positive pressure breathing and racemic epinephrine. Am J Dis Child 1975;129:790.

Tercero-Talavera FI, Rapkin RH. Antibiotic usage in the management of laryngotracheobronchitis. Clin Pediatr 1974;13:1074.

Tunnessen WW, Feinstein AR. The steroid croup controversy: an analytical review of methodological problems. J Pediatr 1980;96:751.

Waisman Y, Klein BL, Boenning DA, et al. Prospective randomized double-blind study comparing L-epinephrine and racemic epinephrine aerosols in the treatment of laryngotracheitis (croup). Pediatrics 1992;89:302.

EPIGLOTTITIS

Berenberg W, Kevy S. Acute epiglottitis in childhood. N Engl J Med 1958;258:870.

Faden HS. Treatment of *Haemophilus influenzae* type b epiglottitis. Pediatrics 1979;63:402.

Greenberg LW, Schisgall R. Acute epiglottitis in a community hospital. Am Fam Physician 1979;12:123.

Johnson GK, Sullivan JF, Bishop LA. Acute epiglottitis—review of 55 cases and suggested protocol. Arch Otolaryngol 1974;100:333.

Mauro RD, Poole SR, Lockhart CH. Differentiation of epiglottitis from laryngotracheitis in the child with stridor. Am J Dis Child 1988; 142:679.

Molteni RA. Epiglottitis. Incidence of extraepiglottic infection: report of 72 cases and review of the literature. Pediatrics 1976;58:526.

Rapkin RH. The diagnosis of epiglottitis: simplicity and reliability of radiographs of the neck in the differential diagnosis of the croup syndrome. J Pediatr 1972;80:96.

Rapkin RH. Tracheostomy in epiglottitis. Pediatrics 1973;52:426.

Valdepena HG, Wald ER, Rose E, et al. Epiglottitis and *Haemophilus influenzae* immunization: the Pittsburgh experience—a five-year review. Pediatrics 1995;96:424–427.

Wetmore RF, Handler SD. Epiglottitis: evolution in management during the last decade. Ann Otol Rhinol Laryngol 1979;88:822.

TRACHEITIS, BACTERIAL

Britto J, Habibi P, Walters S, et al. Systemic complications associated with bacterial tracheitis. Arch Dis Child 1996;74:249–250.

Brook I. Aerobic and anaerobic microbiology of bacterial tracheitis in children. Pediatr Emerg Care 1997;13:16–18.

Jones R, Santos JI, Overall JC. Bacterial tracheitis. JAMA 1979;242:721.

Sofer S, Duncan P, Chernick V. Bacterial tracheitis—an old disease rediscovered. Clin Pediatr 1983;22:407.

BACTERIAL PNEUMONIA

Asmar BI, Slovis TL, Reed JO, et al. *Haemophilus influenzae* type b pneumonia in 43 children. J Pediatr 1978;93:389.

Bechamps GJ, Lynn HB, Wenzl JE. Empyema in children: review of the Mayo Clinic experience. Mayo Clin Proc 1970;45:43.

Forbes GB, Emerson GL. Staphylococcal pneumonia and empyema. Pediatr Clin North Am 1957;4:21.

Goldwater PN, Rice MS. Primary meningococcal pneumonia in a nineteen month old child. Pediatr Infect Dis J 1995;2:155–156.

Harris JA. Antimicrobial therapy of pneumonia in infants and children. Semin Respir Infect 1996;11:139–147.

Hickey RW, Bowman MJ, Smith GA. Utility of blood cultures in pediatric patients found to have pneumonia in the emergency department. Ann Emerg Med 1996;27:721–725.

Klein JO. Diagnostic lung puncture in the pneumonias of infants and children. Pediatrics 1969;44:486.

Leibovitz E, Tabachnik E, Fliedel O, et al. Once-daily intramuscular ceftriaxone in the outpatient treatment of severe community-acquired

pneumonia in children. Clin Pediatr 1990;24:639.

Marshall R, Teele DW, Klein JO. Unsuspected bacteremia due to *Haemophilus influenzae*: outcome in children not initially admitted to hospital. J Pediatr 1979;95:690.

McCarthy PL, Frank AL, Ablow RC, et al. Value of the C-reactive protein test in the differentiation of bacterial and viral pneumonia. J Pediatr 92:454, 1978.

Mimica L, Donoso E, Howard JE, et al. Lung puncture in the etiologic diagnosis of pneumonia. Am J Dis Child 1971;122:278.

Muldoon RL, Jaecker DL, Kiefer HK. Legionnaire's disease in children. Pediatrics 1981;67:329.

Risser WL, Barone JS, Clark PA, et al. Noncomparative, open label, multicenter trial of cefixime for treatment of bacterial pharyngitis, cystitis and pneumonia in pediatric patients. Pediatr Infect Dis J 1987;6:1002.

Rusconi F, Rancilio L, Assael BM, et al. Counterimmunoelectrophoresis and latex particle agglutination in the etiologic diagnosis of presumed bacterial pneumonia in pediatric patients. Pediatr Infect Dis J 1988;7:781.

Ruuskanen O, Nohynek H, Ziegler T, et al. Pneumonia in childhood: etiology and response to antimicrobial therapy. Eur J Clin Microbiol Infect Dis 1992;11:217–223.

Shullteworth DB, Charney EL. Leukocyte count in pneumonia. Am J Dis Child 1971;122:393.

Turner RB, Lande AE, Chase P, et al. Pneumonia in pediatric outpatients. Cause and clinical manifestations. J Pediatr 1987;111:194.

NONBACTERIAL PNEUMONIA

Arth C, Von Schmidt B, Grossman M, et al. Chlamydial pneumonitis. J Pediatr 1978;93:447.

Beem M, Saxon E. Respiratory tract colonization and a distinctive pneumonia syndrome in infants with *Chlamydia trachomatis.* N Engl J Med 1977;296:396.

Beem MO, Saxon E, Tripple MA. Treatment of chlamydial pneumonia of infancy. Pediatrics 1979;63:198.

Brayden RM. Apnea in infants with *Chlamydia trachomatis* pneumonia. Pediatr Infect Dis J 1987;6:423.

Brolin I, Wernstedt L. Radiologic appearance of mycoplasmal pneumonia. Scand J Respir Dis 1978;59:179.

Foy HM, Cooney MK, McMahon R, et al. Viral and mycoplasmal pneumonia in a prepaid medical care group during an eight year period. Am J Epidemiol 1973;97:93.

Grayston TJ. *Chlamydia pneumoniae* (TWAR) infections in children. Pediatr Infect Dis J 1994;13:675–685.

Harrison HR, English MG, Lee CK, et al. *Chlamydia trachomatis* infant pneumonitis. N Engl J Med 1978;298:702.

Paisley JW, Lauer BA, Melinkovich P, et al. Rapid diagnosis of *Chlamydia trachomatis* pneumonia in infants by direct immunofluorescence microscopy of nasopharyngeal secretions. J Pediatr 1986;109:653.

BRONCHIOLITIS

Beem M, Wright FH, Fasam DM, et al. Observations on the etiology of acute bronchiolitis in infants. J Pediatr 1962;61:864.

Chanock RM, Kim HW, Vargoska HG, et al. Respiratory syncytial virus. Virus recovery and other observations during a 1960 outbreak of bronchiolitis, pneumonia, and minor respiratory disease in children. JAMA 1961; 176:647.

Gadomski AM, Aref GH, El Din OB, et al. Oral versus nebulized albuterol in the management of bronchiolitis in Egypt. J Pediatr 1994;124:131–138.

Hall CB, McBride JT, Walsh EE, et al. Aerosolized ribavirin treatment of infants with respiratory syncytial viral infection. N Engl J Med 1983;308:1443.

Hammer J, Numa A, Newth CJL. Albuterol responsiveness in infants with respiratory failure caused by respiratory syncytial virus infection. J Pediatr 1995;127:485–490.

Ho L, Collis G, Le Souef PN. Effect of Salbutamol on oxygen saturation in bronchiolitis. Arch Dis Child 1991;66:1061.

Klassen TP, Rowe PC, Sutcliffe T, et al. Randomized trial of salbutamol in acute bronchiolitis. J Pediatr 1991;118:807.

Menon K, Sutcliffe T, Klassen TP. A randomized trial comparing the efficacy of epinephrine with salbutamol in the treatment of acute bronchiolitis. J Pediatr 1995;126:1004–1007.

Schuh S, Canny G, Preisman JJ, et al. Nebulized albuterol in acute bronchiolitis. J Pediatr 1990;117:633.

Shaw KN, Bell LM, Sherman NH. Outpatient assessment of infants with bronchiolitis. Am J Dis Child 1991;145:151.

PERTUSSIS

Bass JW, Klenk EL, Korherine JB, et al. Antimicrobial treatment of pertussis. J Pediatr 1969;75:768.

Bergquist S, Bernander S, Dahnsjo H, et al. Erythromycin treatment of pertussis: a study of bacteriologic and clinical effects. Pediatr Infect Dis J 1987;6:458.

Brooksaler F, Nelson JD. Pertussis: a reappraisal and report of 190 confirmed cases. Am J Dis Child 1967;67:56.

Christie CDC, Marx ML, Marchant CD, et al. The 1993 epidemic of pertussis in Cincinnati: resurgence of disease in a highly immunized population. N Engl J Med 1994;331:16–21.

Mertsola J, Ruuskanen O, Eerda E, et al. Intrafamilial spread of pertussis. J Pediatr 1987;103:359.

Onorato IM, Wassilak GF. Laboratory diagnosis of pertussis: state of the art. Pediatr Infect Dis J 1987;6:145.

Whitaker JA, Donaldson P, Nelson JD. Diagnosis of pertussis by the fluorescent antibody method. N Engl J Med 1960;263:850.

TUBERCULOSIS

Delacourt C, Poveda JD, Chureau C, et al. Use of polymerase chain reaction for improved diagnosis of tuberculosis in children. J Pediatr 1995;126:703–709.

Doerr C, Starke JR, Ong LT. Clinical and public health aspects of tuberculous meningitis in children. J Pediatr 1995;127:27–33.

Harris VJ, Scharf V, Duda F, et al. Fatal tuberculosis in young children. Pediatrics 1977;63:912.

Hussey G, Chisholm T, Kibel M. Military tuberculosis in children: a review of 94 cases. Pediatr Infect Dis J 1991;10:832.

Kendig EL. Evolution of short-course antimicrobial therapy of tuberculosis in children, 1951–1984. Pediatrics 1985;75:684.

Lincoln EM, Sewell EM. Tuberculosis in Children. New York: McGraw-Hill, 1963.

Vallejo JG, Ong LT, Starke JR. Clinical features, diagnosis, and treatment of tuberculosis in infants. Pediatrics 1994;94:1–7.

HANTAVIRUS

Duchin JS, Koster FT, Peters CJ, et al. Hantavirus pulmonary syndrome: a clinical description of 17 patients with a newly recognized disease. N Engl J Med 1994;330:949–955.

Khan AS, Khabbaz RF, Armstrong LR, et al. Hantavirus pulmonary syndrome: The first 100 U.S. cases. J Infect Dis 1996:173:1297–1303.

Khan AS, Ksiazek TG, Zaki SR, et al. Fatal Hantavirus pulmonary syndrome in an adolescent. Pediatrics 1995;95:276–278.

GASTROENTERITIS/GASTRITIS

Barnes GL, Uren E, Stevens KB, Bishop RF. Etiology of acute gastroenteritis in hospitalized children in Melbourne, Australia, from April 1980 to March 1993. J Clin Microbiol 1998;36:133–138.

Black, RE, Jackson RJ, Tsac T, et al. Epidemic *Yersinia enterocolitica* infection due to contaminated chocolate milk. N Engl J Med 1978;298:76.

Blackow NR, Cugor G. Viral gastroenteritis. N Engl J Med 1981;304:397.

Blair J, Fitzgerald SF. Treatment of nonspecific diarrhea in infants. Clin Pediatr 1974;13:333.

Bonadio W. Efficacy of measuring BUN in assessing children with dehydration due to gastroenteritis. Ann Emerg Med 1989;18:755.

Boyce TG, Swerdlow DL, Griffin PM. *Escherichia coli* O157:H7 and the hemolytic uremic syndrome. N Engl J Med 1995;333:364–369.

Brown KH. Dietary management of acute diarrhea: optimal timing of feeding and appropriate use of milk and mixed diets. Pediatrics 1991; 118:S92.

Burke V, Gracey M, Robinson J, et al. The microbiology of childhood gastroenteritis: Aeromonas species and other infective agents. J Infect Dis 1983;148:68.

Butzler JP, Alexander M, Segers A, et al. Enteritis, abscess, and septicemia due to *Yersinia enterocolitica* in a child with thalassemia. J Pediatr 1978;93:619.

Centers for Disease Control. *Plesiomonas shigelloides* and *Salmonella* serotype Hartford infections associated with a contaminated water supply. MMWR 1998;47:394–396.

Cohen MB. Etiology and mechanisms of acute infectious diarrhea in infants in the United States. J Pediatr 1991;118:S34.

Committee on Nutrition. Use of oral fluid therapy and post-treatment feeding following enteritis in children in a developed country. Pediatrics 1985;75:358.

Delorme J, Laverdiere M. Martinease B, et al. Yersiniosis in children. Can Med Assoc J 1974;110:281.

Deutsch A, Wasserman D, Ruchelli E, et al. An uncommon presentation of *Salmonella*. Pediatr Emerg Care 1996;12:285–287.

Diarrhoeal Diseases Study Group. Loperamide in acute diarrhea in childhood: results of a double blind, placebo controlled multicenter clinical trial. Lancet 1984;289:1263.

Dupont HL, Hornick RB. Adverse affect of Lomotil therapy in shigellosis. JAMA 1973;226:1525.

Evans RG, Dadswell VJ. Human vibriosis. Br Med J 1967;2:240.

Finkelstein JA, Schwartz JS, Torrey S, et al. Common clinical features as predictors of bacterial diarrhea in infants. Am J Emerg Med 1989;7:469.

Guerrant RL, Moore RA, Kirschenfeld PM, et al. Role of toxigenic and invasive bacteria in acute diarrhea of childhood. N Engl J Med 1976; 293:567.

Guerrant RL, Lohr JA, Williams EK. Acute infectious diarrhea. I. Epidemiology, etiology, and pathogenesis. Pediatr Infect Dis J 1986;5:353.

Guerrant RL. Cryptosporidiosis: an emerging, highly infectious threat. Emerging Infect Dis 1997;3:56–63.

Huskins WC, Griffiths JK, Faruque ASG, et al. Shigellosis in neonates and young infants. J Pediatr 1994;125:14–22.

Jacobs JC. *Yersinia enterocolitica* arthritis. Pediatrics 1975;55:236.

Jones NL, Sherman PM. *Helicobacter pylori* infection in children. Curr Opin Pediatr 1998;10:19–23.

Karmal MA, Fleming PC. *Campylobacter* enteritis in children. J Pediatr 1979;94:527.

Khanna B, Cutler A, Israel NR, et al. Use caution with serologic testing for *Helicobacter pylori* infection in children. J Infect Dis 1998;178: 460–465.

Kohl S, Jacobson VA, Nahmias A. *Yersinia enterocolitica* infections in children. J Pediatr 1976;80:77.

Lebenthal E, Lu RB. Glucose polymers as an alternative to glucose in oral rehydration solutions. Pediatrics 1991;118:S62.

Lew JF, Glass RI, Gangarosa RE, et al. Diarrheal deaths in the United States, 1979 through 1987. JAMA 1991;265:3280.

Lyerly DM, Krivan HC, Wilkins TD. Clostridium difficile: its disease and toxins. Clin Microbiol Rev 1988;1:1.

Maki M, Maki R, Besikari T. Faecal leukocytes in Campylobacter associated diarrhea in infants. Acta Pediatr Scand 1979;68:271.

Meropol SB, Luberti AA, DeJong AR. Yield from stool testing of pediatric inpatients. Arch Pediatr Adolesc Med 1997;151:142–145.

Misra S, Diaz PS, Rowley AH. Characteristics of typhoid fever in children and adolescents in a major metropolitan area in the United States. Clin Infect Dis 1997;24:998–1000.

Moskhowitz M, Reif S, Brill S, et al. One-week triple therapy with omeprazole, clarithromycin and nitroimidazole for *Helicobacter pylori* infection in children and adolescents. J Pediatr 1998;102:E14.

Pai CH, Sorger S, Lackman L, et al. Campylobacter gastroenteritis in children. J Pediatr 1979;94:589.

Reid SR, Bonadio WA. Outpatient rapid intravenous rehydration to correct dehydration and resolve vomiting in children with acute gastroenteritis. Ann Emerg Med 1996;28:318–323.

Rodriquez WJ, Kim HW, Arrobia JO, et al. Clinical features of acute gastroenteritis associated with human reovirus-like agents in infants and young children. J Pediatr 1977;91:188.

Rosenstein BH. Salmonellosis in infants and children. J Pediatr 1967;70:1.

Rowe PC, Orrbine E, Lior H, et al. Risk of hemolytic uremic syndrome after sporadic *Escherichia coli* O157:H7 infection. J Pediatr 1998;132:777–782.

Sealy DP, Schuman. Endemic giardiasis and day care. Pediatrics 1983;72:154–158.

Schutze GE, Schutze SE, Kirby RS. Extraintestinal salmonellosis in a children's hospital. Pediatr Infect Dis H 1997;16:482–485.

Skirrow MB. *Campylobacter* enteritis: a new disease. Br Med J 1977;2:9.

Snyder JD. Use and misuse of oral therapy for diarrhea: comparison of U.S. practices with American Academy of Pediatric recommendations. Pediatrics 1991;87:28.

Soriano-Brucher H, Avendano P, O'Ryan M, et al. Bismuth subsalicylate in the treatment of acute diarrhea in children: a clinical study. Pediatrics 1991;87:18.

Torrey S, Fleisher G, Jaffe D. Incidence of *Salmonella* bacteremia in infants with *Salmonella* gastroenteritis. J Pediatr 1986;108:718.

Wolfson JS, Richter JM, Waldron MA, et al. Cryptosporidiosis in immunocompetent patients. N Engl J Med 1985;312:1276.

IMPETIGO

Barton LL, Friedman AD, Sharbey AM, et al. Impetigo contagiosa VII: comparative efficacy of oral erythromycin and topical mupirocin. Pediatr Dermatol 1989;6:134.

Bass JW, Chan DS, Creamer KM, et al. Comparison of oral cephalexin, topical mupirocin and topical bacitracin for treatment of impetigo. Pediatr Infect Dis J 1997;16:709–710.

Dajani AS, Ferrieri P, Wanamaker LW. Natural history of impetigo. J Clin Invest 1972;51:2863.

Dillon HC. The treatment of streptococcal skin infections. J Pediatr 1970;76:675.

Fleisher GR, Wilmott CM, Campos JM. Amoxicillin combined with clavulanic acid for the treatment of soft tissue infections in children. Antimicrob Agents Chemother 1983;24:679.

CELLULITIS

Barkin RM, Todd JK, Amer J. Periorbital cellulitis in children. Pediatrics 1978;62:390.

Ciarallo LC, Rowe PC. Lumbar puncture in children with cellulitis. J Pediatr 1993;122:355–359.

Fleisher GR, Heeger P, Topf S. *Hemophilus influenzae* cellulitis. Am J Emerg Med 1983;1:274.

Fleisher GR, Ludwig S, Campos J. Cellulitis: bacterial etiology, clinical features, and laboratory findings. J Pediatr 1977;94:355.

Santos JI, Jacobson SA, Swensen P, et al. Cellulitis: treatment with cefoxitin compared with multiple antibiotic therapy. Pediatrics 1981;67:887.

Schwartz GR, Wright S. Changing bacteriology of periorbital cellulitis. Ann Emerg Med 1996;28:617–620.

Varma BK. Cellulitis in children—a five-year review. Penn Med 1977;80:43.

Uman SJ, Kunin CM. Needle aspiration in the diagnosis of soft tissue infections. Arch Intern Med 1975;135:959.

OMPHALITIS

Book I. Microbiology of necrotizing fasciitis associated with omphalitis in the newborn infant. J Perinatol 1998;18:28–30.

Cushing AH. Omphalitis: a review. Pediatr Infect Dis J 1985;4:282.

Samuel M, Freeman NV, Vaishnav A, et al. Necrotizing fasciitis: a serious complication of omphalitis in neonates. J Pediatr Surg 1994;29: 1414–1416.

NEONATAL MASTITIS

Bodemer C, Panhans A, Chretien-Rarquest B, et al. Staphylococcal necrotizing fasciitis in the mammary region in childhood: a report of five cases. J Pediatr 1997;131:466–469.

Efrat M, Iujtman M, Eldemberg D, et al. Neonatal mastitis—diagnosis and treatment. Isr J Med Sci 1995;31:558–560.

Rudoy RC, Nelson JD. Breast abscess during the neonatal period. Am J Dis Child 1975;129:1031.

Walsh M, McIntosh K. Neonatal mastitis. Clin Pediatr 1986;25:395.

SEPTIC ARTHRITIS

Del Beccaro MA, Champoux AN, Bockers T, et al. Septic arthritis versus transient synovitis of the hip: the value of screening laboratory tests. Ann Emerg Med 1992;21:1418–1422.

Chusid MJ, Jacobs WM, Stay JR. Pseudomonas arthritis following puncture wounds of the foot. J Pediatr 1979;94:429.

Dagan R. Management of acute hematogenous osteomyelitis and septic arthritis in the pediatric patient. Pediatr Infect Dis J 1993;12:88–93.

Kunnamo I, Kallio P, Pelkonnen P, et al. Clinical signs and laboratory tests in the differential diagnosis of arthritis in children. Am J Dis Child 1987;141:34.

Memon IA, Jacobs NM, Yeh TF, et al. Group B streptococcal osteomyelitis and septic arthritis. Am J Dis Child 1979;133:921.

Molteni RA. The differential diagnosis of benign and septic joint disease in children. Clin Pediatr 1978;17:19.

Nelson JD. The bacterial etiology and antibiotic management of septic arthritis in infants and children. Pediatrics 1972;50:437.

Pittard WB, Thullen JD, Fanaroff AA. Neonatal septic arthritis. J Pediatr 1976;88:621.

Rotbart HA, Glode MP. Haemophilus influenzae type B septic arthritis in children. Report of 23 cases. Pediatrics 1985;75:254.

Schaad UB, McCracken GH, Nelson JD. Pyogenic arthritis of the sacroiliac joint in pediatric patients. Pediatrics 1980;66:375.

Volberg FM, Sumner TE, Abramson JS, et al. Unreliability of radiographic diagnosis of septic hip in children. Pediatrics 1984;74:118.

OSTEOMYELITIS

Capitanio MA, Kirkpatrick JA. Early roentgen observations in acute osteomyelitis. Am J Roentgenol 1970;108:488.

Correa AG, Edwards MS, Baker CJ. Vertebral osteomyelitis in children. Pediatr Infect Dis J 1993;12:228–233.

Dich PQ, Nelson JD, Haltallin KC. Osteomyelitis in infants and children: a review of 163 cases. Am J Dis Child 1975;129:1273.

Edward MD, Baker CJ, Wagner ML, et al. An etiologic shift in infantile osteomyelitis: the emergence of the Group B *Streptococcus.* J Pediatr 1978;93:578.

Fleisher GR, Paradise JE, Plotkin SA, et al. Falsely normal radionuclide scans for osteomyelitis. Am J Dis Child 1980;134:499.

Granoff DM, Sargent E, Jolivette D. *Haemophilus influenzae* type B osteomyelitis. Am J Dis Child 1978;132:488.

Jacobs FR, Adelonan L, Sack CM, et al. Management of *Pseudomonas* osteochondritis complicating puncture wounds of the foot. Pediatrics 1982; 69:432.

Jacobs RF, McCarthy RE, Elser JM. *Pseudomonas* osteochondritis complicating puncture wounds of the foot in children: a 10-year evaluation. J Infect Dis 1989;160:657.

Minnefor AB, Olson MI, Carver DH. *Pseudomonas* osteomyelitis following puncture wounds of the foot. Pediatrics 1971;47:598.

ter Meulen DC, Majd M. Bone scintigraphy in the evaluation of children with obscure skeletal pain. Pediatrics 1987;79:587.

Trevis S, Khettery J, Broker FH, et al. Osteomyelitis: early scintigraphic detection in children. Pediatrics 1976;57:173.

Waldvogel FA, Vasey H. Osteomyelitis: the past decade. N Engl J Med 1980;303:360.

Weissburg ED, Smith AL, Smith DH. Clinical features of neonatal osteomyelitis. Pediatrics 1974;33:505.

URETHRITIS/CERVICITIS

Beck-Sague CM, Farshy CE, Jackson TK, et al. Detection of *Chlamydia trachomatis* cervical infection by urine tests among adolescent clinics. J Adolesc Health 1990;22:197–204.

Brunham RC, Paavonen J, Stevens CE, et al. Mucopurulent cervicitis—the ignored counterpart in women of urethritis in men. N Engl J Med 1984;311:1.

Hammerschlag MR, Doraiswamz B, Alexander ER, et al. Are rectogenital chlamydial infections a marker of sexual abuse in children? Pediatr Infect Dis J 1984;3:100.

Rettig RJ, Nelson JD. Genital tract infection with *Chlamydia trachomatis* in prepubertal children. J Pediatr 1981;99:206.

Saltz GR, Linnemann CL, Brookman RR, et al. *Chlamydia trachomatis* infections in female adolescents. J Pediatr 1981;98:981.

URINARY TRACT INFECTIONS

Bachur R, Caputo GL. Bacteremia and meningitis among infants with urinary tract infections. Pediatr Emerg Care 1995;11:280–284.

Bauchner H, Philipp B, Dashefsky B, et al. Prevalence of bacteriuria in febrile children. Pediatr Infect Dis J 1987;6:239–242.

Crain EF, Gershel JC. Urinary tract infections in febrile infants younger than 8 weeks of age. Pediatrics 1990;86:363–367.

Ginsburg CM, McCracken GH. Urinary tract infections in young infants. Pediatrics 1982;69:409–412.

Hoberman A, Chao HP, Keller DM, et al. Prevalence of urinary tract infection in febrile infants. J Pediatr 1993;123;17–23.

Hoberman A, Wald ER, Reynolds EA, et al. Pyuria and bacteriuria in urine specimens obtained by catheter from young children with fever. J Pediatr 1994;124:513–519.

Hoberman A, Wald ER, Reynolds EA, et al. Is urine culture necessary to rule out urinary tract infection in febrile young children. Pediatr Infect Dis J 1996;15:304–309.

Kuppermann N, Bank DE, Walton EA, et al. Risks for bacteremia and urinary tract infections in young febrile infants with bronchiolitis. Arch Pediatr Adol Med 1997;151:1207–1214.

Lohr JA. Use of routine urinalysis in making a presumptive diagnosis of urinary tract infection in children. Pediatric Infect Dis J 1991;10: 646–650.

Lohr JA, Portilla MG, Geuder TG, et al. Making a presumptive diagnosis of urinary tract infection by using a urinalysis performed in an on-site laboratory. J Pediatr 1993:122;22–25.

Shaw KN, Gorelick M, McGowan KL, et al. Prevalence of urinary tract infection in febrile young children in the emergency department. Pediatrics 1998;102:16e.

Wiswell TE, Roscelli JD. Corroborative evidence for the decreased incidence of urinary tract infections in circumcised boy infants. Pediatrics 1986;78:96–99.

SEXUALLY TRANSMITTED DISEASES

Ackerman AB, Goldfaden G, Cosmides JC. Acquired syphilis in early childhood. Arch Dermatol 1972;106:92.

Bell TA. Major sexually transmitted diseases of children and adolescents. Pediatr Infect Dis J 1983;2:153.

Branch G, Paxton R. A study of gonococcal infection among infants and children. Public Health Rep 1965;80:347.

Finelli L, Crayne EM, Spitalny KC. Treatment of infants with reactive syphilis serology, New Jersey: 1992 to 1996. Pediatrics 1998;102:E27.

Fleisher GR, Hodge D, Cromie W. Penile edema in childhood gonorrhea. Ann Emerg Med 1980;9:314.

Hare M, Mowla A. Genital herpes virus infection in a prepubertal girl. Br J Obstet Gynaecol 1977;84:141.

Henretig FM. Acute urinary retention secondary to severe gonococcal balanoblennorrhea. Pediatrics 1981;67:734.

Ingram DL, Everett VD, Flick LAR, et al. Vaginal gonococcal cultures in sexual abuse evaluations: evaluation of selective criteria for preteenaged girls. Pediatrics 1997;99:E8.

Kaplan KM, Fleisher GR, Paradise E, et al. Social relevance of genital Herpes simplex in children. Am J Dis Child 1984;138:872.

Mascola L, Pelosi R, Blount JH, et al. Congenital syphilis revisited. Am J Dis Child 1985;139:575.

Moyuer VA, Schneider V, Yetman R, et al. Contribution of long-bone radiographs to the management of congenital syphilis in the newborn infant. Arch Pediatr Adolesc Med 1998;152:353–357.

Siegel RM, Schubert CJ, Myers PA, et al. The prevalence of sexually transmitted diseases in children and adolescent evaluated for sexual abuse in Cincinnati: rationale for limited STD testing in prepubertal girls. Pediatrics 1995;96:1090–1094.

Tomeh MO, Wilfert CM. Venereal disease of infants and children at Duke University Medical Center. North Carolina Med J 1973;34:109.

VIRAL SYNDROME

Krugman S, Ward R, Katz S. Infectious Disease of Children. St Louis: Mosby, 1981.

ERYTHEMA INFECTIOSUM

Bell LM, Naides SJ, Stoffman P, et al. Human parvovirus B19 infection among hospital staff members after contact with infected patients. N Engl J Med 1989:321:485–491.

Nunuoe T, Okochi K, Mortimer PP, et al. Human parvovirus (B19) and erythema infectiosum. J Pediatr 1985;107:38–40.

INFECTIOUS MONONUCLEOSIS

Alpert G, Fleisher GR. Complications of EBV infection during childhood. Pediatr Infect Dis J 1984;3:304.

Fleisher GR, Henle W, Henle G, et al. Primary Epstein-Barr virus infections in American infants: clinical and serological observations. J Infect Dis 1979;139:553.

Fleisher GR, Lennete ET, Henle G, et al. Incidence of heterophil antibody responses in children with infectious mononucleosis. J Pediatr 1979; 94:723.

Fleisher GR, Paradise J. Atypical lymphocytosis in children. Ann Emerg Med 1981;10:424.

Fleisher GR, Paradise JE, Lennette ET. The leukocyte response in childhood infectious mononucleosis. Am J Dis Child 1981;135:699.

Peter J, Ray CG. Infectious mononucleosis. Pediatr Rev 1998;19:276–279.

van der Horst C, Joncas J, Ahronheim G, et al. Lack of effect of peroral acyclovir for the treatment of acute infectious mononucleosis. J Infect Dis 1991;164:788.

MEASLES

Barkin RM. Measles mortality: Analysis of the primary cause of death. Am J Dis Child 1975;129:307.

Farizo KM, Stehr-Green PA, Simpson DM, et al. Pediatric emergency room visits: a risk factor for acquiring measles. Pediatrics 1991;87:74–79.

Mason WH, Ross LA, Lanson J, et al. Epidemic measles in the postvaccine era: evaluation of epidemiology, clinical presentation and complications during an urban outbreak. Pediatr Infect Dis J 1993;12:42–48.

ROSEOLA (EXANTHEM SUBITUM)

Lyall EG. Human herpesvirus 6: primary infection and the central nervous system. Pediatr Infect Dis H 1996;15:693–696.

Suga S, Yoshikawa T, Nagia T, et al. Clinical features and virological findings in children with primary human herpesvirus 7 infection. Pediatrics 1997;99:E4.

Yoshikawa T, Suga S, Asano Y, et al. Distribution of antibodies to a causative agent of exanthem subitum (human herpesvirus-6) in healthy individuals. Pediatrics 1989:84:675–677.

VARICELLA

Doctor A, Harper MB, Fleisher GR. Group A beta-hemolytic streptococcal bacteremia: historical overview, changing incidence, and recent association with varicella. Pediatrics 1995;96:428–433.

Dunkle LM, Arvin AM, Whitley RJ, et al. A controlled trial of acyclovir for chickenpox in normal children. N Engl J Med 1991;325:1539.

Fleisher GR, Henry W, McSorley M, et al. Life-threatening complications of varicella. Am J Dis Child 1981;135:896.

Weinstein L. Failure of chemotherapy to prevent the bacterial complications of measles. N Engl J Med 1955;253:679.

BABESIOSIS

Krauss PJ, Telford SR, Pollack RJ, et al. Babesiosis: an underdiagnosed disease of children. Pediatrics 1992;89:1045–1048.

Krauss PJ, Feder HM. Lyme disease and babesiosis. Adv Pediatr Infect Dis 1994;9:183–209.

BOTULISM

Grover W, Peckham G, Berman P. Recovery following cranial nerve dysfunction and muscle weakness in infancy. Dev Med Child Neurol 1979;16:163.

Johnson RO, Clay SA, Arnon SS. Diagnosis and management of infant botulism. Am J Dis Child 1979;133:586.

Long SS, Gajewski JV, Brown LW, et al. Clinical, laboratory, and environmental features of infant botulism in southeastern Pennsylvania. Pediatrics 1985;75:935.

Pickett J, Berg B, Chaplin E, et al. Syndrome of botulism in infancy: clinical and electrophysiologic study. N Engl J Med 1976;295:770.

CAT-SCRATCH DISEASE

Bass JW, Freitas BC, Freitas AD, et al. Prospective randomized double blind placebo-controlled evaluation of azithromycin for treatment of cat-scratch disease. Pediatr Infect Dis J 1998;17:447–52.

Demers DM, Bass JW, Vincent JM, et al. Cat scratch disease in Hawaii: etiology and seroepidemiology. J Pediatr 1995;127:23–26.

Margileth AM. Cat scratch disease: nonbacterial regional lymphadenitis. Pediatrics 1969;42:803–818.

EHRLICHIOSIS

Barton LL, Rathore MH, Dawson JE. Infection with *Ehrlichia* in childhood. J Pediatr 1992;120:998–1001.

Goodman JL, Nelson C, Vitale B, et al. Direct cultivation of the causative agent of human granulocytic ehrlichiosis. N Engl J Med 1996;334:209–215.

Schutze G, Jacobs RF. Human monocytic ehrlichiosis in children. Pediatrics 1997;100:e10.

MALARIA

McCaslin RI, Pikis A, Rodriquez WJ. Pediatric *Plasmodium falciparum* malaria: a ten year experience from Washington, DC. Pediatr Infect Dis J 1994;13:709–715.

White NJ. The treatment of malaria. N Engl J Med 1996;335:800–806.

PARASITES, INTESTINAL

Committee on Infectious Diseases. 1997 Redbook. Elk Grove Village, IL: American Academy of Pediatrics, 1997.

RABIES

Mann JM. Systematic decision-making in rabies prophylaxis. Pediatr Infect Dis J 1983;2:162.

Plotkin SA, Wiktor TJ, Koprowski H, et al. Immunization schedules for the new human diploid cell vaccine. Am J Epidemiol 1976;103:75.

ROCKY MOUNTAIN SPOTTED FEVER

Abramson JS, Givner LB. Should tetracycline be contraindicated for therapy of presumed Rocky Mountain spotted fever in children less than 9 years of age? Pediatrics 1990;86:123.

Bradford WD, Hawkins HK. Rocky Mountain spotted fever in children. Am J Dis Child 1977;131:1228.

Fleisher GR, Lennette ET, Honig P. Diagnosis of Rocky Mountain spotted fever by immunofluorescent identification of *Rickettsia rickettsi* in skin biopsy tissue. J Pediatr 1979;95:63.

Hattwick MAW, Retailliau H, O'Brien RJ. Fatal Rocky Mountain spotted fever. JAMA 1979;240:1499.

Vianna NJ, Himan AR. Rocky Mountain spotted fever in Long Island. Am J Med 1971;51:725.

Wilfert CM, MacCormack JN, Kleeman K, et al. The prevalence of antibodies to *Rickettsia rickettsii* in an area endemic for Rocky Mountain spotted fever. J Infect Dis 1985;151:823.

Woodward TE, Pederson CE, Oster CN, et al. Prompt confirmation of Rocky Mountain spotted fever: identification of rickettsiae in skin tissues. J Infect Dis 1976;134:297.

TETANUS

Brand DA, Acampora D, Gottlieb LD, et al. Adequacy of anti-tetanus prophylaxis in six hospital emergency rooms. N Engl J Med 1983;309:636.

Freelander FC. Tetanus neonatorum. J Pediatr 1951;39:448.

Turner TB, Velasco-Joven EA, Prudovsky S. Studies on the prophylaxis and treatment of tetanus. Bull Johns Hopkins Hosp 1958;102:71.

Weinstein L. Tetanus. N Engl J Med 1973;289:1293.

TOXIC SHOCK SYNDROME

Davis JP, Chesney PJ, Wand PJ, et al. Toxic shock syndrome: epidemiologic features, recurrence, risk factors, and prevention. N Engl J Med 1980;303:1429.

Todd J, Fishant M, Kapral F, et al. Toxic-shock syndrome associated with phage-group-1 staphylococci. Lancet 1978;2:1116.

Wiesenthal AM, Todd JK. Toxic shock syndrome in children aged 10 years or less. Pediatrics 1984;74:112.

CHAPTER 85

Human Immunodeficiency Virus Infection

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EPIDEMIOLOGY

In 1983, the first descriptions of children with acquired immunodeficiency syndrome (AIDS) appeared in the literature. Since then, it has become clear that human immunodeficiency virus (HIV), the etiologic agent for AIDS, has been present in humans for decades. HIV has since been identified and characterized, and our understanding of the clinical manifestations of HIV infection has grown. As of the end of 1997, 90% of pediatric AIDS cases reported in the United States were perinatally acquired. The number of children with perinatally acquired AIDS in the United States peaked in 1992, with the subsequent decline likely a result of the increased use of voluntary HIV testing of pregnant women and the use of antivirals to prevent perinatal transmission. As a worldwide problem, HIV infection, including infection of children, continues to grow. The World Health Organization (WHO) estimates that in 1997, approximately 590,000 children were newly HIV positive, 460,000 children died of HIV, and a cumulative 2.7 million children have died of HIV/AIDS.

AIDS in this country predominantly occurs among homosexual and bisexual men, intravenous (IV) drug users, transfusion recipients and hemophiliacs, and female heterosexual contacts of HIV-infected persons who had additional risk factors themselves such as IV drug use. Rates are highest among African-Americans and Hispanics in the Northeast region of the United States and in certain large metropolitan areas.

To decide whether HIV infection is likely, the emergency physician must consider the prevalence of HIV infection among persons with particular risk behaviors in various parts of the country. Increasingly, mothers are offered testing for HIV as a part of routine antenatal care. If so, the results of testing will identify most children at risk. If the mother was not tested, the physician must ask parents about HIV risk factors (e.g., IV drug use, crack cocaine use, intercourse with a known IV drug user, multiple sex partners). An additional risk factor is a history of blood transfusions in the parent or child between 1978 and 1985, when banked blood began to be routinely tested for HIV. It must be recognized that a high proportion (about one-third) of inner-city patients will report HIV risk behaviors; most of them will not have HIV. If the person who brings the child to the emergency department (ED) is not the parent, inquire about the parents' health and whereabouts. In high-risk areas, many children in foster care or those living with relatives have a parent who has died from an AIDS-related condition.

However, the absence of risk factors does not rule out the possibility of HIV infection. More than one-fourth of infected women cannot report a risk factor, and nearly half of HIV-positive adolescent girls acquire the infection through heterosexual contact. (Often, from the individual's perspective, a series of monogamous relationships is not interpreted as having “multiple partners.”) Because the typical incubation period in adults is about 10 years, most will not develop symptoms of HIV infection during adolescence. However, young adults who develop symptomatic infection often acquired the virus during adolescence. Often, a mother first finds out that she is infected when tested during pregnancy or when her infant is diagnosed as HIV positive. Although all mothers of infants with congenital HIV infection harbor the virus, only a small minority of their infants will be infected (about 25% of infants born to untreated HIV-positive women, or 8% of infants receiving appropriate antiviral perinatal prophylaxis).

PATHOPHYSIOLOGY

The human immunodeficiency virus is an RNA retrovirus. After gaining entrance to the body, it binds through its protein envelope, gp120, to a region of the CD4 molecule on helper T lymphocytes, monocytes, macrophages, dendritic and glial cells, and intestinal endothelial cells. Through this binding process, the virus can fuse with the cell membrane and enter the cell. There, viral RNA is transcribed to DNA by reverse transcriptase and is incorporated into the host cell DNA. The viral DNA may remain dormant for long periods but can be stimulated at any time to transcribe itself into messenger RNA, which, in turn, leads to protein synthesis, assembly, and release of virus and virus particles. How a latent infection is converted into an active one is unknown, but it is thought that the coexistence of other infections stimulates the process.

Abnormalities develop in both the cellular and humoral immune systems as HIV replication continues. Most circulating cells showing HIV infection are CD4 helper lymphocytes and with time the percentage of CD4 helper lymphocytes decreases, and the ratio of helper (CD4) to suppressor (CD8) lymphocytes reverses. In adults, the number of CD4 lymphocytes has been shown to be a useful marker for opportunistic infection; early in childhood, this is not reliably the case. High CD4 counts have been seen in association with *Pneumocystis carinii* pneumonia. The proliferation of T cells in response to antigenic stimulation is impaired, mitogenic responses may be abnormal, and delayed hypersensitivity skin tests (tetanus, *Candida*, measles-mumps-rubella [MMR]) become negative as well. Macrophages, natural killer (NK) cells, CD4 and CD8 lymphocytes, and dendritic cells all show impaired function. In addition, there is an abnormal polyclonal activation of B cells that can result in notable hypergammaglobulinemia; however, these children do not respond with appropriate antibodies to new antigens and often produce autoantibodies.

Mode of Acquisition

The likelihood of acquiring infection depends on the amount of infectious virus in the body fluid and the extent of contact with that body fluid. The amount of virus is highest in the infected individual during the initial acute infection and late in the disease during the symptomatic period when $CD4^+$ cells are depleted. The risk of perinatal acquisition increases with increasing maternal virus load and decreased maternal immunity. Infants may not develop sufficient antiviral cellular immune response to decrease the amount of virus and therefore often have high levels of viremia.

Blood and genital fluids (seminal fluid, vaginal fluid) are the most likely to transmit HIV. Other fluids such as saliva, urine, sweat, amniotic fluid, synovial fluid, feces, and tears contain no virus or only low levels of virus and are not important sources of virus transmission. The likelihood of HIV infection after a single exposure to an HIV-positive source has been estimated to be 0.01 to 1% after sexual intercourse, 0.03% with needlestick injury, 0.5 to 1% with injecting drug use, and greater than 90% with blood transfusion. Breast-feeding is associated with an increased risk of transmission of HIV to the infant. In areas such as the United States where alternatives to breast-feeding are safe, breast-feeding by known HIV-infected women is discouraged. Worldwide it now appears that breast-feeding accounts for about 12% of perinatally acquired infections with the rest occurring in and around the intrapartum period.

Detection of virus can now be accomplished by culture, quantitative HIV RNA polymerase chain reaction (PCR) techniques, and antigen testing. Quantitative HIV RNA PCR techniques allow detection of viral RNA to levels of less than 500 copies per milliliter. Plasma viremia in the acute infection is typically about 5 million per milliliter. During the asymptomatic period, it is in the tens of thousands per milliliter, although the culturable or infectious virus is substantially less (100- to 100,000-fold less).

CLINICAL FINDINGS AND MANAGEMENT

Initial Presentation of Children with HIV

Most pediatric patients are infected perinatally and develop symptoms progressively over time, and although lymphadenopathy, hepatosplenomegaly, and failure to thrive are common clinical features, any organ system can be affected (Table 85.1). Many children show signs of abnormal humoral immune function such as recurrent or persistent bacterial infection. Children lack preexisting antibodies to bacterial pathogens at the time they are infected with the HIV virus; this developmental situation makes them particularly susceptible to infection by common bacterial pathogens. Numerous studies have documented the increased incidence of bacterial infections in HIV-positive children. Some children show early defects in cellular immunity exhibited by persistent candidiasis, chronic diarrhea, or opportunistic infections. Still others may remain relatively asymptomatic for long periods (6% completely asymptomatic at 5 years).

Lymphadenopathy	Recurrent fevers
Hepatomegaly	Splenomegaly
Failure to thrive	Chronic or recurrent diarrhea
Bacteremia	Wasting syndrome
Oral thrush	Developmental delay
Chronic or recurrent parotitis	Acquired microcephaly
Opportunistic infections	Spastic paresis

Table 85.1. Signs and Symptoms of Human Immunodeficiency Virus Infection in Children

Approximately 80% of infants infected perinatally will develop clinical signs or symptoms within the first 12 months of life. However, only 20% experience a rapid progression to profound immunodeficiency. The rate of progression is likely

related to multiple host and virus-specific factors, but in perinatally acquired infection, the rate of disease progression is directly related with the severity of the disease in the mother at the time of delivery.

The initial presentation may actually represent acute HIV infection. Within days to weeks of initial exposure to HIV, a syndrome of fever, fatigue, and maculopapular truncal rash can mark acute disease. Myalgia, arthralgia, pharyngitis, lymphadenopathy, headache, nausea, vomiting, and diarrhea are common. Many also have leukopenia and thrombocytopenia. Many patients with acute HIV infection will seek care; however, few are diagnosed with acute HIV unless a specific history of HIV exposure is given because of the common occurrence of these symptoms with other viruses. The diagnosis of acute HIV infection cannot be made with standard serologic tests (enzyme-linked immunoabsorbent assays [ELISAs] or Western blot) because these tests first become positive 3 to 4 weeks after acute infection. Early detection, when indicated, is possible through the use of p24 antigen testing and plasma viral RNA testing. Follow-up testing and counseling should be arranged.

Fever

As is true for all children with fever, the evaluation of the HIV-infected child with fever requires a careful history, thorough physical examination, and often, laboratory testing. Fever in HIV-infected children can represent simple childhood viral infections, but because of the humoral immunodeficiency of these children, they also commonly suffer from acute bacterial infections. Otitis media, sinusitis, pneumonia, adenitis, bacteremia, and skin and soft-tissue infections all occur commonly. In addition, opportunistic infections must be considered. It is important to inquire about any previous opportunistic infection, or the use of prophylactic medications for the prevention of PCP, *Mycobacterium avium intracellulare* (MAI), or cytomegalovirus (CMV) because these may be markers for poor immune function. Some parents may be able to provide a recent CD4 count, but laboratory data that reflect the status of the immune system usually are not available to the emergency physician. As in any child, the general clinical appearance is an important piece of information in the evaluation of the febrile HIV-positive child. The physician should search carefully for a focus (Fig. 85.1).

FIGURE 85.1. Evaluation of the HIV-positive child with fever. *TMP-SMZ*, trimethoprim–sulfamethoxazole; *CBC*, complete blood count. (Modified from Dorfman D, Crain E, Bernstein L. Care of febrile children with HIV infection in the emergency department. Pediatr Emerg Care 1990;6:308.)

Evaluation of the Febrile, HIV-Positive Child without a Source

Experience has shown that HIV-infected children experience the usual childhood illnesses and are likely to develop the same upper respiratory infections and viral syndromes as immunocompetent children. The clinical appearance of the patient is the starting point for determining ED management (Fig. 85.1).

Evaluation of the Well-Appearing, Febrile HIV-Positive Child

The HIV-infected child who appears well and does not have an obvious source of infection presents a more difficult problem than the child with an obvious localized infection. The first step is to decide on an appropriate evaluation. Several studies have demonstrated that HIV-positive children have an increased incidence of bacteremia. They are also more susceptible to serious viral infections such as disseminated CMV. However, it appears that serious bacterial, viral, or opportunistic infections are relatively uncommon among well-appearing HIV-positive children who present to the ED with fever.

An evaluation similar to that for febrile children at risk for occult bacteremia is reasonable. However, laboratory testing should be obtained even beyond 24 months of age. Thus, even if the older child presents with a temperature of 39°C (102.2°F) or higher, a complete blood count (CBC) with differential and blood culture are recommended. Erythrocyte sedimentation rates are usually elevated in HIV-infected children and therefore are not helpful in their evaluation. If the child is still in diapers, a urine sample should be obtained for analysis and culture. Older children who are toilet-trained usually complain of dysuria or frequency if they have a urinary tract infection. If the child has any respiratory signs or symptoms, including isolated tachypnea, or if the CBC has an elevated leukocyte count with a shift to left regardless of the presence of respiratory signs, pulse oximetry and a chest radiograph should be ordered. The white blood cell (WBC) count is best evaluated in relation to baseline counts because many HIV-infected children have some degree of granulocytopenia. If it is known that the child is not leukopenic or the baseline is not available, a WBC count of 15,000/µL or greater should be considered suggestive of bacterial infection. These patients may be started on antibiotics such as amoxicillin, amoxicillin–clavulinic acid (Augmentin), or intramuscular ceftriaxone (Rocephin), pending culture results. No data support or refute this practice except for the documentation of high rates of bacteremia, predominantly

pneumococcal, as well as recurrent bacteremia in some of these patients.

If the child appears well and the evaluation has not revealed a source for the fever that requires hospitalization, the child may be sent home (if the child's caregiver can be easily contacted and has the means to return if necessary) with instructions to return if symptoms worsen or if the patient develops lethargy or will not take adequate amounts of fluids. A follow-up evaluation by telephone or by a revisit to the child's regular provider or the ED should be scheduled for the next day.

Evaluation of the Ill-Appearing, Febrile, HIV-Positive Child

The HIV-positive child who comes to the ED with fever and appears ill should be treated like other ill-appearing, febrile children because they are likely to be infected with the same types of organisms that infect immunocompetent children. A lumbar puncture is indicated for meningismus, change in mental status, or fever in children whose underlying abnormal mental status makes them difficult to assess. If a child is believed to be so unstable that lumbar puncture is not safe, it can be delayed until later. In either case, the child should be started on parenteral broad-spectrum antibiotics immediately. Ceftriaxone (100 mg/kg per day divided every 12 hours) is an appropriate choice because it covers the organisms that most commonly cause sepsis in children. Especially in young children, because of the possibility of PCP presenting with fever and ill appearance, trimethoprim–sulfamethoxasole (TMP-SMZ) (5 mg/kg per dose every 6 hours) should be added to the antibiotic regimen if there are respiratory symptoms, with or without a positive chest radiograph. Treatment for suspected PCP should not be delayed because of fear of interfering with the diagnostic workup. Fungal infections, with the exception of oral thrush, are uncommon in HIV-infected children. However, candidal sepsis must be considered in hospitalized patients who do not improve with antibiotics.

Evaluation of the HIV-Positive Child with Persistent Fever

Chronic fever is common in HIV-infected children. It can have many causes, and the evaluation of children with fever of unknown origin is often difficult and not always revealing. The major focus of such a workup in the ED is to rule out bacterial infection. A careful history and physical examination should be followed by a CBC; urinalysis; chest and sinus films; and blood, urine, and stool cultures. Recurrent otitis media is commonly seen, and some children may have recurrent parotitis or sinusitis. If no source is obvious and the basic workup is negative, more unusual infections need to be considered. Tuberculosis, although common among HIV-infected adults, is uncommon in children but may be more likely among adolescents. MAI may cause chronic fevers in HIV-infected children. It is often associated with anemia secondary to bone marrow infiltration and can be cultured from blood, stool, and bone marrow. Numerous viruses can cause chronic infections associated with fever in these children. Epstein-Barr virus (EBV) and CMV are among the more common, with CMV often presenting with chronic hepatitis and bloody diarrhea. It may also cause pneumonia and retinitis. A blood buffy coat specimen can be sent for CMV antigen detection. Most HIV-positive children with fever of unknown origin are hospitalized to facilitate the diagnostic process. The possibility that the fever is caused by one of the many drugs the child may be taking must also be considered.

Soft-Tissue Infections

Clinical Presentation

Cervical adenitis and cellulitis are common and may be accompanied by fever. Both cervical adenitis and cellulitis may be secondary to alterations in the child's immune status or may be on a mechanical basis because of superinfection of already enlarged lymph glands (in the case of cervical adenitis) or disruption of the normal skin by other lesions such as condylomata, molluscum, or vesicular and follicular eruptions. Acute bacterial adenitis in these patients is usually caused by *Staphylococcus aureus* or group A streptococci. Other less common causes, including *Bartonella*, the agent of cat-scratch disease, which can also cause bacillary angiomatosis and trench fever in these patients, should be considered.

Parotitis is a common soft-tissue infection that occurs in HIV-positive children who can have chronic enlargement of the parotid glands secondary to lymphocytic infiltration (Fig. 85.2). In these children, the parotid glands are enlarged and firm but nontender, and the overlying skin is not erythematous. With acute suppurative parotitis these children develop fever, tenderness over the parotid, and purulent drainage from Stenson's duct. *S. aureus* is the most likely offending organism.

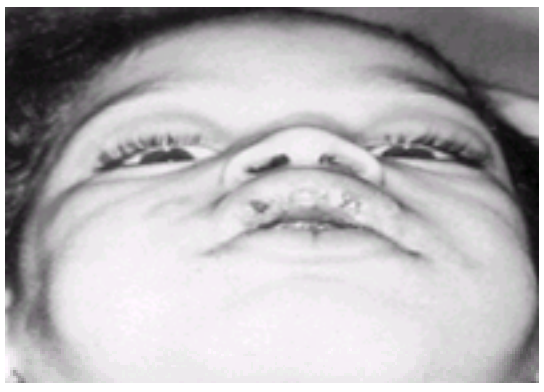

FIGURE 85.2. Chronic parotitis in an HIV-positive child. (Courtesy of Dr. A. Rubenstein.)

Management

If the child appears well and the infection is well circumscribed and does not impinge on a critical structure such as the airway, outpatient antibiotic therapy active against *S. aureus* and *Streptococcus pyogenes* (e.g., cephalexin 60 to 80 mg/kg per day divided, four times daily) is appropriate as long as it appears that the caretaker can adhere to the regimen and the child can be reevaluated within 24 to 48 hours. In most cases, a blood culture should be obtained, particularly if the child has fever, has a history of an opportunistic infection, or does not appear well. In addition, the need for drainage (needle aspiration or surgical) must be considered. Children who are sent home must have close outpatient follow-up.

Pulmonary Infections

Just as respiratory complaints are common among immunocompetent children, so are respiratory conditions common in infants and children with HIV infection. They deserve special attention, however, because they are the most common cause of mortality in these patients. Documentation of oxygenation by pulse oximetry or arterial blood gas, blood culture for bacterial pathogens, nasopharyngeal specimens for rapid viral diagnosis, and viral culture should all be considered in any HIV-positive patient with respiratory symptoms. Chest radiographs should be obtained and can be helpful in determining the cause (Fig. 85.3). Decisions regarding sputum induction or bronchoscopy do not usually need to be made in the ED.

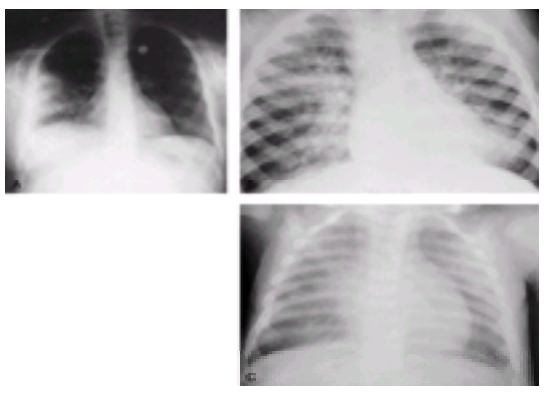

FIGURE 85.3. A. Bacterial pneumonia, lobar pattern. **B.** Increase in hilar structures and a diffuse reticulonodular pattern compatible with LIP/PLH. **C.** Pneumocystis carinii pneumonia. Evidence of cardiomegaly and a diffuse increase in interstitial markings. (Courtesy of Dr. H. Goldman.)

Pneumocystis carinii *Pneumonia*

Background

PCP is the most common serious opportunistic infection in HIV-infected children. Although PCP can occur at any age, in children it develops most commonly between the ages of 2 and 6 months and may be the first presentation of HIV infection. Moreover, the first episode is often acute in onset and may be fatal; overall, the fatality rate is about 40%.

For the most part, the results of immunologic studies are not available to the emergency physician. However, particularly relevant differences between adults and children should be noted in case the results of CD4 lymphocyte counts are known. Among adults, absolute CD4 lymphocyte counts are associated with the risk of acquiring PCP. Prophylaxis against PCP is recommended for adults with CD4 counts lower than 200 cells/µL (or previous oropharyngeal candidiasis). In young children, CD4 counts cannot be used to exclude the risk of PCP. Among HIV-infected children diagnosed with PCP, only 26% of infants 0 to 5 months of age will have a CD4 lymphocyte count of 1500 cells/µL or lower, but by 6 to 11 months of age, this is unusual, although 25% will still have CD4 counts in the range of 750 to 1499 cells/µL. For indications for PCP prophylaxis (and prophylaxis of other infections complicating HIV), see Table 85.2. When indicated, recommended prophylaxis is 150 mg TMP/m^2 per day with 750 mg SMZ/m^2 per day administered as follows: twice a day for 7 days per week; twice a day for 3 consecutive days per week; or a single daily dose for 3 consecutive days per week. For patients who do not tolerate TMP-SMZ, either Dapsone 2 mg/kg (not to exceed 100 mg) orally once per day can be given (as tolerated) or pentamidine can be given. Pentamidine is 300 mg via Respirgard II inhaler monthly for children when tolerated (usually those older than 5 years of age). Pentamidine (4 mg/kg) can also be given intravenously every 2 to 4 weeks.

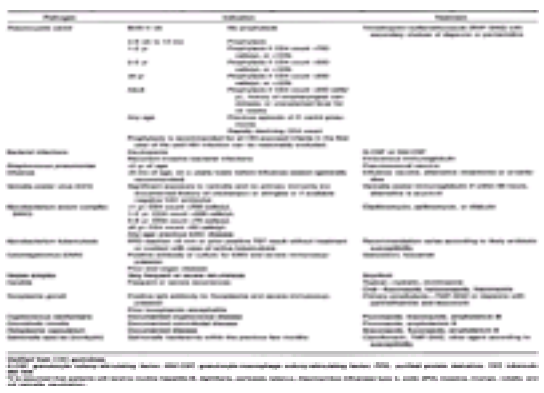

Table 85.2. Prophylaxis of Infections in Human Immunodeficiency Virus (HIV) Infected Patients[a]

Clinical Manifestations

PCP presents as an acute or subacute illness. The infant typically is febrile, with marked tachypnea, wheezing, rhonchi, and diminished breath sounds. Rales are not usually part of the PCP picture, and cough may be absent. When coughing is present, it is typically dry and nonproductive. Over hours to days, the patient develops hypoxia and increased respiratory distress.

Management

When PCP is suspected, the physician should intervene to maintain the airway as necessary, obtain an arterial blood gas or room air pulse oximetry, and provide supplemental oxygen. A chest radiograph and a serum lactate dehydrogenase (LDH) should be ordered. The patient typically has a high (greater than 30 mm Hg) alveolar-arterial (A-a) oxygen gradient and low oxygen saturation, and generally there is marked (greater than 500 IU) elevation of the serum LDH. Radiographic findings typically consist of a diffuse interstitial ("ground glass") pattern, but in infants, there may be patchy infiltrates or complete opacification of the lung fields. Occasionally, however, there may be clear lung fields with hyperinflation suggestive of bronchiolitis, and in 5 to 10% of patients with PCP, the chest radiograph appears normal.

It is often difficult to make the diagnosis of PCP in the ED. If PCP is suspected on the basis of the history, physical examination, or the results of the laboratory investigation, it is appropriate to start IV TMP-SMZ at a dosage of 20 mg/kg per day of TMP divided into 4 doses daily. The child should be hospitalized for close observation and further diagnostic evaluation should it be needed. In general, patients with PCP do not respond rapidly to antibiotic therapy. Patients intolerant of TMP-SMZ can be treated with pentamidine (4 mg/kg per day as a single daily dose) or atovaquone, but these should be considered second-line agents. The addition of corticosteroid therapy in children with severe PCP improves survival and their use is generally recommended. Patients suspected of having PCP should undergo bronchoalveolar lavage (BAL). Results of BAL will remain positive 3 to 4 days after the initiation of TMP-SMZ therapy. Therefore, if PCP is suspected, appropriate therapy should be started immediately and not be withheld pending lavage.

Bacterial Pneumonia

Background and Clinical Manifestations

The HIV-positive child with bacterial pneumonia is most often infected with the usual organisms: *Streptococcus pneumoniae, Hemophilus influenzae,* group A streptococcus, and *Moraxella catarrhalis.* Hospitalized children or those with indwelling devices may be infected with Gram-negative enteric organisms or *S. aureus.* In addition, these children often present with coinfection by respiratory viruses.

Management

A chest radiograph should be part of the evaluation of the HIV-positive child with fever of unknown origin or with respiratory signs or symptoms. The radiograph can help distinguish bacterial pneumonia (Fig. 85.3) from PCP or pulmonary lymphoid hyperplasia (PLH), and in fact, a chest radiograph compatible with bacterial pneumonia in an otherwise well-appearing child suggests that outpatient therapy may be possible if other criteria are met (Table 85.3).

Age >6 months
Able to tolerate oral fluids and medication
Absence of signs of respiratory distress (e.g., flaring or retractions)
Respiratory rate
<45 respirations/minute if age <2 years
<30 respirations/minute if age >2 years
Oxygen saturation ≥93%
Has not already worsened while receiving oral antibiotic
Close clinical follow-up available and patient able to return
Pneumocystis carinii pneumonia should be considered unlikely

Table 85.3. General Guidelines for Outpatient Therapy of Human Immunodeficiency Virus–Positive Children with Pneumonia

The chest radiograph in bacterial pneumonia typically reveals a lobar or segmental infiltrate, and the peripheral WBC count is often above 20,000/mm^3. However, because many of these children are leukopenic when not infected, it may be difficult in the ED to determine what constitutes leukocytosis. For example, a child whose normal WBC count is 2,000/µL may mount a WBC of 8,000 to 10,000/µL in response to a bacterial infection, but this would go unnoticed without knowledge of the child's baseline. Because it is rare to be confident that a pneumonia is not bacterial in origin, children with pulmonary signs and symptoms, especially associated with fever, are commonly given antimicrobial therapy against the common respiratory flora.

Other Pulmonary Infections

Other infections associated with respiratory signs and symptoms in HIV-positive children include viral illnesses, CMV

pneumonia, Mycobacterium tuberculosis, and MAI. Except for M. tuberculosis, which is surprisingly rare in HIV-infected children compared with adults, there has been little documentation of the frequency of the other infections. Adults have also had problems with coccidioidomycosis, blastomycosis, and histoplasmosis.

Wheezing

Background and Clinical Manifestations

Reactive airway disease is the most likely diagnosis in an HIV-positive child with wheezing, with or without fever. If the wheezing is associated with rales, however, the physician needs to consider the possibility of PCP or congestive heart failure. Congestive heart failure is rarely a presenting sign of HIV infection; instead, HIV-positive children with cardiomyopathy can develop congestive heart failure when under additional stress caused by an infection or fever. Physical examination may reveal the constellation of tachycardia, tachypnea, rales, and a palpable liver. However, these findings commonly occur in HIV-positive children without congestive heart failure.

Management

After a rapid but thorough physical examination to evaluate the degree of wheezing and air movement, the presence and location of retractions, and any other focus for fever, note whether the child responds to bronchodilator therapy. If so, reactive airway disease is the likely diagnosis, and the child can be treated accordingly. Pulse oximetry should be performed before the patient is discharged; although an oxygen saturation less than 95% is not an absolute indication for admission, a saturation of 95% or greater is reassuring. Five days of prednisone therapy (2 mg/kg per day divided into two doses, 60 mg/day maximum) can be given if it would be used for an immunocompetent child with the same clinical findings.

If the child has high fever (greater than 39°C [102.2°F]), a chest radiograph should be obtained to look for an infiltrate or evidence of PCP or congestive heart failure. The febrile child with wheezing and rales or evidence of pneumonia on chest radiograph who otherwise appears well enough for outpatient therapy may be given oral amoxicillin (60 to 80 mg/kg per day) or intramuscular (IM) ceftriaxone (50 mg/kg) in addition to bronchodilator therapy. However, no data provide clear support for this practice. These children should be reevaluated within 24 hours.

Children with clinical or radiographic evidence of PCP or congestive heart failure should be hospitalized. A first dose of IV TMP-SMZ should be given to infants suspected of having PCP, and congestive heart failure should be treated with afterload reducers and diuretics in addition to bronchodilators (see Cardiology section).

Lymphocytic Interstitial Pneumonitis

Background

Lymphocytic interstitial pneumonitis (LIP) is a condition that is relatively unique to pediatric HIV infection. It is thought to result from lymphoproliferative responses to Epstein-Barr DNA.

Clinical Manifestations

LIP is an insidious condition that causes a slowly progressive hypoxia typically in children who are older than 1 year of age. Table 85.4 contrasts the findings in LIP with those associated with PCP. Most children present to the ED with cough and mild tachypnea.

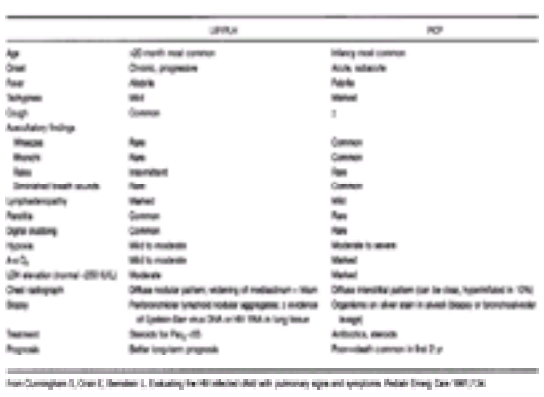

Table 85.4. Findings in Human Immunodeficiency Virus (HIV)–Positive Children with Lymphoid Interstitial Pneumonitis/Pulmonary Lymphoid Hyperplasia (LIP/PLH) vs. *P. carinii* pneumonia (PCP)

Typically, there will be marked lymphadenopathy and sometimes digital clubbing because of the chronic progressive hypoxia. Although in LIP there can be some elevation in the A-a oxygen gradient and LDH, the degree of elevation is usually less than with PCP, and it is not difficult to differentiate between the two entities.

Management

The approach to the evaluation of the HIV-infected child with respiratory signs and symptoms is outlined in Figure 85.4.

FIGURE 85.4. Evaluation of persistent respiratory signs and symptoms. *LDH*, lactate dehydrogenase; *CBC*, complete blood count; *Rx*, treatment; *PCP, Pneumocystis carinii* pneumonia. (Modified from Cunningham SJ, Crain EF, Bernstein LJ. Evaluations of the HIV-infected child with pulmonary signs and symptoms. Pediatr Emerg Care 1991;7:32–37.)

Rarely is intervention required to maintain the airway in children with LIP. Most HIV-positive children with a cough should undergo chest radiography. Chest radiography commonly reveals an interstitial nodular pattern that can be diagnostic. Occasionally, bronchiectasis develops, and these children may become superinfected with bacterial or viral pathogens and have fever. Fever is an important differentiating point in the management of HIV-positive children thought to have LIP. If the Pa_{O_2} is less than 65 mm Hg, LIP is treated with 1 to 2 mg/kg per day of prednisone to a maximum of 60 mg for 2 to 4 weeks and subsequently tapered as necessary to maintain the Pa_{O_2} above 70 mm Hg. If the patient is febrile, tuberculosis or MAI must be ruled out before beginning steroid therapy; these entities can appear similar to PLH both clinically and radiographically, but steroid therapy is contraindicated.

Gastrointestinal Tract

Chronic or recurrent oral thrush or esophageal candidiasis are common and can be treated with nystatin, clotrimazole, or fluconazole.

Diarrhea

Background

The gastrointestinal tract generally shows only subtle changes in histology in HIV infection unless there are secondary infections. In addition to the common causes of diarrhea that affect immunocompetent children, HIV-positive children are prone to parasitic *(Giardia, Microsporidium, Cryptosporidium)*, chronic viral (CMV), mycobacterial, and serious bacterial infections of the gastrointestinal tract (Table 85.5). Malnutrition is a common problem in HIV infection and, in children, requires careful monitoring of growth parameters.

Viral agents	*Shigella*
Rotavirus	*Escherichia coli*
Adenovirus	*Clostridium difficile*
Norwalk agent	*Giardia lamblia*
Cytomegalovirus	*Cryptosporidium*
Salmonella species	*Isospora*
Yersinia enterocolitica	*Microsporidia*
Campylobacter	

Table 85.5. Organisms Associated with Acute Diarrhea in Human Immunodeficiency Virus–Positive Children

Clinical Manifestations and Management

The general evaluation of the child with diarrhea is described in Chapter 19. Because diarrhea can be such a problem in HIV-infected children, the physician should seek to identify the cause. A stool test for blood, a stool smear for polymorphonuclear leukocytes, and a stool culture (for *Salmonella, Shigella, Yersinia, Campylobacter, Escherichia coli*) with consideration given to sending stool to test for parasites and *Clostridium difficile*. The child who is afebrile, appears to be well hydrated, and has no blood or leukocytes in the stool can be treated symptomatically with dietary management and close follow-up. If the child attends a day-care program and is still in diapers, the parents/guardians should be instructed to keep the child home until the illness has resolved and the culture is negative.

In febrile children with gastroenteritis, although viral causes are still most common, *Salmonella* is the primary bacterial pathogen of concern and is a major cause of bacteremia in HIV-positive children. If there is blood or more than 5 leukocytes per high-power field on examination of the stool smear but the child has normal vital signs and looks well, he or she should be treated with TMP-SMZ and reevaluated the next day. Oral ampicillin is an alternative drug, but in many

areas, 20 to 30% of *Salmonella* species are resistant to ampicillin. The child who appears dehydrated or ill should be admitted for IV hydration and parenteral antibiotic therapy.

If the patient remains symptomatic in the face of a negative stool culture and dietary management, a total of at least three stool specimens should be sent for ova and parasite evaluation and two samples tested for *C. difficile* toxin. If no cause is identified, endoscopy or colonoscopy should be considered, depending on the severity of symptoms.

Hematologic

Hematologic abnormalities are common and can be the result of HIV infection itself (occurring early or late in the illness), can be secondary to other concomitant infections (*Mycobacteria*, cytomegalovirus, parvovirus B19, fungal infections) or lymphoma, or can result from medication toxicities. Anemia is the most common, especially with zidovudine therapy; erythropoietin levels may be low, identifying patients likely to benefit from erythropoietin therapy. Nutritional deficiencies are common and may contribute to the anemia as well. Neutropenia is common and may be accentuated by medications (e.g., zidovudine, ganciclovir, TMP-SMZ) but is generally mild. If necessary, granulocyte colony–stimulating factor (G-CSF) or granulocyte-macrophage colony–stimulating factor (GM-CSF) can be given with improvement of the neutropenia. Thrombocytopenia is a typical part of the acute HIV infection and can persist or worsen over time. Improvement is often seen with antiviral therapy. If the thrombocytopenia is severe, it can generally be managed as would the non–HIV-infected patient with idiopathic thrombocytopenia (steroids, Rhogam, or gamma globulin).

Rash

Background and Clinical Manifestations

Of all the categories of dermatologic manifestations of HIV infection in children, seborrheic dermatitis and infections are by far the most common. This is in contradistinction to HIV-positive adults in whom neoplasms, particularly Kaposi's sarcoma, are seen more often. Measles can be particularly severe in HIV-positive children. The illness may be associated with the characteristic clinical signs and symptoms of generalized rash, coryza, conjunctivitis, cough, and Koplik's spots, or it may occur without the typical rash. The HIV-infected child with measles must be evaluated carefully for signs of dehydration and respiratory distress. If the child is taking liquids well and breathing comfortably, he or she may be sent home with careful instructions to return for reevaluation if status worsens. All HIV-positive children who have been exposed to measles should receive gamma globulin (0.5 mL/kg with maximum of 15 mL given intramuscularly), whether or not they have been vaccinated against measles.

Varicella can also cause severe illness in the immunocompromised host. Varicella-zoster immunoglobulin should be given to HIV-infected children after exposure to chickenpox (1 vial containing 125 U for each 10 kg of body weight with any opened vial used completely, maximal dose is 5 vials). Once clinical illness has started, these children should initially be treated with IV acyclovir (10 mg/kg every 8 hours). Children with local zoster infection may be treated with oral acyclovir (20 mg/kg per dose given every 6 hours). These children must be followed closely to ensure that the infection does not disseminate.

More than 10% of children infected with HIV will have thrombocytopenia associated with high levels of circulating immune complexes and antiplatelet antibodies that may manifest as petechiae or easy bruising. Patients with fewer than 50,000 platelets/µL should be considered for admission and treatment. Febrile or toxic-appearing HIV-positive children with petechiae must be considered to have septicemia (see Chapter 84). After a rapid assessment of the airway, breathing, and circulation, these patients should undergo a full evaluation for sepsis, including lumbar puncture, and they should receive parenteral antibiotics pending culture results.

Syphilis screening should be done for any HIV-infected child whose syphilis serology at birth is unknown because women with HIV have a high rate of coinfection with syphilis.

Neurologic Manifestations

Background

Once transmitted to the central nervous system, a neurotropic HIV strain emerges. In the early 1980s, a progressive dementia was reported in adults with AIDS. A syndrome analogous to the adult AIDS dementia complex was described in HIV-infected children in 1985 and was called AIDS encephalopathy. As it turns out, AIDS encephalopathy is common and does not require other manifestations of full-blown AIDS. More than 85% of children with HIV infection have neurologic involvement, and the possibility of HIV infection should be considered in the differential diagnosis of developmental delay or loss of milestones.

Clinical Manifestation.

These children exhibit developmental delay or developmental regression, acquired microcephaly, and pyramidal tract signs. Although growth and development are often affected by any serious illness in a child, AIDS encephalopathy may occur in patients with no signs of opportunistic infections and few signs of immunodeficiency. AIDS encephalopathy may manifest itself as a static, progressive, or indolent encephalopathy with periods of plateaus in cognitive and motor development. Static encephalopathy is defined by the presence of nonprogressive cognitive or motor deficits. Progressive encephalopathy usually begins within 2 months to 5 years after initial exposure to HIV, usually in the perinatal period. It is characterized initially by deterioration of play and progressive apathy. Developmental delay ensues, with loss of developmental milestones, including deficits in socially adaptive language and in fine and gross motor skills. As the condition progresses, there may be spastic diplegia and quadriparesis. Patients develop extrapyramidal and cerebellar signs, including rigidity, dystonic posturing, and ataxia. Seizures, although uncommon, may occur.

Another group of children present with indolent encephalopathy. These patients experience variable plateaus in their development during which there is little or no further cognitive growth. Either new milestones are not obtained or the rate of acquisition of new skills deviates from the norm and the child's initial rate of developmental progress. Many of these children may go on to develop the progressive form of AIDS encephalopathy.

Physical examination of the patient often reveals microcephaly. Younger children will be hypotonic with persistence of the Moro or tonic neck reflexes after 4 months of age. Older children may have symmetric ankle clonus and extensor plantar responses. As the condition progresses, pyramidal signs of varying severity, including a pure spastic quadriparesis with signs of pseudobulbar palsy, dysphagia, and dysarthria, are seen. Ataxia may be seen in children old enough to walk.

Managemen.

The diagnosis of AIDS encephalopathy involves obtaining a history suggestive of developmental delay or regression in an HIV-infected child. Management is more complicated when these children come to the ED with fever because it may be difficult to evaluate their mental status. In general, a lumbar puncture is necessary to rule out bacterial meningitis, unless the physician can be confident on a clinical basis that the child is behaving at baseline and that the fever is not secondary to central nervous system infection.

Other Neurologic Manifestation.

HIV-infected children are at increased risk of meningitis (including cryptococcal although less commonly than in adults, and tuberculous meningitis), encephalitis (including that caused by *Toxoplasma*), stroke and cerebral infarcts, progressive multifocal leukoencephalopathy (caused by human polyomavirus JC), and a variety of vasculopathies.

Cancers

Cancers have been seen less commonly in children than in adults. The most common malignancy of children with AIDS are non-Hodgkin's lymphomas; the second most common are leiomyomas and leiomyosarcomas in the gastrointestinal tract, and these are associated with EBV infection. Some other malignancies have also been linked to specific viral infections. Kaposi's sarcoma is associated with human herpes virus-8 infection, some peripheral lymphomas and most primary to the brain contain EBV, and anal cancers and cervical carcinomas are linked with human papilloma virus infections.

MANAGEMENT OF HUMAN IMMUNODEFICIENCY VIRUS

Overview of Anti-HIV Medications

Many effective anti-HIV medications are now available (Table 85.6). At present, these are in two general classes.

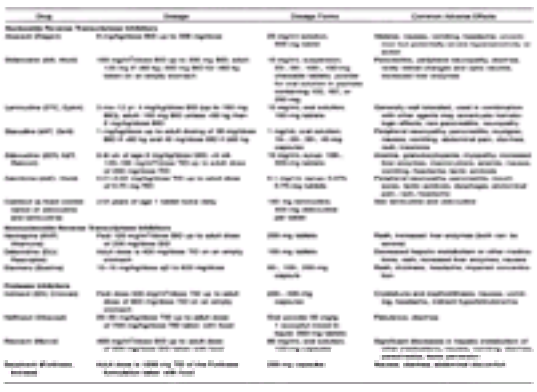

Table 85.6. Anti–Human Immunodeficiency Virus Medications

Reverse transcriptase inhibitors are nucleoside analogs that compete with the viral reverse transcriptase such as AZT (Retrovir, zidovudine), ddI (Videx, didanosine), ddC (Hivid, zalcitabine), d4T (Zerit, stavudine), abacavir (Ziagen), and 3TC (Epivir, lamivudine) or nonnucleoside reverse transcriptase inhibitors such as nevirapine (Viramune), delaviradine (Rescriptor), or etavirenz (Sustiva).

Protease inhibitors such as nelfinavir (Viracept), saquinavir (Invirase), ritonavir (Norvir), and indinavir (Crixivan) work by blocking a protease enzyme responsible for cleaving viral proteins into functional units.

Because HIV has the capacity to rapidly develop resistance to individual antiviral agents, combination therapy using three to four agents has now become the treatment standard.

Unfortunately, drug therapy is significantly limited. None of the drugs currently available has been shown to eradicate infection. The efficacy of these antivirals in various tissues that 1) may harbor virus, 2) be important in spread of virus, or 3) be important to symptomatology (e.g., lymph nodes, brain, testes, mucosal surfaces) is not always known. These drugs also commonly cause significant side effects and drug interactions that may bring patients to the ED for attention. These most commonly include rash, headache, nausea, diarrhea, pancreatitis, fatigue, anemia, granulocytopenia, peripheral neuropathy, renal stones, decreased absorption of other medications, and increased or decreased metabolism

of other medications.

Prevention of HIV Acquisition

Development of an effective and safe vaccine for the prevention of HIV is a high priority but will be unavailable for several years. Current strategies for preventing HIV infection take advantage of the finding that it may take several hours after exposure (or possibly in some cases days) for HIV infection to become established. During this time, antiviral medications can be given to prevent the transmitted virus from causing an established infection. Prevention of perinatal acquisition through the use of anti-HIV medications during the last trimester of pregnancy and the first few weeks of infancy (only AZT has been studied) has been successful in significantly reducing the risk of infection. The introduction of AZT therapy has largely been credited with the reduction in pediatric HIV cases currently seen in the United States. Unfortunately, the cost of this treatment is prohibitive in the countries where 90% of HIV infections occur. In addition, antiviral medications have significant side effects, and when considered for use to prevent transmission of HIV, the risk of drug toxicity must be weighed against the risk of HIV acquisition and the potential benefit of therapy. The risk of acquisition from exposures to HIV are listed in Table 85.7.

Exposure	Risk of Infection (per 1000)
Transfusion with positive blood unit	950
Intravenous drug use	7
Percutaneous exposure (needlestick)	3
Receptive anal intercourse	3
Receptive vaginal intercourse	1
Receptive oral sex	Low but reported
Perinatal exposure without zidovudine	250
Perinatal exposure with zidovudine	80
Breast-feeding (not single exposure)	120

Table 85.7. Approximate Risk of Human Immunodeficiency Virus Acquisition after a Single Exposure Listed by Source

Postexposure Prophylaxis

Community Exposures

Sexual contact is the most common means of transmitting HIV infection, and reducing exposure is the mainstay of public health efforts. Prophylaxis after sexual intercourse or sharing needles could potentially decrease transmission, although efficacy has not yet been demonstrated. A patient with a significant exposure to HIV, such as receptive or penetrative anal or vaginal sex, receptive oral intercourse with ejaculation, or needle sharing involving a partner who is HIV positive or who is in a known risk group should be considered for postexposure prophylaxis (PEP). At the time of this writing, no consensus guidelines have been published regarding PEP for sexual assault victims. The possibility of HIV transmission with such contacts should be discussed with the victim, the risk of transmission assessed, and PEP offered or recommended if appropriate. Risk becomes more than low risk if there is ejaculation (to risk area), multiple assailants, injuries that involve blood, evidence to suggest illicit drug use on the part of the assailant(s), and/or threats or suggestion that the assailant(s) is HIV positive. The patient who has had consensual sex with an individual known to have or to be at high risk of having HIV in which safe sex practices were not followed (or failed, e.g., broken condom) should also be offered HIV prophylaxis and risk reduction counseling.

Accidental community-acquired needlestick exposures also require attention. HIV has been detected and infectious virus has been recovered from syringes obtained from high-risk community sources. Most discarded syringes will not have any recoverable HIV, and if complete drying of the syringe has occurred, there will be no infectious HIV. Thus far, no report exists of transmission of HIV from a discarded syringe left in a public place. Therefore, the risk of transmission of HIV from an accidental needlestick from a needle/syringe found in a public place is likely low. Prophylaxis should not be routinely recommended, but because the risk is not zero, careful discussion with the family should include the need for subsequent monitoring and testing of the patient. Voluntary sharing of needles among IV drug abusers would pose a substantially higher risk because immediate use of the needle would result in a greater likelihood of infectious virus being transmitted. A prophylaxis strategy in this situation would demand the patient remove themselves from ongoing exposures, placing them at risk of acquiring HIV.

When considered appropriate (Fig. 85.5), PEP should be instituted as quickly as possible after exposure. Most would limit PEP to exposures within 72 hours of exposure (preferably less than 24 hours). Patients receiving PEP should receive first doses of medication in the ED and will need follow-up with clinicians knowledgeable about antiviral therapies and able to provide intensive emotional and behavioral counseling to help cope with the immediate event and to help them avoid situations likely to result in future exposure. The medications, which must be taken for 4 weeks, can have significant side effects.

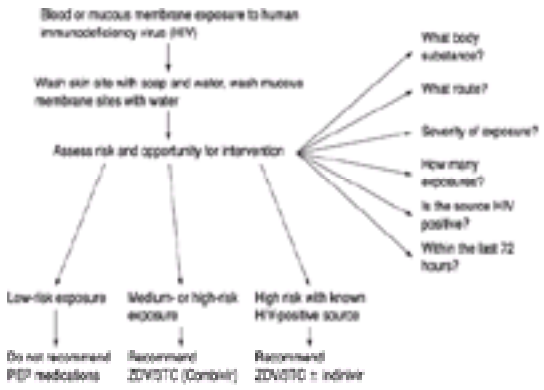

FIGURE 85.5. Postexposure prophylaxis for possible community acquired exposures to human immunodeficiency virus. When available, additional information regarding the source individual may alter recommendations. *PEP*, postexposure prophylaxis.

Health Care Worker Exposures

An increased risk for HIV infection after percutaneous exposures to HIV-infected blood is associated with the quantity of blood, as indicated by visible contamination of the device with the patient's blood, its use directly in a vein or artery, or a deep injury to the health care worker. It is likely to increase with a higher viral load in the patient's blood. Finally, the risk of acquiring HIV is decreased an estimated 80% by the prompt postexposure use of effective antiretrovirals. Each health care institution should have a system to evaluate their employees occupational exposures and offer treatment. As of December, 1997, the Centers for Disease Control and Prevention was aware of 54 health care workers that had documented seroconversion after occupational exposures and another 132 HIV-positive health care workers who are likely to have acquired infection after an occupational exposure.

Acknowledgments

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Suggested Readings

GENERAL

American Academy of Pediatrics. Committee on Pediatric AIDS. Evaluation and medical treatment of the HIV-exposed infant. Pediatrics 1997;99:6, 909–917.

Kahn JO, Walker BD. Acute human immunodeficiency virus type 1 infection. N Engl J Med 1998;339:1, 33–39.

Levy JA. HIV and the Pathogenesis of AIDS. 2nd ed. Washington DC: American Society for Microbiology, 1998.

Shearer WT, Quinn TC, LaRussa P, et al. Viral load and disease progression in infants infected with human immunodeficiency virus type 1. Women and Infants Transmission Study Group. N Engl J Med 1997;336:19, 1337–1342.

EVALUATION OF THE FEBRILE CHILD

Andiman WA, Mezger J, Shapiro E. Invasive bacterial infections in children born to women infected with human immunodeficiency virus type 1. J Pediatrics 1994;124:846–852.

Bye MR. HIV in children. Clin Chest Med 1996;17:4, 787–796.

Dayan PS, Chamberlain JM, Arpadi SM, et al. Streptococcus pneumoniae bacteremia in children infected with HIV: presentation, course, and outcome. Pediatr Emerg Care 1998;14:194–197.

Dorfman DH, Crain EF, Bernstein LJ. Care of febrile children with HIV infection in the emergency department. Pediatr Emerg Care 1990;6:305.

Ellaurie M, Burns ER, Bernstein LJ, et al. Thrombocytopenia and human immunodeficiency virus in children. Pediatrics 1988;82:905.

Farley JJ, King JC Jr, Nair P, et al. Invasive pneumococcal disease among infected and uninfected children of mothers with human immunodeficiency virus infection. J Pediatr 1994;124:853–858.

Gesner M, Desiderio D, Kim M, et al. Streptococcus pneumoniae in human immunodeficiency virus type 1-infected children. Pediatr Infect Dis J 1994;13:697–703.

Krasinski K, Borkowsky W, Bonk S, et al. Bacterial infections in human immunodeficiency virus-infected children. Pediatr Infect Dis J 1988;7:323.

Mao C, Harper M, McIntosh K, et al. Invasive pneumococcal infections in human immunodeficiency virus-infected children. J Infect Dis 1996;173:870–6.

The National Institute of Child Health and Human Developments Intravenous Immunoglobulin Study Group. Intravenous immune globulin for the prevention of bacterial infections in children with symptomatic human immunodeficiency virus infection (See comments). N Engl J Med 1991;325:73–80.

1995 Revised guidelines for prophylaxis against Pneumocystis carinii pneumonia for children infected with or perinatally exposed to human immunodeficiency virus. MMWR 1995;44(RR-4):1–11.

Principi N, Marchisio P, Tornaghi R, et al. Occurrence of infections in children infected with human immunodeficiency virus. Pediatr Infect Dis J 1991;10:190.

NEUROLOGIC MANIFESTATIONS

Agostini HT, Ryschkewitsch CF, Mory R, et al. JC virus (JCV) genotypes in brain tissue from patients with progressive multifocal leukoencephalopathy (PML) and in urine from controls without PML: increased frequency of JCV type 2 in PML. J Infect Dis 1997;171:1.

Zuckerman G, Metrou M, Bernstein LJ, et al. Neurologic disorders and dermatologic manifestations in HIV-infected children. Pediatr Emerg Care 1991;7:99.

PREVENTION OF HIV ACQUISITION

Case-Control study of HIV seroconversion in health-care workers after percutaneous exposure to HIV-infected blood—France, United Kingdom, and United States, January 1988–August 1994. MMWR 1995;44(50):929–933.

Daily JP. Postexposure prophylaxis for HIV. AIDS Clin Care 1997;9:8, 59–61, 66.

Public Health Service Guidelines for the management of health-care worker exposures to HIV and recommendations for postexposure prophylaxis. MMWR 1998;47(RR-7):1–34.

Public Health Service Task Force recommendations for the use of antiretroviral drugs in pregnant women infected with HIV-1 for maternal health and for reducing perinatal HIV-1 transmission in the United States. MMWR 1998;47(RR-2).

Zamora AB, Rivera MO, Garcia-Algar O, et al. Detection of infectious human immunodeficiency type 1 virus in discarded syringes of intravenous drug users. Pediatr Infect Dis J 1998;17:7655–7657.

ANTIVIRAL MEDICATIONS

Drugs for HIV infection. Med Lett 1997;39:1015.

Drug Therapy: HIV-Protease Inhibitors. N Engl J Med 338:18, 1281–1292.

Guidelines for the use of antiretroviral agents in pediatric HIV infection. MMWR 1998;47(RR-4).

Report of the NIH panel to define principles of therapy of HIV infection and guidelines for the use of antiretroviral agents in HIV-infected adults and adolescents. MMWR 1998;47(RR-5).

USEFUL WEB SITES

Centers of Disease Control, Division of HIV/AIDS Prevention, National Center for HIV, STD, and TB Prevention (NCHSTP). http://www.cdc.gov/nchstp/hiv_aids/

HIV InSite: Gateway to AIDS Knowledge. http://hivinsite.ucsf.edu/

National Aids Treatment Information Project. http://www.kff.org/archive/aids_hiv/natip/html/

CHAPTER 86

Renal and Electrolyte Emergencies

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DEHYDRATION

Isotonic Dehydration

Background

Isotonic dehydration reflects a loss of total body water coupled with a loss of sodium (Na) of an equal magnitude. Clinically significant dehydration occurs commonly in children. Several factors predispose infants and children to dehydration, including 1) the frequent development of acute infections with high fevers, 2) a tendency to develop vomiting and diarrhea with both nonenteral and enteral infections, and 3) the greater ratio of surface area to mass of the child.

Children may lose fluids through the gastrointestinal (GI) tract, the kidneys, and/or the skin. In the otherwise healthy child, GI losses account for most instances of dehydration, the majority of these being caused by viral or bacterial enteritis.

Pathophysiology

An understanding of dehydration in the child requires a familiarity with fluid balance in the healthy person, as well as the pathophysiology of excessive fluid losses. The daily maintenance fluid requirement is directly related to caloric expenditure. Because surface area correlates best with caloric expenditure, this parameter often serves as a guide to fluid therapy. However, the physician either must memorize estimated surface areas for children of various ages or calculate the area for each child by using a nomogram based on height and weight. Alternatively, the weight alone can rapidly provide a basis for establishing maintenance requirements. Because surface area increases at a slower rate than body mass (weight), the increment in maintenance fluids per kilogram decreases as the child grows in size. The maintenance requirement can be calculated from the following formulas:

Weight (kg)	Fluid/24 hours
<10 kg	100 mL/kg
11–20 kg	100 mL × 10 + 50 mL for each kg over 10 kg
>20 kg	100 mL × 10 + 50 mL × 10 + 20 mL for each kg over 20 kg

The maintenance requirement for electrolytes directly relates to the caloric expenditure, which also determines the requirement for water. For Na, this is 2 to 3 mEq/100 mL, and for potassium (K), 2 mEq/100 mL. As an example, a 30-kg child would have a daily maintenance requirement for fluid of 1700 mL (100 mL/kg × 10 kg + 50 mL/kg × 10 kg + 20 mL/kg × 10 kg). The requirement for Na would be 34 to 51 mEq, and for K, 34 mEq.

Isotonic dehydration is defined as a loss of total body water with the maintenance of a normal serum [Na] ranging from 130 to 145 mEq/L. The final [Na] in the extracellular fluid (ECF) is a function not only of what is lost from the body but also of the composition of replacement fluids administered at home and transcellular shifts of fluid. The initial loss of fluid from the body depletes the ECF; gradually, water shifts from the intracellular space to maintain the ECF, and this fluid is lost if dehydration persists. If dehydration is acute (the duration of the illness is fewer than 3 days), approximately 80% of the fluid loss is from the ECF and 20% is from the intracellular fluid (ICF). Additional fluid moves out of the ICF if the loss continues for 3 days or more. In such cases, the proportion of fluid lost from the two compartments is 60% from ECF and 40% from ICF. ECF contains predominantly Na at a concentration of 140 mEq/L, and ICF contains K at a concentration of 150 mEq/L.

Clinical Manifestations

A careful history taken from the parents of a child with isotonic dehydration helps establish the cause for the fluid loss and to estimate the degree of depletion. Specific inquiries must be made about the volume of the child's intake and output, focusing on GI and renal function. The physician should ask about the amount and duration of vomiting, diarrhea, and other abnormal losses, as well as about the adequacy of urine flow. In infants, the absence of tears with crying and a decrease in the number of wet diapers suggest the presence of dehydration.

The physical examination should begin with a careful weighing of the child—the most accurate clinical indicator of dehydration. The signs of dehydration become increasingly severe as the degree of dehydration progresses (see Table 18.4). With a 5% loss of body weight, the skin and mucous membranes feel dry, but there are no signs of vascular instability. Tachycardia and hypotension appear as the loss of fluid exceeds this mark, and the skin turgor shows deterioration. The development of acidosis leads to tachypnea and contributes to poor peripheral perfusion. If the child has lost more than 10% of body weight in a brief time, signs of shock, including a rapid, thready pulse; marked hypotension; and cold, clammy skin, may appear (see Chapter 3).

I. Increased Total Body Na or Increased Total Body Na Greater Than Increased Total Body Water
A. Na poisoning (accidental; Na bicarbonate therapy)
B. Hyperaldosteronism (rare in children)
II. Normal Total Body Na; "Pure" Water Loss
A. Insensible losses—respiratory and skin
B. Renal (central and nephrogenic diabetes insipidus)
C. Inadequate access to water
III. Decreased Total Body Na Less Than Decreased Total Body Water
A. Extrarenal (gastrointestinal)*
B. Renal (osmotic diuretics; glucose, mannitol, urea)
C. Obstructive uropathy
IV. Normal Total Body Na and Water with Abnormal Central Osmotic Regulation of Water Balance
A. Essential hypernatremia

Na, sodium.
*In diarrheal states, hypernatremia usually results from a combination of relatively greater water than Na losses coupled with relatively greater Na than water replacement.

Table 86.4. Causes of Hypernatremia

By definition, the [Na] falls within the normal range, 130 to 145 mEq/L, in isotonic dehydration. In mild dehydration, the serum bicarbonate is usually 15 to 20 mEq/L, but it may drop to 6 to 10 mEq/L with more severe losses. The [K] is usually normal (3.5 to 5.0 mEq/L), although transcellular shifts from acidosis may elevate the serum level (p. 821). The blood urea nitrogen (BUN) rises proportionately to the degree of fluid loss, varying from 20 to 30 mg/dL in mild dehydration to 50 to 100 mg/dL in more severe cases. However, the BUN may show less of an increase than expected from the clinical estimate of dehydration if the child's protein intake has been significantly limited in the preceding 24 to 48 hours.

Urine flow is usually scant and may even be absent, but an effort should be made to obtain a specimen. In older children with dehydration, the specific gravity rises above 1.020, often reaching 1.035. Infants, however, have a relative lack of renal concentrating ability. Even in the face of severe dehydration in the child in the first 3 months of life, the urine specific gravity may be only 1.020.

Management

In all children with significant dehydration (5% or greater), an intravenous (IV) infusion should be started and blood sent to the laboratory for measurement of electrolytes and BUN. The child with shock demands immediate fluid resuscitation and monitoring as described in Chapter 3. An initial fluid bolus of 20 mL/kg physiologic saline given rapidly in 30 to 60 minutes is the treatment of choice. With lesser degrees of dehydration, the optimal type and rate of fluid infusion are

calculated on the basis of the child's estimated ideal weight and degree of dehydration. Sufficient fluids are administered in the first 24 hours to fulfill the maintenance requirement and correct the total deficit; half the deficit is replaced in the first 8 hours.

Example

A 5-month-old infant has a 3-day history of diarrhea and a decreased oral intake. One week previously, she weighed 5.0 kg in the pediatrician's office. Now she weighs 4.5 kg and has a temperature of 37.0°C (98.6°F), pulse of 120 beats/minute, respiratory rate of 30 breaths/minute, and blood pressure (BP) of 30/40 mm Hg. The child's skin and mucosa are dry, and the skin shows “tenting” if lifted. The urine specific gravity is 1.028, and the serum electrolytes are as follows: [Na] 135 mEq/L; [K] 4.0 mEq/L; [Cl] 90 mEq/L; and HCO_3] 9.0 mEq/L. The BUN is 30 mg/dL.

This infant has lost 10% of her body weight. Because acute decreases in weight reflect fluid loss, the fluid deficit is 500 mL. Dehydration has occurred over 3 days, indicating that 60% of the loss is from the ECF and 40% from the ICF. Thus, the Na and K deficits are calculated as follows:

$$\text{Na deficit} = 135\ \text{mEq/1000 mL} \times 0.6 \times 500\ \text{mL} = 40\ \text{mEq}$$

$$\text{K deficit} = 150\ \text{mEq/1000 mL} \times 0.4 \times 500\ \text{mL} = 30\ \text{mEq}$$

The maintenance requirements for this infant are 500 mL of fluid (100 mL/kg × 5 kg), 15 mEq of Na (3 mEq/100 mL × 500 mL), and 10 mEq of K (2 mEq/100 mL × 500 mL) for each 24-hour period.

Half of the fluid and Na deficits are replaced during the first 8 hours of rehydration. Thus, the infant would receive 250 mL of deficit fluid with 20 mEq of Na. In addition to this, one-third of the maintenance requirement, or 175 mL of fluid with 5 mEq of Na, would be given during that 8-hour period, for a total of 425 mL of fluid with 25 mEq of Na.

Only half of the K deficit is corrected during each of the first 2 days, and this is done at a constant rate. Because the maintenance K for this infant is 15 mEq and the deficit is 30 mEq, the amount of K administered would be 30 mEq daily (or 10 mEq for each 8-hour period) for the first 48 hours. The solution would also need to contain an appropriate amount of bicarbonate (p. 820). A maintenance amount of bicarbonate is 2 mEq/kg per day.

Hypotonic Dehydration

The pathophysiology and clinical findings for hypotonic (hyponatremic) dehydration are discussed in the section under Disorders of Sodium Homeostasis: Hyponatremia, below. An example follows.

Example

The 5-month-old infant described under the previous section on isotonic dehydration might well have presented with more pronounced clinical manifestations of dehydration and the following serum electrolytes: [Na] 128 mEq/L; [K] 4.0 mEq/L; [Cl] 90 mEq/L; and [HCO_3^-] 9.0 mEq/L. The BUN is 30 mg/dL.

Deficits of Na and K are calculated in the same fashion as given in the previous example of isotonic dehydration. Maintenance requirements are also the same. What is different about the calculations in this example is the added Na requirement in the deficit fluids to bring the serum Na from 128 to 135 mEq/L. The calculation should be as follows:

1. $\text{Additional mEq of Na} = [135 - 128\ \text{mEq/L}]_{\text{“ideal” – “observed”}} \times \underbrace{5.0}_{\text{“healthy” body weight (kg)}} \times \underbrace{0.6}_{\text{Na space}}$
2. $\text{Additional mEq of Na} = 7.0\ \text{mEq/L} \times 5\ \text{kg} \times 0.6 = 7.0 \times 3.0\ \text{L (kg)}$
3. $\text{Additional mEq of Na} = 21\ \text{mEq}$

This extra Na is added to the previously calculated 40 mEq Na in the deficit fluids, but the rate of repair may be staged in the same fashion as for isotonic dehydration.

Hypertonic Dehydration

The background, pathophysiology, clinical manifestations, and management of hypertonic (hypernatremic) dehydration are discussed under Hypernatremia (p. 816).

ELECTROLYTE DISORDERS

Disorders of Sodium Homeostasis: Hyponatremia

Background

Hyponatremia is defined as a measured serum [Na] of less than 130 mEq/L. This common electrolyte abnormality is commonly encountered in the emergency department (ED). The multiple causes of hyponatremia are grouped into four

categories (Table 86.1).

I.	Normal Total Body Water and Na (Hyperosmolar Hyponatremia)
	A. Hyperglycemia[a]
	B. Mannitol, glycerol therapy
II.	Increased Total Body Water and Na (Edema-Forming States)
	A. Congestive heart failure
	B. Nephrosis
	C. Cirrhosis
	D. Acute renal failure
III.	Decreased Total Body Water and Na (Hypovolemic States)
	A. Gastrointestinal losses (vomiting, diarrhea, fistulas)
	B. Renal losses (diuretics, renal tubular acidosis, primary interstitial disease)
	C. Adrenal (mineralocorticoid deficiencies)
	D. Third-space losses (ascites, burns, pancreatitis, peritonitis)
IV.	Increased Total Body Water but Normal Total Body Na
	A. Syndrome of inappropriate antidiuretic hormone secretion
	B. Water intoxication
	C. Miscellaneous (reset osmostat, hypothyroidism, glucocorticoid deficiency)
V.	Pseudohyponatremia
	A. Extreme hyperlipidemia or hyperproteinemia

Na, sodium.
[a]For every 100 mg/dL rise in plasma glucose concentration above normal, there is a correspondingly decrease in plasma sodium concentration of approximately 1.6 mEq/L.

Table 86.1. Causes of Rhabdomyolysis

Pathophysiology

Basic Mechanisms

Two fundamental principles regulating Na and water balance must be reviewed. First, total body Na determines ECF volume. This is because water moves freely throughout all body compartments to restore a disturbed osmotic equilibrium, and Na is the predominant ion of the ECF space. Second, the kidney normally defends against hyponatremia by its ability to dilute the urine. Any disorder that impairs urinary dilution will lead to hyponatremia (e.g., syndrome of inappropriate antidiuretic hormone [SIADH]). Hyponatremia may result after the restoration of osmotic equilibrium in response to an absolute or relative gain of water or an absolute or relative loss of salt from the body. Total body Na can be normal, low, or even increased in the face of hyponatremia.

Applications

In the edema-forming states (Table 86.1, II), a decreased effective circulating plasma volume is sensed by the kidney as hypoperfusion. Sodium is maximally reabsorbed, the urine is concentrated (ADH is secreted in excess), and hyponatremia often ensues. Acute reduction in glomerular filtration rate (GFR) and decreased delivery of fluid to the distal tubular diluting site often result in hyponatremia in acute renal failure (ARF); this is often exacerbated by patients ingesting hypotonic fluids.

Extrarenal losses of Na and water seen with diarrhea and or vomiting lead to negative Na balance and a diminished ECF volume (Table 86.1, III). Under these hypovolemic conditions ADH is released; the activation of the volume receptors leads to increased ADH secretion even in the face of hyponatremia. Water reabsorption is maximal, the urine is concentrated, and hyponatremia results. Preservation of the ECF volume and maintaining perfusion takes precedence over avoiding hyponatremia. Renal and adrenal Na wasting may be disease or drug induced (Table 86.1, III). Structural renal disease also impairs Na and chloride reabsorption at the diluting site, leading to hyponatremia. A negative Na balance and ECF volume contraction occur in the face of inappropriately high urine Na concentrations. ADH secretion is stimulated, and renal water excretion falls. Hyponatremia occurs in states with a normal total body Na, but abnormally increased water intake or excess ADH secretion (Table 86.1, IV).

Pseudohyponatremia (Table 86.1,V) develops when significantly elevated lipid or protein concentrations expand the nonaqueous plasma volume, and the laboratory reports Na concentrations in liters of plasma and not plasma water. Extreme hyperproteinemia and/or hyperlipidemia may result in an apparently low Na concentration in the ECF by decreasing the percentage of water contained in a unit volume of plasma (normally 93%). This displacement is given by the formula:

$$\text{mL of } H_2O \text{ per 100 mL of plasma} = 91.1 - [(1.03 \times \text{total lipid in g/dL}) + (0.73 + \text{total protein in g/dL})].$$

The most common causes of hyponatremia seen in the ED are GI losses and water intoxication. The latter occurs particularly in infancy and has been increasingly reported in recent years.

Clinical Manifestations

Symptoms and signs of hyponatremia are related to the absolute level and the rate of fall of serum Na from the normal range, but they tend to be somewhat nonspecific (Table 86.2). A child may be dramatically symptomatic at a serum Na of 125 mEq/L if the Na had fallen 15 mEq/L in only 1 to 2 hours and equilibrium had not yet been restored. In contrast, another child might be totally asymptomatic at a serum Na of 120 mEq/L if the Na had fallen 20 mEq/L in 2 to 3 days and osmotic equilibrium had been reestablished. Signs and symptoms are usually seen at serum Na lower than 120 mEq/L, but specific symptoms and signs do not correlate with specific levels of serum Na. The clinical examination can help in limiting the possible diagnoses to explain the hyponatremia. It is especially critical to note the presence of edema (Table 86.1, II) and hypovolemia (Table 86.1, III).

Symptoms	Signs
Anorexia	Clouded sensorium
Nausea	Decreased tendon reflexes
Muscle cramps	Pathologic reflexes
Lethargy	Cheyne-Stokes respiration
Apathy	Hypothermia
Disorientation	Pseudobulbar palsy
Agitation	Seizures
Acute respiratory failure	

Table 86.2. Symptoms and Signs of Hyponatremia

Complications of hyponatremia that require urgent diagnosis and treatment include Cheyne-Stokes respirations and seizures. However, a clouded sensorium and pathologic reflexes are often warning signs of seizures. The laboratory studies required to initiate the assessment of most children with symptomatic hyponatremia are given in Table 86.3.

I. Blood	II. Urine
A. Electrolytes (Na, K, Cl, HCO3)	A. Urinalysis, including specific gravity
B. BUN, creatinine	B. Urine Na
C. Liver function tests	C. Urine creatinine
D. Osmolality	D. Urine osmolality

Na, sodium; K, potassium; Cl, chloride; HCO3, bicarbonate.

Table 86.3. Laboratory Evaluation of Hyponatremia

Management

Armed with a working knowledge of pathophysiology, the clinical history and examination, and the few simple laboratory tests as outlined in Table 86.3, the emergency physician should be able to diagnose rapidly the specific cause of hyponatremia in most cases. A specific diagnosis is necessary because therapies differ significantly depending on the cause of the hyponatremia. A working schema is outlined in Figure 86.1. If measurement artifacts are ruled out, one must determine whether underlying disease, such as congestive heart failure, is present. If none of these conditions are present, the presence or absence of edema will be helpful; if no edema is appreciated, one must look for evidence of third spacing as in the case of pancreatitis. In the absence of third spacing, relevant historical questions may point in the direction of dehydration. If the patient is not dehydrated but hyperkalemia is present, this may suggest congenital adrenal hyperplasia or Addison's disease. The absence of hyperkalemia may point to polydipsia or inappropriately prepared formula as a cause. If not, and the urine osmolality is greater than serum osmolality, SIADH is possible. If the urine osmolality is normal, mild dehydration, occult water intoxication, or underlying disease should be considered (Table 86.1).

FIGURE 86.1. Diagnostic approach to hyponatremia. *SIADH,* syndrome of inappropriate secretion of antidiuretic hormone.

In the patient with hyponatremia in the face of obvious contraction of ECF volume (diarrhea or vomiting), reexpansion with isotonic saline is appropriate. The volume and rate of infusion are dictated by estimates of fluid loss (i.e., weight loss) made from the history and physical examination. In shock, 20 mL/kg isotonic saline can be administered rapidly over 1 hour or more as needed and then repeated as necessary until BP and peripheral circulation return to normal. Underlying diseases, such as renal tubular acidosis (RTA) and adrenal insufficiency, can be treated most effectively by

specific replacement therapy. Diuretics, if previously given, should be discontinued promptly.

In water intoxication, restriction of daily free water administration by 25 to 50%, depending on the chronicity and severity of the hyponatremia, is the treatment of choice. In the acutely ill patient with neurologic symptoms and signs, immediate relief may be accomplished temporarily by rapidly elevating the serum Na by 10 mEq/L up to 125 mEq/L with the IV administration of 3% sodium chloride as follows: mL 3% NaCl (0.5 mEq/mL) to give = 10 mEq/L × body weight (kg) × 0.6 (ECF space). Alternatively, 10 to 12 mL/kg of 3% NaCl can be infused over 1 hour. In adults, the rapid overcorrection of hyponatremia (e.g., an increase in serum Na of more than 2 mEq/L per hour) may be dangerous, producing the crippling or even fatal osmotic demyelination syndrome. The risk of this is greatest when a rapid overcorrection is made in a case of chronic hyponatremia.

In simple water intoxication, the normal kidney responds with maximal urinary dilution, which, when coupled with restriction of water intake, rapidly restores Na concentration to normal. In SIADH, water restriction is the initial treatment of choice but is not always effective. In the edema-forming states and ARF, hyponatremia is usually mild and water restriction usually suffices. In some patients, diuretics may be necessary to treat the underlying disease. In such situations, free water excretion is increased but at the risk of inducing ECF volume contraction through increased Na excretion. Admission is recommended for any patient with symptomatic hyponatremia or hyponatremia per se (less than 130 mEq/L) when the cause is not obvious.

Disorders of Sodium Homeostasis:Hypernatremia

Background

Hypernatremia is defined as a measured serum Na concentration of greater than 145 mEq/L in the ECF. As with hyponatremia, the causes of hypernatremia can be grouped into four major categories (Table 86.4) on the basis of net changes in total body water and Na.

Pathophysiology

In hypernatremia, either an absolute or relative water deficit can occur in the face of normal, increased, or even decreased total body Na (Table 86.4). In Na poisoning, water moves into the ECF in response to an increase in Na, and the initial physical findings are usually those of obvious expansion of the ECF (Table 86.4, I). Eventually, some of this “new” ECF water is lost when the kidney excretes the extra Na.

An important cause of hypernatremia in pediatrics is the ADH-deficiency syndrome (Table 86.4, II.B), caused by partial or complete central diabetes insipidus and partial or complete nephrogenic diabetes insipidus. These patients with “pure” renal water losses never manifest signs of ECF expansion or depletion, unless they are denied free access to water or have an associated hypothalamic (e.g., third center) disturbance, in which case signs of ECF volume depletion supervene. Normally, the maximum stimulus for urinary concentration is a body weight loss of 3 to 5%; this response is blunted or absent in diabetes insipidus, and the urine is inappropriately hypotonic. Compulsive water drinking is sometimes confused with pituitary diabetes insipidus but can be easily separated. Occasionally, a child loses large quantities of water through hyperventilation or sweating in a hot, humid environment without adequate water replacement, leading to hypernatremia. Here, too, the normal renal response concentrates the urine.

When total body Na is reduced through abnormal GI fluid losses, the physical signs are those of contraction of the ECF with recent weight loss, dry mucous membranes, and once fluid losses are significant, poor skin turgor (Table 86.4, III.A). The renal response is directed toward restoring the ECF volume with maximum conservation of Na and water. Thus, the urine is concentrated and contains little Na. During an osmotic diuresis, more water than Na is lost, resulting in hypernatremia and volume depletion. Excessive solute, not reabsorbed in the proximal tubule, obligates water delivery distally and impairs free water generation. This leads to a reduced renal concentrating capacity, enhancing urinary Na and water losses (Table 86.4, III.B).

The most common cause of hypernatremia encountered in the ED is hypernatremic dehydration secondary to diarrhea.

Clinical Manifestations

Most symptoms and signs of hypernatremia result from cellular dehydration as water moves into the ECF space to lower osmolality. Brain cells are the most vulnerable to water loss, especially if the loss is acute. Symptoms and signs range from lethargy and irritability to muscle weakness, convulsions, and coma. However, if hypernatremia develops more slowly, brain cells defend their volume by manufacturing additional intracellular solute, so-called idiogenic osmoles such as the amino acid taurine, that reduces water loss to the ECF. Therefore, symptoms and signs are related not only to the level of serum Na concentration but also to its rate of rise. By the same token, therefore, rapid restoration of ECF osmolality to normal after the slow development of a hyperosmolar (e.g., hypernatremic) state does not permit brain cells to inactivate idiogenic osmoles, and cerebral edema may result. Finally, because ECF volume is defended early in the course of a dehydrating illness associated with hypernatremia, the classic physical sign of decreased skin turgor is absent until total fluid losses are severe (»10 to 15% of body weight).

The major complications of hypernatremia that require urgent diagnosis are seizures and coma. Central nervous system (CNS) signs and symptoms correlate with the degree of hypernatremia, but long-term neurologic sequelae do not, although they may be seen more often than previously thought in patients with initial serum Na values greater than 160 mEq/L.

The laboratory studies required to initiate the assessment of most children with symptomatic hypernatremia are the same as those given for hyponatremia in Table 86.3.

Management

As in the hyponatremic states, the emergency physician should be able to reach a specific category of disease or diagnosis armed with a working knowledge of pathophysiology, clinical evaluation of the status of ECF volume, and a few simple serum and urine tests.

Because emergency therapies vary considerably among the disorders noted in Table 86.4, accuracy of interpretation is important. The algorithm in Figure 86.2 provides a working schema. If there is no known underlying disease, one should probe for a history of excess sodium intake. If signs of dehydration are present, hypertonic dehydration is likely; in the absence of signs of dehydration, other conditions, such as hyperaldosteronism, should be considered.

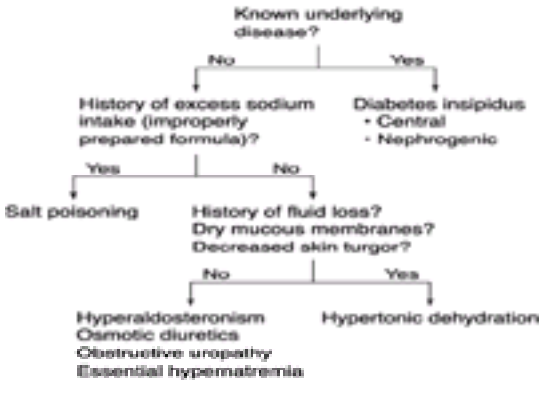

FIGURE 86.2. Diagnostic approach to hypernatremia.

In patients who are severely dehydrated and in shock, reexpansion with isotonic saline is the appropriate initial therapy (see Hyponatremia). Osmotic diuretics, if previously given, must be stopped. Replacement of water and Na losses with hypotonic electrolyte solutions is appropriate, but in the case of diarrhea, serum Na should be lowered slowly (usually no more than 10 to 15 mEq/L per 24 hours) to guard against brain edema. In hypertonic dehydration, a free-water deficit of 4 mL/kg for every 1 mEq/L of serum Na greater than 145 mEq/L should be replaced over 48 to 72 hours.

For example, a 5-month-old infant has a 3-day history of diarrhea. Last week she weighed 5.0 kg but now she weighs 4.5 kg. She has been receiving undiluted skim milk and “salt water” orally. Vital signs are normal. Electrolytes are as follows: [Na] 160 mEq/L; [Cl] 120 mEq/L; [CO_2] 10 mEq/L; and [K] 3.5 mEq/L. This patient has lost 10% of her body weight but is asymptomatic because of relative preservation of the ECF space in hypernatremia. That percentage of the total fluid deficit (i.e., 500 mL) that is to be replaced as solute-free water is calculated as follows:

$$\text{Free water (mL)} = [160 - 145\ \text{mEq/L}] \times 4\ \text{mL/kg} \times 5\ \text{kg} = 300\ \text{mL}$$

This is given slowly over 2 days to lower serum Na by approximately 10 mEq/L. The other 200 mL of the deficit are also given slowly over 2 days as solute-containing fluid. Calculation of the Na deficit (i.e., the amount of Na to put in the remaining 200 mL of volume deficit) can be made from any one of a number of general formulas based on certain fundamental assumptions about fluid and electrolyte physiology as discussed elsewhere (see Chapter 18). If one views the solute-containing fluid deficit as coming 40% from the ICF and 60% from the ECF, then deficits of Na and K are as follows:

1. ECF: $60\% \times 200\ \text{mL} \times 145\ \text{mEq/L}$, $[\text{Na}] = 17\ \text{mEq}$
2. ICF: $40\% \times 200\ \text{mL} \times 150\ \text{mEq/L}$, $[\text{K}] = 12\ \text{mEq}$

In patients with increased insensible water losses, simple free water replacement with glucose solutions is all that is required. Here, monitoring weight and serum Na concentrations are useful guides to the adequacy of therapy. Correction can usually be given over several days.

The emergency treatment of the diabetes insipidus syndromes is free water replacement, monitoring vital signs and clinical signs of dehydration, and serum Na concentration as guides to the rate and volume of replacement. There is no place for the various antidiuretic agents that promote ADH release or mimic its actions in central diabetes insipidus or for thiazide diuretics in nephrogenic diabetes insipidus as initial therapies.

Children who are victims of acute salt poisoning and who are severely symptomatic can safely have serum Na concentrations lowered rapidly, either by a combination of loop diuretics (furosemide 0.5 to 2 mg/kg every 12 hours) and glucose water administration or, rarely, peritoneal dialysis against a Na-free dialysate. The latter procedure should be carried out only in consultation with a nephrologist. Admission is recommended for any patient with symptomatic hypernatremia or severe hypernatremia per se (greater than 160 mEq/L) when the cause is not obvious.

Disorders of Potassium Homeostasis: Hypokalemia

Background

Hypokalemia is defined as a measured serum K concentration of less than 3.5 mEq/L. The three most common causes of hypokalemia seen in the pediatric ED are 1) gastric alkalosis from vomiting (e.g., pyloric stenosis), 2) chronic use of loop diuretics for conditions such as congenital heart disease and bronchopulmonary dysplasia, and 3) uncontrolled diabetic ketoacidosis (DKA). No specific environmental factors are known to result in hypokalemia. A rare genetic cause of hypokalemia that affects children is familial periodic paralysis. The causes of hypokalemia are varied. With the exception of transcellular shifts of K, most causes of hypokalemia result in increased renal K excretion, either as a primary or secondary event. The four major categories of hypokalemia are shown in Table 86.5.

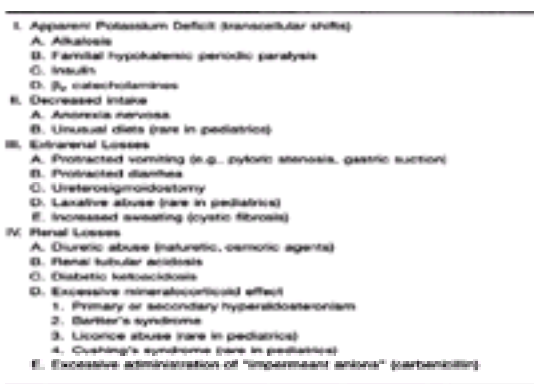

Table 86.5. Causes of Hypokalemia

Pathophysiology

Basic Mechanisms

When confronted with unexpected hypokalemia, the emergency physician is dealing with one of two pathophysiologic situations: 1) K shifts into cells in exchange for hydrogen ion (H^+), or 2) extrarenal or renal losses. Potassium shifts into cells in response to alkalosis and out of cells in response to acidosis; for every 0.1-unit rise or fall in pH, there is a reciprocal change in ECF K concentration of approximately 0.6 mEq/L. Insulin facilitates cellular uptake of K. In view of these observations and with the knowledge that K is primarily an intracellular cation, it can be appreciated that ECF K concentration does not reflect the status of total body K. Therefore, when evaluating total body K in light of disturbances in ECF K concentrations, one must know the plasma pH.

The mechanisms that govern renal K excretion are outlined in Table 86.6. Potassium is filtered at the glomerulus and reabsorbed in the proximal tubule (65%) and the ascending loop of Henle (25 to 30%). The K that appears in the urine is the result of distal tubular secretion. Hyperaldosteronism increases Na-K exchange in the distal tubule. Conditions that increase Na delivery to the distal tubule, such as volume expansion, also lead to increased K excretion.

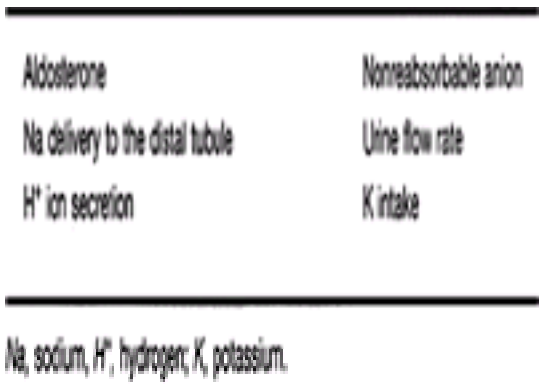

Table 86.6. Factors Governing Renal Potassium Excretion

Applications

Increased renal secretion of K is seen in 1) alkalosis, which increases distal tubular delivery of bicarbonate, 2) high cellular K concentration in response to elevated systemic pH, 3) increased urinary flow rate, and 4) increased K intake.

Potassium depletion and the resultant hypokalemia that occur with vomiting cannot result primarily from the K lost in the vomitus itself because gastric fluid concentration of K is only 5 to 10 mEq/L. Rather, renal losses account for most of the K deficit seen with vomiting. ECF volume depletion leads to secondary hyperaldosteronism, and alkalosis leads to increased bicarbonate delivery to the distal tubule. Both phenomena increase urinary losses of K. In addition, with continued volume depletion, alkalosis is maintained because the proximal tubule preferentially reabsorbs Na with bicarbonate to restore ECF volume (filtered chloride is reduced and K shifts into cells). Early in the course of vomiting, hypokalemia can be corrected merely by restoring ECF volume with isotonic saline without K supplements, providing

convincing evidence of the role of volume contraction in causing renal K wasting. Chronic diarrhea results in large K losses. Ureterosigmoidostomy can lead to K and bicarbonate secretion in exchange for Na and chloride (Cl) reabsorption, resulting in a hyperchloremic, hypokalemic acidosis. In cystic fibrosis, volume contraction can result from excessive cutaneous losses of water, Na, and Cl through increased sweating in the summer months. This may lead to renal K wasting and alkalosis.

One of the hallmarks of RTA is hypokalemia, which is paradoxical in view of the corresponding acidosis but occurs because renal K wasting results from secondary hyperaldosteronism and bicarbonaturia. The marked glycosuria of DKA increases urine flow rate and distal Na delivery, thus enhancing K excretion. Bartter's syndrome is an uncommon, poorly understood cause of hypochloremic, hypokalemic metabolic alkalosis. Hypokalemia is often profound and resistant to replacement therapy. The mechanism is probably either a primary renal chloride or K leak.

Clinical Manifestations

The cause of hypokalemia can usually be suspected as belonging to one particular diagnostic category after obtaining a careful history. For example, in familial periodic paralysis, the weakness comes on gradually over a few hours and may last 48 to 72 hours. It may be heralded by short episodes of weakness in one or more extremities. It usually occurs during periods of rest after vigorous exercise or a carbohydrate load.

Potassium depletion can result in widespread disturbances in cellular physiology and function, although symptoms are usually not seen at serum K concentrations above 3 mEq/L. The major abnormalities and their clinical consequences are listed in Table 86.7. The most important clinical manifestations of hypokalemia relate to abnormal neuromuscular function. Impulse formation and propagation and the resultant muscle contraction are impaired in both striated and smooth muscle, leading to ileus, tetany, skeletal muscle weakness, and if severe enough, paralysis and areflexia.

Muscle cell dysfunction (rhabdomyolysis)
Cardiac cell dysfunction (myocardiopathy, arrhythmias)
Neuromuscular dysfunction (weakness/paralysis, ileus, tetany, encephalopathy with underlying liver disease)
Renal (polydipsia, polyuria, concentration defect)

Table 86.7. Pathophysiological (Clinical) Consequences of Hypokalemia

Hypokalemia may cause rhabdomyolysis with myoglobinuria. Alteration of the cardiac action potential by slowing the rate of repolarization leads to conduction abnormalities and arrhythmias (see Chapter 82). However, although the signs and symptoms of hypokalemia generally parallel its rate of development and its severity, electrocardiogram (ECG) changes often fail to correlate with serum K. They are helpful if present but not reassuring if absent. In the presence of digitalis, however, hypokalemia is much more likely to produce cardiac arrhythmias.

Complications that require urgent diagnosis include acute respiratory failure from muscle paralysis, cardiac arrhythmias, and myoglobinuria, which can lead to ARF.

Laboratory and radiologic evaluations are outlined in Table 86.8. Generally, in situations of total body K depletion, a 1 mEq/L fall in serum K concentration reflects a 100 to 200 mEq K deficit. This figure may be somewhat lower in young children. The blood glucose rises in diabetes mellitus and creatinine phosphokinase with rhabdomyolysis. An increased BUN reflects contraction of ECF volume. If the electrolytes reveal a hyperchloremic hypokalemic metabolic acidosis with a normal anion gap and an alkaline urine pH, RTA should be suspected. When there is a hypochloremic metabolic alkalosis, the urine electrolytes are helpful. A urine chloride less than 10 mEq/L suggests vomiting, cystic fibrosis, or diuretic abuse as the cause of hypokalemia. A urine chloride greater than 20 mEq/L points to one of the disorders that lead to mineralocorticoid excess. When the urinary K is less than 10 mEq/L, several conclusions can be drawn. First, the K deficiency has probably been present for at least 2 weeks. Second, the kidney can be excluded as the route of K depletion. An elevated urinary K concentration, however, suggests either K wasting of short duration or a primary renal loss. In similar fashion, a urinary concentrating defect that persists in the face of a stimulus to concentrate bespeaks chronic K depletion.

I.	Blood
	A. Electrolytes (Na, K, Cl, HCO_3)
	B. Blood urea nitrogen, creatinine
	C. Glucose
	D. Arterial blood gas
	E. Creatine phosphokinase
II.	Urine
	A. Urinalysis
	B. Urine Na, K, Cl
	C. Urine pH
	D. Urine osmolality
III.	Other*
	A. Electrocardiogram
	B. Plain abdominal radiography
	C. Upper gastrointestinal series or ultrasound

Na, sodium; K, potassium; Cl, chloride; HCO_3, bicarbonate.
*Selection of studies depends on the suspected diagnoses.

Table 86.8. Laboratory Evaluation of Hypokalemia

Management

To effectively manage hypokalemia in the ED, one must delineate the source of the condition, as shown in Figure 86.3. An apparent K deficit (with normal total body K) caused by transcellular shifts may occur with alkalosis. Known underlying disease, such as cystic fibrosis and RTA, also causes hypokalemia. If the patient is acidotic, the emergency physician should consider DKA with a severe potassium deficiency, whereas alkalosis suggests conditions such as pyloric stenosis or cystic fibrosis. A normal pH indicates unusual diets (anorexia nervosa), Cushing's syndrome, or hyperaldosteronism.

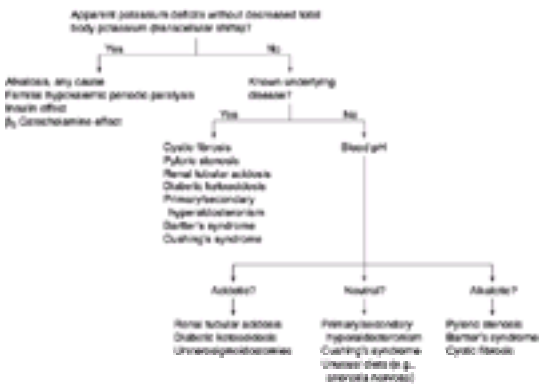

FIGURE 86.3. Diagnostic approach to hypokalemia.

When hypokalemia results from simple transcellular shifts in response to alkalosis without an accompanying K deficit, correction of the pH is all that is required. It is estimated that for every 0.1-unit change in pH, there is an average inverse change in the serum potassium of 0.6 mEq/L. In periodic paralysis, K supplementation with 2 to 6 mEq/kg per day is recommended with careful monitoring of serum K to avoid hyperkalemia as paralysis subsides. In most circumstances, K repletion should be slow (over days) and given by the oral route once urine flow is confirmed. IV loading should be avoided except under special conditions. Despite the fact that ECF K concentration does not reflect accurately total body K deficits, serum K concentration is the only practical way of assessing adequacy of replacement and avoiding unwanted complications. An estimate of the K deficit is generally obtained from the degree of hypokalemia and the blood pH, and it can be replaced over 2 to 3 days (assuming no ongoing losses) (Table 86.9). If IV replacement must be used, no more than 40 mEq/L of K should be given by peripheral vein and 80 mEq/L by central vein. In terms of the quantitative rate of repair, this should be no more than 0.2 to 0.3 mEq K/kg per hour. However, if potentially life-threatening cardiac arrhythmias or respiratory paralysis are evident, up to 1 mEq/kg per hour can be given by infusion pump with continuous ECG monitoring. Finally, the selection of the specific K salt used in repairing deficits is important. Generally, potassium chloride should be used if there is alkalosis, and potassium bicarbonate or its equivalent should be used if there is acidosis. In states of ECF volume depletion from any cause, volume replacement with isotonic saline is as important as K replacement in normalizing serum K and turning off renal K wasting.

Serum K (mg/L)	Plasma pH	K Deficit (%)
2.9	7.6	18
2.5	7.5	15
3.0	7.4	10
3.5	7.3	10

K, potassium.

Table 86.9. Twenty-kg Child; Total Body Potassium = 40–50 mEq/kg or 800–1000 mEq

The child with symptomatic hypokalemia requires admission for therapy and monitoring (and possibly for diagnostic workup), as do most children with a serum K of less than 3.0 mEq/L.

Disorders of Potassium Homeostasis: Hyperkalemia

Background

Hyperkalemia is defined as a measured serum K concentration greater than 5.5 mEq/L. No specific environmental factors are known to result in hyperkalemia. The causes of hyperkalemia are varied. With the exception of transcellular shifts of

K, most of the common causes of hyperkalemia result from impaired renal excretion because of decreased glomerular filtration, low urine flow, or decreased tubular secretion. Occasionally, exogenous or endogenous (i.e., from cell breakdown) K loading is responsible for the observed hyperkalemia. The categories of hyperkalemia are shown in Table 86.10.

I. Pseudohyperkalemia (Hemolysis, extreme leukocytosis, or thrombocytosis)
II. Apparent K Excess (Transcellular shifts)
A. Acidosis
III. Increased Intake
A. Endogenous (rhabdomyolysis, massive hemolysis)
B. Exogenous (suicide attempt with K salts)
IV. Decreased Excretion
A. Acute or chronic renal failure (oliguria)
B. Adrenal corticoid deficiency (acute adrenal insufficiency, hyporeninemic hypoaldosteronism)
C. Use of K-sparing diuretics in renal failure or in conjunction with dietary K supplements
D. β-Blockers converting enzyme inhibitors

K, potassium.

Table 86.10. Causes of Hyperkalemia

Pathophysiology

Pseudohyperkalemia is seen when blood is drawn after prolonged application of a tourniquet but is easily diagnosed by repeating the K measurement after drawing blood without a tourniquet. It may also occur with extreme leukocytosis or extreme thrombocytosis. In these situations, the diagnosis is made by measuring the white blood cell (WBC) or platelet count or plasma K. Hyperkalemia that results from transcellular K shifts in response to acidosis can be seen with normal or decreased total body K. In the former case, simple correction of the pH is all that is required. However, hyperkalemia from acidosis usually occurs in the face of total body K deficits such as diarrheal dehydration or DKA. In such a case, it is vitally important to begin K replacement as the pH is being returned to normal, thus avoiding a sudden fall in serum K concentration as it shifts back into cells.

The pathophysiology of hyperkalemia in cases of endogenous K release from cells is straightforward. Cellular catabolism occurs in the face of negative nitrogen balance from any cause; in pediatrics, this usually results from dietary protein restriction. Injury from trauma or burns can accelerate K delivery into the ECF, but life-threatening hyperkalemia rarely ensues if renal function is intact. The most common cause of hyperkalemia encountered in the ED is probably metabolic acidosis followed by reduced renal excretion. The latter occurs commonly in ARF from any cause, especially when there is concomitant oliguria. (See Table 86.6 for factors that control urinary K excretion.) In adrenal corticoid deficiency states or in patients who receive K-sparing diuretics, distal tubular secretion of K is impaired.

Clinical Manifestations

The predominant symptoms and signs are neuromuscular. Paresthesias are followed by weakness and even flaccid paralysis. Major toxicity is reflected in the ECG. The earliest change is symmetric peaking of the T wave, then widening of the P-R interval. First-degree heart block, loss of the P wave, ventricular arrhythmias, and cardiac standstill may follow. Cardiac arrest is more commonly seen with hyperkalemia than with hypokalemia. In general, the ECG changes parallel the degree of hyperkalemia when it has developed acutely. The presence of any ECG changes associated with hyperkalemia mandates urgent diagnosis and therapy.

The laboratory evaluation of hyperkalemia is outlined in Table 86.11. Although hypokalemia may cause rhabdomyolysis, hyperkalemia is often an early and life-threatening consequence of rhabdomyolysis from other causes. An elevated BUN and creatinine point to ARF. The urinary electrolyte pattern of the untreated patient can be helpful if adrenal corticoid deficiency is suspected. In acute adrenal insufficiency, urine Na concentration is inappropriately high and urine K concentration is inappropriately low for their respective serum concentrations.

I. Blood	II. Urine
A. Electrolytes (Na, K, Cl, HCO_3)	A. Urinalysis
B. Blood urea nitrogen, creatinine	B. Urine Na, K, Cl
C. Glucose	C. Urine pH
D. Arterial blood gas	D. Urine osmolality
E. Creatine phosphokinase	III. Other
	A. Electrocardiogram

Na, sodium; K, potassium; Cl, chloride; HCO_3, bicarbonate.

Table 86.11. Laboratory Evaluation of Hyperkalemia

Management

Determining the origin of hyperkalemia is crucial before managing this life-threatening condition. Figure 86.4 provides an algorithm for ascertaining the cause. If the patient has apparent K surplus, conditions such as hemolysis or metabolic acidosis may be responsible. Conversely, if there is no evidence of K surplus, a known underlying disease such as acute adrenal insufficiency may be present. When there is no known underlying disease and the blood pH indicates acidosis, acute or chronic oliguric renal failure may offer an explanation. Alkalosis points to excessive potassium intake, rhabdomyolysis, or ingestion of K-sparing diuretics, b-blockers, or angiotensin-converting enzyme (ACE) inhibitors. A normal pH suggests a hyporeninemic state.

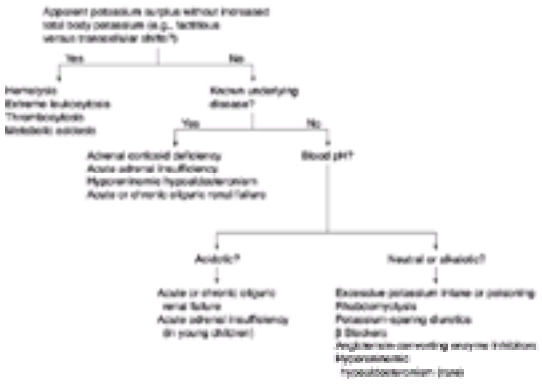

FIGURE 86.4. Diagnostic approach to hyperkalemia.

Three general techniques are used (Table 86.12) to lower serum K levels to normal: 1) reverse the membrane effects, 2) transfer K into cells, and 3) enhance renal excretion of K. If patients are asymptomatic, serum K is less than 6.5 mEq/L, and the ECG is normal or reveals only peaked T waves, all that may be required is discontinuation of K intake, removal of K-sparing diuretics if they are being used, and treatment of acidosis. Exceptions to this occur in acute oliguric renal failure and rhabdomyolysis, in which the serum K level may rise to much higher levels precipitously, and a more aggressive therapeutic approach is indicated.

Technique	Agent	Dose	Route of Administration	Onset/Duration of Action	Comment
Reversal of membrane effects	10% Calcium Gluconate	0.5 mL/kg	3-5 min IV	Min/30-60 min	ECG monitor; discontinue if pulse rate <100
Movement of K into cells	Na bicarbonate, 7.5% (1 mEq = 1 mL)	2-4 mL/kg	30-60 min	30 min/1-4 hr	May use in the absence of acidosis
	Glucose 50% plus insulin (regular)	1 unit for every 3-4 g glucose	Bolus	Bolus	Monitor blood glucose
Enhanced excretion of K	Kayexalate	1 g/kg	Can be given in 10% glucose (1 g/3-4 mL) every 4-6 hr	Hours/variable	Can be given PO or by rectum

IV, intravenous; ECG, electrocardiogram; K, potassium; Na, sodium; PO, orally

Table 86.12. Emergency Treatment of Hyperkalemia

When ECG changes are more widespread and/or serum K is greater than 7.0 mEq/L, several available therapies are designed to move K into cells acutely, including glucose and insulin combination and Na bicarbonate. The latter agent has recently been shown to be effective even in the absence of acidosis. With the onset of cardiac arrhythmias or a serum K level greater than 8.0 mEq/L, urgent therapy is needed. Under continuous ECG monitoring, IV calcium is given first to reverse potentially life- threatening arrhythmias without altering serum K. Calcium accomplishes this by restoring a more normal differential between the threshold and resting transmembrane potentials. This then may be followed by glucose and insulin and Na bicarbonate.

For more long-term control of hyperkalemia, the cation exchange resin Na polystyrene sulfonate (Kayexalate) can be administered. Finally, in patients with oliguric renal failure, peritoneal dialysis removes potassium, although the immediate fall in serum levels may reflect redistribution caused by the alkalinizing effect of dialysis and the glucose load in the dialysate itself. The dosages of drugs used to treat hyperkalemia, the recommended rates of administration, and onset of action are detailed in Table 86.12.

Any child with symptomatic hyperkalemia or a serum K level greater than 6.5 mEq/L on a nonhemolized sample deserves admission for therapy and additional workup.

Disorders of Calcium Homeostasis: Hypocalcemia

Background

Hypocalcemia beyond the neonatal period is defined as a measured total serum calcium (Ca) concentration of less than 9.0 mg/dL. There are three major categories of hypocalcemia in infants and children, as noted in Table 86.13.

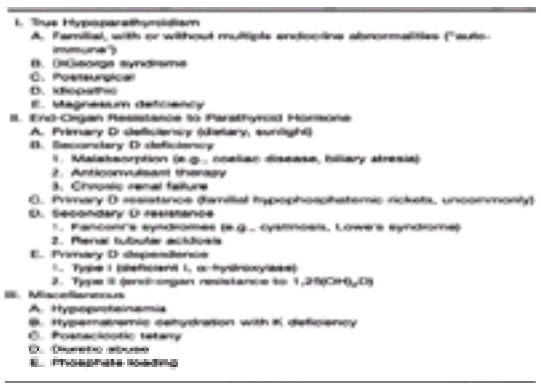

Table 86.13. Causes of Hypocalcemia

Pathophysiology

Basic Mechanisms

The skeleton contains 99% of total body Ca; the remaining 1% is distributed in intravascular, interstitial, and intracellular fluids. Most of the skeletal Ca is in a nonexchangeable pool and is unavailable for moment-to-moment regulation of Ca homeostasis. In addition, there is an as yet unclear but reciprocal relationship between serum Ca and serum inorganic phosphorus; factors that raise phosphorus result in a lowering of Ca and vice versa. The active form of vitamin D [1,25(OH)$_2$D] promotes Ca and phosphorus absorption from the gut when present in physiologic concentrations. It also enhances the parathyroid hormone (PTH)-dependent mobilization of Ca from mineralized bone and has a small but important action on the renal conservation of Ca. Thus, 1,25(OH) $_2$D is a Ca-promoting hormone that serves to raise serum Ca in response to hypocalcemia and/or increased tissue demands for Ca. PTH secretion is enhanced by hypocalcemia and suppressed by hypercalcemia. Serum Ca is raised in response to increased PTH by at least two mechanisms: 1) increased Ca resorption from bone and 2) increased renal excretion of phosphorus. Calcitonin acts in response to hypercalcemia by inhibiting bone resorption and thereby lowering Ca delivery into the ECF. The action of 1,25(OH)$_2$D and PTH is through receptor binding and stimulation of mediators such as cyclic adenosine monophosphate (cAMP). Therefore, hypocalcemia could result from an absolute deficiency of one or both of these compounds, end-organ resistance caused by a lack of receptors or abnormal receptor binding, or impaired formation or action of the mediators.

Applications

Examples of all of these potential abnormalities have now been described and are outlined in Table 86.13. The causes of hypocalcemia listed under Table 86.13, I and II are rarely encountered in the ED.

In simple or primary vitamin D deficiency, impaired Ca absorption and bone resorption cause the observed hypocalcemia, although 1,25(OH)$_2$D may be normal or even elevated in blood. The anticonvulsant agents diphenylhydantoin and phenobarbital probably induce hepatic microsomal enzymes that increase the conversion of 25(OH)D in the liver to inactive metabolites. In addition, diphenylhydantoins also interfere with intestinal calcium absorption. Hypocalcemia is uncommon in children who take anticonvulsant agents unless they are also receiving a poor Ca intake and have little sunlight exposure. When chronic renal insufficiency is moderate to severe (i.e., GFR of 50% of normal or lower), a decrease in renal mass leads to decreased production of 1,25(OH) $_2$D. In addition, decreased renal excretion of phosphorus causes hyperphosphatemia and a reciprocal fall in serum Ca. Secondary hyperparathyroidism then ensues. Dietary intake of Ca is often decreased in these patients. For all of these reasons, hypocalcemia is common and often resistant in this condition. Occasionally, hypocalcemia is seen in ARF, especially when acidosis is vigorously treated with alkali.

Hypocalcemia is not commonly seen but may occur in familial hypophosphatemic rickets. In RTA, the combination of hypercalciuria secondary to obligatory bicarbonaturia and acidosis and probable impairment in 1,25(OH) $_2$D production may lead to hypocalcemia. Hypocalcemia is often profound and refractory to therapy in the various forms of vitamin D dependence. The characteristic blood profile in this and the other abnormalities of vitamin D metabolism are outlined in Table 86.14.

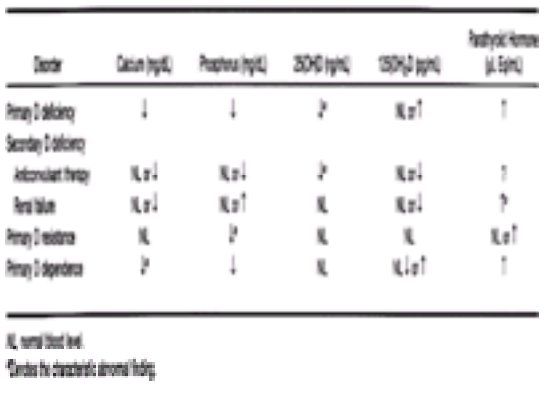

Table 86.14. Blood Profile in Disorders of Vitamin D Metabolism

Hypocalcemia that results from a lowered protein-bound fraction is commonly associated with the hypoproteinemia of nephrotic syndrome or protein-losing enteropathy, although ionized Ca is normal. The hypocalcemia is mild, usually in the range of 7.5 to 9.0 mg/dL, and rarely causes symptoms. Hypernatremic dehydration in association with a K deficit results in hypocalcemia for reasons that are unclear. Overvigorous correction of acidosis, especially in the dehydrated child, will drive Ca into bone, resulting in hypocalcemic tetany. Diuretic abuse with the potent loop blockers (ethacrynic acid, furosemide) may cause massive hypercalciuria and hypocalcemia, especially in the face of a hypochloremic alkalosis. Phosphate loading, either exogenous or endogenous (e.g., lysis of tumor cells), results in hyperphosphatemia, with a corresponding fall in serum Ca until the extra phosphorus is excreted.

Clinical Manifestations

The signs and symptoms of hypocalcemia per se are primarily neuromuscular in origin. Nonspecific findings (vomiting, muscle weakness, and irritability) are common. In addition to the characteristic tetany and positive Chvostek's and Trousseau's signs, there may be frank seizures or laryngospasm with upper airway obstruction. Rickets is characterized by thinning of the inner table of the skull (craniotabes), enlarged costochondral junctions (rachitic rosary), and thickening of the wrists and ankles. The ECG may reveal a prolonged Q-T interval.

Complications that require urgent diagnosis include frank tetany, laryngospasm, and seizures. Laboratory studies that should be included in the initial evaluation are listed in Table 86.15. The diagnosis of primary hypoparathyroidism is established by finding hypocalcemia, hyperphosphatemia, undetectable PTH, and a normal renal and skeletal (calcemic) response to exogenous PTH. Hypomagnesemia may also result in functional hypoparathyroidism. The characteristic laboratory profiles of the various vitamin D abnormalities are outlined in Table 86.14. In this latter group of diseases, symptomatic hypocalcemia is often preceded by poor linear growth and clinical rickets.

I. Blood	II. Urine
A. Calcium (total and ionized), phosphorus, alkaline phosphatase	A. Calcium, phosphorus
B. Magnesium	B. Creatinine
C. Total protein, albumin	III. Other[a]
D. Blood urea nitrogen, creatinine	A. Electrocardiogram
E. Parathyroid hormone	B. Skull, chest radiograph
F. pH	

[a]Selection of studies depends on the suspected diagnosis.

Table 86.15. Laboratory Evaluation of Hypocalcemia

Management

Specific therapy varies by diagnosis (Fig. 86.5). If there is an apparent Ca deficit (without evidence of hypocalcemia), hypoalbuminemia must be considered. When calcium deficiency is suspected, the serum phosphate must be measured. If this value is normal, entities such as anticonvulsant therapy and malabsorptive syndromes may explain the hypocalcemia. If the serum phosphate level is elevated, true hypoparathyroidism should be considered. A low serum phosphate level suggests primary vitamin D deficiency, RTA, or other conditions (Fig. 86.5).

FIGURE 86.5. Diagnostic approach to hypocalcemia.

The emergency treatment of choice for hypocalcemia from any cause other than hypomagnesemia is IV calcium. It should be given under cardiac monitoring at a starting dose of 0.5 to 1.0 mL/kg as 10% Ca gluconate administered over 3 to 5 minutes. The infusion should stop if the heart rate falls below 60 beats/minute. Once symptoms are relieved, Ca gluconate can then be added to the IV solution (100 mg of elemental Ca/kg per 24 hours) or Ca may be administered orally. When magnesium deficiency is suspected or confirmed as the cause of hypocalcemia, magnesium should be administered, usually by the intramuscular (IM) route. One gram of 50% magnesium sulfate contains 99 mg of magnesium, or roughly 8 mEq. The dose of magnesium is 0.5 mEq/kg (6 mg/kg) or 0.125 mL/kg of the 50% solution.

Children with unanticipated hypocalcemic tetany, seizures, or laryngospasm should be admitted for treatment and workup for the underlying cause of the hypocalcemia.

Disorders of Calcium Homeostasis: Hypercalcemia

Background

Hypercalcemia is defined as a measured total serum Ca concentration of greater than 11.0 mg/dL. This discussion excludes hypercalcemia in the newborn. The major causes of hypercalcemia in infants and children are outlined in Table 86.16. Hypercalcemia can result from increased Ca absorption from the gut or increased Ca resorption from bone. Hypercalcemia occasionally results from a massive increase in dietary Ca intake or a reduction in the renal excretion of Ca.

I. Primary Hyperparathyroidism (rare in pediatrics)
  A. Sporadic
  B. Familial (with or without associated endocrine abnormalities)
II. Infantile Hypercalcemia
III. Vitamin D Intoxication
IV. Immobilization
V. Malignant Disease
  A. Bony metastases (especially associated with lymphoreticular malignancy)
  B. Ectopic production of parathyroid hormone or other bone-resorbing factors
VI. Miscellaneous
  A. Sarcoidosis (rare in pediatrics)
  B. Thiazide diuretics
  C. Hypervitaminosis A

*This list is not meant to be comprehensive but includes the important causes in children.

Table 86.16. Causes of Hypercalcemia[a]

Pathophysiology

In hyperparathyroidism, increased bone resorption of Ca results in hypercalcemia, and decreased renal reabsorption of phosphorus results in hypophosphatemia. Because PTH stimulates bone turnover, alkaline phosphatase is elevated. PTH is inappropriately elevated for the level of serum Ca and confirms the diagnosis.

The vitamin D toxicity syndromes can usually be suspected from the history; hypercalcemia is the result of increased Ca absorption. If the intoxicating compound was conventional vitamin D, the hypercalcemia may be prolonged because of the storage of this compound in adipose tissue.

Immobilization hypercalcemia occurs typically in the adolescent who is growing rapidly. Acute injury or illness that requires prolonged immobilization (especially in traction) leads first to hypercalciuria and then to hypercalcemia. The presumed cause is increased bone resorption in the face of decreased or arrested bone mineralization.

Lymphoreticular malignancies in childhood may be associated with hypercalcemia from one of several mechanisms: 1) bony metastases with localized bone resorption; 2) rarely, the elaboration of a PTH-like peptide by tumor cells; and 3) release of other factors that promote bone resorption, such as prostaglandin E or osteoclast activating factor. In sarcoidosis, there is a heightened sensitivity to the Ca-absorbing effects of vitamin D, but the precise mechanism is unclear. Thiazide diuretics reduce renal Ca excretion probably by two mechanisms: 1) ECF contraction that leads to enhanced proximal tubular reabsorption of Ca with Na, and 2) increased renal tubular sensitivity to PTH-induced Ca reabsorption. Hypervitaminosis A can increase skeletal resorption of Ca, leading to hypercalcemia. Associated and characteristic findings are failure to thrive, dry skin and rash, poor hair texture, papilledema, and headache.

Clinical Manifestations

Symptoms and signs of hypercalcemia are listed in Table 86.17 and are grouped by organ system. Mild hypercalcemia (Ca 11 to 13 mg/dL) usually produces headache, irritability, and GI upset. When serum Ca rises above 14 to 15 mg/dL abruptly, a life-threatening hypercalcemic crisis may occur, consisting of severe vomiting, hypertension, polyuric dehydration, ARF, and coma. Laboratory studies that should be included in the initial evaluation are shown in Table 86.18. In any patient with unexplained hypercalcemia, an appropriate workup for hidden malignancy should be initiated once the hypercalcemia is controlled.

I. Neurologic
  A. Headache, irritability, lethargy, fatigue
  B. Weakness, seizures, coma
  C. Hyporeflexia, behavioral changes
II. Gastrointestinal
  A. Anorexia, nausea, vomiting, constipation
  B. Dehydration
III. Cardiovascular
  A. Bradycardia, hypertension, short QTc interval (electrocardiograph)
IV. Renal
  A. Polydipsia, polyuria
  B. Hypokalemia, aminoaciduria, nephrocalcinosis, nephrolithiasis
V. Dermatologic
  A. Pruritus
  B. Band keratopathy, ectopic calcification

Table 86.17. Signs and Symptoms of Hypercalcemia

I. Blood
A. Calcium (total and ionized), phosphorus, alkaline phosphatase
B. Total protein, albumin
C. Blood urea nitrogen, creatinine
D. Parathyroid hormone
E. Vitamin D[a]
II. Urine
A. Calcium, phosphorus
B. Creatinine
III. Other[b]
A. Electrocardiograph
B. Skull, abdominal radiograph
C. Skeletal survey
D. Intravenous pyelogram

[a]If vitamin D intoxication is suspected, blood for 25(OH)D should be drawn acutely and sent to the appropriate reference laboratory.
[b]Selection of studies depends on the suspected diagnosis.

Table 86.18. Laboratory Evaluation of Hypercalcemia

Management

Treatment of hypercalcemia is facilitated by knowing the underlying cause of the derangement. Figure 86.6 provides an algorithmic approach to delineating the cause. Entities such as malignancy and sarcoidosis may result in hypercalcemia; if there is no known underlying disease and the serum phosphorus is normal, immobilization, thiazide diuretics, or hypervitaminosis A may be responsible. If the serum phosphorus level is low, primary hyperparathyroidism must be considered. If the phosphate level is elevated, vitamin D intoxication is a possibility.

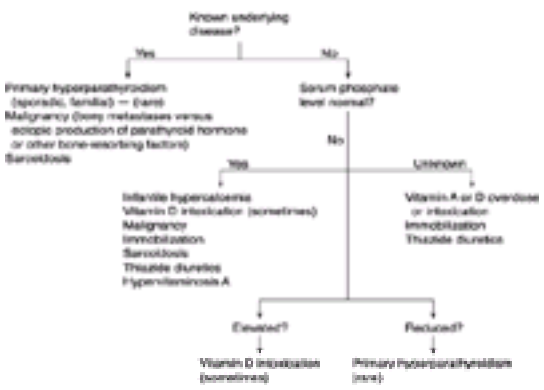

FIGURE 86.6. Diagnostic approach to hypercalcemia.

The choice of therapy depends on whether the kidneys are functioning normally. The initial emergency treatment for symptomatic hypercalcemia is designed to enhance Ca excretion by saline infusion at a rate of twice maintenance followed by bolus injections of furosemide, 1 to 2 mg/kg every 6 to 8 hours. The subsequent amount and rate of saline to be administered depends on the state of hydration and presence or absence of hypertension or preexisting cardiac disease, but in an otherwise normal patient, saline flow rates of two to three times daily maintenance would be appropriate until the serum Ca returns to normal. Treatment of a hypercalcemic crisis depends on the underlying cause, the level of serum Ca, and the severity of signs and symptoms. It always requires hospitalization in an intensive care setting. In acute oliguric renal failure, peritoneal or hemodialysis against a low Ca dialysate is usually effective, albeit slowly administered over hours. Any child in hypercalcemic crisis or with a serum Ca greater than 13 mg/dL should be admitted for therapy and diagnostic evaluation.

Disorders of Magnesium Homeostasis: Hypomagnesemia

Background

Serum magnesium (Mg) levels range from 1.5 to 2.2 mEq/L and do not vary with age. Balance studies indicate that approximately 50% of ingested Mg is absorbed in the intestine, with an average adult diet containing 200 to 700 mg/day. Normally, the kidney reabsorbs 95% of the filtered Mg, but virtual exclusion of Mg from the urine can occur in states of dietary Mg deprivation or extrarenal losses after 2 to 3 days. Diuretics and volume expanders all enhance urine Mg excretion. Mg deficiency can occur in the hospitalized child and is of particular importance in the intensive care unit, where up to 20% of patients may experience hypomagnesemia.

Pathophysiology

Hypomagnesemia is defined as a serum level less than 1.5 mEq/L. The major causes are related to GI and renal losses and are outlined in Table 86.19. GI losses account for most cases of hypomagnesemia in children. Upper GI fluids contain only 1 to 2 mEq/L Mg, but diarrheal fluid can contain up to 15 mEq/L. Renal excretion parallels urine flow, as well

as Na and Ca excretion; factors that increase Ca and/or Na excretion also enhance Mg excretion. Many drugs induce renal Mg wasting through proximal tubular cell injury.

I. Gastrointestinal Disorders
A. Acute or chronic diarrhea
B. Malabsorption states
C. After extensive bowel resection
D. Enteric fistulas
E. Prolonged nasogastric suction
II. Renal Loss
A. Osmotic diuresis (glucose, mannitol)
B. Chronic parenteral fluid therapy
C. Alcoholism
D. Hypercalcemia
E. Chronic renal disease (e.g., tubulointerstitial)
F. Drugs
1. Diuretics
2. Aminoglycosides, amphotericin B
3. Cisplatin, cyclosporin
III. Endocrine-Metabolic
A. Diabetes mellitus
B. Phosphate depletion
C. Hyperparathyroidism or hypoparathyroidism
D. Primary aldosteronism

Table 86.19. Causes of Hypomagnesemia

Clinical Manifestations

The clinical signs and symptoms of hypomagnesemia relate to signs of neuromuscular irritability, like those seen with low serum Ca. Chvostek's and Trousseau's signs may be seen along with carpopedal spasms. Ataxia, vertigo, athetoid, choreiform movements, tremors, nystagmus, and seizures are manifestations of severe deficiency. If Mg deficiency is subacute or chronic, muscle weakness and atrophy may be noted. ECG changes (prolonged P-R interval, long Q-T interval, tachyarrhythmias) may be noted.

Management

Figure 86.7 presents an approach to diagnosing the causes of hypomagnesemia and determining subsequent therapy. Known underlying disease, such as short gut syndrome and malabsorptive states, can lead to this condition. In the absence of known diseases, chronic diarrhea, enteric fistulae, and chronic diuretic use are possibilities.

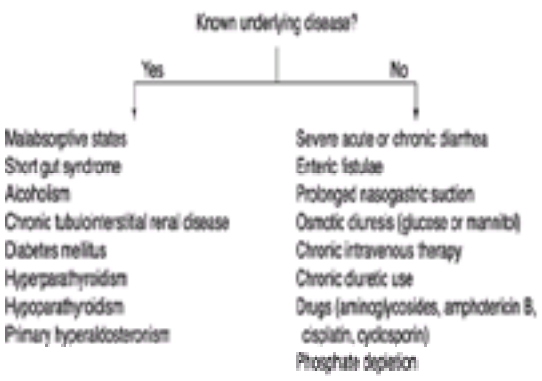

FIGURE 86.7. Diagnostic approach to hypomagnesemia.

Patients with symptoms and signs of Mg deficiency should be treated, although the magnitude of the deficiency is difficult to estimate. IM injections of $MgSO_4$ are particularly painful in children. Therefore, an IV injection or infusion is preferred in the ED, particularly in cases of seizures or cardiac arrhythmias. An initial dose of 25 to 50 mg/kg can be given either as the 10 or 50% solution (100 or 500 mg/mL), then repeated every 4 to 6 hours as necessary. Alternatively, a constant infusion of between 100 to 200 mg/kg per day may occasionally be needed to keep serum Mg normal.

Disorders of Magnesium Homeostasis: Hypermagnesemia

Background

Hypermagnesemia is uncommon in pediatrics but, as in adults, is usually related to accidental overdose. This is particularly true in patients with impaired renal function.

Pathophysiology

Blood levels exceed 2.2 mEq/L in these patients. Antacids, enemas, purgatives, or IV solutions that contain Mg usually account for the source of the intoxication.

Clinical Manifestations

Neuromuscular symptoms and signs are the earliest and most predominant presentations of hypermagnesemia and roughly parallel blood levels. BP may fall at levels greater than 5 mEq/L. Reflexes are lost early (e.g., 4 to 5 mEq/L), followed by depressed respirations and even apnea (e.g., 8 to 10 mEq/L). ECG changes include increased P-R, QRS,

and Q-T intervals (also noted at Mg levels greater than 5 mEq/L). At levels greater than 15 mEq/L, heart block may be noted. Coexisting hypocalcemia is often noted in patients with hypermagnesemia, possibly reflecting a suppressive effect of high Mg levels on PTH secretion.

Management

The first principle of therapy is to discontinue any form of Mg intake. If renal function is normal, hydration and diuresis are indicated. The signs and symptoms of acute, severe Mg intoxication may be antagonized initially by IV infusion of Ca (e.g., 0.5 mL/kg of 10% Ca gluconate). Patients with renal insufficiency and magnesium intoxication should undergo acute peritoneal dialysis or hemodialysis using a low Mg bath.

Disorders of Acid-Base Homeostasis: Metabolic Acidosis

Background

Definition

Of the four primary acid-base disorders, only metabolic acidosis is discussed in this section because it is the most commonly encountered acid-base disturbance in pediatric emergencies. *Metabolic acidosis* is defined as a net gain in H^+ ions or a net loss of bicarbonate HCO_3^- ions in the ECF. Clinically, this is reflected by a fall in plasma or serum HCO_3^- (or in some laboratories total, CO_2 or CO_2 that is approximately 95% HCO_3^-). The lower limit of normal for infants and children is 20 mEq/L.

Epidemiology

Most of the common causes of metabolic acidosis are acquired. However, certain inborn errors of metabolism may present clinically with severe metabolic acidosis in the first few months of life. An important epidemiologic consideration in all children with unexplained metabolic acidosis is accidental or intentional poisoning with any number of agents.

Etiology

The causes of metabolic acidosis are conveniently grouped into two major categories: those with a normal anion gap (also known as the “delta” or “R” fraction) and those with an increased anion gap. The anion gap is determined from the serum electrolytes by the following formula:

$$\text{Anion gap} = \underset{\text{mEq/L}}{\text{Serum [Na] mEq/L}} - \underset{\text{mEq/L}}{[Cl^- + HCO_3^-]}$$

Potassium is not included in this formula because it is present only in relatively low concentrations that vary slightly compared with the other ions. We use 16±4 mEq/L as the normal range for patients less than 2 years old. The causes of metabolic acidosis are outlined in Table 86.20. An algorithmic guide (Fig. 86.8) to the causes of metabolic acidosis is based on the value of the anion gap. If the anion gap is elevated and there is known disease, possibilities include DKA, acute or chronic renal failure, or diarrheal dehydration. If the anion gap is elevated without a history of known disease, it is valuable to check the serum lactate. If this is elevated, hypoxia and sepsis must be considered. Normal serum lactate points to conditions such as poisoning. If the anion gap is normal, renal disease such as RTA and tubulointerstitial disease are possibilities. When the anion gap is normal and there is no known underlying disease, entities such as hypernatremic dehydration, enteric fistulae, and drug must be considered.

I. Elevated Anion Gap Acidosis
   A. Diarrheal dehydration
   B. Diabetic ketoacidosis
   C. Renal failure (acute or chronic)
   D. Inborn errors of metabolism
   E. Poisons (e.g., salicylates, ethanol, ethylene glycol)
   F. Lactic acidosis (e.g., hypoxia, sepsis, idiopathic)
II. Normal Anion Gap Acidosis
   A. Hypernatremic dehydration (older children)
   B. Renal tubular acidosis
   C. Hyperalimentation
   D. Enteric fistulas (e.g., pancreatic) or enterostomies
   E. Ureterosigmoidostomy
   F. Drugs (e.g., Sulfamylon, ammonium chloride, amphotericin, acetazolamide)
   G. Early renal failure (chronic interstitial nephritis)
   H. Dilution (rapid volume expansion)

Table 86.20. Cause of Metabolic Acidosis

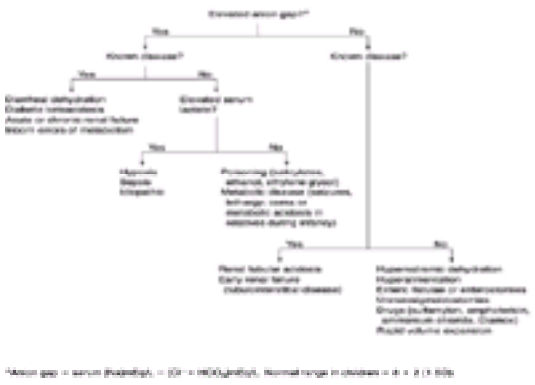

FIGURE 86.8. Diagnostic approach to metabolic acidosis (reduced serum bicarbonate for age). [a]Anion gap = serum [Na]mEq/L – [Cl^- + HCO_3]mEq/L. Normal range in children =16±4 mEq/L.

Pathophysiology

Basic Mechanisms

Metabolic acidosis results from one of three general pathophysiologic mechanisms: 1) increased H^+ ion delivery into the ECF, 2) increased HCO_3^- loss from the ECF (GI or renal), and 3) decreased renal H^+ ion excretion. The result is a fall in serum tCO_2 or HCO_3^-; the pH that is the negative logarithm of the [H^+] is usually below normal (less than 7.37) unless there is a second acid-base disturbance that drives the pH in the opposite direction.

ECF [H^+] or pH is maintained within narrow limits by a series of buffers, the most important of which is the bicarbonate buffer system. It operates in the ECF (35% of total buffering) and ICF of red blood cells (RBCs) (18% of total buffering) and provides immediate defense against life-threatening acidemia. It is an excellent and versatile buffer system for two reasons: 1) the carbonic acid-bicarbonate system represents a weak acid-strong conjugate base system that readily accepts protons of H^+ ions from nonvolatile acids in the usual range of plasma pH, and 2) the product that results from adding H^+ to this system is CO_2, which can be blown off by the lungs, thus blunting the net change in H^+ or pH.

The overall buffering mechanisms in response to metabolic acidosis are listed in Table 86.21. Of practical importance is the length of time it takes each mechanism to operate in returning pH to normal. Eventual full correction of metabolic acidosis requires renal excretion of excess H^+ ions, which takes several days to accomplish. Normally, children produce twice as much acid as adults per day (i.e., 2 to 3 mEq/kg). Because the final pH that results from metabolic acidosis is a reflection of the extent of the physiologic compensatory (i.e., adaptive) response, as well as the magnitude of the initiating disturbance, we must briefly consider this adaptive response. The compensatory response to metabolic acidosis is hyperventilation. This process produces a respiratory alkalosis, defined as a P_{CO_2} lower than normal (e.g., 40 mm Hg), which returns the pH *toward,* but *not to,* normal. This response is immediate and, for all practical purposes, is completed by the time the emergency physician sees the child with a metabolic acidosis (e.g., more than 12 to 24 hours after the onset of the inciting illness). The stimuli to hyperventilation are probably 1) through peripheral chemoreceptors that immediately sense the fall in plasma pH or rise in H^+ and, somewhat later, 2) the respiratory center that senses similar changes in the cerebrospinal fluid (CSF). These later changes do not occur immediately because P_{CO_2} diffuses across the blood-brain barrier faster than HCO_3^-, and initially the CSF pH may actually rise. A useful formula that predicts the normal adaptive response to metabolic acidosis is as follows:

1. Extracellular buffering (instantaneous)	3. Intracellular buffering (2–4 hr)
$H^+ + Buf^- \rightarrow HBuf$	Diffusion of H^+ into cells
$H^+ + HCO_3^- \rightarrow H_2CO_3$	H^+ + proteinate$^-$ → H protein
2. Respiratory buffering (10–15 min)	H^+ + hemoglobinate$^-$ →
$H^+ + HCO_3^- \rightleftharpoons H_2CO_3 \rightleftharpoons H_2O +$	H hemoglobin
$CO_2 \uparrow$	$H^+ + PO_4^- \rightarrow HPO_4^-$

Table 86.21. Buffering Mechanisms in Metabolic Acidosis

$$P_{CO_2} = 1.5 \times [HCO_3^-] + 8 \pm 2$$

If the P_{CO_2} is greater than expected from the calculations, a second acid-base disturbance, respiratory acidosis, should be suspected. In other words, the compensatory response is inadequate. If, however, there is an exaggerated compensatory response with a P_{CO_2} lower than expected from the calculations, a primary respiratory alkalosis should be suspected. Note that this formula probably does not apply to pH values less than 7.00.

Net renal acid excretion is accomplished by two mechanisms: 1) H^+ ion secretion by the renal tubule in exchange for HCO_3^- ion, and 2) H^+ ion binding to ammonia. Negligible H^+ ion is quantitatively excreted in the free state.

Applications

Elevated Anion Gap Let us first consider the causes of an elevated anion gap acidosis (Table 86.20). Diarrheal dehydration in infants and young children is the most common cause of metabolic acidosis. It tends to produce an elevated anion gap because of the early development of tissue catabolism and starvation ketosis. These result in increased H^+ ion production in association with release of increased amounts of organic anions into the ECF. DKA results in the increased production of b-hydroxybutyric acid and acetoacetic acid, both of which raise the anion gap. In severe renal failure, normally occurring anions, including phosphates, sulfates, and creatinine, accumulate. In addition, tissue catabolism ensues early in the face of oliguria and decreased caloric intake. The acidosis itself is also contributed to by decreased ammonia production by the damaged kidney.

A number of the rarely seen inborn errors of metabolism may present with severe metabolic acidosis after the institution of milk (protein) feeds. Various poisons, but especially salicylates, cause metabolic acidosis, either by their metabolic conversion to acids that are fully ionized at body pH or by liberation of endogenous acids as a consequence of interference with normal metabolic pathways. Suspicion of occult poisoning can be derived from a laboratory clue—an increased osmolal gap. The normal difference between the measured and calculated serum osmolality is 10 mOsm/L or less. The calculated osmolality equals 2[Na] + BUN over 2.8 + glucose over 18. Alcohols such as methanol or ethylene glycol are often responsible for a raised osmolal gap. Lactic acidosis is probably a more common cause of metabolic acidosis than is currently recognized in pediatrics. It most typically develops in the setting of acute circulatory and/or respiratory failure with shock, hypoxia, and poor tissue perfusion. A common predisposing event is sepsis with Gram-negative organisms. Hepatic failure, drugs and toxins, type I glycogen storage disease, and pulmonary embolus are other causes seen in children. In a significant number of patients, the cause is unknown. Primary hyperventilation produces moderate elevations in serum lactate but rarely leads to symptomatic acidosis.

Normal Anion Gap We can now turn to a consideration of the common causes of a normal anion gap acidosis (Table 86.20). In most cases, this results from a relative or absolute hyperchloremia. In hypernatremic dehydration secondary to diarrhea seen in adults and children, increased Cl^- reabsorption in the large bowel may account in part for the relative hyperchloremia. In addition, in extreme hypertonic states (i.e., serum Na greater than 165 mEq/L), Na^+ may be underestimated relative to Cl^- if electrolytes are being measured by the autoanalyzer technique. The hyperchloremia seen in both the proximal and distal forms of RTA is thought to result from a renal loss of HCO_3^- without a corresponding loss of chloride. The Na^+ deficit occasioned by the loss of $NaHCO_3$ and other Na^+ salts in turn leads to volume contraction, stimulating the renal tubular reabsorption of NaCl. Effectively, $NaHCO_3$ is replaced by NaCl in the ECF. Another clue to the diagnosis is an inappropriately alkaline urine pH (greater than 5.5) in the face of systemic acidosis.

Other more rare causes of a normal anion gap acidosis include therapy with hyperalimentation solutions, amphotericin B, and Sulfamylon.

Enteric fistulas and ureterosigmoidostomies probably produce a normal anion gap acidosis through enhanced Cl^--HCO_3^- exchange in the bowel.

Chronic interstitial nephritis with early renal failure (GFR 25 to 50% of normal) may result in a hyperchloremic acidosis. The pathophysiologic picture is one of RTA with renal bicarbonate wasting, but an additional component is reduced ammonia production and ammonium excretion.

Clinical Manifestations

The clinical manifestations of metabolic acidosis usually reflect the predisposing illness and are not unique in themselves. Nonetheless, in some patients, the presenting complaints appear to result primarily from the acid-base disturbance, in that they resolve after bicarbonate therapy. The signs and symptoms include tachypnea with or without hyperventilation, abdominal pain, vomiting, unexplained fever, and lethargy. Tachypnea and hyperpnea are characteristic of severe lactic acidosis, and coma may ensue if the pH is significantly depressed. There is often an associated but unexplained leukocytosis.

The urgency to diagnose metabolic acidosis is linked to the clinical imperative of defending the blood pH within a life-sustaining range. Factors that mandate rapid diagnosis (therapy) include 1) a severely depressed blood pH (less than 7.15 to 7.20), indicating marked acidemia; 2) a critically ill patient with multisystem disease, especially pulmonary and/or renal disease; 3) inability to treat the underlying disease effectively; and 4) the combination of hypoxia and acidemia that together can cause myocardial depression.

The laboratory studies required to diagnose and characterize metabolic acidosis are given in Table 86.22. A measurement of pH is needed to assess the potential urgency of alkali therapy, and the remainder of the arterial blood gas analysis is needed to assess the adequacy of respiratory compensation. The simultaneous measurement of blood and urine pH provides a clue to the diagnosis of RTA; this diagnosis is also suspected in the patient who has hypokalemia rather than normokalemia or hyperkalemia.

I. Blood	E. Toxic screen[b]
A. Electrolytes (Na, K, Cl, HCO_3)[a]	F. Lactate, pyruvate[c]
B. Arterial blood gases	II. Urine
C. Blood urea nitrogen, creatinine	A. Dipstick (pH, glucose, protein)
D. Glucose	

Na, sodium; *K*, potassium; *Cl*, chloride; HCO_3, bicarbonate.
[a]Calculate the anion gap.
[b]Or measurements of specific drugs if suspected of causing the acidosis.
[c]If available.

Table 86.22. Laboratory Evaluation of Metabolic Acidosis

Management

The choice of therapy is alkali, and the preferred agent is almost always $NaHCO_3$. Sodium lactate, given as lactated Ringer's solution, is an acceptable alternative, provided that liver function is normal and lactic acidosis is ruled out. Patients require treatment if the serum HCO_3^- is less than 15 mEq/L and/or the pH is less than 7.20, unless the underlying disorder is simple diarrheal dehydration. In that case, discontinuing oral intake and administering IV fluids are usually the only therapies required. The diarrhea usually stops, and the kidney corrects the acidosis.

Of equal importance to the choice of alkali therapy is the amount of bicarbonate to use and the rate of repair. The bicarbonate or buffer deficit requires some estimate of the “bicarbonate space,” which in health equals the ECF space of 20% of body weight in liters. However, recent experimental studies in dogs have suggested that the bicarbonate space is increased in severe metabolic acidosis to as much as 50% and, in lactic acidosis, even to 100%. The proposed reason for this is the movement of excess H^+ ions out of the ECF into other body compartments. Calculations of the HCO_3^- deficit therefore may be as follows:

Mild/moderate acidosis (pH 7.20 to 7.37):

$$HCO_3^- \text{ deficit in mEq} = (\text{“Normal” serum } [HCO_3^-] - \text{“Observed” serum } [HCO_3^-]) \times 20\% \text{ of total body weight in liters}$$

Severe acidosis (pH less than 7.20):

$$HCO_3^- \text{ deficit in mEq} = (\text{“Normal” serum } [HCO_3^-] - \text{“Observed” serum } [HCO_3^-]) \times 50\% \text{ of total body weight in liters}$$

If the volume of infused solution must be limited, 7.5% $NaHCO_3$ (1 mEq/mL) is used; otherwise, lesser concentrations should be used. Full correction of serum HCO_3^- should never be attempted; a reasonable goal is to increase serum HCO_3^- in increments of 5 to 10 mEq/L until a level of 15 to 18 mEq/L is achieved or a pH of 7.25 or greater. At this point, maintenance HCO_3^- therapy can be continued at roughly 2 mEq/kg per day unless the underlying cause of the acidosis has been successfully treated.

Requirements for alkali can vary because acid production may continue and/or the distribution space for bicarbonate theoretically could change. Frequent checks of serum HCO_3^- must accompany therapy. Overzealous alkali therapy is risky and can lead to a variety of complications, as outlined in Table 86.23. In some patients, such as those with uremic acidosis, chronic bicarbonate therapy may not be indicated because stabilization occurs with only mildly positive H^+ ion balance (HCO_3^- greater than 15 mEq/L), and the Na load occasioned by additional alkali therapy may aggravate preexisting hypertension or congestive heart failure. Similar reasoning would apply to patients who are hypernatremic or hyperosmolar. Any child who requires IV alkali therapy should be admitted to the hospital.

I. Hypokalemia	C. Endogenous manufacture of HCO_3^-
A. K^+ losses as part of the disease process (e.g., renal tubular acidosis, diabetic ketoacidosis)	III. Cerebrospinal Fluid Acidosis
	A. Delay in equilibrium of HCO_3^- across the blood-brain barrier
B. K^+ shifts into cells	IV. Sodium Overload
II. Alkalosis	V. Hypocalcemic Tetany
A. Overcorrection	A. Ca^{2+} binding to protein
B. Persistent hyperventilation	B. Ca^{2+} incorporation into bone

K^+, potassium; HCO_3^-, bicarbonate; Ca^{2+}, calcium.

Table 86.23. Complications of Alkali Therapy in Metabolic Acidosis

SPECIFIC RENAL SYNDROMES

Nephrotic Syndrome

Background

Definition

Nephrotic syndrome is the clinical expression for a variety of primary and secondary glomerular disorders, the hallmarks of which are 1) hypoproteinemia (serum albumin less than 3.0 g/dL); 2) heavy proteinuria, initially or at some point in the illness (more than 40 mg/m^2 per hour in a 24-hour urine); 3) edema; and less consistently 4) hyperlipidemia (predominantly triglycerides and cholesterol). *Primary nephrotic syndrome* is the term applied to diseases limited to the kidney. They are further classified according to the response to corticosteroid therapy and histology on renal biopsy. *Secondary nephrotic syndrome* is the term applied to multisystem disease in which the kidney is involved. Occasionally, nephrotic syndrome develops as a consequence of exposure to environmental agents, including heavy metals and bee venom. Table 86.24 lists the most important disorders in each category.

Syndrome and Histologic Pattern	Usual Response to Corticosteroids
I.Primary	
A. Minimal change (also lipoid nephrosis, "nil" disease)	S
B. Focal segmental sclerosis	R
C. Membranoproliferative nephritis	R
D. Membranous nephropathy	rS
E. Proliferative nephritis	S
1. Mesangial	R(?)
2. Focal	R(?)
3. Diffuse	
II. Secondary	S
A. Lupus nephritis	R
B. Sickle cell anemia	R
C. Henoch-Schönlein purpura	R
D. Hereditary nephritis	R
E. Drugs, toxins	R
F. Infections	R
G. Miscellaneous	

S, sensitive; R, resistant.

Table 86.24. Nephrotic Syndrome

Epidemiology

Nephrotic syndrome is worldwide in distribution. It tends to occur more commonly in boys than in girls. The mean age of onset tends to be earlier (younger than 6 years) in primary nephrotic syndrome than in secondary nephrotic syndrome (older than 6 years). Incidence and prevalence figures show some geographic variation: in the United States, the figures are 1.3 to 2.8 per 100,000 and 14 to 16 per 100,000 children, respectively. Although the disease is usually sporadic, a familial incidence is clearly established with a polygenic inheritance pattern. The infantile or congenital form of nephrotic syndrome is particularly common in children of Finnish extraction and is inherited as an autosomal-recessive trait.

Etiology

The causes of the primary and most of the secondary glomerular disorders associated with the nephrotic syndrome are largely unknown. By far, the most common form of nephrotic syndrome is that associated with “nil disease,” or minimal change on renal biopsy. It is also called idiopathic nephrosis of childhood and accounts for 80% of pediatric cases. There is no definite association with antecedent bacterial (e.g., streptococcal) or viral infections, although the presenting illness and episodes of clinical relapse are often associated with upper respiratory or GI infections. An immunologic basis for nephrotic syndrome has been suggested from a number of studies that have reported 1) persistently elevated serum IgM levels, 2) circulating immune complexes, 3) spontaneous remissions with natural measles infections (which are known to induce suppression of cell-mediated immunity), 4) suppression of lymphocyte proliferative responses in vitro by serum from patients with nephrotic syndrome, 5) hyperactivity of lymphocytes from patients when exposed to renal antigens in vitro, and 6) response of some patients to immunosuppressive agents. The strongest argument against an immunologic factor as a cause of nephrotic syndrome has been the failure to find immune reactants or inflammation in kidney biopsies, despite repeated studies of these patients.

The typical age of presentation of primary nephrotic syndrome is 18 months to 5 or 6 years. When nephrotic syndrome appears in the neonatal period, it likely is the congenital or Finnish type, which is steroid resistant and generally carries a fatal prognosis. Conversely, nephrotic syndrome that presents in a teenager is more likely to be associated with a primary or secondary form of underlying nephritis, and renal biopsy is generally indicated.

Pathophysiology

The hallmark of nephrotic syndrome is edema, signaling salt and water retention. Although the mechanisms of edema formation are incompletely understood, altered Na handling by the kidney always occurs. The site within the kidney may depend on the state of ECF volume. It is generally believed that the initiating factor is a large glomerular leak of proteins, predominantly albumin, leading to hypoalbuminemia. The leak is probably related to some noninflammatory immunologic or metabolic process that reduces the negative charges in the glomerular basement membrane. These charges are primarily represented by sialic acid residues that ordinarily repel albumin and other negatively charged plasma proteins. With loss of serum albumin and other plasma proteins, the intravascular oncotic pressure falls, and fluid moves out of the vascular and into the interstitial spaces, in accordance with Starling's principles. Because the liver ordinarily has a large

synthetic capacity for albumin, the persistent hypoalbuminemia noted in most nephrotics is probably not simply the result of urinary losses. Other contributing factors suggested in the literature, but not proved, are 1) decreased protein intake, 2) decreased synthesis, and 3) increased catabolism. Once plasma oncotic pressure falls, extracellular volume is reduced, which the kidney “reads” as a decreased effective circulating arterial volume. Proximal tubular sodium reabsorption is increased in response to this stimulus. The renin–angiotensin system is also stimulated, aldosterone secretion rises, and distal tubular sodium reabsorption intensifies. This secondary hyperaldosteronism perpetuates the edema-forming state. However, some patients may have normal or increased extracellular volume in the face of edema, further emphasizing the complexities of renal Na handling in this syndrome. The increased lipid turnover seen in nephrotic syndrome is characterized by elevations is serum triglycerides and cholesterol. The stimulus to this increased synthesis is unknown but is related to the degree of hypoproteinemia. Limited studies have pointed to a reduction in lipoprotein lipase and/or other circulating lipolytic factors and a selective retention of large-molecular-weight lipoproteins.

Clinical Manifestations

Presentations

The major presenting complaint is edema, which may be localized or diffuse. A typical story is that of a 2- or 3-year-old child with puffy eyes who was treated for allergies but did not improve. A rapidly changing belt, trouser, or shoe size may be indicative of rapid weight gain before edema is detectable. The rate and degree of edema formation vary from child to child and appear to be directly related to the degree of hypoalbuminemia. This, in turn, reflects the degree of albuminuria and the dietary protein intake. Pleural effusions and ascites, for example, are typically seen when serum albumin is below 1.5 g/dL. The degree of edema also depends on and varies inversely with the urine output, which is typically reduced in the full-blown case. In fact, in some patients true oliguria (less than 300 mL/m $_2$ per day) may be seen, although it almost never signifies ARF. Rarely, salt and water retention is abrupt and massive, leading to respiratory distress because of a combination of hydrothorax and ascites with elevation of the diaphragm. Ascites may also be associated with various abdominal complaints such as anorexia, nausea, and vomiting, which are thought to result from edema of the intestinal wall because they disappear with successful treatment of the edema.

Complications

The acute complications of nephrotic syndrome occur in two groups of patients: 1) those who present de novo or in relapse but not taking steroids, and 2) those who present in relapse or remission while still receiving pharmacologic doses of steroids (Table 86.25). Bacterial infections are noted with increased frequency in both groups, although they are more common in the steroid-treated children. The types of infections include cellulitis, peritonitis, sepsis, pneumonia, meningitis, and arthritis. More recently, Gram-negative organisms have been reported as often as Gram-positive organisms, the most common of which is *Streptococcus pneumoniae.* The typical signs and symptoms of infection may be masked in the steroid-treated nephrotic child, especially when the dose of steroid is high (e.g., 2 mg/kg per day prednisone). Even peritonitis may occur without local abdominal signs in the child with ascites because the accumulated fluid prevents painful contact between the inflamed visceral and parietal layers of the peritoneum.

I. Without Steroid Therapy	C. Hypercoagulability[a]
A. Bacterial infection	D. Respiratory embarrassment
B. Hypovolemia	E. Hypertension
C. Hypercoagulability (thrombo-embolic phenomena)	F. Altered behavior
D. Respiratory embarrassment	G. Steroid withdrawal (benign intracranial hypertension)
II. With Steroid Therapy	
A. Bacterial infection[a]	
B. Hypovolemia[a]	

[a]These complications occur more often after steroid therapy.

Table 86.25. Acute Complications of Nephrotic Syndrome

Symptomatic hypovolemia, which can progress to shock despite the presence of edema, results from injudicious fluid restriction, excess diuretic administration, or a combination of both. This complication should rarely happen once the patient is under medical management. The problem is not total body water or salt depletion but intravascular depletion that results from the abnormal distribution of what amounts to excess total body salt and water in the interstitial spaces. The signs and symptoms are those common to any child with hypovolemic shock.

Hypercoagulability, the tendency to form venous thromboses and thromboemboli, stems from many factors in nephrotic syndrome, including hyperlipidemia that leads to hyperviscosity, thrombocytosis, and increased levels of circulating fibrinolytic inhibitors. Renal vein, pulmonary artery, and peripheral pulmonary emboli are particularly devastating manifestations of hypercoagulability in nephrotic syndrome. The addition of steroid therapy enhances this risk by mechanisms that are unclear, although it is believed that prednisone exerts some antiheparin effect in humans. For these reasons, nephrotic children should never have femoral or other deep venipunctures unless no alternative vascular access exists.

As mentioned already, massive ascites may rarely lead to acute respiratory embarrassment, the treatment for which includes emergency paracentesis.

In steroid-treated children, acute rises in BP with symptoms of headache, blurred vision, or frank encephalopathy may

occur at any point in the clinical course. The diagnosis of hypertensive encephalopathy does not require a specific level of systolic and/or diastolic BP. Rather, it is the degree of BP change and rate of rise that cause symptoms. Acute mood changes, ranging from euphoria to depression, are associated with the introduction, sudden increase, or decrease of steroid therapy. Symptomatic complaints include irritability with a low frustration level, hyperwakefulness at night, and emotional lability with crying and withdrawal. Abrupt reductions in steroids may lead to benign intracranial hypertension characterized by headaches, vomiting, and occasional papilledema, which are not associated with arterial hypertension.

Although hyponatremia and hypocalcemia are often noted in the laboratory (see Laboratory and Radiologic Studies), they are rarely associated with acute symptoms.

Laboratory and Radiologic Studies

Laboratory studies can be grouped into three general categories: 1) those required to confirm nephrotic syndrome, 2) those designed to categorize nephrotic syndrome as primary or secondary, and 3) those designed as aids to medical management. These studies are outlined in Table 86.26.

I. Diagnostic Tests to Confirm Nephrotic Syndrome
  A. Serum proteins (albumin, globulin)
  B. Serum cholesterol
  C. Urine protein
    1. Qualitative (dipstick: albumin)
    2. Quantitative (24-hr collection)
II. Diagnostic Tests to Distinguish Primary from Secondary Nephrotic Syndrome
  A. Urinalysis (evidence of nephritis)*
  B. Screening test for sickle cell anemia
  C. Serum immunoglobulins
  D. Serum C3 complement
  E. Serum antinuclear antibody, DNA binding
  F. Hepatitis B surface antigen
III. Management Tests
  A. Complete blood count, especially hematocrit
  B. Serum Na, K, CO_2, Cl, Ca, uric acid
  C. Serum creatinine, blood urea nitrogen

Na, sodium; K, potassium; CO_2, carbon dioxide; Cl, chloride; Ca, calcium.
*Hematuria plus proteinuria generally indicates nephritis, especially if there are cellular (e.g., red blood cell) casts in the sediment.

Table 86.26. Laboratory Tests in Nephrotic Syndrome

Hypoalbuminemia is defined as a serum albumin less than 3.0 g/dL and occurs in virtually every child with nephrotic syndrome. The measurement of the urinary protein concentration varies somewhat with urinary volume but is usually 3 to 4+ (300 to 1000 mg/dL) in the untreated patient. Heavy proteinuria is the most reliable indicator of nephrotic syndrome and is defined as a 24-hour urine protein excretion of more than 40 mg/m^2 per hour, or approximately 1 g in a 30-kg child. The urinalysis occasionally shows RBCs and casts, suggesting an underlying nephritis, although this will not distinguish between causes of primary and secondary nephrotic syndrome. In most nephrotics, the urine-specific gravity is high, generally greater than 1.020.

The child with nephrotic syndrome often has an elevated hematocrit that results from intravascular dehydration. It is typical for nephrotic children to have depressed serum Na levels, usually in the range of 120 to 135 mEq/L. This rarely causes symptoms and does not require specific treatment. Hypocalcemia is also common but usually asymptomatic; the fall in Ca usually parallels that of albumin. Although the baseline uric acid is normal, many of these children later receive diuretic agents that can cause hyperuricemia. An initial elevation of the BUN in the range of 20 to 40 mg/dL is not at all uncommon and may reflect a reduction in GFR because of low plasma volume. Persistent azotemia can result from persistent reduction in GFR (an ominous prognostic sign) or, more commonly, from any combination of injudicious fluid restriction, increased catabolism caused by poor dietary intake and/or infection, and/or steroid therapy. Serum creatinine should serve to differentiate these two categories and either confirm or deny a true impairment in renal function caused by intrinsic renal damage.

Management

Acute management of nephrotic syndrome can be divided into two categories: specific and supportive. In the ED, the primary goal is usually to restore and preserve intravascular volume or to treat symptomatic edema.

Despite the presence of peripheral edema, shock is treated in the usual way, with 20 mL/kg per hour of normal saline until circulation is restored (see Chapter 3). If the child is clinically dehydrated and hemoconcentrated (hematocrit more than 50%) but not in shock, a trial of Na-deficient fluids orally at twice maintenance is preferable to an immediate start of hypotonic IV solutions (i.e., 5% dextrose in 0.25 N salt solution). Fluids should be given in small amounts (1 to 4 oz) at frequent intervals (1 to 4 hours) to avoid vomiting caused by an edematous gut. Although Na restriction is indicated for an edematous nephrotic child, water restriction is rarely indicated and only further decreases a usually low urine output. The ongoing state of intravascular hydration can be assessed by serial hematocrit tests.

If the patient is well hydrated but symptomatic from massive edema, a trial of diuretics is warranted. Symptoms include difficulty in ambulating, abdominal discomfort, skin breakdown, and respiratory distress. Furosemide, 1 to 2 mg/kg per day in two divided oral doses, can be used. If there is no response, additional diuretics that act at other sites in the tubule (thus enhancing the diuretic effect) may be added. Commonly used agents are spironolactone and hydrochlorothiazide, both starting at 1 mg/kg per day in two doses. Diuretics do not usually work, however, if the serum albumin concentration is less than 1.5 g/dL. When it appears urgent to remove some edema fluid, a combination of albumin infusions followed 30 minutes later by IV furosemide is often effective. The dose of albumin is 0.5 to 1.0 g/kg given as 25% salt-deficient albumin followed by 0.5 to 1.0 mg/kg of furosemide. Paracentesis is rarely indicated but may bring prompt relief of severe respiratory distress from massive ascites.

Prednisone is generally begun at a dosage of 2 mg/kg per day in two or three divided doses after the workup is initiated and a tuberculin test is placed. If the patient has previously been responsive to prednisone and is on either no drug or a maintenance program, a return to full therapy is indicated, provided frank relapse is obvious. If not, a quantitative 24-hour urine test should be ordered. Concurrent administration of a low-sodium antacid may reduce the risk of gastric irritation.

Antibiotics are not administered prophylactically but are used when a bacterial infection is suspected and the physician is awaiting results of appropriate cultures. This is particularly true if the patient is receiving Cytoxan or chlorambucil, as well as prednisone. Any child with active nephrotic syndrome and an unexplained fever must be considered to have a bacterial infection until proved otherwise. A blood culture is indicated, and a diagnostic paracentesis for Gram stain and culture is appropriate in the presence of obvious ascites. Penicillin has been the appropriate first choice in the past to treat *S. pneumoniae.* However, in view of reports of increased Gram-negative bacterial infections in nephrotic children, it may be necessary to broaden this initial coverage with ampicillin, one of the newer cephalosporins, and/or an aminoglycoside. In the presence of documented infection, high-dose steroid treatment should be reduced but not discontinued. Reducing the daily or alternate-day dose (if 2 mg/kg or more) by half is appropriate—but no lower than 10 to 15 mg/day. Finally, good hygiene and a balanced high-protein diet are important adjuncts to therapy.

Indications for emergency admission of a child with active nephrotic syndrome are listed in Table 86.27. As in diabetes mellitus, the admission of a newly diagnosed child is as much for patient and parental education as it is for further workup and treatment. If a patient is more than 10% dehydrated, has orthostatic hypotension, and/or has a hemoglobin greater than 16 g/dL or a hematocrit more than 50%, admission is advised for IV rehydration therapy with close observation of vital signs and urine output.

Newly diagnosed patient	Refractory edema (e.g., respiratory distress)
Severe dehydration (e.g., poor intake, persistent vomiting)	Peritonitis
Unexplained fever (e.g., suspected bacterial infection)	Renal insufficiency (e.g., elevated serum creatinine)

Table 86.27. Indications for Admission for Nephrotic Syndrome

Hypertension

Background

Definition

For the purposes of this chapter, *hypertension* (see Chapter 35) is defined as a systolic and/or diastolic BP higher than two standard deviations above the mean for age and sex. This definition implies that BP measurements have been taken carefully several times during the course of evaluation and that the child is not diagnosed as being hypertensive until the mean values from two or three such evaluations have been established over several weeks. The emergency physician is not afforded this opportunity for continued surveillance and is often dealing with symptomatic children who are severely hypertensive. Nonetheless, as with all patients who are diagnosed as being hypertensive, strict attention must be paid to the technical details of the measurement. The apparatus must calibrate to “zero,” the cuff should be one-half to two-thirds the width of the upper arm, the inflatable bladder should encircle the arm, and note should be taken of undue patient anxiety, pain, or muscle contraction, all of which can elevate the systolic and diastolic readings.

It can be useful to group children into four major age categories when defining upper limits of systolic and diastolic BPs (Table 86.28). In addition, it is useful to think of hypertension in terms of its four different presentations, each of which is discussed under Clinical Manifestations: 1) asymptomatic or minimally symptomatic hypertension, 2) malignant hypertension, 3) accelerated hypertension, and 4) hypertensive encephalopathy.

	Upper Limit (mm Hg)	
Age (yr)	Systolic	Diastolic
0–2	110	65
3–6	120	70
7–10	130	75
11–15	140	80

Table 86.28. Hypertension

Epidemiology

Hypertension is a major worldwide public health concern; in the United States alone, it is estimated that 40 million adults are hypertensive, or roughly twice the number reported in yearly surveys. It has only been in recent years that pediatricians have recognized hypertension as a widespread and common health problem among children. Hypertension occurs throughout childhood and shows no sexual preference. As with adults, it probably occurs more often in African-Americans than in Caucasians. There appear to be certain predisposing factors in genetically susceptible children, including dietary sodium intake, physical inactivity, and obesity. In addition, genetic studies have also shown “clustering” of hypertension in certain families with strongly positive histories. Finally, infants who have high-normal or frankly elevated BP readings tend to “track” along the same BP percentiles as they progress through childhood.

Etiology

Now that essential hypertension is acknowledged as a cause of hypertension in childhood, pediatric hypertension should be divided into primary and secondary causes (Table 86.29). Generally, the endocrinologic, cardiac, neurologic, and miscellaneous causes of hypertension produce relatively mild and asymptomatic increases in BP and affect systolic more than diastolic readings. The one exception is pheochromocytoma, a rare disease in children. Steroid-induced hypertension usually requires several weeks of pharmacologic doses to develop (e.g., 2 mg/kg per day prednisone). Occasionally, primary (essential) hypertension produces symptoms such as headaches, abdominal pain, and/or visual disturbances but rarely leads to malignant hypertension with or without encephalopathy. Most children with severe elevations of diastolic BP (e.g., higher than 95 mm Hg for those less than 10 years old; higher than 100 mm Hg for those more than 10 years old) and hypertensive crises have a secondary renal or renovascular cause for their hypertension. In several large surveys of severe diastolic hypertension reported from tertiary referral centers, the four most common causes in order of decreasing frequency were 1) chronic glomerulonephritis, 2) chronic (primary or secondary) pyelonephritis, 3) coarctation of the aorta, and 4) renal artery stenosis. The younger the child and the higher the BP, the more likely it is that an underlying cause for hypertension will be found.

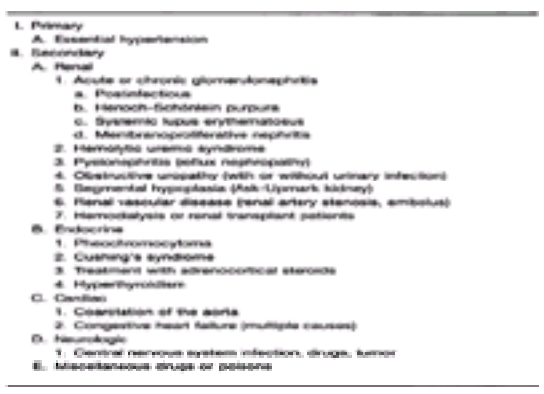
I. Primary
A. Essential hypertension
II. Secondary
A. Renal
1. Acute or chronic glomerulonephritis
a. Postinfectious
b. Henoch-Schönlein purpura
c. Systemic lupus erythematosus
d. Membranoproliferative nephritis
2. Hemolytic uremic syndrome
3. Pyelonephritis (reflux nephropathy)
4. Obstructive uropathy (with or without urinary infection)
5. Segmental hypoplasia (Ask-Upmark kidney)
6. Renal vascular disease (renal artery stenosis, embolus)
7. Hemodialysis or renal transplant patients
B. Endocrine
1. Pheochromocytoma
2. Cushing's syndrome
3. Treatment with adrenocortical steroids
4. Hyperthyroidism
C. Cardiac
1. Coarctation of the aorta
2. Congestive heart failure (multiple causes)
D. Neurologic
1. Central nervous system infection, drugs, tumor
E. Miscellaneous drugs or poisons

Table 86.29. Causes of Hypertension

Pathophysiology

Measured BP results from the interaction of physiologic mechanisms that regulate vascular volume and cardiac output on the one hand and peripheral vascular tone on the other hand. Because pressure equals flow times the resistance to flow, BP equals cardiac output (CO) times total peripheral resistance (TPR), or:

$$BP = CO \times TPR$$

Factors that influence CO, either directly or through changes in vascular volume, include circulating aldosterone levels, the autonomic nervous system, renal regulation of sodium balance, and small vessel compliance (i.e., capacitance and resistance).

The distribution of the blood volume is as important as the total blood volume itself in affecting BP. Normally, volume is distributed partly in “resistance” vessels on the arterial side of the circulation. When blood volume is increased primarily in the resistance vessels, CO and BP are much more likely to rise than when the increase is primarily in the capacitance vessels. Factors that influence TPR by inducing vasoconstriction or vasodilation include activity of the autonomic nervous system, levels of circulating catecholamines, and levels of circulating angiotensin II (Fig. 86.9).

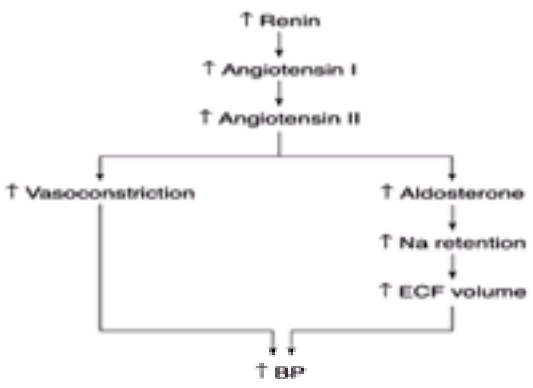

FIGURE 86.9. Mechanisms generating hypertension. *Na,* sodium; *ECF,* extracellular fluid; *BP,* blood pressure.

Obviously, hypertension occurs when an increase in CO is not matched by a physiologic and reciprocal fall in TPR, and vice versa. In clinical practice, however, hypertension is only rarely the result of a single pathophysiologic abnormality, such as activation of the renin–angiotensin system, leading to vasoconstriction and thereby to increased TPR, or excessive salt and water retention that leads to increased blood volume and CO. Rather, hypertension can be thought of as developing in two phases: 1) an initial or generating phase, which may be caused primarily by increased CO or vasoconstriction; and 2) a maintenance phase that is almost always the result of increased CO from any number of causes. Generally, the history and physical examination provide reliable guides in assessing the relative roles of CO and TPR in the acutely hypertensive child. Using this information, the emergency physician can make a rational selection of initial antihypertensive drugs.

The pathophysiology of hypertensive encephalopathy is controversial. What is generally agreed on is that autoregulation of cerebral blood flow is disrupted, leading either to overregulation with exaggerated vasospasm and ischemic injury or to underregulation with breakthrough of the circulation, increased cerebral flow, and cerebral edema.

Clinical Manifestations

As already outlined, hypertension usually presents in one of four ways (Table 86.30). First, there is the asymptomatic or mildly symptomatic child with mild to moderate elevations in BP (130 to 150 mm Hg systolic; 80 to 94 diastolic). Acute symptoms are usually nonspecific and may include headaches, abdominal pain, epistaxis, and irritability. If the hypertension is chronic, failure to thrive, irritability, personality changes, and deteriorating school performance may be prominent complaints.

Asymptomatic or mildly symptomatic	Accelerated hypertension
Malignant hypertension	Malignant hypertensive encephalopathy

Table 86.30. Clinical Presentations of Hypertension

Malignant hypertension is characterized by marked elevations in systolic and/or diastolic BP (e.g., 160 mm Hg or higher systolic for those less than 10 years of age; 170 mm Hg or higher systolic for those more than 10 years; 105 mm Hg or higher diastolic for those less than 10 years; 110 mm Hg or higher diastolic for those more than 10 years) and is often associated with spasm and tortuousity of the retinal arteries, papilledema, and hemorrhages and exudates on funduscopic examination. This condition is much more commonly seen in adults than in children. *Accelerated hypertension* is defined as an acute rise in systolic and/or diastolic BP superimposed on previously existing hypertension. In both malignant hypertension and accelerated hypertension, the patient may present with dramatic symptoms and signs such as heart murmur, congestive heart failure (CHF), lower motor neuron facial palsy, and hematuria.

Hypertensive encephalopathy is often seen in malignant hypertension and consists of a combination of symptoms and signs that often vary from patient to patient (Table 86.31). No single symptom or sign is diagnostic of this syndrome; the diagnosis is confirmed by demonstrating a rapid improvement in the symptoms and signs after the BP is lowered. Although there is no generally agreed-on level of systolic and/or diastolic BP at which encephalopathy occurs, most investigators believe that the rate of rise in the BP is as important as the actual level itself. In this, as in all forms of severe hypertension (which are almost always secondary), the presenting complaints are usually attributable to the hypertension itself and not to the underlying disease.

Nausea, vomiting
Headaches
Altered mental status (neuropsychiatric symptoms, confusion, stupor, coma)
Visual disturbances (blurry vision, decreased visual acuity, diplopia)
Seizures, stroke

Table 86.31. Malignant Hypertensive Encephalopathy

Ascertaining the cause for increased BP in the acutely hypertensive child with an abnormal neurologic examination presents a difficult challenge (Table 86.32). In general, when there is primary neurologic disease with secondary hypertension, the hypertension is usually mild and predominantly systolic. To determine whether the hypertension caused the neurologic abnormalities or vice versa, the physician must first observe the neurologic response to lowering the BP. If signs and symptoms clear rapidly, he or she is probably dealing with true hypertensive encephalopathy. In addition, primary neurologic disease can be screened for by a spinal tap (if a mass lesion is not suspected) or computed tomography (CT) scan (if a mass lesion or intracranial bleeding is suspected).

Head trauma	Brain tumor
Cerebral hemorrhage or infarction	Uremic encephalopathy
Meningitis, encephalitis	

Table 86.32. Differential Diagnosis of Hypertensive Encephalopathy

The urgency of prompt treatment of malignant hypertensive encephalopathy is attested to by the fact that fully one-third of severely hypertensive children develop neurologic abnormalities that may be sudden in onset yet may leave permanent deficits. These include cortical blindness, infarction of the optic nerve, and hemiplegia.

Finally, in patients with an acute exacerbation of long-standing hypertension, overzealous antihypertensive therapy can produce relative hypotension and, paradoxically, can lead to some of the neurologic abnormalities previously cited.

When confronted with newly diagnosed hypertension in the child, the physician should ask three important questions: 1) Is the hypertension primary or secondary? 2) Is there evidence of target organ injury? and 3) Are there associated risk factors that would worsen the prognosis if the hypertension were not treated or were treated unsuccessfully? The laboratory and radiologic workup can then be divided conveniently into three categories, as outlined in Table 86.33. The laboratory and radiology studies listed in the table are intended to be comprehensive, covering children with the full range of secondary causes of hypertension. Obviously, the emergency physician can usually establish a strong working diagnosis from the history and physical examination and selected laboratory tests.

I. Diagnosis: Primary or Secondary
A. Laboratory
1. UrinalysIs
2. Urine culture
3. Urinary catecholamines
4. Complete blood count with platelet count and blood smear
5. Serum Na, K, Cl, CO_2, Ca, P,
6. Serum blood urea nitrogen, creatinine
7. Serum C3 complement, antistreptolysin O titer, antinuclear antibody
8. Plasma renin
B. Radiology
1. Chest radiograph
2. Intravenous pyelogram
3. Voiding cystourethrogram
4. Cardiac catheterization
5. Renal ultrasound
6. Renal scan
7. Renal arteriogram
C. Other
1. Electrocardiogram
II. Tests for Target Organ Injury
A. Urinalysis
B. Chest radiograph
C. Electrocardiogram
III. Tests for Associated Risk Factors
A. Serum lipid profile (e.g., lipoprotein electrophoresis)
B. Serum uric acid

Na, sodium; K, potassium; Cl, chloride; CO_2, carbon dioxide; Ca, calcium; P, phosphorus.

Table 86.33. Laboratory Tests in Hypertension

Management

Acute Management of Hypertension

The spectrum of hypertension that presents to the pediatric ED ranges from mild and asymptomatic to a true hypertensive emergency (see Chapter 35). A brief but careful history and physical examination aim to classify the severity of the hypertension encountered. When the hypertension is severe, this evaluation should progress only after the ABCs (airway, breathing, and circulation) of resuscitation have been accomplished. Features of the history to be ascertained include duration of hypertension, details of its onset, and degree of compliance with any current drug therapy. The possibility of renal disease should be explored: has there been any history of urinary tract infections, failure to thrive, hematuria, edema, or umbilical artery catheterization? In addition, does the patient have a history of joint pain, swelling, palpitations, weight loss, flushing, weakness, drug ingestion, or a family history of renal disease or hypertension?

After several determinations of BP, a focused physical examination should be performed immediately. Emphasis should be placed on the neurologic examination, searching for any evidence of dysfunction. Funduscopic examination may reveal hemorrhage, papilledema, or infarcts. Any signs of CHF and discrepant upper and lower extremity BP measurements should be noted. Palpable kidneys or peripheral edema may suggest a renal origin, and an abdominal bruit, if present, suggests the possibility of renovascular hypertension. Initial laboratory studies should include a complete blood count (CBC), electrolytes, BUN, creatinine, urinalysis, chest radiograph, and ECG.

It is the presence or absence of acute end-organ dysfunction discovered in the history, physical examination, or laboratory studies and not the height of the BP that distinguishes a hypertensive urgency from a hypertensive emergency (Fig. 86.10). The division into these categories determines the site of treatment (i.e., ED, intensive care unit, routine hospital unit). The patient's clinical profile may provide clues regarding derangements in CO and/or TPR. These, along with a suspected origin, will guide the mode of therapy. Classifying the hypertensive episode as urgent or emergent governs the approach to treatment.

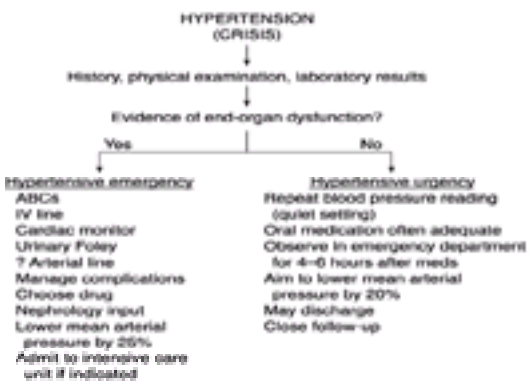

FIGURE 86.10. Diagnostic approach for management of acute hypertension. *ABCs,* airway, breathing, circulation; *IV,* intravenous.

Hypertensive Emergency

In a hypertensive emergency, an IV line should be placed immediately to allow administration of medications and fluid resuscitation if indicated. It is important to note that many patients in hypertensive crisis have volume depletion most likely caused by vomiting, diarrhea, or a diuresis of unclear origin. A cardiac monitor and urinary catheter should be used from the outset. Continuous BP monitoring must be provided, preferably by arterial catheter. Any serious complications must be managed before or as the hypertension is treated (e.g., anticonvulsants should be administered to a seizing patient along with hypertensive medications). A number of medications are available for hypertensive emergencies, and the drug(s) chosen will depend on several factors. To be considered are the patient's clinical condition, the presumed cause, whether there is a change in CO (propanolol would be contraindicated in the presence of CHF) or TPR, and whether there is end-organ involvement (nifedipine would be contraindicated in the presence of intracerebral bleeding).

In a hypertensive emergency, the goal is to lower the BP promptly but gradually. The mean arterial pressure should be lowered by 25% over several minutes to several hours, depending on the nature of the emergency. For example, a patient who is seizing or herniating must have the BP reduced immediately. This is not so for a patient who presents with headache or vomiting. Once antihypertensive therapy is begun, the patient's condition must be assessed frequently, giving special attention to the BP and neurologic status. Precipitous decreases in BP can lead to avoidable neurologic deficits. Therefore, the preferred drugs are those that allow for close monitoring of BP reduction (e.g., those given in incremental infusions). Most hypertensive emergencies can be controlled given the availability of new classes of potent antihypertensive agents and the expertise to use them. In general, physicians treat hypertensive emergencies most effectively by becoming expert in the use of a few agents. Each of the most commonly used medications offers distinct advantages and disadvantages. Each clinical situation dictates the precise mode of therapy, but some general guidelines are usually applicable.

The medications described next are recommended for use in the emergency setting (Table 86.34). Sodium nitroprusside is an arteriolar and venous vasodilator that is invariably effective. It reduces peripheral resistance and cardiac filling pressure. BP therefore decreases with little change in CO, and reflex tachycardia does not occur. It is largely metabolized to cyanide in the RBCs and cyanide reacts with thiosulfate in the liver to form thiocyanate. Sodium

nitroprusside is administered by constant infusion. Its effect is immediate and lasts only as long as the infusion is continued. Its use requires intensive observation and therefore may not be indicated in the ED. The drug requires 10 minutes to prepare and is sensitive to light. The other disadvantages are its extreme potency and the risk of thiocyanate and cyanide toxicity, which are time-dependent phenomena. Thiocyanate is excreted by the kidneys; therefore, the risk of early toxicity is increased in patients who present with renal insufficiency.

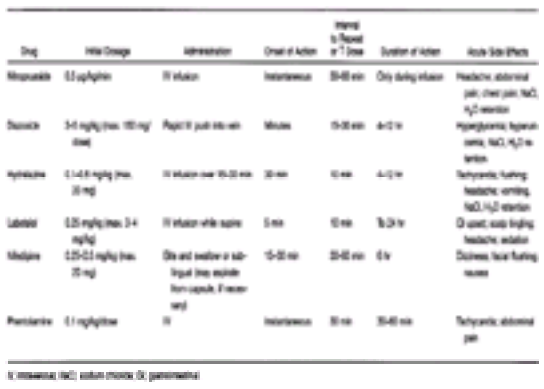

Table 86.34. Drugs Used in Hypertensive Emergencies

Diazoxide is an arteriolar vasodilator that is a potent hypotensive agent. It has little effect on capacitance vessels and no direct cardiac effect. Its onset of action is 1 to 5 minutes, and it can be administered in frequent small boluses to avoid hypotension. It may provide a long duration of BP control (8 to 12 hours). It causes marked salt and water retention, and in patients with edema, it should be followed by a diuretic agent. It also causes a reflex tachycardia and hyperglycemia.

Hydralazine is an arteriolar vasodilator that is not as potent as diazoxide and sodium nitroprusside. It is administered by IV, and the onset of action is usually within 30 minutes. Reflex tachycardia often occurs and may require the introduction of a b blocker.

Labetalol is an a_1- and nonselective b-adrenergic blocker. Blockade of the a_1 receptor results in vasodilation and is thought to be responsible for the acute antihypertensive action. The b blockade prevents reflex tachycardia. It is available in both oral and IV forms. b Blockade may result in hyperglycemia and/or hyperkalemia. Dosing is independent of renal function. It has been reported to be effective in the management of severe hypertension that results from pheochromocytoma and coarctation of the aorta and is a reasonable alternative in the treatment of hypertensive crises in patients with end-stage renal disease.

Nifedipine, a calcium channel blocker, causes direct vasodilation of the arterioles, leading to a reduction in peripheral vascular resistance. It does not affect CO. It can be administered sublingually, but biting the capsule and swallowing its contents achieves measurable blood levels more rapidly than the sublingual route. Its action begins within 15 minutes, and the peak activity occurs at 30 to 60 minutes. A mild increase in heart rate may occur. The patient may complain of headache and flushing. Its use depends on the patient's state of consciousness.

Phentolamine is a pure a-adrenergic blocker used almost exclusively for the treatment of catecholamine crisis (as seen in patients with pheochromocytoma or ingestion of sympathomimetic agents such as cocaine). It is administered as an IV bolus, and the effect is immediate. The response lasts approximately 15 minutes. Side effects include tachyarrhythmia and angina.

After antihypertensive therapy has been instituted and the patient's end-organ disease has been stabilized, the patient must be admitted to the intensive care unit for close monitoring and further hypertensive management.

Hypertensive Urgency

A *hypertensive urgency* is defined as severe hypertension without evidence of end-organ involvement. Patients with known hypertension who present in an urgent hypertensive crisis may not require hospitalization if the therapy in the ED is successful and adequate follow-up can be ensured. Often, oral antihypertensive agents will be sufficient (Table 86.35), although there are occasions when parenteral therapy is indicated.

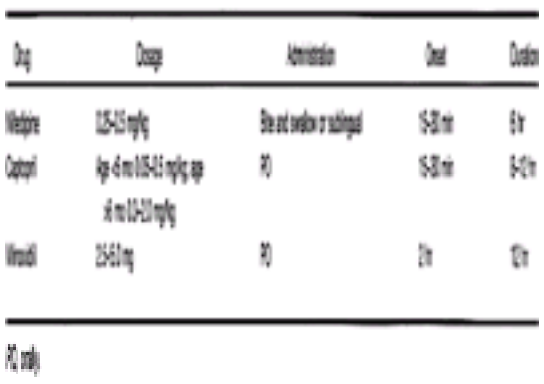

Table 86.35. Drugs Used in Hypertensive Urgencies

Nifedipine is an appropriate and effective agent for hypertensive urgencies. Its features have already been discussed. Captopril is a rapidly acting, powerful inhibitor of the ACE. It is absorbed rapidly after an oral dose and has an onset of action in 30 minutes. Its use does not result in a change in CO or heart rate. Minoxidil is a potent arteriolar dilator that can be given orally. It blocks calcium uptake through the cell membrane. Its onset of action is within 2 hours. Reflex tachycardia often occurs, along with fluid retention that requires a diuretic.

A 4- to 6-hour period of observation should follow the administration of the antihypertensive agent in the ED. This should be done to identify any untoward effects of the medication such as orthostasis. Patients should be discharged on the same medication used in the ED.

When hypertension is discovered by accident and is not the reason for the patient's visit, medical follow-up for repeated BP measurements is indicated before therapy is begun, especially if the elevation is mild (no more than 5 to 10 mm Hg above the upper limits of normal for systolic and diastolic pressures given in Table 86.28). If the BP is moderately elevated but the patient is asymptomatic, two options exist. Arrangements can be made for an outpatient workup in the future and a thiazide diuretic or a b blocker may be initiated at a low dosage. Alternatively, the patient may be admitted to begin an evaluation and therapy under hospital observation.

What follows are illustrative cases of hypertensive crises and their management.

Case 1. A 13-year-old obese African-American male has occasional bifrontal headaches and a BP of 155/95 mm Hg (mean of three readings). Funduscopic and cardiovascular examinations are normal. The family history is positive for hypertension in an obese mother and maternal grandmother, and the diet contains excessive fried food.

Comment. This story is classic for essential hypertension, and the only diagnostic studies that are required immediately are a urinalysis and urine culture, although many physicians would do a CBC and tests for creatinine and electrolytes. Studies for target organ damage and associated risk factors should be done at a later time.

Case 2. An 11-year-old girl presents with a gradual onset of headaches and weight loss and the acute onset of abdominal pain with vomiting and blurry vision. She has a history of three episodes of “cystitis” and cystoscopy with urethral dilation. Examination reveals a thin girl in obvious pain with a BP of 180/120 mm Hg (mean of three readings) and arteriolar spasm and papilledema on funduscopic examination. Urinalysis reveals clear urine with a specific gravity of 1.005, but otherwise, it is negative.

Comment. This scenario represents a hypertensive emergency: severe hypertension with target organ disease. The likely diagnosis is pyelonephritis-induced reflux nephropathy given the history of urinary tract infections, low urine specific gravity, and the level of the BP. This patient will require an IV line and the prompt reduction of her BP. The treatment choices are diazoxide, hydralazine, or nifedipine. Nitroprusside would be contraindicated if the patient was known to have severe renal insufficiency. BP and neurologic status should be monitored frequently. This patient will require admission, probably to the intensive care unit. Appropriate renal studies should be performed after stabilization and during hospitalization. A nephrology consult should be obtained.

Case 3. A 10-year-old, previously well boy has a gradual onset of aggressive, sullen behavior followed by a severe headache, decreased visual acuity, and a generalized seizure. Family and patient medical histories are negative. BP is 220/150 mm Hg (mean of three readings); there is florid papilledema with hemorrhages and exudates in the retina, cardiac enlargement, and a bruit over the left flank.

Comment. The presumptive diagnosis is hypertensive encephalopathy caused by renal artery stenosis. The patient has a hypertensive emergency. After the ABCs are monitored and stabilized, the patient will require parenteral antihypertensive therapy. Anticonvulsant drugs should be readily available in the event that the seizure activity recurs in the ED. Either diazoxide or sodium nitroprusside may be used. If the patient is known to have renal insufficiency and presents with signs or symptoms of hypertensive encephalopathy, nitroprusside should be used as a last resort and only for a brief period. The BP should be reduced over 2 to 3 hours by not more than 25% of the mean arterial pressure. This patient is a candidate for an intra-arterial catheter to measure the BP frequently and accurately. Intensive care unit admission is required. When the patient is stable, a renal scan and arteriogram should be performed.

Case 4. A 6-year-old boy presents with obvious symptoms and signs of CHF and red–brown urine. Urine output has been “low.” He had a sore throat 10 days ago. The BP is 160/110 mm Hg (mean of three readings), there is obvious CHF with peripheral edema, and the urine reveals 3+ protein, large blood, and 40 to 50 RBCs, 10 to 20 WBCs, 3 to 6 granular, and 0 to 2 RBC casts per high-power field.

Comment. This patient has acute glomerulonephritis (presumably poststreptococcal infection) with acute hypertension and oliguria leading to salt and water retention and CHF. His BP requires immediate therapy. An IV line, cardiac monitor, and urinary Foley must be placed as soon as possible. Electrolytes, BUN, and creatinine should be sent to the laboratory immediately. The preferred antihypertensive agent is hydralazine or diazoxide. He will also require diuretic therapy. After the BP has been stabilized, the patient will require admission. Appropriate serologic and urinary studies can be sent when time permits.

Acute Renal Failure

Background

Definition

ARF is an acute reduction in renal function, the hallmark of which is a decreased GFR. It may occur de novo or superimposed on preexisting renal disease. Cardinal features are solute retention, demonstrated by a rise in serum BUN and/or creatinine concentrations (i.e., azotemia) and oliguria or, more rarely, anuria. Oliguria has been variably defined in the pediatric patient as 1) urine output 300 mL/m^2 per day or less, 2) urine output 400 mL/day or less, 3) urine output 0.5 or 1 mL/kg per hour or less for an undefined period, and 4) urine output 0.5 or 1 mL/kg per hour or less for 12 to 24 hours. In the ED, precise measurements of previous urine output are rarely available, so it is convenient to use the figure 1 mL/kg per hour or less for an unspecified period. True oliguria can then be confirmed later after 24 hours of hospital observation.

Although less common in children than in adults, ARF can occur in the face of normal or near normal urine output, particularly after burns or exposure to nephrotoxins. Finally, ARF can and should also be defined anatomically in terms of the localization of the insult. Thus, azotemia and oliguria may be caused by prerenal, renal (i.e., parenchymal), or postrenal (i.e., obstructive) factors. These are important clinical distinctions to make because if recognized promptly, prerenal and postrenal causes of ARF can often be reversed with appropriate therapy.

Epidemiology

ARF is generally sporadic in nature. There are no clearly defined environmental factors that play a role in the common causes of pediatric ARF, other than various infectious agents that have been linked with hemolytic uremic syndrome (HUS). Rarely, myoglobinuric ARF is secondary to a genetic abnormality in muscle metabolism. The incidence of ARF in hospitalized children beyond the neonatal period is unknown. At The Children's Hospital of Philadelphia, approximately 5 to 10 children have ARF on admission, and 30 to 40 are seen in consultation in the hospital each year.

Etiology

As suggested in the definition of ARF, an anatomic approach to the various causes is useful (Table 86.36). Prerenal factors include 1) decreased CO that results from various hemodynamic abnormalities that cause left ventricular failure or cardiogenic shock; 2) true hypovolemia that results from excessive salt and water losses or from hemorrhage; and 3) “relative” hypovolemia that results from an altered distribution of salt and water out of the ECF space (i.e., “third-spacing”) as seen in nephrotic syndrome, cirrhosis, pancreatitis, and burns.

I. Prerenal
A. Decreased cardiac output (cardiogenic shock)
B. Decreased intravascular volume (hemorrhage, dehydration, “third-spacing”)
II. Renal
A. Primary renal parenchymal disease
1. Vascular (acute glomerulonephritis, HUS)
2. Interstitial (pyelonephritis, drug-induced)
B. Acute tubular necrosis
1. Ischemic injury (see I.B, above)
2. Nephrotoxic injury (antibiotics, uric acid)
3. Pigmenturia (myoglobinuria, hemoglobinuria)
III. Postrenal
A. Obstructive uropathy
1. Posterior urethral valves
2. Intraabdominal tumor
3. Nephrolithiasis (rare)
B. Renal vein thrombosis (rare outside of the neonatal period)

HUS, hemolytic uremic syndrome.
[a]Major pediatric causes of acute renal failure are listed in parentheses.

Table 86.36. Causes of Acute Renal Failure[a]

Primary renal (parenchymal) causes of ARF can be divided into two general categories: 1) inflammatory diseases that are usually immunologic or infectious in nature, such as acute poststreptococcal glomerulonephritis, methicillin-induced hypersensitivity interstitial nephritis, and pyelonephritis, and 2) ischemic or nephrotoxic injuries that have been grouped under the general heading of acute tubular necrosis. Of increasing pediatric interest and perhaps importance are nephrotoxins that have long been known to play an important role in adult ARF. Most prominent in this regard are drugs, including several commonly used antibiotics (aminoglycosides and cephalosporins), heavy metals (mercury, lead, cisplatinum), and diuretics (thiazides and furosemide). When administered together, the antibiotics and diuretics appear to act synergistically in their nephrotoxic effect. Uric acid may also have a direct nephrotoxic effect, and it also may cause renal tubular (luminal) obstruction.

Postrenal or obstructive factors include, most commonly, posterior urethral valves and abdominal lymphomas or rhabdomyosarcomas that cause extraureteral or extravesical compression. Rarely, a large kidney stone will obstruct the bladder outlet or the urethra.

The frequency of various causative factors in ARF differs significantly between adults and children, as seen in Table 86.37. Medical causes are much more common in children; postsurgical ARF is much more common in adults. Although the incidence of nephrotoxic ARF in children is low (Table 86.37), it reflects the data from several older published series and focuses more on the newborn and younger infant than on the older child.

Category	Approximate Incidence (%)[a]	
	Adults (N = 2476)	Children (N = 362)
Medical	24	85
Postsurgical	43	11
Toxin	10	<1
Trauma	14	?
Miscellaneous	9	4

[a]Adult data are based on two large series; pediatric data are based on six series, three of which focus on the newborn and infant less than 1 year old.

Table 86.37. Categories of Acute Renal Failure

Pathophysiology

Early concepts of the pathophysiology of parenchymal ARF in humans suggested that various predisposing factors led to a final common pathway of renal ischemia that, in turn, resulted in tubular injury. This theory gave way to the theory of "vasomotor nephropathy," which suggested that ARF could be divided into an initial phase, mediated either by ischemia or nephrotoxic injury; a maintenance phase, which resulted from increased intrarenal renin release and consequent intrarenal vasoconstriction; and a recovery phase. However, recent studies have thrown doubt on this largely hemodynamic model as satisfactory to explain oliguria and/or solute retention in all patients with ARF. In some animal models, vasoconstriction and renal ischemia may lead to a parallel decrease in renal blood flow and GFR in the initial phase of ARF, but renal blood flow spontaneously returns to normal (or is restored with vasodilators) despite a persistently low GFR with oliguria in the maintenance phase of ARF. It is more likely that parallel changes in renal blood flow and GFR explain the solute retention and oliguria of prerenal ARF rather than intrinsic ARF. What differentiates prerenal from intrinsic ARF is preservation of tubular function in the former, especially maximum urinary concentration and sodium reabsorption (see Clinical Findings), and response of azotemia and oliguria to volume replacement.

In addition to hemodynamic mediators of renal injury, nephronal and cellular mediators play important roles in the induction of ARF. Proximal tubular cell injury leads to epithelial cell death, loss of tubular integrity with fluid and solute "backleak," and the release of cellular debris that leads to tubular luminal obstruction. Cellular and metabolic alterations include production of oxygen free radicals when renal blood flow is restored, Ca influx into the cell that leads to changes in tubule cell volume, and an energy crisis within renal tubular cells caused by depletion of adenine nucleotides. Studies in several animal models have led to the conclusion that four major factors are probably operative alone or in combination in all forms of human intrinsic ARF. These are decreased renal blood flow, decreased glomerular filtration (i.e., decreased glomerular permeability, K_f, and/or glomerular surface area), tubular luminal obstruction, and tubular backleak. A summary of one proposed schema for the pathogenesis of ARF in humans is outlined in Figure 86.11. What is important to emphasize is that in most patients with ARF, more than one pathophysiologic abnormality is operating at any given time.

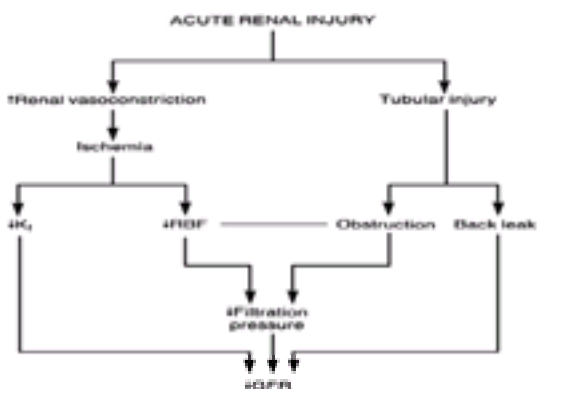

FIGURE 86.11. Acute renal failure pathogenesis. K_f, glomerular capillary ultrafiltration coefficient; *RBF*, renal blood flow; *GFR*, glomerular filtration rate. (Adapted from Hostetter JH, et al. Mechanisms of impaired glomerular filtration in acute renal failure. In: Brenner BM, Stem JH, eds. Contemporary Issues in Nephrology: Acute Renal Failure. New York: Churchill Livingstone, 1981:6.)

Clinical Manifestations

Presentation

The presentation of ARF varies and usually relates to the underlying disorder. Typical symptoms and signs are given in Table 86.38, together with the likely diagnosis. This list is by no means comprehensive but emphasizes those disorders likely to be encountered. Most children are oliguric or give a history of "decreased urination." If solute retention is severe and has persisted for days to weeks before seeking medical attention, the clinical manifestations of uremia may ensue and obscure, for the moment, the underlying diagnosis (Table 86.39). One consideration that must always be raised in a patient with suspected ARF is whether it has occurred de novo or is superimposed on preexisting chronic renal failure. Clinical clues that may lead to the latter diagnosis are failure to thrive, a history of polyuria/polydipsia, continued good urine output despite historical and physical evidence of dehydration, and physical evidence of renal rickets. Relevant

laboratory data to support the diagnosis of chronic renal disease are reviewed later in this chapter.

Symptoms	Signs	Likely Diagnosis
Nausea, vomiting	—	Gastroenteritis (ATN)
Diarrhea	Dehydration, shock	Gastroenteritis (ATN)
Hemorrhage	Shock	ATN
Fever	Petechiae, bleeding	Sepsis, DIC (ACN)
Melena	—	HUS
Sudden pallor	—	HUS
Grand mal seizures	—	HUS
Fever, chills	Flank tenderness	Pyelonephritis
Fever, skin rash	Erythema multiforme, purpura	AIN HSP nephritis
Sore throat	Hypertension	PSGN
Pyoderma	Edema	PSGN
Grand mal seizures	Congestive heart failure	PSGN
Trauma	Muscle tenderness	Myoglobinuria
Myalgia	Myoedema	Myoglobinuria
Antibiotics, diuretics	—	Nephrotoxic acute renal failure
Variable urine output	Suprapubic mass	OU

ATN, acute tubular necrosis; *DIC*, disseminated intravascular coagulation; *ACN*, acute cortical necrosis; *HUS*, hemolytic uremic syndrome; *AIN*, acute interstitial nephritis ("hypersensitivity nephritis"); *HSP* Henoch–Schönlein purpura nephritis; *PSGN*, poststreptococcal glomerulonephritis; *OU*, obstructive uropathy.

Table 86.38. Acute Renal Failure Presenting Symptoms and Signs

I. Gastrointestinal
- A. Nausea, vomiting, diarrhea
- B. Hiccoughs, fetid odor
- C. Hematemesis, melena

II. Cardiovascular
- A. Pericarditis

III. Dermatologic
- A. Pruritus
- B. Uremic "frost"

IV. Neurologic
- A. Apathy, fatigue
- B. Psychiatric disturbance
- C. Seizures
- D. Asterixis
- E. Coma
- F. Peripheral neuropathy

Table 86.39. Acute Renal Failure: Clinical Uremia

Complications

It is the immediate or, occasionally, the delayed complications of ARF, not ARF itself, that confront the emergency physician with the most important diagnostic and therapeutic challenges. The major complications in terms of frequency and threat to life are disturbances in serum tonicity and water balance; severe hyperkalemia with impending or actual cardiac arrhythmia; CHF with pulmonary edema, usually secondary to hypertension; malignant hypertensive encephalopathy with seizures; urinary tract infection with associated urinary obstruction; and metabolic seizures.

As previously discussed, serum tonicity and serum osmolality measure different phenomena. The former reflects solutes that do not traverse cell membranes, thereby regulating transcellular water movement. The latter reflects all solutes, regardless of their cell membrane permeability. ARF is an excellent example of this distinction. Because urea freely crosses all cell membranes, it raises body fluid osmolality without affecting tonicity or transcellular water movement. A child with a serum Na of 140 mEq/L, glucose of 90 mg/dL, and BUN of 100 mg/dL is hyperosmolar but isotonic. Water balance is normal. If that same child had a serum Na of 125 mEq/L, he or she would be considered isoosmolar but hypotonic and would require water restriction to forestall all swelling.

Hyperkalemia occurs in association with many causes of ARF but particularly in association with myoglobinuria and after open heart surgery. It is aggravated by hemolysis, acidosis, infection, and catabolic stress. CHF and malignant hypertensive encephalopathy typically occur in many forms of acute glomerulonephritis but especially in poststreptococcal glomerulonephritis. The risks of rapid destruction of renal parenchyma by bacterial organisms is dramatically increased in the face of obstructive uropathy. Such patients usually have systemic symptoms and signs of infection, including fever, chills, vomiting, abdominal pain, and costovertebral angle tenderness. Metabolic seizures are most often the result of uremia and not hyponatremia and/or hypocalcemia, which are often present in ARF. Hyponatremia is mild and usually develops slowly. Hypocalcemia usually does not cause tetany in the untreated patient because of coexistent metabolic acidosis.

Laboratory and Radiologic Studies

Of particular importance in differentiating the three anatomic forms of ARF are the urinalysis and the so-called urinary indices. In the critically ill patient who has not passed urine, a sterile straight catheterization of the bladder is appropriate to obtain a sample. The urinalysis is most helpful in separating glomerulonephritis from the other causes of ARF. In the typical case of acute glomerulonephritis, the dipstick shows large amounts of blood and protein, and RBCs, granular, and cellular (i.e., RBC) casts are in the spun sediment. Patients with prerenal ARF and those with acute tubular necrosis typically have little blood or protein by dipstick and an unremarkable sediment save for hyaline casts. Occasionally, the latter group will have prominent numbers of renal tubular epithelial cells and epithelial cell casts in the sediment, but this is generally unhelpful.

The major differentiation of prerenal ARF from acute tubular necrosis is by urine concentration as measured by specific gravity. Typically, the patient with prerenal ARF has a concentrated urine (specific gravity greater than 1.025), whereas the patient with acute tubular necrosis tends to have an isosthenuric urine (specific gravity 1.005 to 1.015). Hematuria by

dipstick examination without corresponding RBCs in the sediment suggests hemoglobinuria or myoglobinuria as the cause of ARF, especially if pigmented granular casts are also seen. Renal tubular and bladder epithelial cells and epithelial cell casts are commonly seen in nephrotoxic ARF or drug-induced (hypersensitivity) acute interstitial nephritis. Eosinophils on a Wright's stained urine sediment make the latter diagnosis much more likely. Marked pyuria, leukocyte casts, and a positive Gram stain of the urine all support the diagnosis of acute pyelonephritis, with or without coexistent obstruction.

Urinary indices refer to the ratios of the simultaneously measured solutes sodium, creatinine, and urea, and osmolality in “spot” samples of blood and urine. The primary purpose of these indices is to assist in differentiating prerenal ARF from acute tubular necrosis. These indices have generally replaced the BUN:creatinine ratio and “spot” urine sodium concentrations, which are too variable to reliably separate prerenal from renal ARF. They are outlined in Table 86.40.

Indices	Acute Renal Failure	
	Prerenal	Intrinsic[b]
Older Children and Adults		
U/P urea nitrogen	>8	<3
U/P creatinine	>40	<20
U/P osmolality	>500 mOsm/kg H_2O: >1.5	<350 mOsm/kg H_2O: <1.5
FE_{Na} (%)[c]	<1.0	>1.0
Neonates and Infants		
U/P urea nitrogen	Variable	Variable
U/P creatinine	Variable	Variable
U/PO osmolality	>1.0	<1.0
FE_{Na} (%)[c]	>2.5	<2.5

[a] U_x/P_x refers to simultaneously measured urine and plasma concentrations of x.
[b] Refers to classical acute tubular necrosis from various causes.
[c] Fractional excretion or filtered sodium = $(U/P)_{Na}/(U/P)_{creatinine} \times 100$.

Table 86.40. Acute Renal Failure: Urinary Indices[a]

Other diagnostic studies are outlined in Table 86.41. HUS is typically characterized by thrombocytopenia, microangiopathic hemolytic anemia with RBC fragmentation, and oliguria. However, early in the course of HUS, the only abnormal findings may be a rising BUN and falling hemoglobin, or bloody diarrhea alone, suggesting inflammatory bowel disease. The coagulation profile is often abnormal in HUS and always so in disseminated intravascular coagulation (DIC). Low C3 complement is seen in 90% of children with poststreptococcal glomerulonephritis, and elevated streptococcal serologies occur in 85 to 95% of such children, depending on the number of antibodies measured. A typical patient with poststreptococcal glomerulonephritis is presented in the section on hypertension (Case 4). Elevated antinuclear antibody titers are found in virtually all patients with lupus nephritis, albeit a rare cause of ARF. High IgE levels and absolute eosinophilia suggest acute interstitial nephritis. Elevated creatine phosphokinase (CPK) is an invariable accompaniment of rhabdomyolysis and myoglobinuria. In patients with massive hemolysis, serum haptoglobin falls to undetectable levels and elevated plasma hemoglobin may impart a pink–red color to the serum. An elevated 24-hour urine excretion of uric acid supports the diagnosis of uric acid nephropathy as the primary cause of ARF. A potentially useful screening test is the ratio of urine uric acid to creatinine on a “spot” urine specimen; if the ratio is 1.0 or greater, it supports this diagnosis. Most of the other causes of ARF result in a raised serum but not urine uric acid.

Test	Diagnosis
Blood	
Platelet count	HUS, DIC
Blood smear	HUS, DIC
Coagulation profile	HUS, DIC
Blood culture	DIC, acute pyelonephritis
Streptococcal serologies	PSGN
C3 complement	PSGN
Antinuclear antibody	Systemic lupus erythematosus-nephritis
IgE, eosinophil count	AIN
Aminoglycoside level	Nephrotoxicity
Creatine phosphokinase (CPK)	Myoglobinuria
Haptoglobin, “pink” plasma	Hemoglobinuria
Urine	
Culture	Acute pyelonephritis
Protein (24 hr)	Acute nephritis
Uric acid (24 hr or $U_{uric}/U_{creatinine}$ ratio)	Uric acid nephropathy
Radiology	
Renal ultrasound	Obstructive uropathy
Intravenous pyelogram	Obstructive uropathy, pyelonephritis
Voiding cystourethrogram (VCUG)	Underlying chronic renal disease, obstructive uropathy
Renal flow scan	Acute tubular necrosis (cortical necrosis), renal vascular insult

HUS, hemolytic uremic syndrome; DIC, disseminated intravascular coagulation (bacterial sepsis); PSGN, poststreptococcal glomerulonephritis; AIN, acute interstitial nephritis (“hypersensitivity” nephritis).

Table 86.41. Acute Renal Failure Laboratory Tests for Diagnosis

Renal ultrasound has replaced intravenous pyelogram (IVP) as the initial radiograph study of choice to differentiate postrenal (obstructive) from intrinsic renal causes of ARF. This is particularly true in critically ill patients because the ultrasound evaluation is a noninvasive procedure without the risk of IV injection of contrast. The ultrasound examination does not usually show the specific site of obstruction, but the IVP should not be performed until the patient is cardiovascularly stable and well hydrated. The IVP subsequently provides data on renal size, position, and to some extent, function. Information gained may include evidence of obstruction; pyelonephritic scarring; and small, contracted kidneys that suggest underlying chronic disease. An early but persistent nephrogram phase of the IVP, without normal concentration of the dye in the pelvocaliceal system, supports the diagnosis of acute tubular necrosis with tubular backleak. The vesicoureterogram (VCUG) is often underused in the initial evaluation of suspected causes of postrenal ARF. It is the best test to diagnose posterior urethral valves (the most common obstructive cause of ARF) and provides additional information about the presence or absence of coexistent vesicoureteral reflux. The renal scan is best used to assess blood flow to and within the kidneys. In experienced hands, the scan can also assess renal size and differentiate intrinsic renal from postrenal causes of ARF. It may be used in conjunction with ultrasound in the child whose clinical status rules out the immediate use of IVP dye. An additional advantage of the scan over IVP is that it does not require a

minimal level of estimated GFR to be performed (i.e., serum creatinine 4.0 mg/dL or greater).

Management

When confronted in the ED with a child who has ARF, the examining physician should always ask four questions about therapy: 1) Is this prerenal ARF and can parenchymal ARF be prevented by the appropriate fluid therapy? 2) Are there any life-threatening complications evident at this time that must be treated immediately? 3) Is there urinary tract infection with associated obstruction that must be relieved immediately? and 4) Are there indications for immediate peritoneal dialysis or hemodialysis? An initial set of laboratory tests provides a guide for subsequent management (Table 86.42):

Blood	Other
Hemoglobin, hematocrit	Chest radiograph
Blood urea nitrogen, creatinine	Electrocardiogram
Electrolytes (Na, K, Cl, HCO_3)	Echocardiogram
Blood gas (optional)	
Ca, P, Mg, uric acid	

Table 86.42. Acute Renal Failure: Laboratory Tests for Management

1. If prerenal ARF is suspected from the clinical history, physical examination, and urinary indices, fluid resuscitation should be used. Confirmation of this diagnosis requires a resumption of normal urine flow and a decrease in solute retention after restoration of euvolemia. An approach to fluid resuscitation is outlined in Table 86.43. A single exception to this approach might be the patient who is euvolemic or even hypervolemic but in cardiogenic shock. In the critically ill, unconscious, or uncooperative patient with an uncertain urine output, placement of an indwelling urinary catheter helps monitor urine output accurately. One clinical condition that demands the use of mannitol and furosemide is myoglobinuria or hemoglobinuria. Here the purpose of therapy is to prevent tubular obstruction by pigmented proteins after ECF volume is restored. The order of therapy in Table 86.43 is revised; 1 to 2 mg/kg of furosemide IV is given initially followed 5 to 10 minutes later by 0.5 g/kg mannitol. After urine flow is established, an infusion of 5% mannitol in one-quarter strength saline can be administered as milliliter-for-milliliter replacement of urine until the pigmenturia has resolved. Finally, as indicated in Table 86.43, failure to respond to a fluid challenge or a fluid plus diuretic challenge has one of three explanations: 1) volume losses have been underestimated, 2) there is coexistent urinary obstruction, or 3) the patient has already developed parenchymal ARF. The major risk of mannitol occurs in a parenchymal ARF because if not excreted, it will recirculate and may cause ECF volume expansion.

I. Dehydration with Shock
A. 20 mL/kg/hr of crystalloid solution[a] until vital signs stable and urine flow reestablished (0–10 mL/kg/hr)
B. Repeat hourly if necessary for 1–2 doses
C. After hour 2 or 3, if no urine flow, catheterize
D. If no urine in bladder, give furosemide, 2 mg/kg IV[b]
E. If no urine flow, treat as parenchymal ARF
II. Hemorrhage with Shock
A. 20 mL/kg/hr of plasma, or if unavailable, crystalloid solution as listed in I.A
B. Transfuse when blood available (whole fresh blood or packed red blood cells plus fresh-frozen plasma)
C. After hour 2 or 3, if no urine flow, catheterize
D. If no urine in bladder, give furosemide, 2 mg/kg IV[b]
E. If no urine flow, treat as parenchymal ARF

[a]Normal saline; 5% dextrose in normal saline; 10% dextrose in one-quarter strength sodium chloride plus one-quarter strength sodium bicarbonate (37.5 mEq/L each).
[b]Mannitol can be substituted for furosemide at a dose of 0.5 g/kg (2.5 mL/kg of a 20% solution) infused over 10–20 min. A urine flow of 6–10 mL/kg/hr should be established in the first several hours.

Table 86.43. Acute Renal Failure (ARF): Immediate Therapy of Prerenal ARF

2. Regarding life-threatening complications of ARF, the therapy of hypertensive emergencies has been discussed previously (p. 839). Hyponatremia is common but rarely symptomatic in ARF unless it is the result of ECF volume depletion. In euvolemic or clinically edematous patients, the treatment is fluid restoration and no extra sodium. Hypocalcemia is also common but rarely symptomatic in ARF and should not be treated with supplemental calcium until and unless the serum phosphorus concentration is known. Failure to take this precaution may result in raising the $Ca \times P_i$ product and risk ectopic calcification or further renal damage. Metabolic acidosis does not need correction unless the serum bicarbonate is less than 15 mEq/L, and only then with slow replacement with 1 mEq/kg per day of bicarbonate and frequent monitoring. “Overshoot” alkalosis can easily occur in the face of a rapidly changing GFR and urine flow. Also, a sudden shift of the pH toward normal or an alkaline range can convert asymptomatic hypocalcemia into frank tetany. Treatment of hyperkalemia is often the most urgent goal in ARF. Specifics of therapy for varying levels of serum K are outlined in Table 86.44.

1. Serum [K] 5.5–7.0 mEq/L (normal ECG): Kayexalate 1 g/kg PO or per rectum*
2. Serum [K] >7.5 mEq/L or >7.0 mEq/L with abnormal ECG†: Step 1. Calcium gluconate 0.5 mL/kg as 10% solution over 2–4 min with ECG monitoring; stop when pulse rate falls 20 beats/min or to <100 beats/min Step 2. Sodium bicarbonate 3.3 mL/kg of 7.5% solution Step 3. Glucose 1 mL/kg as 50% solution; hyperkalemia persists, infuse a 20–30% glucose solution with 0.5 unit regular insulin/kg; keep blood sugar <300 mg/dL
3. Serum [K] persistently >6.5 mEq/L: Dialysis

ECG, electrocardiogram.
*Kayexalate exchanges 1 mEq K for 1 mEq Na and lowers serum [K] by approximately 1 mEq/L within 4 hr. It can be administered p.o. with food or beverage, by nasogastric tube, or per rectum in 10% glucose/water (1 g in 4 ml) or in 20% sorbitol (50–100 ml). It must be retained for at least 30 min.
†Serum [K] >7.0 with a normal ECG can be treated as outlined in Step 1.

Table 86.44. Acute Renal Failure: Emergency Treatment of Hyperkalemia

3. If the clinical picture, urinalysis, and Gram stain suggest urinary tract infection, then coexistent obstructive uropathy must be ruled out rapidly. It can be suspected immediately because acute pyelonephritis in the unobstructed patient rarely causes ARF. Absence of a history of difficulty voiding or failure to palpate an enlarged bladder does not rule out obstruction, and a renal ultrasound should be obtained.
4. The indications for dialysis are outlined in Table 86.45. Generally, peritoneal dialysis is favored over hemodialysis, although the latter may be more efficient at removing certain nephrotoxins and potassium. The reasons for favoring peritoneal dialysis are its ready availability, the relatively simple technique used, and its safety in terms of preserving cardiovascular stability or minimally disturbing cardiovascular instability. It is generally not recommended in patients with generalized vasculitis, heat stroke, or recent abdominal surgery. An exciting and newer modality of acute renal replacement therapy is continuous arteriovenous hemofiltration (CAVH). It is of particular value in removing plasma water and small middle solutes in hemodynamically unstable patients. Any child with suspected or proven ARF from any cause deserves hospitalization.

Uremic syndrome
Blood urea nitrogen >100 mg/dL
Persistent hyperkalemia (serum [K] >6.5 mEq/L)
Persistent metabolic acidosis (serum [HCO_3] <10 mEq/L)
Persistent congestive heart failure
Oliguric acute renal failure secondary to hemolytic uremic syndrome or rhabdomyolysis with myoglobinuria

Table 86.45. Acute Renal Failure: Indications for Peritoneal Dialysis

Hemolytic Uremic Syndrome

Background

Definition

HUS is a multisystem disorder that affects primarily infants and young children. It is the most common cause of renal failure in young children and has an incidence of 0.3 to 10 cases per 100,000 during childhood. The definition includes renal dysfunction, nonimmune hemolytic anemia, and thrombocytopenia. Extrarenal manifestations such as neurologic injury may occur and indicate that the process is not solely limited to the renal system.

Epidemiology

Caucasian children are affected more often than African-American children, and HUS has been reported in teenagers and adults. HUS occurs worldwide, but there are several well-defined regions in which HUS is endemic (Quebec province in Canada, Buenos Aires, and surrounding environs of Argentina). Causative factors include various infectious agents, both bacterial and viral. HUS appears to occur sporadically and in epidemics, and recent work has clearly established several familial patterns to confirm the role of genetics.

Subtypes

Two principal subgroups are defined in children: diarrhea associated (D +) and non–diarrhea-associated (D–). The diarrhea-associated type includes typical cases that present with a diarrheal illness of sudden onset. It is most often associated with *Shigella*-like toxin (SLT) producing *Escherichia coli* or *Shigella dysenteriae* type 1. The incidence of diarrhea peaks in summer and early fall and affects all races and genders equally. The nondiarrheal type may include a genetic predisposition, may relapse, does not show a seasonal variation, and has an overall worse prognosis. It is not associated with SLT producing *E. coli* or *S. dysenteriae* type 1. However, *S. pneumoniae* has been implicated as a causative agent. Other causes of nondiarrheal HUS are pregnancy, malignancy, and oral contraceptives.

Pathophysiology

In diarrhea-associated HUS, the usual causative organism is an enterohemorrhagic strain of *E. coli* of the serotype O157:H7. Serotype O157:H7 produces large amounts of toxins (verotoxins or Shiga-like toxins). There are two main types of Shiga toxins, 1 and 2, which are produced by the *E. coli* O157:H7 strain. This strain is responsible for approximately three-quarters of the cases of HUS. The major reservoir for this pathogen is the intestinal tract of domestic animals. The usual route of contamination in humans is via ingestion of improperly cooked meats and unpasteurized milk. Other potential sources are vegetables, unpasteurized apple cider, and swimming pools. *Shigella* also produces a toxin that is similar structurally to the *E. coli* toxin. Various other bacteria and viruses have been implicated in the initiation of HUS, but the only proven links are *E. coli* O157:H7 and some non-O157 strains, *Shigella,* and *S. pneumoniae.* Once an individual is infected, he or she can transmit the pathogen to others.

After the verotoxin is absorbed by the intestine, it is thought to initiate glomerular endothelial cell injury, which initiates the pathogenesis of HUS. This process then leads to intravascular and intraglomerular fibrin clot formation, which is enhanced by simultaneous activation of the coagulation system, leading to vascular occlusion and sclerosis of the glomeruli. Vessel narrowing leads to mechanical damage of erythrocytes, resulting in a microangiopathic hemolytic anemia. Intrarenal platelet adhesion occurs and causes thrombocytopenia. Platelet aggregation factors appear to play an important role in this process. Endothelial cell prostacyclin, a platelet antiaggregating factor, is depleted in HUS because of toxin-induced endothelial cell damage. Renal dysfunction results from decreased glomerular filtration because blood flow is reduced through the stenotic vessels.

Clinical Manifestations

Historical features of diarrhea-associated HUS include abdominal pain and diarrhea. Fever and vomiting may also be present, but it should be noted that *E. coli* O157:H7 infection rarely results in fever. There may also be blood in the stool, and this is seen more often in *E. coli* O157:H7 infections. Within the first week after the development of these symptoms, the patient will experience an abrupt onset of pallor, listlessness, irritability, and oliguria. The onset of HUS is relatively explosive, particularly the change in skin color. Physical examination reveals a sallow complexioned, listless, dehydrated child who may have edema hypertension, petechiae, and/or hepatosplenomegaly. Neurologic manifestations may be striking in HUS; some degree of encephalopathy is present in most patients. Specific findings include obtundation, hemiparesis, seizures, and brainstem dysfunction. The initial impression may be that of a surgical abdomen, primary colitis, or intussusception when hematochezia is the predominant complaint; in these cases, a barium enema is often the first diagnostic test ordered.

Diagnosis of HUS is based on the clinical profile of hemolytic anemia, thrombocytopenia, and ARF. The anemia is severe in many cases with a hemoglobin of 5 to 9 g/dL. The reticulocyte count is mildly elevated, and the platelet count can drop as low as 20,000/mm^3. The peripheral smear shows helmet cells, burr cells, and schistocytes, confirming the microangiopathic process. The urinalysis reveals hematuria, sometimes gross, with variable degrees of proteinuria and leukocyturia. Granular and hyaline casts are often seen in the urine sediment. Chemical studies reveal azotemia, metabolic acidosis, hyperbilirubinemia, and an increased lactate dehydrogenase (LDH). Routine stool cultures will not reveal *E. coli* O157:H7. The presence of O157 antigen must be detected with a specialized antiserum directed at the antigen. The laboratory evaluation of HUS is summarized in Table 86.46. Other causes of microangiopathic hemolytic anemia and renal failure that should be considered are systemic lupus erythematosus and malignant hypertension (Table 86.47).

I. Blood	E. Electrolytes (Na, K, Cl, HCO_3)
A. Complete blood count, reticulocyte count	F. Blood urea nitrogen, creatinine
B. Platelet count	G. Liver function tests
C. Blood smear	II. Urine
D. Coagulation screen (PT, PTT)	A. Urinalysis
	III. Stool

PT, prothrombin time; *PTT*, partial thromboplastin time.
A, antigen studies for *E. coli* 0159:H7.

Table 86.46. Laboratory Evaluation of Hemolytic Uremic Syndrome

I. Sporadic	B. Type 2
Infant–young child	1. Low prostaglandin levels
Typical prodrome	2. Atypical prodrome (URI)
Good prognosis	3. Older children
II. Familial	4. Sporadic occurrence; sibs develop much later
A. Type I	5. Poor prognosis
1. Typical prodrome	C. Type 3
2. Occurs in endemic areas	1. Autosomal-dominant trait
3. Siblings develop HUS within 1–2 wk	2. Older patient
4. Good prognosis	3. Pregnancy associated
	4. High mortality

URI, upper respiratory infection.

Table 86.47. Clinical Subtypes of Hemolytic Uremic Syndrome (HUS)

Management

The cornerstone of treatment remains early recognition and supportive care. Oliguric ARF is best managed by dialysis when any one or a combination of the following complications occurs: 1) BUN more than 100 mg/dL; 2) CHF; 3) encephalopathy; and 4) hyperkalemia, particularly if associated with an arrhythmia. Peritoneal dialysis has been shown to be as effective as hemodialysis. Treatment of the intrarenal coagulation with heparin or streptokinase is discouraged. The microangiopathic process is managed with transfusions of blood and platelets as clinically indicated. There has been no efficacy demonstrated in the use of fresh-frozen plasma (FFP) or plasmapheresis in the diarrhea-associated form of HUS. However, success has been reported after infusion of FFP in patients with the nondiarrheal form of HUS.

Aggressive management of HUS yields a greater than 90% survival rate. Return to normal renal function occurs in 65 to 85% of these patients. Older patients without a diarrheal syndrome, pregnant women, and familial cases have a much worse prognosis. In addition, patients with severe hypertension and arteriolar changes on renal biopsy and those with recurrent disease fare less well over time.

Acute Glomerulonephritis

Background

Definition

Acute glomerulonephritis refers to a syndrome characterized by the sudden appearance of smoky, tea-colored, or grossly bloody urine in which heavy proteinuria and hematuria is found along with the appearance of RBC casts, as well as hyaline and granular casts. The urine is typically concentrated and acidic.

Epidemiology

The paradigm form of acute glomerulonephritis is postinfectious or poststreptococcal nephritis. Specific strains of group A, b-hemolytic streptococci stimulate an immune response that is nephritogenic in character. In the temperate climates, type 12 b-hemolytic streptococci induce the typical “strep throats” noted in these patients before the onset of the urinary abnormalities. Such children are typically school-age and male. In the southern and tropical climates, type 49 streptococci induce pyoderma as the antecedent infection, usually in younger preschool children. In certain populations, the disease appears to be endemic, and cycles of epidemic acute glomerulonephritis have been noted (Red Lake Minnesota Indians; Maracaibo, Venezuela; Trinidad). Curiously, the prognosis appears to be more favorable in the epidemic than in the sporadic form of the disease.

Etiologies

Various causes may lead to acute glomerulonephritis. These causes can be divided into conditions that affect the kidney and conditions that are more systemic in nature that may present with renal manifestations (Fig. 86.12).

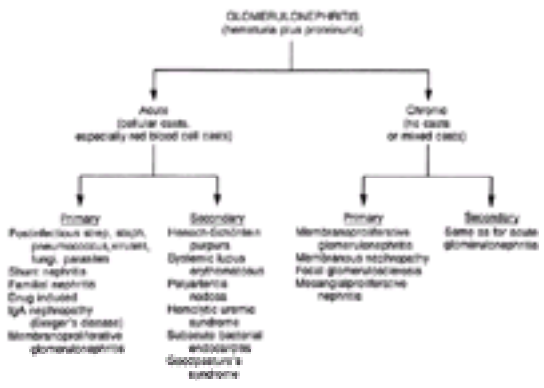

FIGURE 86.12. Categorization of glomerulonephritis.

Pathophysiology

It is believed that postinfectious acute glomerulonephritis results from the formation of soluble immune complexes either in the circulation or in situ within the kidneys in response to infection with streptococci or other agents. These complexes then deposit along the glomerular basement membrane and activate the complement system, thereby leading to the release of more inflammatory mediators and recruitment of acute inflammatory cells to the glomeruli. Localization of immune complexes is a function of their size, the activity of the reticuloendothelial system, hemodynamic factors, and the activity of glomerular mesangial cells in clearing and/or transporting these complexes. Similar pathogenetic mechanisms appear to be operative for shunt nephritis, lupus nephritis, and drug-induced nephritis. In Goodpasture's syndrome, rarely seen in children, antibodies are formed against pulmonary and glomerular basement membranes, leading to an

inflammatory reaction.

Clinical Manifestations

Generally, the diagnosis of acute glomerulonephritis is not difficult to make in the ED. The typical story is that of a 5- or 6-year-old boy who, 1 to 2 weeks after a sore throat, develops the sudden onset of brown, tea-colored, or grossly bloody urine in association with peripheral edema, particularly around the eyes, and a decreased urinary output. There may be associated cough and congestion. On physical examination, hypertension, both systolic and diastolic, is found. Some children are completely asymptomatic and present merely with abnormally colored urine. Rarely, patients may develop acute CHF or acute malignant hypertensive encephalopathy, dramatic complications of a sudden rise in BP caused by an acute reduction in GFR, with consequent retention of salt and water. These patients are particularly challenging for the emergency physician because the complications often mask the underlying disease. The history may or may not be positive for an antecedent infection. Generally, however, there is a latent period of 10 to 14 days after a sore throat and 14 to 20 days after pyoderma in poststreptococcal glomerulonephritis; 75% of children have edema and abnormally colored urine. This edema is usually firm in texture. The incidence of hypertension and oliguria varies from 25 to 33% of patients. ARF and nephrotic syndrome occur, but much less commonly.

The emergency physician must always consider an underlying chronic nephritis with an acute exacerbation when making the diagnosis of acute glomerulonephritis de novo. Underlying chronic nephritis may be suspected initially by the absence of a latent period between the antecedent infection and the onset of urinary abnormalities, an anemia disproportionate to the degree of renal glomerular insufficiency, and changes in the optic fundi of chronic hypertension. Poor linear growth is also a clue to the duration of glomerulonephritis and attendant renal insufficiency.

IgA nephropathy and membranoproliferative glomerulonephritis may be difficult to differentiate from the typical form of poststreptococcal glomerulonephritis if there is not good evidence for an antecedent strep infection. Familial nephritis is often associated with eye and hearing abnormalities. Secondary forms of glomerulonephritis are diagnosed on the basis of other systemic manifestations; these diseases are covered elsewhere in this text.

The laboratory database is outlined in Table 86.48. The findings on urinalysis are the single most important in categorizing glomerulonephritis. Proteinuria, although a function of urinary concentration, is almost always greater than 2 on the dipstick, indicating that the protein present is not merely a result of the hematuria itself. RBC casts are the hallmark of acute glomerulonephritis from any cause. Leukocyturia may also be seen. In girls, urinary tract infection can present with proteinuria and hematuria, so a urine culture should always be obtained in the initial workup. Routine electrolytes may reveal hyponatremia secondary to water retention, hyperkalemia secondary to oliguria, and of course, azotemia with a raised BUN and creatinine. In nephrotic patients, serum albumin is reduced. Hemoglobin is usually normal. Serologic studies are important in confirming a streptococcal source. Antibodies to any or all five extracellular streptococcal antigens may be measured; the more antibodies measured, the more likely abnormal results will be found. For patients who sustain strep throats, antistreptolysin O (ASO) and antihyaluronidase (AH) titers are measured. Antistreptolysin peaks at 10 to 14 days and AH at 3 to 4 weeks. For patients who sustain pyoderma, anti DNAse-B antibodies are measured. Antibiotic therapy blunts the antibody responses to these streptococcal antigens. If systemic lupus erythematosus is suspected as the cause of acute glomerulonephritis, an antinuclear antibody titer should be measured. The C3 complement is depressed in 90% of patients with poststreptococcal glomerulonephritis and in a number of other types of acute glomerulonephritis. Occasionally, group A b-hemolytic streptococci will be recovered from the throat or the skin when the patient presents with urinary abnormalities. Therefore, cultures should be routinely obtained. A chest radiograph is advised in hypertensive patients with or without signs and symptoms of CHF. Renal biopsy reveals swollen glomeruli with proliferation of the extracapillary cells; deposition of immunofluorescent-positive granules of IgG, IgM, and C3; and subepithelial electron-dense deposits. The latter two findings represent immune complexes.

I. Urine	E. Streptococcal serologies (ASO, anti-DNAse-B)
A. Urinalysis	F. C3 complement
B. Urine culture	G. Antinuclear antibody
II. Blood	III. Other
A. Complete blood count	A. Throat culture
B. Electrolytes (Na, K, Cl, HCO_3)	B. Skin culture (if pyoderma is present)
C. Blood urea nitrogen, creatinine	C. Chest radiograph
D. Total protein, albumin, and globulin	

Table 86.48. Laboratory Evaluation of Acute Glomerulonephritis

Management

The goals of the emergency physician are to recognize the diagnosis and to treat life-threatening emergencies secondary to hypertension. If CHF (p. 838) is noted, the head-up position, supplemental oxygen, and 0.5 to 1.0 mg/kg Lasix administered by IV are recommended. Hypertension may be treated with 0.25 to 0.5 mg/kg sublingual nifedipine, 2.5 to 5.0 mg/kg IV diazoxide by push, or 0.5 mg/kg hydralazine given by IV over several minutes. Nitroprusside is rarely required. Encephalopathy usually requires antihypertensive therapy only if the link between severe hypertension and the neurologic abnormalities is noted immediately.

Any child with acute glomerulonephritis who is oliguric or hypertensive should be admitted for close observation. In the mildly affected patient, discharge from the ED is reasonable if the patient is instructed to follow a low-sodium diet. A follow-up appointment with the primary physician is advised within 48 to 72 hours, and instructions are given to the family on following urine output and weight and observing for the signs and symptoms of hypertension.

Poststreptococcal glomerulonephritis carries an excellent prognosis in children; more than 80% recover spontaneously without apparent sequelae. A particularly ominous variant of acute glomerulonephritis, known as rapidly progressive glomerulonephritis, is heralded by progressive oligoanuria and azotemia, often leading to uremia. Extracapillary crescents of proliferating cells are noted in the glomeruli on renal biopsy in these patients. Persistent hypertension, edema, nephrotic-range proteinuria, azotemia, and hypocomplementemia that occur in a patient who, on first evaluation, appears to have acute glomerulonephritis may be indications of a more chronic form of the disease and are indications for renal biopsy.

Henoch-SchöAdonlein Purpura

Background and Pathogenesis

Henoch-SchöAdonlein purpura is considered one of the collagen vascular diseases in which a vasculitis of small blood vessels give rise to the triad of abdominal pain, arthritis, and purpura. Renal involvement with an acute glomerulonephritis is often present. The disease occurs in school-age children and young adults and is more common in Caucasians and males. The cause is unclear. Patients are often found to have elevated IgA levels in the blood in the acute phase, and both IgA-containing circulating immune complexes and glomerular mesangial deposition of IgA by immunofluorescence microscopy are additional markers of the disease. The IgA-containing immune complexes may also be detected in the skin lesions in these patients.

Clinical Manifestations

Antecedent upper respiratory tract infections are noted in one-third to three-quarters of patients. The abdominal pain is described as colicky but often severe. The arthritis is migratory, affecting the larger joints. Swelling may not be present around these joints. The rash is symmetric and purpuric and most noticeable over the extensor surfaces of the arms, legs, and buttocks. Other rashes, including erythema multiforme, may also be noted. Renal involvement is variable but typically occurs in the first month of illness. Asymptomatic microhematuria occurs most commonly in 80% of affected patients. Azotemia is usually transient. The older the child and the later the onset of renal involvement, especially if there is nephrotic-range proteinuria and/or renal insufficiency, the worse the prognosis. Hypertension is uncommon.

Routine laboratory studies that include CBC, electrolytes, serum proteins, and C3 complement are usually normal. The diagnosis is primarily based on clinical findings. Henoch-SchöAdonlein purpura nephritis is difficult to differentiate from IgA nephropathy in the absence of the skin rash and joint findings.

Management

There is no specific therapy for Henoch-SchöAdonlein purpura other than the use of corticosteroids used acutely to treat the severe abdominal pain or arthritis that occurs in selected patients. Signs and symptoms may wax and wane for a period of several months. Prognosis is generally excellent but depends on the severity and extent of renal involvement.

Renal Tubular Acidosis

RTA is a syndrome characterized by a persistent hyperchloremic nonanion gap metabolic acidosis. In this disorder, the renal regulation of the bicarbonate reabsorption and/or regeneration is deranged. Although RTA is rare, the emergency physician may occasionally encounter children with this abnormality; therefore, knowledge of its presentation and treatment is indicated.

A child with RTA may confront the emergency physician in one of several clinical settings: 1) an acute urinary tract infection, 2) muscle weakness and/or an adynamic ileus of the bowel secondary to profound hypokalemia, and least common, 3) acute renal colic secondary to the passage of renal calculi. RTA is divided into four types (Table 86.49).

Types	• Proximal RTA
• Distal RTA	Type 2 RTA
Type 1 or classic RTA	• Hybrid RTA
Type 4 or hyperkalemic RTA	

Table 86.49. Renal Tubular Acidosis (RTA)

Patients with all types of RTA present with a nonanion gap hyperchloremic metabolic acidosis. The GFR is almost always normal. The disorder may be primary (sporadic or familial) or secondary to various systemic disorders. Specific features facilitate the categorization of RTA types.

The physiologic characteristics of type 1 (distal) RTA are related to a reduced net rate of hydrogen ion secretion of the distal nephron. There may also be mild to moderate bicarbonate wasting. Patients with type 1 RTA are not able to lower their urine pH below 6 regardless of the severity of the acidosis. Proximal tubular functions are normal. The presentation may include acidosis, failure to thrive, hypokalemia (at times severe), nephrocalcinosis, or rickets.

Hyperkalemic, or type 4, RTA is thought to be the most common occurrence in children. This abnormality is caused by a disturbance in the renal handling of hydrogen and potassium ions. Patients with type 4 RTA are usually able to make a mildly acidic urine.

Type 2, or proximal, RTA is a defect in the reabsorption of filtered bicarbonate in the proximal tubule. This represents a derangement in the sodium-hydrogen ion exchange mechanism in this tubule segment. Distal mechanisms of acidification are intact. Many patients with this tubular disturbance manifest the problem as part of the Fanconi's syndrome. Patients with type 2 RTA have severe bicarbonate wasting when plasma bicarbonate is normal and an alkaline urine pH, but during acidotic states, the bicarbonaturia is decreased and the urine pH may be 5 to 5.5.

Once a patient is found to have a nonanion gap hyperchloremic metabolic acidosis, it is important to rule out other possible sources of the acidosis. For example, diarrhea with bicarbonate wasting and urinary tract infection with a urea-splitting organism should be considered. The arterial pH and P CO_2 should be measured several times, and all urine specimens passed should be tested for pH. A serum creatinine should be checked to confirm normal glomerular function. Several serum potassium determinations should be performed, and these may delineate the type of RTA present.

Management

The immediate treatment of RTA in the ED depends on the severity of signs and symptoms of hypokalemia (see section on Hypokalemia). Therapy of this disorder is best carried out in conjunction with a nephrologist. Administration of alkali is the treatment of choice for most types of RTA. However, overzealous IV alkali therapy should not be attempted because a rise in blood pH often lowers serum potassium and may exacerbate symptoms of weakness and/or cardiac arrhythmia.

The most commonly used alkalis are sodium bicarbonate and sodium citrate. In patients with hypokalemic RTA, a portion of the alkali can be given as $KHCO_3^-$ or K^- citrate. Plasma bicarbonate and serum potassium levels should be determined every 2 to 4 days initially. In patients with hyperkalemic RTA (type 4), furosemide is occasionally required to return the potassium to normal. Rarely, exchange resins are indicated.

Bartter's Syndrome

Bartter's syndrome was first described in 1962 and represents a constellation of renal tubular disorders characterized by profound hypokalemia, hypochloremic metabolic alkalosis, hyperreninemia with secondary hyperaldosteronism, but normal BP and renal glomerular function. The normal hypertensive response to IV angiotensin II infusion is blunted.

The pathogenesis of this disorder is controversial and probably involves a variety of tubular disturbances in K, Na, Cl, and Mg transport. The hallmark of Bartter's syndrome is urinary potassium wasting, but salt wasting is also common. Urinary prostaglandin excretion is increased, and a central role for prostaglandin overproduction is supported by the tendency of the biochemical abnormalities of this disorder to respond to the administration of prostaglandin synthetase inhibitors such as aspirin, indomethacin, and ibuprofen. In some patients, renal biopsy reveals hyperplasia of the juxtaglomerular apparatus, the site of renin production in the kidney.

The clinical presentation in infants and young children is characteristic and often reminiscent of RTA. Growth failure is severe, and episodes of severe vomiting and diarrhea that lead to dehydration often punctuate an early history of anorexia, polydipsia, and polyuria. There may be muscle weakness, aggravated by the bouts of dehydration. Such weakness is a constant complaint in older children. Unlike the more common forms of metabolic alkalosis that result from vomiting and overzealous diuretic therapy, sodium chloride therapy does not reverse the alkalosis and hypochloremia, and potassium chloride supplements rarely raise serum potassium levels to normal.

The differential diagnosis includes primary causes of hyperaldosteronism and renin-secreting tumors, in which hypotension is a prominent clinical finding, and pseudo-Bartter's syndrome is characterized by sodium, chloride, and potassium losses. Pseudo-Bartter's syndrome may be caused by cystic fibrosis with excessive sweating, diuretic abuse, chloride diarrhea, and the ingestion of a chloride-deficient formula.

The urinalysis reveals a concentrating defect but is otherwise unremarkable, save for the high concentration of electrolytes. Potassium repletion should be accomplished slowly unless there is life-threatening hypokalemia. As in severe RTA, mechanical ventilation may be necessary for extreme hypoventilation with associated respiratory acidosis. There is no role for ammonium chloride or a prostaglandin synthetase inhibitor in the emergency setting. Volume depletion should be managed in the conventional manner, with IV normal saline.

Urolithiasis

Background

Urolithiasis refers to calculi formation in the kidneys, ureters, or bladder. Once thought to be primarily an adult disease, it

is known to affect children and adolescents as well. It is important that physicians who treat pediatric patients in ED recognize those children who are at greatest risk of urolithiasis and develop an approach to its management.

Epidemiology

Urolithiasis occurs at a rate of 140 per 100,000 adults in the United States, and the incidence in children is approximately $^{1}/_{50}$ of that seen in adults. The incidence of pediatric admissions caused by urinary calculi varies from 1 per 7,600 to 1 per 1,066, depending on geographic area. It is estimated that at least an equal number of children are treated for stone disease as outpatients. Urolithiasis is endemic to specific areas of the United States, most notably portions of the southeastern states also known as the “stone belt” and southern California. Geographic location worldwide also plays a part in the cause of urinary calculi. In European children, infection-related stones comprised 75% of the diagnoses, whereas in Southeast Asia, endemic uric acid bladder stones are most common. In North America, metabolic causes account for more than 50% of diagnoses, and the stones contain calcium for the most part.

Among children, boys and girls are affected almost equally, unlike the male preponderance seen in adult stone patients. Urolithiasis is rare in African-American children, with 94% of urinary calculi occurring in Caucasians. The mean age of diagnosis is approximately 9 years.

Pathophysiology

Stone formation is a complex process that involves multiple physiochemical and anatomic factors (Fig. 86.13). Urinary crystallization is strongly related to the free concentrations of the lithogenic ions such as calcium and oxalate. The activity product is equal to the product of the concentrations of the ions in question, and it often exceeds the solubility product. Thus, urine is often supersaturated with calcium and oxalate ions at concentrations that, under laboratory conditions, would crystallize in vitro. Yet this does not happen in the urine because in most people inhibitors of nucleation are present. Such inhibitors may include urinary citrate, glycosaminoglycans, and more recently, glycoproteins such as nephrocalcin and uropontin.

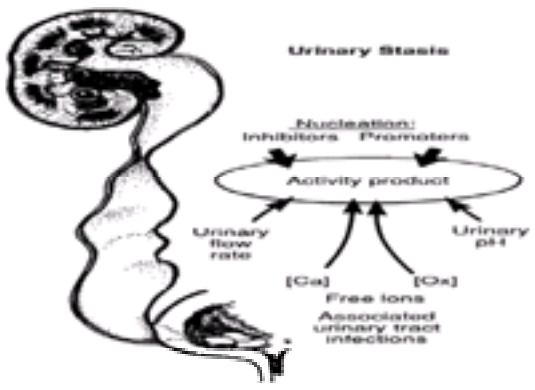

FIGURE 86.13. Pathogenesis of urinary calculi formation.

Certain factors act to promote urinary stone formation such as stasis of urine. Anatomic causes of bladder stasis may include megaureter, ureteropelvic junction (UPJ) obstruction, or a neurogenic bladder. Crystallization can be initiated on damaged urothelium or on a foreign body, and these are examples of heterogeneous nucleation. Certainly one factor in stone disease is dehydration because it raises the urinary free ion concentrations beyond a point at which even the inhibitors can prevent nucleation. Maintaining a dilute urine is the first step in preventing recurrence of nephrolithiasis.

Types of Calculi

Urinary calculi are often divided into four major categories: calcium, infection-related, cystine, and uric acid stones. Approximately 60% of urinary stones are calcium oxalate or calcium phosphate. Hypercalciuria is the most common noninfectious cause of urolithiasis in children. It generally correlates with a daily urinary calcium excretion greater than 4 mg/kg, and a random Ca:Cr ratio greater than or equal to 0.21 when the patient is on an undefined diet. Hypercalciuria usually occurs in the absence of an elevated serum calcium, and the idiopathic form is most commonly seen in children. Idiopathic hypercalciuria, even in the absence of calculus formation, has been associated with hematuria. This is attributable to calcium oxalate crystals that injure the urothelium. Some other causes of calcium urolithiasis are hyperparathyroidism; hypercalcemia; distal RTA; medications such as furosemide; and hyperoxaluria, hyperuricosuria, and hypocitraturia, and the idiopathic form that may result from increased absorption or renal leak of calcium.

Infection-induced stones are made of struvite (magnesium, ammonium, phosphate) and carbonate apatite. The bacterial enzyme urease ultimately creates an environment that favors the formation of struvite. Organisms that produce urease are *Proteus, Pseudomonas, Klebsiella, Serratia, Mycoplasma,* and *Staphylococcus.* Infection-induced stones are usually discovered before age 5 and 80% are found in boys. Typically, affected children have staghorn calculi that fill the renal calyces or pelvis. Urinary sediment will reveal pyuria, bacteriuria, and struvite crystalluria.

Cystine calculi account for approximately 4% of pediatric urolithiasis. Cystinuria is a recessively inherited disorder of amino acid transport manifested as excessive urinary excretion of cystine, arginine, lysine, and ornithine and by formation of urinary calculi. Cystine stones are usually radiopaque because of the sulfur ions present. Cystine crystals in the urine can be identified by their flat hexagonal shape.

Uric acid calculi account for 3 to 5% of pediatric urolithiasis. Most uric acid calculi result from precipitation of uric acid from supersaturated urine. Lithiasis may be idiopathic or associated with hyperuricemia, hyperuricosuria, or chronic excessive fluid losses. The most common cause of uric acid urolithiasis in children in the hyperuricemia/hyperuricosuria that results from increased purine synthesis as in patients with myeloproliferative disorders. Uric acid stones are generally multiple and are radiolucent.

Clinical Manifestations

Children with urolithiasis rarely present with the excruciating pain of stone passage as seen in adults. Pain occurs in the abdomen or flank in 50% of patients. In infants, such pain may be confused with colic, and occasionally, a diaper that is stained or shows crystals will provide a clue. Hematuria, either microscopic or macroscopic, occurs in at least 90% of children with urolithiasis. Other symptoms include urinary frequency, dysuria, and at times, urinary retention. A history of urinary tract infection is variably present. A family history of urolithiasis can be elicited in 50% of patients. Colic and hematuria episodes are most characteristic of calcium stones.

The physical examination may reveal tachycardia and an increase in BP because of pain; fever may be seen in 15% of patients. Costovertebral angle and/or flank tenderness may be present.

Management

If the history and physical examination are suggestive of urolithiasis, one should proceed to the detection of the calculus (Fig. 86.14). A urinalysis is an essential diagnostic screen for stone disease. In most affected children, there will be microscopic or macroscopic hematuria; pyuria and bacteriuria may also be found. Crystal formation, if present in the urinary sediment, is an additional supportive and often diagnostic finding. Definitive detection of a radiopaque calculus relies on an imaging study. It is estimated that 95% of urinary calculi are radiopaque and can therefore be identified on a plain abdominal radiograph. Ultrasonography is an adjunctive study that can detect upper tract calculi reliably and is also particularly useful for delineating obstructing calculi. The IVP is helpful in locating a ureteral or bladder stone and urinary obstruction.

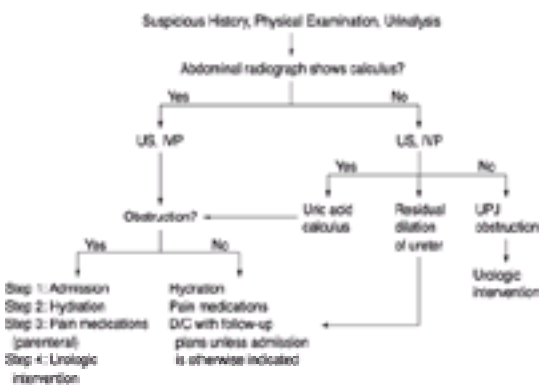

FIGURE 86.14. Diagnostic approach for evaluation for urinary calculi. *US,* ultrasound; *IVP,* intravenous pyelogram; *D/C,* discharge; *UPJ,* ureteropelvic junction.

Once a calculus has been detected in the ED, attention should be focused on the recognition and treatment of the potential acute complications that may accompany urolithiasis. Such complications include pain (at times severe), urinary tract infection, and/or urinary obstruction. If a urinary calculus is not found with the aforementioned studies but the history is suspicious of calculus disease, it is likely that the patient has already passed the stone and presently is suffering from the aftermath of stone passage (i.e., spasm and dilation of the ureter). In this scenario, one must carefully examine the urine for crystalluria and hematuria, as well as search for an increased Ca:Cr ratio.

For severe pain, relief should be provided promptly. Narcotic analgesics such as morphine sulfate or meperidine may be necessary. Nonsteroidal anti-inflammatory medicines such as ibuprofen may have an important role in pain management if the patient can tolerate oral medications. Some patients may be unable to drink, and IV hydration may be required to ensure an adequate urine flow rate. If there is no evidence of urinary obstruction or renal insufficiency, fluids should be run at twice the maintenance requirement. When an associated urinary tract infection is suspected, appropriate antimicrobials should be initiated after culture. If urinary obstruction is entertained, a sonogram should be performed urgently and a urologic consultation should be obtained. Immediate treatment of an obstruction-inducing calculus includes extracorporeal shock wave lithotripsy, percutaneous nephrostolithotomy, or open stone surgery. All urine should be strained to assist in collecting gravel or stone particles for analysis. If a patient is known to have renal insufficiency, the management of superimposed urolithiasis should be done in conjunction with a pediatric nephrologist. Numerous indications exist for the admission of patients with urolithiasis (Table 86.50). If the patient does not fit into one of these categories, outpatient management is appropriate. Because the rate of recurrence of calculi formation is high, a stone patient should be referred to a pediatric urologist or nephrologist to ensure a detailed follow-up that focuses on stone analysis and long-term management. Prevention relies heavily on consistently high fluid intake.

Urinary obstruction	Solitary kidney
Intractable pain	Renal insufficiency
Dehydration	Inability to tolerate oral fluids

Table 86.50. Urolithiasis—Indications for Admission

Chronic Renal Failure

Background

Chronic renal failure (CRF) in children is not a rare entity; studies suggest an incidence of 16 per million population in the United States. Many children who have reached end-stage renal failure (ESRF) are receiving chronic hemodialysis or, more recently, chronic ambulatory peritoneal dialysis (CAPD), often in the home. In all pediatric patients with ESRF, renal transplantation is the ultimate therapeutic goal. The causes of ESRF vary with age: for those younger than 5 years, congenital structural lesions predominate, along with obstructive uropathies; for those older than 5 years, acquired lesions are more commonly seen. A summary list of causes for ESRF is shown in Table 86.51.

I. Congenital Structural Lesions
A. Hypoplasia, dysplasia
B. Malformations (cystic diseases)
II. Obstructive Uropathies
A. Posterior urethral valves
B. Bilateral ureteropelvic junction obstruction
C. Bilateral vesicoureteral reflux with infection (e.g., reflux nephropathy)
III. Acquired Nephropathies
A. Chronic glomerulonephritis
B. Hemolytic uremic syndrome
C. Acute tubular or cortical necrosis
D. Hereditary nephritis

[a]This list is not intended to be comprehensive; it lists the most common causes of end-stage renal failure.

Table 86.51. Causes of End-Stage Renal Failure[a]

Pathophysiology and Clinical Findings

Once a critical level of renal functional deterioration occurs from any cause, eventual progression to ESRF is inevitable. This “threshold level” has not been defined, and the rate of progression varies, although it appears highest at around the time of puberty. Many factors can precipitate this deterioration: ongoing immunologic injury, urinary tract infection, hypertension, extra cellular volume depletion, urinary obstruction, and hypercalciuria with or without nephrolithiasis. Many patients are not known to have CRF when they present acutely to the ED. Clues in the history are excessive fatigue, anorexia, vomiting, short stature, skeletal pain, polyuria, and polydipsia. On physical examination, signs of anemia, a fetid breath, chronic changes of hypertension, asterixis, and peripheral neuropathy are tell-tale clues. Signs and symptoms of CRF usually begin at a GFR 20% of normal or less, and virtually always when the GFR reaches 10% of normal. Rapid falls in GFR may exacerbate the clinical picture.

Management

Disturbances in fluid electrolyte and acid-base balance, calcium and vitamin D metabolism, and cardiovascular and neurologic function predominate in ESRF and are outlined in Table 86.52. Anemia in ESRF has been managed successfully with a combination of effective dialysis and recombinant erythropoietin injections. CHF may be aided by improving the hemoglobin level and restricting or removing extra salt and water, either with diuretics or dialysis. Uremic pericarditis is less commonly seen in children than in adults and appears to correlate with the level of serum creatinine. The careful monitoring of water intake is required to avoid hyponatremia and hypernatremia because the kidneys' ability to modulate urinary water excretion is greatly reduced. Some patients may also exhibit an Na-wasting state despite the low GFR. Potassium retention is a significant risk in ESRF, particularly in patients who have not yet started dialysis. Uremia may block transcellular K transport in these patients. Accumulation of organic acids and the inability of the damaged kidney to regenerate new bicarbonate buffer explain the metabolic acidosis of ESRF. Modest doses of alkali normally correct this problem unless the patient is “Na-sensitive”; in that case, dialysis is indicated to avoid fluid overload.

I. Anemia
A. Decreased erythropoietin production
B. Hemolysis
C. Blood loss (bleeding tendency)*
II. Cardiovascular
A. Congestive heart failure*
B. Uremic pericarditis*
III. Fluid, Electrolyte, Acid-Base Balance
A. Reduced free water clearance, obligatory isothermia*
B. K balance lost when glomerular filtration rate <10 mL/min, hyperkalemia common*
C. Metabolic acidosis (increased anion gap)*
IV. Vitamin D/Ca Metabolism
A. Hypocalcemia, hyperphosphatemia*
B. Secondary hyperparathyroidism
C. Osteomalacia (aluminum bone disease)
V. Immune Function
A. Increased risk of infection*
1. Impaired host defense (white blood cell function)
VI. Neurologic Function*
A. Inability to concentrate, loss of memory
B. Headache, drowsiness, coma
C. Weakness, tremors, seizures
D. Peripheral neuropathy
E. Autoimmune dysfunction (sweating, swings in blood pressure)

*Improved with dialysis.

Table 86.52. Matabolic and Clinical Abnormalities in End-Stage Renal Failure

Renal osteodystrophy is the term used to describe the myriad changes that occur in Ca, vitamin D, and bone metabolism in ESRF. The combination of a reduced phosphate intake and/or phosphate binders, supplemental calcium (when serum phosphate has been normalized), and 1,25-dihydroxyvitamin D therapy often improves patients with renal osteodystrophy before dialysis is required. The use of antacids and dialysis baths that contain aluminum has provided a particularly devastating form of osteomalacia and dementia not amenable to the usual therapies.

The uremic state clearly impairs WBC function; neutrophil chemotaxis and mononuclear cell chemotaxis are reduced, increasing susceptibility to infection.

The neurologic disturbances noted in ESRF are what usually define the “uremic state.” Clinical manifestations are diverse but often respond dramatically and rapidly to efficient dialysis. Of particular note is the dialysis disequilibrium syndrome, characterized by headache, nausea and vomiting, visual disturbances, disorientation, wide swings in BP, and seizures. This condition is less common in children than in adults but occurs when initial dialysis (usually hemodialysis) lowers a significantly elevated BUN (150 mg/dL or greater) too rapidly, allowing water to move into brain cells and cause cerebral edema. Rapid infusion of mannitol is often effective in reversing these signs and symptoms.

Rhabdomyolysis

Background

Rhabdomyolysis represents a disruption of the skeletal muscle leading to leaking of intracellular contents. It is uncommon in pediatrics, affecting adolescents more commonly than children. A number of disorders, including trauma, intoxications, seizures, infections, endocrinopathies, and metabolic defects, may lead to significant injury of skeletal muscle (Table 86.53). Influenza infections are perhaps the most common precipitating event overall. Of particular note in otherwise healthy adolescents is exertional rhabdomyolysis, which may develop after strenuous exercise.

Trauma	Amphetamines
Extensive muscle injury	Aspirin
Crush injury	Neuroleptic agents
Compartment syndrome	Monoamine oxidase inhibitors
Strenuous exertion	Succinylcholine
Infections	Envenomations
Influenza	Endocrinopathies
Sepsis	Hyperthyroidism
Toxic shock syndrome	Hypothyroidism
Rocky Mountain spotted fever	Diabetic ketoacidosis
Tetanus	Inherited disorders of muscle enzymes
Hyperthermia	Miscellaneous
Prolonged seizure	Polymyositis
Toxins/medications	
Ethanol	
Cocaine	

Table 86.53. Causes of Rhabdomyolysis

Pathophysiology

Regardless of the specific insult, the final common pathway in rhabdomyolysis is injury to skeletal muscle leading to myolysis. As the muscle cells break down, they release their intracellular contents, including myoglobin, CPK, glutamic oxaloacetic transaminase, lactate dehydrogenase, potassium, and phosphate, into the plasma. The circulating myoglobin is subsequently excreted in the urine.

Clinical Manifestations

The classic triad of complaints in rhabdomyolysis consists of myalgias, weakness, and dark urine. In mild cases, particularly early in the course, myalgias may be the predominant manifestation, with or without mild weakness. The emergency physician should inquire about preceding viral infections, exercise, environmental conditions, injuries, bite wounds, ingestions, and medication use. Important considerations in the medical history include seizures, thyroid disorders, and diabetes.

On examination, findings specific to rhabdomyolysis include tenderness of the muscles to palpation, decreased strength, and less commonly, edema. The vital signs may be revealing of the cause in the case of hyperthermia. In some cases, trauma may be apparent, as in the form of a crush injury; however, patients with muscle injury secondary to vigorous exercise may manifest no local signs or minimal tenderness and edema.

The most reliable test for rhabdomyolysis is elevation of the CPK level to at least five times the upper limit of normal. The release of CPK occurs rapidly after injury to the muscle, peaks at 24 to 36 hours, and persists for several days. Levels as high as 50,000 to 100,000 units/mL are not unusual. The urine from patients with rhabdomyolysis may appear dark and test positive for blood with a reagent strip, but RBCs are not increased on microscopy. Other laboratory abnormalities include hyperphosphatemia, hypocalcemia, acidosis, hyperuricemia, and elevations of BUN and creatinine.

The major potential complication of rhabdomyolysis is ARF, resulting at least in part from myoglobin casts obstructing renal tubules. Renal failure manifests with oliguria or anuria and worsens the biochemical profile of the patient by increasing the plasma levels of hydrogen ions, phosphate, potassium, BUN, and creatinine.

Management

In addition to general supportive care of critically ill patients, when possible, steps should be taken to eliminate the inciting event. As an example, anticonvulsants are administered to interrupt status epilepticus and cooling is indicated for the patient with hyperthermia. Initial measurement of muscle enzymes and electrolytes is appropriate, and vital signs should be monitored. Therapy is directed at restoring vascular volume, when compromised, and facilitating blood flow to the kidneys in an effort to preserve renal function. Management begins with the delivery of a 20 mL/kg bolus of normal saline. In more severe cases, diuresis is achieved either with mannitol (1 g/kg) or furosemide (1 mg/kg). Depending on their severity, acidosis and electrolyte disorders may need specific treatment; occasionally, dialysis is required. Patients with significant rhabdomyolysis, as defined by markedly elevated levels of CPK and/or myoglobinuria, require admission to the hospital.

Suggested Readings

FLUID AND ELECTROLYTE DISTURBANCES: ISOTONIC DEHYDRATION, HYPONATREMIA, HYPERNATREMIA

Berry PL, Belsha CW. Hyponatremia. Pediatr Clin North Am 1990;37:351.

Boineau FG, Lewy JE. Estimation of parenteral fluid requirements. Pediatr Clin North Am 1990;37:257.

Conley SB. Hypernatremia. Pediatr Clin North Am 1990;37:257.

Editorial. Excess water administration and hyponatremic convulsions in infancy. Lancet 1992;1:153.

Humes HD, Narins RG, Brenner BW. Disorders of water balance. Hosp Pract 1979;14:133.

Kallen RJ. The management of diarrheal dehydration in infants using parenteral fluids. Pediatr Clin North Am 1990;37:265.

Kallen RJ, Lonergan M. Fluid resuscitation of acute hypovolemic hypoperfusion states in pediatrics. Pediatr Clin North Am 1990;37:287.

Moses AM, Miller M. Drug-induced dilutional hyponatremia. N Engl J Med 1984;291:1234.

Narins RG. Therapy of hyponatremia. N Engl J Med 1986;314:1573.

Norman ME. The physiology of fluid-electrolyte and acid-base balance in children. Part II. In: Honig PJ, Tunnessen WW Jr, eds. Clinical Diagnosis Quiz. Syracuse, NY: Creative Medical Publications, 1980;5(6).

Schrier RW. New treatments for hyponatremia (Editorial). N Engl J Med 1978;298:214.

Trachtman H. Sodium and water homeostasis. Pediatr Clin North Am 1995;42.

HYPOKALEMIA, HYPERKALEMIA

Brem AS. Disorders of potassium homeostasis. Pediatr Clin North Am 1990;37:419.

Choen JJ. Disorders of potassium balance. Hosp Pract 1979;14:119.

Editorial. Familial hypokalemic periodic paralysis. Lancet 1981;1:1140.

Gabow P. Disorders of potassium metabolism. In: Schrier RW, ed. Renal and Electrolyte Disorders. Boston: Little, Brown, 1976.

Reineck HJ. The control of potassium homeostasis. Kidney 1979;12:13.

HYPOCALCEMIA, HYPERCALCEMIA

Root AW, Harrison HE. Recent advances in calcium metabolism. I. Mechanisms of calcium homeostasis. J Pediatr 1976;88:1.

Root AW, Harrison HE. Recent advances in calcium metabolism. II. Disorders of calcium homeostasis. J Pediatr 1976;88:177.

METABOLIC ACIDOSIS

Brewer ED. Disorders of acid-base balance. Pediatr Clin North Am 1990; 37:429.

Foreman JW. Metabolic acid-base disorders. In: Cohn RW, Roth K, Segal S, eds. Metabolic Disease. A Guide to Early Recognition. Philadelphia: WB Saunders, 1983.

Hanna JA, Scheinman JI, Chan JC. The kidney in acid-base balance. Pediatr Clin North Am 1995;42(6):1365.

Winters RW. Pathophysiology of acid-base disorders. In: Winters RW, ed. The Body Fluids in Pediatrics. Boston: Little, Brown, 1973.

NEPHROTIC SYNDROME

Barnett HL, Edelmann CM, Bernstein J, et al. The nephrotic syndrome. In: Edelmann CM Jr, ed. Pediatric Kidney Disease. Boston: Little, Brown, 1992;II:1247–1266.

Giangiacomo JG, Cleary TG, Cole BR, et al. Serum immunoglobulins in the nephrotic syndrome. N Engl J Med 1975;293:8.

Habib R, Kleinknecht C. The primary nephrotic syndrome of childhood: classification and clinicopathologic study of 406 cases. In: Somers SC, ed. Pathology Annual. New York: Appleton-Century-Crofts, 1971:165.

Levinsky RJ, Malleson PN, Barratt TM, et al. Circulating immune complexes in steroid-responsive nephrotic syndrome. N Engl J Med 1978;298:126.

Toghman-Adham M, Siegler RL, Pysher TJ. Acute renal failure in idiopathic nephrotic syndrome. Clin Nephrol 1997;47:76–88.

Tulassay T, Raschek W, Scherer K. Intra- and extrarenal factors of oedema formation in the nephrotic syndrome. Pediatr Nephrol 1989;3:92.

Vernier RL. Urinary nephrotic syndrome. In: Holliday MA, Barratt TM, Vernier RL, eds. Pediatric Nephrology. 2nd ed. Baltimore: Williams & Wilkins, 1987.

White RHR, Glasgow EF, Mills RJ. Clinicopathologic study of nephrotic syndrome in childhood. Lancet 1970;1:1353.

HYPERTENSION

Balfe JW, Rance CP. Recognition and management of hypertensive crises in childhood. Pediatr Clin North Am 1978;25:159.

Bunchman TE, Lynch RE, Wood EG. Intravenously administered labetalol for treatment of hypertension in children. J Pediatr 1991;120:140–144.

Calhoun DA, Oparil S. Treatment of hypertensive crisis. N Engl J Med 1990;323:1177–1183.

Cetta F, Hurley M, Hatch D, et al. Malignant hypertension. J Pediatr 1991;118:981–986.

Farine M, Arbus GS. Management of hypertensive emergencies in children. Pediatr Emerg Care 1989;5:51–55.

Fleischmann LE. Management of hypertensive crises in children. Pediatr Ann 1977;6:72.

Gifford RW. Management of hypertensive crises. JAMA 1991;266:829–835.

Gill DG, daCosta M, Cameron JS, et al. Analysis of 100 children with severe and persistent hypertension. Arch Dis Child 1976;51:951.

Groshong T. Hypertensive crisis in children. Pediatr Ann 1996;25:368–376.

Hanna JD, Chan JC, Gill JR. Hypertension and the kidney. J Pediatr 1991;118:327–339.

Hulse JA, Taylor DSI, Dillon MJ. Blindness and paraplegia in severe childhood hypertension. Lancet 1979;2:553.

Kaplan NM. Management of hypertensive emergencies. Lancet 1994;344:1335–1338.

Lloyd-Still J, Cottom D. Severe hypertension in childhood. Arch Dis Child 1967;42:34.

Mendoza S. Hypertension in infants and children. Nephron 1990;54:289–295.

Murphy C. Hypertensive emergencies. Emerg Med Clin North Am 1995;13:973–1007.

Ram CVS. Hypertensive encephalopathy. Arch Intern Med 1978;138:1851.

Report of the Task Force on Blood Pressure Control in Children. Pediatrics 1977;59(Suppl):797.

Report of the Task Force on Blood Pressure Control in Children. Pediatrics 1987;79:1–25.

Sinaiko AR. Hypertension in children. N Engl J Med 1996;335:1968–1973.

Tarazi RC. Management of the patient with resistant hypertension. Hosp Pract 1981;16:49.

ACUTE RENAL FAILURE

Anderson RJ, Schrier RW. Clinical spectrum of oliguric and nonoliguric acute renal failure. In: Brenner BM, Stem JH, eds. Contemporary Issues in Nephrology. Acute Renal Failure. New York: Churchill Livingstone, 1981:6.

Bastl CP, Rudnick MR, Narins RG. Diagnostic approaches to acute renal failure. In: Brenner BM, Stem JH, eds. Contemporary Issues in Nephrology. Acute Renal Failure. New York: Churchill Livingstone, 1981:6.

Ellis D, Gartner JC, Galvis AG. Acute renal failure in infants and children: diagnosis, complications and treatment. Crit Care Med 1981;9:607.

Gaudio KM, Siegel NJ. Pathogenesis and treatment of acute renal failure. Pediatr Clin North Am 1987;34:771.

Hostetter JH, Wilkes BM, Brenner BM. Mechanisms of impaired glomerular filtration in acute renal failure. In: Brenner BM, Stem JH, eds. Contemporary Issues in Nephrology. Acute Renal Failure. New York: Churchill Livingstone, 1981:6.

Mathew OP, Jones AS, James E, et al. Neonatal renal failure: usefulness of diagnostic indices. Pediatrics 1980;65:57.

Miller TR, Anderson RJ, Linas SL, et al. Urinary diagnostic indices in acute renal failure. Ann Intern Med 1978;89:47.

Ng RCK, Suku W. Treatment of acute renal failure. In: Brenner BM, Stem JH, eds. Contemporary Issues in Nephrology. Acute Renal Failure.

New York: Churchill Livingstone, 1981:6.

Schrier RW. Acute renal failure: pathogenesis, diagnosis and management. Hosp Pract 1981;16:93.

Smotens P, Stein JH. Pathophysiology of acute renal failure. Am J Med 1981;70:479.

Thadhani R, Pascual M, Bonventre J. Acute renal failure. N Engl J Med 1996;334:1448–1460.

Trainin EB, Spitzer A. Treatment of acute renal failure. In: Edelmann CM Jr, ed. Pediatric Kidney Disease. Boston: Little, Brown, 1978.

HEMOLYTIC UREMIC SYNDROME

Milford DV, Taylor CM. New insights into the haemolytic uraemic syndromes. Arch Dis Child 1990;65:713.

Neild G. The hemolytic uremic syndrome: a review. Q J Med 1987;241:367.

Remuzzi G, Ruggenenti P. The hemolytic uremic syndrome. Kidney Int 1998;66:554–557.

Rondeau R, Peraldi MN. *Escherichia coli* and the hemolytic uremic syndrome. N Engl J Med 1996;335:660–662.

Siegel NJ. Acute renal failure. In: Tune BM, Mendoza SA, eds. Pediatric Nephrology. New York: Churchill Livingstone, 1984:306.

ACUTE GLOMERULONEPHRITIS

Gauthier B, Edelmann CM Jr, Barrett HL, eds. Clinical acute glomerulonephritis. In: Nephrology and Urology for the Pediatrician. Boston: Little, Brown, 1982:109.

Hoedemacker DLJ. Immunopathogenesis of glomerular diseases. In: Brodehl J, Ehrich JHH, eds. Paediatr Nephrology. Berlin: Springer-Verlag, 1983:17.

Jordan SC, Lemir JM. Acute glomerulonephritis. Pediatr Clin North Am 1982;29:857.

Siegel NJ. Acute renal failure. In: Tune BM, Mendoza SA, eds. Pediatric Nephrology. New York: Churchill Livingstone, 1984:306.

White RHR. Shunt Nephritis. In: Brodehl J, Ehrich JHH, eds. Paediatric Nephrology. Berlin, Springer-Verlag, 1983:221.

HENOCH-SCHöNLEIN PURPURA

Counahan R, Cameron JS. Henoch-SchöAdonlein nephritis. Contrib Nephrol 1977;7:143.

Kaku Y, Nohara K, Honda S. Renal involvement in HSP: a multivariate analysis of prognostic factors. Kidney Int 1998;53:1755–1759.

McEnery PT, Strife CF. Nephropathy associated with Henoch-SchöAdonlein (anaphylactoid) purpura. In: Tune BM, Mendoza SA, eds. Pediatric Nephrology. New York: Churchill Livingstone, 1984:239.

Meadow SR. The prognosis of Henoch-SchöAdonlein nephritis. Clin Nephrol 1987;9:87.

Orfila C, Lepert JC, Modesto A, et al. HSP and membranoproliferative-like glomerulonephritis. Nephron 1996;74:209–213.

RENAL TUBULAR ACIDOSIS

Donckerwolcke R. Renal tubular acidosis. Pediatr Clin North Am 1982; 29:898.

Gauthier B, Edelmann CM Jr, Barrett HL, eds. Management of tubular and related disorders. In: Nephrology and Urology for the Pediatrician. Boston: Little, Brown, 1982:287.

Norman ME. Renal tubular acidosis: differential diagnosis, proximal or distal. In: Schwartz AB, Lyons H, eds. Acid Base and Electrolyte Disorders. New York: Grune & Stratton, 1977.

Smulders YM, Frissen PH, Slaats EH, et al. Renal tubular acidosis: pathophysiology and diagnosis. Arch Intern Med 1996;156:1629–1636.

BARTTER'S SYNDROME

Donckerwolcke RA. Diagnosis and treatment of renal tubular disorders in children. Pediatr Clin North Am 1982;29:895.

Gauthier B, Edelmann CM Jr, Barrett HL, eds. Sodium chloride-resistant metabolic alkalosis. In: Nephrology and Urology for the Pediatrician. Boston: Little, Brown, 1982:59.

Gitelman HJ. Unsolved issues in the pathogenesis of Bartter's syndrome and its variants. Curr Opin Nephrol Hypertension 1994;3:471–474.

Stein J. The pathogenetic spectrum of Bartter's syndrome. Kidney Int 1985;28:85.

UROLITHIASIS

Garcia CD, Miller LA, Stapleton FB. Natural history of hematuria associated with hypercalciuria in children. Am J Dis Child 1991;145:1204–1207.

Lemann J. Composition of the diet and calcium kidney stones. N Engl J Med 1993;328:880–882.

Nakano M, Kacnor G, Forman JW, et al. Renal Calculi. In: Barakat AY, ed. Renal Disease in Children. Clinical Evaluation and Diagnosis. 1990: 341–355.

National Institutes of Health. Consensus Development Conference Statement. Prevention and Treatment of Kidney Stones. New York: Springer-Verlag, 1988;7:1–8.

Noe HN, Stapleton FB. Pediatric stone disease. In: Rous SN, ed. Stone Disease: Diagnosis and Management. Orlando, FL: Grune & Stratton, 1987:347–379.

Polinsky MS, Kaiser DI, Baluarte HJ. Urolithiasis in childhood. Pediatr Clin North Am 1987;34:687–711.

Stapleton FB. Nephroliathiasis in children. Pediatr Rev 1989:11:21–30.

CHRONIC RENAL FAILURE

Chantier C, Holliday M. Progressive loss of renal function. In: Holliday M, Barratt TM, Vernier RL, eds. Pediatric Nephrology. 2nd ed. Baltimore: Williams & Wilkins, 1987.

Evans E, Greenbaum L, Ettinger R. Principles of renal replacement therapy in children. Pediatr Clin North Am 1995;42(6):1579–1602.

Foreman JW, Chan JCM. Chronic renal failure in infants and children. J Pediatr 1988;113:793.

SchöAdarer K, Miller-Wiefel DE. Hematologic complications of renal failure. In: Holliday M, Barratt TM, Vernier RL, eds. Pediatric Nephrology. 2nd ed. Baltimore: Williams & Wilkins, 1987.

SchöAdarer J, Ulmer H. Cardiovascular complications of renal failure. In: Holliday M, Barratt TM, Vernier RL, eds. Pediatric Nephrology. 2nd ed. Baltimore: Williams & Wilkins, 1987.

Trompeter RSL. Neurologic problems of renal failure. In: Holliday M, Barratt TM, Vernier RL, eds. Pediatric Nephrology. 2nd ed. Baltimore: Williams & Wilkins, 1987.

RHABDOMYOLYSIS

Chamberlain MC. Rhabdomyolysis in children: a 3-year retrospective study. Pediatr Neurol 1991;7:226–228.

Moghtader J, Brady WJ, Bonadio W. Exertional rhabdomyolysis in an adolescent athlete. Pediatr Emerg Care 1997;13:382–385.

Schwengel D, Ludwig S. Rhabdomyolysis and myoglobinuria as manifestations of child abuse. Pediatr Emerg Care 1985;1:194–197.

CHAPTER 87

Hematologic Emergencies

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Hematologic emergencies arise in children who have been previously well, who have known blood diseases, or who have systemic diseases. Although the particular setting in which a serious blood abnormality occurs affects some facets of emergency care, the initial measures of support, diagnosis, and treatment are based on general principles that often cross the boundaries between the usual categories of blood disorders. This chapter emphasizes these principles as they apply to disorders of red cells, white cells, platelets, and coagulation. The initial evaluation and treatment of life-threatening disorders are described in detail. When controversy exists regarding specific management problems, alternative approaches are presented.

DISORDERS OF RED BLOOD CELLS

Severe anemia is a pediatric emergency that requires rapid evaluation and treatment to prevent hypoxia, congestive heart failure, and death. The classification of causes of anemia according to 1) blood loss, 2) increased red cell destruction, and 3) decreased red cell production is familiar to most physicians and provides an excellent starting point for the evaluation of the anemic child. In Chapter 59, these categories are used for the differential diagnosis of hematologic causes of pallor and for the appropriate selection of initial laboratory studies. In the section that follows, the same classification is applied to the emergency management of specific hematologic disorders.

Blood Loss

Trauma is the leading cause of major hemorrhage in children. Every emergency physician must be prepared to act quickly and systematically when confronted with an actively bleeding child. The initial approach often requires the joint effort of a team of doctors and nurses to accomplish numerous tasks simultaneously. Within the first few minutes, the nature of the accident, an estimate of blood loss, and the presence of major current or chronic illnesses, including bleeding disorders, should be determined. The adequacy of the airway must be ensured. Vital signs should be measured frequently to detect early signs of hypovolemic shock. All clothing should be removed, and the child should be examined for sites of bleeding other than those found on initial inspection. Intravenous (IV) access should be established, preferably in a large vein, and blood samples should be drawn for a complete blood count (CBC) (including platelet count), screening coagulation studies (prothrombin time and partial thromboplastin time), and the cross-matching of donor blood. A spun hematocrit should be measured immediately in the emergency department (ED) if a CBC cannot be obtained quickly. If bleeding is brisk and sustained or if there is any suggestion of hypovolemic shock, volume expanders should be infused. Both colloid preparations, such as 5% albumin, and crystalloids, such as saline or Ringer's lactate, are effective for the maintenance of intravascular volume, but the latter solutions may be more readily available and should be used initially.

After immediate stabilization has been completed and external hemorrhage has been slowed or stopped, the child should be evaluated for internal hemorrhage. This evaluation is especially important when the nature of the trauma is unclear or when multiple areas of the body may have been involved, as in automobile or bicycle accidents. The importance of

locating internal hemorrhage is underscored by the occurrence of hypovolemic shock and death in the child whose skin lacerations were sutured but whose ruptured spleen went undetected. Suspicion of internal bleeding should be raised in the presence of a continuously falling hematocrit or continuing signs of hypovolemic shock despite control of external bleeding and the replacement of seemingly adequate volume. Respiratory compromise, a protuberant abdomen, or changing sensorium may be further clues to the presence of internal hemorrhage. Further studies, including radiographs of the chest and abdomen, peritoneal lavage, and computed axial tomography (CAT) of the head or abdomen, should be instituted when appropriate.

Gastrointestinal (GI) bleeding and other forms of nontraumatic hemorrhage can also be life-threatening. In some cases, the severity of bleeding is accentuated by the combination of an anatomic lesion and a related bleeding disorder, such as esophageal varices with a coagulopathy caused by liver failure. Unexplained severe anemia requires a careful search for bleeding in the GI tract, retroperitoneal space, or elsewhere.

The approach to blood transfusion (Table 87.1) can be divided into three levels of intervention, depending on the clinical findings and the laboratory data:

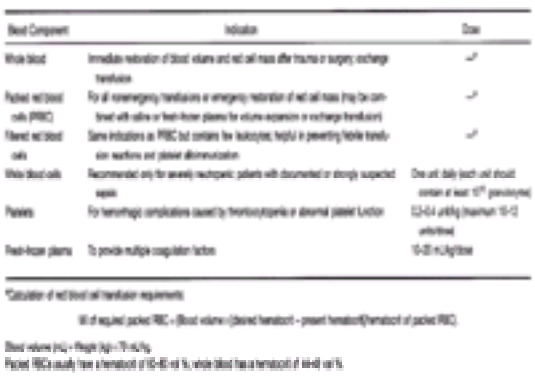


Table 87.1. Guidelines to Transfusion Therapy

1. If bleeding has been controlled, vital signs are stable, the hematocrit remains above 20%, and further bleeding is considered unlikely, the initially crossmatched blood should be held for at least 24 hours and then released for other use if no longer required for this patient.
2. If bleeding has led to hypovolemic shock but tissue oxygenation is not critically affected, intravascular volume should be supported with crystalloid or colloid solutions until a crossmatch has been performed and compatible donor blood is available. If necessary, group and Rh type-specific but non–cross-matched blood can be used. A similar approach should be used if the hematocrit slowly falls to a level less than 15 to 20% or if the hematocrit remains stable at a low level but further bleeding is considered likely (e.g., esophageal varices).
3. Only when bleeding is accompanied by life-threatening hypoxia should non–cross-matched group O, Rh-negative blood be administered. Transfusion of blood with minor blood group incompatibilities may result in immediate hemolysis and renal failure or, more commonly, may result in sensitization of the recipient to red cell antigens, making future blood compatibility testing difficult. The determination of the patient's ABO or Rh blood group can be performed within a few minutes, so selection of ABO- and Rh-compatible donor units is almost always feasible.

A common pitfall in the assessment and treatment of the bleeding patient is the underestimation of the amount of blood loss. Neither the history of bleeding nor the initial hemoglobin level may accurately reflect the severity of hemorrhage. For example, a child with upper GI bleeding may have a modest amount of hematemesis or melena and a hemoglobin of 8 g/dL when initially evaluated in the ED. However, within an hour, the child may pass a large amount of tarry stool and the hemoglobin level may fall to 3 g/dL. Tachycardia or hypotension in a patient with only a moderate degree of anemia should serve as a warning that intravascular blood loss is out of proportion to the hemoglobin level and that early replenishment of intravascular volume is essential.

Increased Red Cell Destruction

Membrane Disorders

The anemia in disorders of the red cell membrane (hereditary spherocytosis, hereditary elliptocytosis, stomatocytosis, liver disease) is rarely severe enough to constitute a hematologic emergency. However, the hemoglobin level may fall even further when red cell destruction increases (hemolytic crisis) or red cell production slows (aplastic crisis). Hemolytic crises are usually associated with acute infections and are self-limiting. Most aplastic crises accompany parvovirus infection; anemia may be the only manifestation of the infectious process.

The hemoglobin level and reticulocyte count should be routinely checked when children with known disorders of the red cell membrane develop increasing jaundice or pallor associated with an infectious illness. The hemolytic crisis is characterized by worsening jaundice, falling hemoglobin level, and increasing reticulocyte count. In contrast, the aplastic crisis is associated with slowly increasing pallor, worsening anemia, and low or absent reticulocytes. In children whose underlying red cell membrane disorder is associated with brisk hemolysis (hemoglobin level less than 8 to 9 g/dL and reticulocytes greater than 5 to 7%), these crises may produce acute symptoms of anemia. If the hemoglobin level falls below 3 to 4 g/dL or if cardiovascular stability is threatened, red cell transfusions may be necessary. One unit or less of red cells is usually sufficient to support the patient until the hemolytic or aplastic crisis is over.

For some children, an aplastic crisis may be the first clinical manifestation of an undiagnosed membrane disorder or

other chronic hemolytic anemia. The low hemoglobin level and reticulocyte count may suggest a pure problem of red cell production such as transient erythroblastopenia of childhood or Diamond-Blackfan anemia. However, a careful history for features such as neonatal jaundice or splenectomy in other family members, a thorough physical examination to assess spleen size, and a review of the peripheral smear to look for spherocytes or other abnormalities may identify the underlying hemolytic anemia.

The need for transfusions in an aplastic crisis should be considered carefully because the relatively slow development of the anemia usually allows adequate time for compensatory physiologic responses to the anemia. An increase in cardiac output keeps the patient hemodynamically stable even at very low hemoglobin levels. Moreover, many patients with aplastic crises are already beginning to resume red cell production when the crises are recognized, and the reemergence of reticulocytes in the peripheral blood or the presence of mature erythrocyte precursors in the bone marrow often precludes the need for red cell transfusions.

Older children and adolescents with red cell membrane disorders may develop gallstones because of increased red cell destruction and bilirubin release. Cholelithiasis or cholecystitis in affected patients should be managed the same way as in patients without underlying hematologic disease (see Chapter 93).

Metabolic Abnormalities

Like the red cell membrane disorders, erythrocyte metabolic abnormalities usually do not cause severe anemia. However, episodes of acute and sometimes life-threatening hemolysis can occur in many variants of glucose-6-phosphate dehydrogenase (G6PD) deficiency, including the A $^{-}$ variant found in 10% of African-American boys, after exposure to drugs or chemicals (Table 87.2) or during an infectious illness. Ingestion of naphthalene-containing mothballs is the most common cause of severe hemolysis in American children with G6PD deficiency, and parents should be asked about the presence of mothballs as part of the evaluation of any child with an acute hemolytic anemia. The acute intravascular hemolysis of G6PD deficiency usually occurs within 1 to 3 days of oxidant exposure and is characterized by pallor, malaise, fever, scleral icterus, abdominal and back pain, and dark urine. The anemia is accompanied by an increased reticulocyte count, and diagnostic blister cells are present on the peripheral smear. Hematologic changes may be minimal or absent in the first 24 hours after ingestion. Careful monitoring of the patient should continue for at least another day. Treatment should include removal of the offending agent and fluid administration to prevent renal tubular damage. When hemolysis is severe, red cell transfusions may be required. However, if the diagnosis is uncertain, a pretransfusion blood sample should be saved for measurement of specific enzyme levels. Because enzyme levels are higher in younger red cells, the diagnosis of G6PD or other enzyme deficiencies may be obscured at the time of acute hemolysis and a high reticulocyte count.

Antimalarials (primaquine)
Sulfonamides (including sulfasalazine and trimethoprim-sulfamethoxazole)
Nalidixic acid and nitrofurantoin
Naphthalene (moth balls)
Fava beans
Aspirin (does not cause acute hemolysis with G6PD deficiency in African-Americans when used in therapeutic doses)

Table 87.2. Drugs and Substances Associated with Acute Hemolysis in Children with Glucose-6-Phosphate Dehydrogenase (G6PD) Deficiency

Aplastic crises may occur in more severe variants of G6PD deficiency and other red cell metabolic disorders such as pyruvate kinase deficiency that are associated with chronic hemolysis. Diagnosis and treatment of this complication are the same as described in the previous section regarding membrane disorders.

Autoimmune Hemolytic Anemia

Background

One of the most serious causes of severe anemia in children is autoimmune hemolytic anemia (AIHA). This antibody-mediated disorder occurs most commonly in young children. Affected erythrocytes are lysed intravascularly or removed prematurely from the circulation by macrophages of the reticuloendothelial system. AIHA may be associated with infections, drugs, inflammatory diseases, or malignancies, but a specific cause is rarely identified in pediatric patients.

Clinical Manifestations

Although this disorder may occasionally be indolent and may go undetected for days or weeks, AIHA is usually associated with the sudden onset of pallor, jaundice, and dark urine. The hemoglobin level may be as low as 1 to 2 g/dL at the time of diagnosis. When the anemia is this severe, the child may appear moribund and desperately ill. Signs of congestive heart failure may be prominent.

The anemia is usually accompanied by reticulocytosis, although the reticulocyte count may be below 5% during the first

few days of the illness. Occasionally, patients remain reticulocytopenic for prolonged periods. Spherocytes are often found on the peripheral smear, and red cell fragments may sometimes be present. Free hemoglobin in the urine produces a positive dipstick reaction for blood in the absence of red cells on microscopic urinalysis. When hemolysis is severe enough to exceed the renal clearance of hemoglobin, the plasma will be pink, and careful inspection of the plasma layer of a spun hematocrit may provide an early diagnostic clue. The direct Coombs test using broad-spectrum Coombs serum (IgG, IgM, and complement) is usually positive in childhood AIHA. Acute hemolysis is most commonly associated with IgG antibody and/or complement but may also occur with IgM-mediated disease. Although the antibody may appear to have specificity in vitro (usually in the Rh system), the shortened survival of “compatible” blood suggests the presence of wider activity of the identified antibody or the presence of additional undetected antibodies in many cases. However, certain specific antibodies are associated with infectious causes of AIHA, such as *Mycoplasma* (anti-I) or infectious mononucleosis (anti-i).

Management

The management of the child with AIHA should be aggressive because the hemoglobin level may fall precipitously (Table 87.3). Hospitalization for careful observation and treatment is usually necessary. The immediate institution of corticosteroid therapy (prednisone 2 to 4 mg/kg per day or equivalent doses of parenteral preparations) may prevent or reduce the need for red cell transfusions. Alternatively, the patient may be treated with g-globulin 1 g/kg by IV infusion. For life-threatening AIHA, the use of steroids and g-globulin should be considered. Patients with cold-reacting antibodies (most IgM and some IgG antibodies) do not respond as favorably to steroids and g-globulin as those with warm-reacting antibodies (most IgG antibodies), but a trial of either therapy is still warranted in the severely anemic patient. The response to steroids or g-globulin in AIHA usually occurs within a few hours or days.

Maintain normal or increased urine output with intravenous (IV) fluids.
Immediately begin corticosteroid therapy with prednisone 2 mg/kg/day or a parenteral preparation in an equivalent dose. Alternatively, administer γ-globulin 1 g/kg by IV, alone or in combination with corticosteroid.
Administer red cell transfusions when severe anemia is accompanied by signs of hypoxia or cardiac failure.
Give first 5 mL in 10–15 min and observe for symptoms of acute hemolysis.
Check plasma layer of a spun hematocrit for pink color indicative of hemolysis of the transfused red cells.
If symptoms or signs of worsening hemolysis are present, try a different unit of red cells.
If hemoglobin level does not increase after transfusion:
Increase steroid dosage to 4 mg/kg/day; or
Administer IV γ-globulin 1 g/kg; or
Begin plasmapheresis or exchange transfusion; or
Perform splenectomy.

Table 87.3. Treatment of Severe Autoimmune Hemolytic Anemia

Red cell transfusions are hazardous in patients with AIHA and should be reserved for children with severe anemia and signs of hypoxia or cardiac failure. The presence of a nonspecific antibody in the patient's serum makes it difficult to find a unit of donor blood compatible in the major cross-match (donor cells and patient serum). The finding of an apparently compatible unit may pose even greater danger because the physician is lured into a false sense of confidence when, in fact, an undetected antibody may still cause a severe hemolytic transfusion reaction. The use of the “least incompatible” unit is a common practice, although data to support this approach are lacking. The best policy is to avoid transfusion when possible. If red cells are required and a compatible donor unit can be found, this unit should be used. Otherwise, ABO- and, if possible, Rh-compatible units should be administered despite the incompatibility in vitro. The recognition of the risks of transfusion in children with AIHA should not lead to the withholding of “incompatible” blood when transfusion therapy is required to prevent severe morbidity or death.

Whether the unit of red cells appears compatible or incompatible on the basis of serologic studies, special precautions should be taken during the actual transfusion. The first 5 mL should be administered in 10 to 15 minutes, and the patient should be observed closely for malaise, back pain, fever, and other signs of acute hemolysis. The plasma layer of the spun hematocrit should be carefully inspected for the pink color of free hemoglobin. If any of these findings is present, the transfusion should be stopped and normal saline should be administered until a new unit can be prepared. If the patient is asymptomatic and the plasma is clear, the remainder of the unit should be given with continuing close observation. Blood administered to patients with cold antibodies should be infused through a warmer.

In rare instances, the hemoglobin level continues to fall despite steroids, g-globulin, and red cell transfusion, necessitating alternative therapeutic attempts to sustain life. Plasmapheresis may remove sufficient antibody to reduce the destruction of the patient's erythrocytes and to allow improved survival of transfused red cells. If this measure fails, emergency splenectomy may be required.

Nonimmune Acquired Hemolytic Anemia

Acute hemolytic anemia in children may be caused by infections, chemicals, or drugs that damage the red cell directly. These disorders resemble AIHA in their clinical presentation and should be considered in the child with acquired hemolytic anemia and a negative Coombs test. Infectious agents that may induce hemolytic anemia include malaria (which is of particular importance in Southeast Asian and African immigrants), other protozoa, and a wide variety of Gram-positive and Gram-negative organisms. Chemicals and drugs that cause oxidative hemolysis include naphthalene, nitrofurantoin, and sulfasalazine. Treatment is directed at elimination of the offending agent. Red cell transfusions are usually unnecessary unless anemia is severe (hematocrit less than 15%) or accompanied by signs of cardiovascular compromise.

Erythrocyte Fragmentation Syndromes

Red cells undergo fragmentation and lysis when subjected to excessive physical trauma within the cardiovascular system. Hemolytic anemias as a result of red cell fragmentation have been associated with abnormalities of the heart (valve homografts and synthetic prostheses, uncorrected valvular disease), great vessels (coarctation of the aorta), and small vessels (hemolytic uremic syndrome, thrombotic thrombocytopenic purpura, collagen vascular disease, hemangiomas). Physical findings are related to the underlying disorder. The presence of red cell fragments on the peripheral smear strongly suggests mechanical damage to the erythrocyte. When small vessels are involved, thrombocytopenia may also be present. Hemolytic anemia associated with valvular or great vessel disease rarely causes severe anemia. However, iron deficiency as a result of intravascular lysis and urinary excretion of hemosiderin in renal tubular epithelial cells may aggravate the hemolytic anemia. Oral iron supplementation may obviate the need for transfusions. When hemolysis is a result of small vessel disease, treatment of the underlying disorder (e.g., collagen vascular disease) or primarily affected organs (e.g., renal failure in hemolytic uremic syndrome) is the first priority. Red cell transfusions should be reserved for the treatment of symptomatic anemia. Because the hemolysis is caused by extracorpuscular factors, survival of transfused cells may be markedly shortened. The management of intravascular coagulation associated with several of these disorders is discussed later in this chapter.

Decreased Red Cell Production

Disorders of red cell production, unless accompanied by shortened red cell survival, are characterized by a slowly progressive anemia. Consequently, the physician does not often encounter many of the difficulties associated with acute, life-threatening hemolysis or severe bleeding. However, the insidious onset of anemia when erythropoiesis is impaired may delay recognition of the disorder, and severe anemia and cardiac failure may be present at the time of diagnosis. Tissue oxygenation may be inadequate because of the low hemoglobin level, and conditions that increase the cardiac rate or output (fever, exercise) may precipitate congestive heart failure in the previously compensated patient. In addition, anemia secondary to diminished red cell production may be associated with an underlying, severe illness such as leukemia, neuroblastoma, or aplastic anemia in which other life-threatening hematologic abnormalities (severe neutropenia or thrombocytopenia) may be present. Thus, the patient with impaired production of erythrocytes may be as ill as the patient with acute hemolysis.

The important role of the history, physical examination, and laboratory studies in the initial evaluation of the child with decreased red cell production is described in Chapter 59. Initial management should include basic support of cardiorespiratory function and identification and treatment of conditions such as fever, which may be compounding the problems of severe anemia. The patient with hypoxia or cardiac failure requires red cell transfusions. As described earlier, the urgency of the clinical situation rarely dictates the need to abbreviate the standard cross-matching procedures. A pretransfusion anticoagulated blood sample and a serum sample should always be saved for further diagnostic studies as well as for the determination of the patient's red cell antigen profile should chronic transfusion therapy be necessary. The initial transfusion should be given as a small aliquot of packed red cells. In many instances, the symptoms of severe anemia will be relieved after the hemoglobin level has risen only 1 or 2 g/dL. The administration of additional blood is rarely necessary in the early stages of therapy. Furthermore, the added volume may precipitate cardiac failure in the face of a preexisting high output state. A helpful rule is to administer a number of milliliters per kilogram of packed red cells equivalent to the hemoglobin level. For example, in a child with aplastic anemia, a hemoglobin level of 3 g/dL and early signs of cardiovascular compromise would indicate that 3 mL/kg of packed red cells be given. Some physicians routinely administer diuretics (e.g., furosemide 1 mg/kg per dose) during the transfusion of a severely anemic patient. An alternative approach is to reserve diuretic therapy for those patient who develop signs of increasing cardiac compromise during the transfusion.

Aplastic and Hypoplastic Anemias

The differential diagnosis of aplastic and hypoplastic anemias is discussed in Chapter 59. Most of these disorders have a protracted course and, after initial stabilization of the patient, require intensive diagnostic evaluation and careful assessment of chronic therapy rather than emergency management. Transfusion should be used with particular caution in the initial management of patients with hypoplastic and aplastic anemias because exposure to human leukocyte antigen (HLA) and other antigens may adversely affect engraftment of transplanted bone marrow in patients who might otherwise have benefited from this procedure. If transfusions are required for severe anemia (hemoglobin less than 3 to 4 g/dL) and signs of cardiac failure or poor oxygenation, the goal of treatment should be relief of symptoms, not restoration of a normal hemoglobin level. When possible, filtered red cells should be used to decrease exposure to donor white cells, which contribute significantly to refractoriness to platelet transfusions. Related family members should not be used as blood donors. If the patient is cytomegalovirus (CMV) seronegative, the use of filtered blood or blood from CMV-seronegative donors may decrease the likelihood of CMV-related complications if a bone marrow transplant is later performed.

For patients with a hypoplastic anemia suggestive of transient erythroblastopenia of childhood, a bone marrow aspirate may be helpful in predicting the course of the disease during the next few days and, in particular, the likelihood that red cell transfusions will be required later. For example, a patient with transient erythroblastopenia of childhood has a hemoglobin level of 4 g/dL and absent reticulocytes at the time of diagnosis. If examination of the bone marrow reveals only an occasional pronormoblast, a further decrease of the hemoglobin concentration should be anticipated and red cell transfusions will almost certainly be required. However, if the bone marrow aspirate shows numerous erythrocyte precursors progressing through all levels of red cell maturation, a peripheral reticulocytosis can be expected within 24 hours and red cell transfusions will be unnecessary (Fig. 87.1).

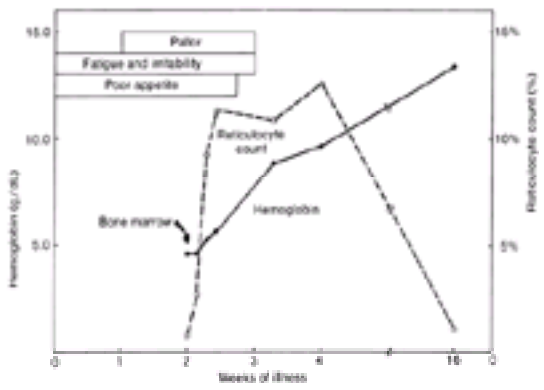


FIGURE 87.1. Clinical course of transient erythroblastopenia of childhood (TEC) in a 2-year-old girl. There was a 1- to 2-week history of pallor, fatigue, and decreased appetite. A bone marrow aspirate showed an active erythroid series that was reflected in the subsequent reticulocytosis and full hematologic recovery.

Nutritional Anemias

Nutritional anemias in children constitute more of a public health problem than a hematologic emergency. However, on occasion, the hemoglobin level may be very low at the time of diagnosis. Severe iron deficiency occurs mainly in 1- to 2-year-old children who drink 1 quart or more of cow's milk daily and have little room for other foods richer in iron. Adolescent girls make up another group at high risk for iron deficiency because a diet normally marginal in iron content becomes totally inadequate in the face of menstrual blood losses. The presenting complaint in severe iron deficiency anemia is usually pallor, lethargy, irritability, or poor exercise tolerance. In megaloblastic anemias such as vitamin B_{12} deficiency in an infant exclusively breast-fed by a vegetarian mother or in folic acid deficiency caused by impaired folate absorption, nonhematologic symptoms such as diarrhea, slowed development, or coma may be more prominent than the symptoms of anemia.

Stabilization and improvement can usually be achieved with replacement of the deficient nutrient. Nucleated red cells or reticulocytes usually appear within 48 hours of replacement therapy in folic acid or vitamin B_{12} deficiency and within 72 hours of therapy in severe iron deficiency anemia. Because of this rapid response, red cell transfusions are rarely required unless symptoms associated with the anemia pose a serious threat. A response to replacement therapy should not preclude further investigation of the origin of the anemia, especially when the dietary history is inconclusive. For example, iron deficiency anemia may result from repeated small pulmonary hemorrhages or chronic bleeding from an intestinal lesion rather than from inadequate iron intake. Similarly, megaloblastic anemias may be caused by deficient intrinsic factor or abnormalities of folic acid transport rather than from a seriously altered diet.

Iron replacement therapy consists of 3 to 6 mg/kg per day of elemental iron given orally as ferrous sulfate in two or three divided doses. Parenteral iron is painful and dangerous. Moreover, the hematologic response to intramuscular or IV iron dextran is no faster than the response to oral iron. Replacement doses of 1 mg of folic acid and 100 µg of vitamin B_{12} daily are undoubtedly excessive, but their common use reflects the safety and the concentrations of the available compounds.

The administration of supplemental iron, vitamin B_{12}, or folic acid should not be considered a substitute for adequate dietary intake when nutritional deficiency is recognized. Unlike most hematologic emergencies, the rapid improvement after treatment of these disorders may reduce the likelihood of further visits despite attempts to ensure adequate follow-up care. Therefore, a strong effort to restructure the diet should begin at the time of the initial contact.

DISORDERS OF HEMOGLOBIN STRUCTURE AND PRODUCTION

The disorders of hemoglobin structure and production that are most often encountered in a pediatric ED are the sickle hemoglobin syndromes (e.g., sickle cell anemia, hemoglobin sickle cell disease, hemoglobin S-b-thalassemia). Although many physicians are familiar with these diseases, the frequency with which affected patients are seen may lead to a false sense of complacency, leaving subtle findings undetected. Thalassemia major and methemoglobinemia occur much less commonly than the sickling disorders. Lack of familiarity with these diseases may delay recognition of serious illness, resulting in severe morbidity and even death. In the section that follows, particular attention is paid to the recognition of unusual but serious diseases and the management of the many and diverse complications associated with the hemoglobinopathies.

Sickle Hemoglobin Disorders

Background

The sickling disorders are responsible for a large percentage of hematologic emergencies and a major proportion of total visits for any reason in many urban pediatric EDs. Although the basic molecular lesion in these disorders is well defined, the mechanisms responsible for the numerous complications remain poorly understood and treatment is often unsatisfactory. Nonetheless, early recognition and aggressive management of specific problems may alleviate unnecessary suffering and prevent much of the morbidity and mortality associated with the sickling disorders. Optimal long-term care should be provided at a center with specialists who are familiar with sickle cell disease and its complications.

Clinical Manifestations/Management

Presentation

Newborn screening for sickling disorders is now common in the United States. However, newborns may occasionally elude testing or may rarely be misidentified as having sickle trait. In some instances, the information regarding the newborn screening is not available at the time of the ED visit.

It is important to identify the ill child with an undiagnosed or unrevealed sickling disorder so that appropriate therapy is instituted. The diagnosis of sickle cell disease should be considered in African-American children with unexplained pain or swelling (especially of the hands or feet), pneumonia, meningitis, sepsis, neurologic abnormalities, splenomegaly, or anemia. The hemoglobin level and reticulocyte count are inadequate screening tests for the sickle hemoglobinopathies because values in affected patients (especially those with hemoglobin sickle cell disease and S-b-thalassemia) may overlap with normal values. Similarly, the peripheral smear may be devoid of sickled cells. Definitive testing for sickling disorders can be accomplished quickly by hemoglobin electrophoresis, isoelectric focusing, or high-pressure liquid chromatography (HPLC). If these tests are not available, standard solubility tests can be used to identify the presence of sickle hemoglobin. However, solubility tests do not distinguish patients with sickle cell trait, hemoglobin sickle cell disease, or other sickle variants from patients with sickle cell anemia (hemoglobin SS). Therefore, the results of solubility screening tests must be considered in the context of the clinical presentation and other laboratory studies. In addition, whether the screening test is positive or negative, confirmatory testing by hemoglobin electrophoresis or another method is mandatory in all patients with hematologic or nonhematologic emergencies that may be related to sickle cell anemia.

Sepsis

A combination of impaired immunologic functions, including early loss of normal splenic activity, contributes to the significantly increased frequency of sepsis in patients with sickle cell disease and the fulminant nature of this complication. The risk of bacterial sepsis in the patient with sickle cell disease is increased several hundredfold in comparison with the normal population. *Streptococcus pneumoniae* and *Haemophilus influenzae* are the most common pathogens in young children, although *Escherichia coli* and *Salmonella* are more frequent causes of bacteremia in older children. The period of greatest risk is between the ages of 6 months and 3 years, when development of protective antibodies is limited and splenic function is diminished or absent.

The incidence of bacteremia in children with hemoglobin SS during the first 3 years of life is 7.98 per 100 patient-years. Approximately one-fifth of these children die. The comparable incidence rate for children with hemoglobin sickle cell disease in the first 3 years of life is 3.54 per 100 patient-years, although mortality is not as high as in patients with hemoglobin SS. Immunization with pneumococcal and *H. influenzae* vaccines and administration of prophylactic penicillin help prevent serious infections but certainly do not eliminate this complication of sickling disorders.

The common occurrence of fever with no obvious source in young children with sickle cell disease makes the distinction between serious bacterial infections and benign, self-limiting viral disorders a particularly frustrating problem. Unfortunately, no single physical finding or laboratory test (other than blood culture) can accurately identify the septic patient. The physician consequently must choose among several options including routine admission, prolonged observation, or outpatient management. No matter which option is selected, the goal is to be certain that all children with sickle cell disease and sepsis receive appropriate antimicrobial therapy. Thus, the cornerstone of management in the ED is rapid initiation of antibiotics after obtaining appropriate cultures. Differences in subsequent management should not detract from the importance of this early step.

The treatment of the very ill-appearing child with sickle cell disease and probable sepsis should include the rapid institution of antibiotic therapy and aggressive management of septic shock. As in other patients with reduced or absent splenic function, clinical deterioration may be extremely rapid. The patient who is alert on arrival to the ED may be moribund and hypotensive 30 minutes later. Because of the emergence of penicillin-resistant strains of *S. pneumoniae,* vancomycin may be added. Septic shock should be treated in the same way as in patients without hematologic disorders (see Chapter 3). Simple red cell transfusions or exchange transfusions may be needed to correct severe anemia or to reduce the likelihood of secondary organ damage caused by massive sickling in the presence of hypoxia, stasis, and acidosis.

In many centers, children with sickle cell disease and fever who do not appear to be seriously ill, but who nonetheless are at increased risk for sepsis, continue to be admitted to the hospital for at least 48 hours. Ampicillin or a third-generation cephalosporin is usually administered intravenously until the cultures are confirmed to be negative.

As an alternative to conventional inpatient management, other centers now treat selected children with sickle cell disease and fever as outpatients. This approach is usually restricted to children who do not appear acutely ill and who, on physical examination, do not have findings such as pallor, rales, or increased spleen size that indicate additional problems. Some centers employ additional criteria such as age, previous history, temperature, and white blood cell count. Young children are at higher risk of bacterial sepsis and may be more difficult to assess for early signs of sepsis than older children. A history of bacterial infection may be a risk factor for a subsequent episode. Temperatures greater than 39 to 40 degrees Centigrade have been associated with an increased likelihood of sepsis in some studies of children with sickle cell disease, although lesser degrees of elevation do not guarantee negative blood cultures.

Those children who do not appear to be seriously ill and who, on the basis of physical findings and results of laboratory tests, are judged to be at low risk for bacteremia, are treated in the ED with a long-acting cephalosporin such as ceftriaxone (75 mg/kg) and then discharged. A compromise between outpatient therapy and inpatient care is the use of a short-stay unit. With this approach, the patient receives a long-acting cephalosporin and is observed for 4 to 24 hours.

Newer culture systems detect most cases of bacteremia with *S. pneumoniae* within this time frame. Further therapy after discharge from the ED or short-stay unit varies among centers but may include 1 to 3 days of an oral antibiotic such as amoxicillin. A key component of the outpatient management of children with sickle cell disease and fever is a return visit or telephone report within 24 hours after discharge from the ED or short-stay unit. Inpatient care should be strongly considered for children whose families are unlikely to comply with this followup.

Because unexplained fever is uncommon in older children in general, the diagnosis of bacterial sepsis should be strongly considered in the older child with sickle cell anemia and fever. A careful assessment of the child's clinical condition should take into account the factors noted earlier. If the child appears toxic, admission for antibiotic treatment is advisable even in the absence of high fever or leukocytosis. Once again, good follow-up care must be ensured if the patient is managed as an outpatient.

Other Infections

Children with sickle cell disease are affected more often with infections other than sepsis in comparison with their hematologically normal counterparts. Meningitis, pneumonia, septic arthritis, and osteomyelitis may be responsible for substantial morbidity and mortality unless promptly recognized and appropriately treated. The level of suspicion for meningitis should be particularly high in the young, irritable child with sickle cell disease and unexplained fever. Antibiotic therapy of meningitis is similar to that recommended for hematologically normal children with this disorder (see Chapter 84). In particular, the possibility of resistant *S. pneumoniae* should guide treatment. Exchange transfusion to lower the percentage of sickle hemoglobin may reduce the risk of intracerebral sickling and infarction in areas of local swelling and possible red cell sludging. This procedure may also help resolve the conflict between the need for maintenance or greater fluid therapy to prevent vaso-occlusion and the need to restrict fluids in the face of cerebral swelling and possible inappropriate antidiuretic hormone secretion. When hemoglobin S is less than 30% of the total hemoglobin, sickling is unlikely and fluid management can be dictated by the central nervous system findings.

Acute chest syndrome, which includes pneumonia as well as pulmonary infarction, is one of the most common reasons for hospital admission for children with sickle cell anemia. The affected patient is usually tachypneic, even after antipyretic therapy. Rales, rhonchi, and physical findings of lobar consolidation may be present. However, in some children, particularly those who are somewhat dehydrated, physical findings may be far less striking. Rales may be heard only after several hours of rehydration. Because acute chest syndrome may escape detection on physical examination, a chest radiograph should be obtained in children with sickle cell disease and unexplained fever or chest pain. A decrease in oxygen saturation, readily measured in the ED and compared with baseline values, may identify patients with early acute chest syndrome.

The problem of identifying a responsible pathogen in patients with sickling disorders and acute chest syndrome is similar to that encountered in hematologically normal children with pneumonia (see Chapter 84). Although pneumonia caused by *Mycoplasma pneumoniae, S. pneumoniae, Chlamydia trachomatis,* and Gram-negative organisms is more common in sickle cell disease, a causative organism is rarely identified in cultures of the blood and sputum or in counterimmunoelectrophoresis or latex agglutination studies of the blood and urine. The initial white count and differential are usually not helpful in distinguishing patients with bacterial pneumonia from those with viral pneumonia or pulmonary infarction. The hemoglobin level is more likely to fall and the fever is more likely to persist during the course of bacterial pneumonia, but this information is not yet known, of course, when the patient is first seen in the ED.

Because a responsible organism for acute chest syndrome is nearly never known at the outset, treatment is begun with IV-administered ampicillin or a third generation of cephalosporin and modified according to the clinical response. In the very ill child, the identification of the causative organism should be pursued more vigorously with tracheal aspirate, aspiration of pleural fluid when present, or aspiration of lung tissue. Initial therapy of the child with severe acute chest syndrome should also include erythromycin. Oxygen should be administered to children with acute chest syndrome who have evidence of respiratory distress or hypoxia. Red cell transfusions or exchange transfusion should be used very early in the course when the patient is severely anemic (e.g., hemoglobin less than 5 g/dL), is hypoxic, or has radiologic or other evidence of severe or rapidly progressive disease.

Septic arthritis and osteomyelitis present particularly difficult diagnostic problems in children with sickle cell disease because the clinical findings so closely resemble those found in infarctions of the bone. A careful physical examination and judicious use of laboratory tests help the physician weigh the relative likelihood of infection and infarction. If symptoms are of recent onset (less than 3 days), a ^{99m}Tc-diphosphonate bone scan in conjunction with a ^{99m}Tc-sulfur colloid bone marrow scan may be helpful in distinguishing the two processes. In osteomyelitis, the bone scan shows increased uptake and the bone marrow scan is normal. In bone infarction, the bone scan is normal but the bone marrow scan shows decreased uptake. Magnetic resonance imaging (MRI), gallium scans, and radiolabeled white cell scans may also be of diagnostic help, but the value of these techniques in distinguishing bone infection from infarction remains to be proven. Closed or open bone aspiration should precede the institution of antibiotic therapy in the patient with suspected osteomyelitis. Similarly, aspiration of an affected joint should be performed if septic arthritis is strongly suspected. In most instances, swollen, warm, and tender joints are caused by local infarction. The presence of other sites of concurrent infarction and the patient's description of the pain as typical “crisis pain” may be helpful in identifying the cause as vaso-occlusion. The total white cell count and differential count of the joint fluid may be similar in both septic arthritis and sterile effusion secondary to infarction. Therefore, the Gram stain and culture are especially important. Septic arthritis of the hip deserves special mention because delayed intervention may result in necrosis of the femoral head. Children with this complication usually appear quite ill and hold the limb in a “frog-leg” position. Confirmation of septic arthritis by joint aspirate should be followed as soon as possible by surgical decompression.

Vaso-Occlusion

Infarction of bone, soft tissue, and viscera may occur as a result of intravascular sickling and vessel occlusion. Physiologic or environmental factors that initiate the process of vaso-occlusion and pain are rarely identified, although

swimming in cold water may be one important cause of painful crisis. Children may have only pain or may have symptoms related to the affected organ (e.g., hematuria in papillary necrosis, jaundice in hepatic infarct, seizures or weakness in central nervous system ischemia, respiratory distress in pulmonary infarction). Initial management usually centers around control of pain, general supportive measures, and differentiation of vaso-occlusion and disorders unrelated to the hematologic abnormality.

The treatment of the child with a painful crisis requires an objective assessment of the severity of the discomfort and an appropriate use of analgesic therapy (Table 87.4). Once nonsickling disorders have been ruled out, hydration should be undertaken with D5¼ normal saline solution (NSS) or D5½ NSS at a rate of 1.5 maintenance fluid requirements (see Chapter 18 and Chapter 84). The choice of analgesic is aided by familiarity with the patient's previous crises. Hesitancy to use parenteral narcotics may result in inadequate pain relief, mounting anxiety, and a loss of trust between physician and patient. This is a particularly common occurrence when the patient has had repeated visits to the ED and physicians are suspicious of the stated degree of discomfort. The use of the placebo saline injection is to be decried as an insensitive and potentially cruel test of the patient's pain level. For moderate or severe vaso-occlusive pain, morphine sulfate (0.10 to 0.15 mg/kg) should be administered by IV, and further therapy should be based on the degree of pain and the duration of pain control. Admission to the hospital is necessary if continuing parenteral analgesic therapy is required, fluid intake is inadequate, or the child has had several visits for the same problem. Repeated prolonged stays in the ED often leave the child and family exhausted and rarely prevent hospital admission.

Mild or Moderate Pain
Hydration—1½ × maintenance with oral fluids or IV D5¼ normal saline solution (NSS) or D5½ NSS
Analgesia—Acetaminophen with or without codeine
Disposition—Admit if pain worsens, oral fluid intake is inadequate, or repeat visits to the emergency department have occurred
Severe pain
Hydration—1½ × maintenance with IV D5¼ NSS or D5½ NSS
Analgesia—Morphine sulfate, 0.10–0.15 mg/kg IV
Disposition—Admit unless pain is markedly reduced and patient can take oral fluids

Table 87.4. Management of Vaso-Occlusive Crisis in Sickle Cell Crisis

Several specific areas of vaso-occlusion deserve special attention. Between 6 and 24 months of age, dactylitis is a common manifestation of sickle cell disease. Infarction of the metarcarpals and metatarsals results in swelling of the hands and feet. These episodes recur frequently. Pain usually resolves after several days, but swelling may persist for 1 or 2 weeks. Treatment is similar to that described for a painful crisis.

Because it is difficult to distinguish pulmonary infarction from pneumonia on the basis of physical findings and noninvasive laboratory studies, these two disorders are classified together under the heading of acute chest syndrome. Pulmonary infarction may be associated with severe respiratory distress and substantial morbidity in children with sickle cell disease. The patient's clinical condition should be monitored closely. Pulse oximetry may help determine the need for arterial blood gas measurement and additional therapy. Treatment of acute chest syndrome is described on page 867.

Infarction of abdominal and retroperitoneal organs may produce clinical findings that closely resemble the findings in a variety of nonhematologic diseases. The distinction between occlusion of the mesenteric vessels and appendicitis or other causes of an acute abdomen is, at times, particularly difficult. Physical findings and laboratory studies are remarkably similar. The onset and quality of the pain may be familiar to the patient and readily recognized as typical “crisis pain.” The patient may describe the symptoms as distinctly different from episodes of infarction, however, giving support to the diagnosis of an acute abdomen. Because painful crises occur far more often than appendicitis and other causes of acute abdomen, a period of careful observation is warranted unless the patient is severely ill (e.g., perforated appendix). Repeated assessment of the abdominal examination and the clinical response to fluid therapy help identify the child with an acute abdomen and reduces unnecessary and risky emergency surgical procedures in children with sickle cell disease. The hours required for transfusion before surgery provide an additional period of observation, during which time symptoms may abate.

Hepatic infarction may also create a diagnostic dilemma because the acute onset of jaundice and abdominal pain that characterize this disorder are similar to the symptoms of hepatitis, cholecystitis, and biliary obstruction. In addition, vaso-occlusion elsewhere in the abdomen that causes right upper quadrant pain may mimic biliary tract disease. The distinction between infarction and cholecystitis or biliary obstruction is particularly important because recurrent gallbladder disease is an indication for cholecystectomy. In both hepatic infarction and biliary obstruction, the alanine aminotransferase (ALT) and direct bilirubin levels may be increased. Ultrasonography of the abdomen often shows a dilated common bile duct or the presence of stones in the duct in children with biliary tract disease when the study is performed shortly after the onset of symptoms. In many instances, however, biliary tract disease and vaso-occlusion cannot be definitively distinguished, and the clinician must depend on a pattern of recurrence for additional information. The initial management of these disorders is similar to that described for vaso-occlusive crises (i.e., fluids, analgesics). A nasogastric tube may relieve abdominal discomfort caused by distension.

The major emergencies related to vaso-occlusion within the genitourinary tract are hematuria that results from renal papillary necrosis and priapism. The hematuria of papillary necrosis is usually painless and often persistent. A history of recent trauma, streptococcal infection, or recurrent urinary tract infection should alert the physician to other causes of hematuria. Similarly, hypertension suggests the presence of nephritis rather than simple vaso-occlusion. In papillary

necrosis, microscopic examination of the urine shows numerous red cells, but red cell casts are rarely seen. Pyuria and proteinuria in excess of what might be attributed to the blood in the urine are not found in papillary necrosis but may indicate nephritis. The hematocrit or hemoglobin level should be measured because the hematuria, if persistent or severe, may markedly worsen the chronic anemia. For the patient who is otherwise well, diagnostic studies can be accomplished on an outpatient basis, and a trial of increased oral fluids (twice maintenance) should be undertaken. In many instances, however, admission to the hospital is required for IV hydration. Alkalinization of the urine may reduce bleeding but is difficult to accomplish and usually unnecessary. Administration of antifibrinolytic drugs such as epsilon aminocaproic acid (Amicar) (100 mg/ kg every 6 hours) or tranexamic acid (25 mg/kg every 6 to 8 hours) may stop bleeding but carries a risk of ureteral clot formation. When hematuria is severe, red cell transfusions are sometimes required for treatment of anemia. Transfusions or exchange transfusions may also be useful in shortening the course of hematuria.

Priapism is an unusually painful and frightening form of vaso-occlusion. The penis becomes swollen, edematous, and very tender. Urination may be difficult. The initial treatment consists of fluid therapy and analgesics (Table 87.5). Once again, red cell transfusions or exchange transfusion may promote resolution, but these forms of therapy should be reserved for patients without a rapid response to other measures. An increased risk of complications of the central nervous system, including stroke, has been associated with exchange transfusion for priapism. Early aspiration of the corpora has been recommended to abort the course of priapism and preserve later potency. The relationship between duration of priapism and later potency in boys with sickle cell disease is still unclear, adding to the uncertainty of when to use particular therapies. However, the trend is toward more aggressive treatment to promote earlier resolution.

Hospitalize if erection persists or if pain is severe.
Intravenous hydration with D5¼ normal saline solution (NSS) or D5½ NSS at 1½–2 x maintenance for 24–48 hr.
Consider aspiration of the corpora.
If swelling does not decrease, transfuse with red cells to raise hemoglobin level to 9–10 g/dL.
If no improvement after simple transfusion, institute exchange transfusion to reduce HbS to less than 30% of total hemoglobin.
Reserve shunting procedures for patients who have failed other forms of therapy in the first 72 hr.

Table 87.5. Management of Priapism in Sickle Cell Anemia

Infarction of the central nervous system is a catastrophic form of vaso-occlusion. The initial presentation varies from the mild and fleeting symptoms of a transient ischemic attack to seizures, hemiparesis, coma, and death. Physical findings usually define, and MRI usually confirms, the area of cortical infarction. Supportive therapy should be instituted immediately (Table 87.6). A 1.5 or 2 volume exchange transfusion should begin as soon as the blood is ready. This procedure reduces the likelihood of further intravascular sickling and may prevent extension of cortical damage.

1. Obtain computed tomography scan or magnetic resonance imaging to identify an area of infarction or to rule out a ruptured cerebral aneurysm or other intracranial bleed.
2. Immediately begin 1½–2 volume exchange transfusion to reduce HbS below 30% of total hemoglobin.
a. Use whole blood less than 3–5 days old or use packed red cells less than 3–5 days old reconstituted with fresh-frozen plasma.
3. Reserve pretransfusion blood sample for characterization of red cell antigens in preparation for chronic transfusion program.

Table 87.6. Management of Stroke in Sickle Cell Anemia

Cerebral aneurysms occur with increased frequency in patients with sickle cell disease. The origin of this complication, which is usually detected in teenagers or adults, remains obscure but may be related to local vessel occlusion or ischemia. Unfortunately, the aneurysm often escapes detection until after major, and often fatal, subarachnoid or intracerebral bleeding. The severe morbidity and high mortality associated with ruptured cerebral aneurysms require careful evaluation of the patient with sickle cell disease and headaches or neurologic findings. If the aneurysm is accessible and bleeding persists, surgical intervention should follow radiologic confirmation.

Less common areas of infarction that may be particularly confusing to the physician include the orbits and cervical spine. Vaso-occlusion that involves the bones of the orbit may produce findings similar to those of orbital cellulitis. Tenderness and fever may be present in bony infarction but are sometimes less remarkable than when found in cellulitis. A bone marrow scan using ^{99m}Tc-sulfur colloid may demonstrate decreased uptake in the affected area, confirming vaso-occlusion and avoiding the need for prolonged antibiotic therapy. Vaso-occlusion that presumably occurs in the cervical spine may cause meningismus. A lumbar puncture is sometimes necessary to rule out meningitis. As in other situations, the patient's evaluation of the pain may be helpful in distinguishing vaso-occlusion processes from more

serious disorders.

Splenic Sequestration Crisis

The sudden enlargement of the spleen with resulting sequestration of a substantial portion of the blood volume is a life-threatening complication of sickle cell disease. Because this crisis requires the presence of vascularized splenic tissue, it usually occurs before 5 years of age in patients with homozygous sickle cell disease but may occur much later in children with milder sickling disorders such as sickle cell disease or S-b $^{+}$-thalassemia. The patient undergoing a severe sequestration crisis may first complain of left upper quadrant pain (Table 87.7 and Table 87.8). Within hours, the patient becomes very pale, lethargic, and disoriented, and appears ill. The physical examination shows evidence of cardiovascular collapse; hypotension and tachycardia are often present. The level of consciousness falls. The hallmark of a severe sequestration crisis is a spleen that is significantly enlarged in comparison with previous examinations and is unusually hard. The hematocrit or hemoglobin level is much lower than during routine visits, and the reticulocyte count is usually increased (Fig. 87.2). Mild neutropenia or thrombocytopenia may be present.

Symptoms	Laboratory Findings
Left upper quadrant pain	Severe anemia
Pallor	Increased reticulocytes
Lethargy	Mild to moderate thrombocytopenia and neutropenia
Signs	
Hypotension	Management
Tachycardia	Immediate volume replacement
Markedly enlarged and firm spleen	Transfusion with packed red cells or whole blood

Table 87.7. Splenic Sequestration Crisis

	Sequestration Crisis	Aplastic Crisis	Hemolytic Crisis
Onset	Sudden	Gradual	Sudden
Pallor	Present	Present	Present
Jaundice	Normal	Normal	Increased
Abdominal pain	Present	Absent	Absent
Hemoglobin level	Very low	Low or very low	Low
Reticulocytes	Unchanged or increased	Decreased	Increased
Marrow erythroid activity	Unchanged or increased	Decreased	Increased

Table 87.8. Comparison of Findings in Sequestration, Aplastic, and Hemolytic Crises in Sickle Cell Disease

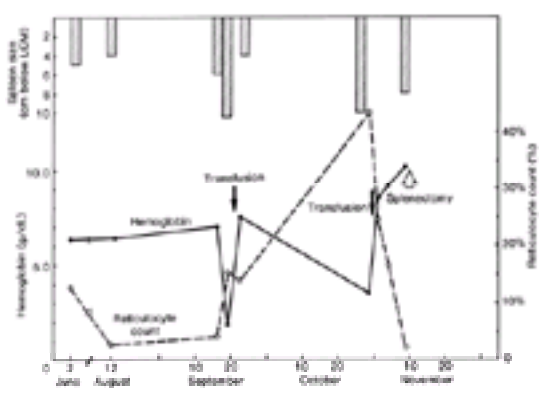


FIGURE 87.2. Clinical course of a 6-year-old girl with hemoglobin S-b°-thalassemia and two splenic sequestration crises that were characterized by abdominal pain, increased splenic size, and a rapid fall in hemoglobin concentration.

Recognition of this complication should be immediate so that lifesaving therapy begins without delay. The rapid infusion of large amounts of normal saline or albumin is necessary to restore intravascular volume. Although a sufficient number of red cells to relieve tissue hypoxia may be released by the spleen after initial fluid resuscitation, transfusion with packed red cells (2 to 10 mL/kg) is often required in more severe cases. Whole blood transfusion may help relieve the dual problems of intravascular volume depletion and impaired tissue oxygenation. Reversal of shock and a rising hematocrit signal improvement of a sequestration crisis. The spleen gradually becomes less firm and smaller.

Aplastic Crisis

Increased bone marrow erythroid activity (as reflected by the elevated reticulocyte count and presence of nucleated red

cells in the peripheral blood) partially compensates for the shortened red cell survival in sickle cell anemia and other hemolytic disorders. If erythropoiesis slows or ceases, this precarious balance is disturbed, and the hemoglobin level may gradually fall (Table 87.8). The event that most commonly causes erythroid aplasia is a parvovirus infection. Progressive pallor is unaccompanied by jaundice or other signs of hemolysis. Severe anemia may result in dyspnea and changes in level of consciousness. The hemoglobin level is unusually low, and reticulocytes are decreased or absent (Fig. 87.3). In the early phase of an aplastic crisis, the bone marrow has a paucity of erythroid activity. During the recovery stages, erythroid activity increases and the cells steadily mature. The level of red cell maturity can be used to predict the appearance of reticulocytosis in the peripheral blood, and this information may be helpful in determining whether the patient needs a red cell transfusion or can await the recovery of the bone marrow without further compromising oxygen delivery. If a red cell transfusion is required, a small aliquot is usually sufficient to raise the hemoglobin concentration to a level that ensures adequate oxygenation until red cell production recovers.

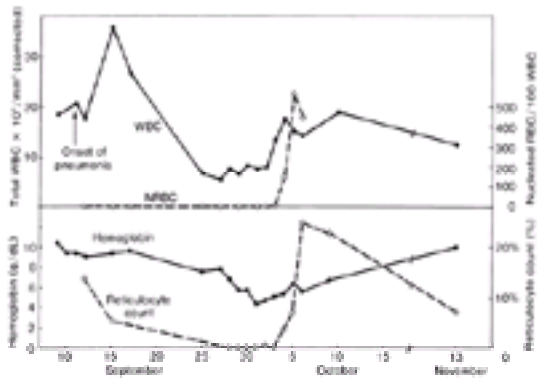


FIGURE 87.3. Aplastic crisis in a 5-year-old girl with sickle cell anemia and *Mycoplasma pneumoniae* pneumonia. Her aplastic crisis was characterized by a low hemoglobin level and reticulocytopenia. The white blood cell count was also transiently decreased. Recovery was characterized by reticulocytosis, marked increased in the number of nucleated red blood cells, and a rise in hemoglobin level.

Hemolytic Crisis

Worsening anemia and increasing reticulocytosis may accompany viral and bacterial infections in children with sickle cell disease (Table 87.8). Scleral icterus is more prominent than usual. The findings are consistent with an increasing degree of active hemolysis. The hemoglobin level rarely falls low enough to require specific therapy. Hematologic values return to the usual level as the infection process resolves.

Comment

The long-term management of many of the complications of sickle cell disease is beyond the scope of this chapter but has been discussed in detail in review articles and textbooks. In most instances, a thorough understanding of the extended care is necessary for correct management of the initial stages of hematologic emergencies. The clinical course and previous laboratory data of the patient should be familiar to someone involved in the care of the acute problem. Therefore, the treatment of hematologic emergencies in children with sickle cell disease is accomplished best in a center that also provides comprehensive care to affected patients.

Thalassemia Major (Cooley's Anemia)

Background

The thalassemias are disorders characterized by an inability to synthesize sufficient amounts of the globin component of hemoglobin. In b-thalassemia major, the most common of the homozygous thalassemia syndromes, the affected child produces little or no hemoglobin A and is usually transfusion-dependent from early childhood. The b-thalassemia gene occurs commonly in countries that border the Mediterranean Sea as well as in Southeast Asia. Other thalassemic disorders that may be associated with severe anemia include hemoglobin E-b°-thalassemia and hemoglobin H disease (absence of three of the four normal a-globin genes). Although most of the problems associated with these disorders are the result of long-term transfusion therapy, the severe anemia at the time of diagnosis may constitute a hematologic emergency.

Clinical Manifestations

Children with thalassemia major usually develop a sallow complexion and increasing fatigue between the ages of 6 and 24 months. Weight gain and linear growth may be retarded. Physical examination shows pallor and enlargement of the liver and spleen. The hemoglobin level may be as low as 3 or 4 g/dL, and the mean corpuscular volume (MCV) is usually low. The red cells are hypochromic and microcytic with striking variation in size and shape; nucleated red cells are present in the peripheral smear. Thalassemia major is readily distinguishable from severe nutritional iron deficiency. In the latter disorder, the dietary history is grossly abnormal, organomegaly is uncommon, changes in red cell morphology are less impressive, and nucleated red cells are rarely seen in the peripheral smear. The diagnosis of thalassemia major should be considered in a child with severe microcytic anemia and an appropriate ethnic background. Although severe anemia is extremely rare in heterozygous thalassemia disorders, thalassemia trait and concomitant iron deficiency may be particularly difficult to distinguish from a homozygous thalassemia disorder.

Management

The moderate anemia usually apparent at presentation allows sufficient time for a careful diagnostic evaluation and outpatient transfusion therapy. However, when anemia is severe and congestive heart failure is present or imminent, the need for red cell transfusion may be urgent. In such instances, pretransfusion blood should be saved for appropriate diagnostic studies (hemoglobin electrophoresis) and initial red cell antigen typing. If transfusion is necessary, small aliquots of red cells (2 to 3 mL/kg) should be given. The administration of a rapid-acting diuretic (furosemide 1 mg/kg per dose) may diminish the risk of fluid overload. Partial exchange transfusion has also been recommended for patients with severe anemia to prevent further increases in intravascular volume and myocardial stress. Because patients with thalassemia major and severe anemia invariably have a lifelong dependence on red cell transfusions, the use of non–cross-matched blood should be scrupulously avoided at the time of presentation to prevent sensitization to foreign red cell antigens.

Methemoglobinemia

Background

Methemoglobinemia is an uncommon cause of cyanosis in infants and children but is capable of causing severe problems and even death. Cyanosis results from a disproportionate amount of heme iron being present in the ferric rather than ferrous state. Under these conditions, oxygen binding of hemoglobin is severely impaired. The diagnosis of methemoglobinemia should be considered when cyanosis occurs in the absence of demonstrable cardiac or pulmonary disease.

The disturbance in the usual balance between ferrous and ferric iron may be a result of alterations of hemoglobin structure (hemoglobin M), abnormalities of red cell enzymes (methemoglobin reductase), or exposure to oxidant drugs or chemicals (Table 87.9). Infants are particularly susceptible to acute methemoglobinemia because of the relative immaturity of the enzyme system required to maintain hemoglobin iron in a reduced state. Acute infectious illnesses such as gastroenteritis may cause symptomatic methemoglobinemia in infants. The inherited forms of methemoglobinemia may be characterized by chronic cyanosis. However, in the absence of a specific oxidant stress, further symptoms are uncommon and treatment is given primarily for cosmetic reasons. When acute methemoglobinemia results from an oxidant stress, oxygen delivery may be severely compromised and the patient becomes acutely ill. If the agent acts as a direct oxidant, the onset of symptoms is rapid. However, if methemoglobin formation is caused by a metabolite of the original compound or secondary alterations in red cell metabolism, symptoms may be delayed. For example, methemoglobinemia is seen 12 to 15 hours after exposure to nitrobenzene.

Drugs
Sulfonamide antibiotics
Quinones
Phenacetin
Benzocaine
Domestic and Environmental Substances
Foods containing nitrates or nitrites
Well water containing nitrates
Aniline dyes (certain marking inks, dyes for some clothing and shoes, some crayons)
Naphthalene (mothballs)
Soap enemas
Certain industrial compounds (nitrobenzenes, nitrous gases, organic amines)

Table 87.9. Substances and Drugs Implicated in the Formation of Methemoglobin in Children

Clinical Manifestations

Symptoms depend on the concentration of methemoglobin (Table 87.10). When methemoglobin constitutes approximately 10 to 30% of total hemoglobin, only cyanosis occurs. As the level rises to 30 to 50%, dyspnea, tachycardia, dizziness, fatigue, and headache may be noted. Severe lethargy and stupor are often present when the methemoglobin concentration exceeds 50%, and death may occur at concentrations greater than 70%. If anemia is present, oxygen delivery is further compromised and toxicity may be more severe at lower concentrations of methemoglobin.

Methemoglobin Level	Symptoms
10–30%	Cyanosis
30–50%	Dyspnea, tachycardia, dizziness, fatigue, headache
50–70%	Lethargy, stupor
>70%	Death

Table 87.10. Symptoms and Signs According to Severity of Methemoglobinemia

Accurate diagnosis and rapid therapy prevent serious damage. The diagnosis should be strongly suspected when oxygen administration fails to affect the cyanosis. To eliminate an anatomic abnormality as a cause of oxygen-unresponsive cyanosis, an attempt should be made to oxygenate the patient's blood in vitro. As a rapid screening test, a drop of blood is placed on filter paper. After the filter paper is waved in the air for 30 to 60 seconds, normal blood appears bright red, whereas blood from a patient with methemoglobinemia remains reddish-brown. Arterial blood oxygen saturation is low when measured directly by blood oximetry rather than calculated, even though P_{O_2} is normal. Although blood oximetry measures oxyhemoglobin as a percent of total hemoglobin, including methemoglobin that is nonfunctional, pulse oximetry devices measure oxygen saturation of only that hemoglobin that is available for saturation. Thus, a patient with methemoglobinemia and obvious cyanosis may have normal oxygen saturation as measured by pulse oximetry. Spectrophotometric assays can be used for confirmation of methemoglobinemia as well as for determination of the level of methemoglobin.

Management

The treatment of methemoglobinemia depends on the clinical severity (Table 87.11). In all cases, an attempt should be made to identify an oxidant stress and, once identified, to remove the causative substance. If symptoms are mild after oxidant exposure, therapy is unnecessary. Red cells with normal metabolism will reduce the methemoglobin in several hours. If the symptoms are severe, 1 to 2 mg/kg of methylene blue as a 1% solution in saline should be infused over 5 minutes. A second dose can be given if symptoms are still present 1 hour later. Because methylene blue can act as an oxidant at high dosages, the total dosage should not exceed 7 mg/kg. Failure of methylene blue to improve the course of methemoglobinemia may be a result of concomitant G6PD deficiency because the therapeutic effect requires an intact hexose monophosphate shunt. For patients with G6PD deficiency, ascorbic acid (500 mg orally) may be of some value, but if symptoms are severe, exchange transfusion or hyperbaric oxygen may be required. Even if treatment with methylene blue or ascorbic acid in the ED is successful, any child with symptomatic methemoglobinemia should be admitted to the hospital for close observation and further evaluation of the underlying abnormality or causative agent.

Methemoglobin Level	Treatment
<30%	Not needed
30–70%	Methylene blue, 2 mg/kg of a 1% solution, infused intravenously over 5 min[a]
Severely ill and no response to methylene blue	Hyperbaric oxygen or exchange transfusion

[a]If no response to two doses of methylene blue in a noncritically ill patient or a patient with known G6PD deficiency, use ascorbic acid 500 mg orally.

Table 87.11. Treatment of Methemoglobinemia

DISORDERS OF WHITE BLOOD CELLS

Infection is the most significant complication associated with quantitative or qualitative white cell disorders. In some children, death may follow a single episode of acute, overwhelming sepsis. In others, repeated local infections may cause severe organ damage or may culminate in a fatal, disseminated fungal infection. The appropriate emergency management of the child with white cell abnormalities and fever or other signs of infection may have a profound impact on the length and quality of the patient's life.

Neutropenia

The most common forms of neutropenia and abnormal neutrophil function are listed in Table 87.12. Neutropenia is usually defined as an absolute neutrophil count below 1000 to 1500/mm^3. When the neutrophil count falls below 500/mm^3, the patient exhibits an increased susceptibility to infections caused by normal skin, respiratory, or GI flora. Between 500 and 1000/mm^3, susceptibility to infection is less significant but the host's ability to combat more typical infections is impaired. However, management of the patient cannot be based on the absolute neutrophil count alone because other, often unknown factors contribute to the severity of the clinical course. For example, serious, recurrent bacterial infections are common in Kostmann's neutropenia, and in the absence of treatment with growth factors, death often occurs in early childhood. In contrast, the clinical course of chronic benign neutropenia is much milder. Yet, the absolute neutrophil counts in the two disorders sometimes overlap. Similarly, serious infection is unusual in immune-mediated neutropenia, although the absolute neutrophil count may be less than 500/mm^3, a level at which severe morbidity and substantial mortality may be found in children with leukemia undergoing chemotherapy.

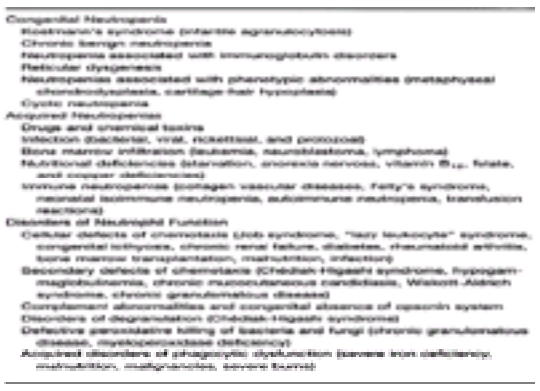


Table 87.12. Causes of Neutropenia and Disorders of Neutrophil Function in Children

The management of localized infection or unexplained fever in the child with neutropenia depends in large part on the underlying disorder and on the patient's history of infection. In neutropenic states associated with repeated, severe infections, an aggressive attempt to identify a causative organism should be undertaken. Blood and urine cultures, along with appropriate cultures from identified areas of infection (e.g., skin abscess, cellulitis), should be obtained. The cerebrospinal fluid should be examined and cultured when central nervous system infection is suspected. If the child appears ill or toxic, broad-spectrum IV antibiotic therapy should be instituted with modification of therapy when culture results are available. Initial treatment should include antibiotics effective against *Staphylococcus aureus* and other Gram-positive organisms as well as gram-negative bacteria, including *Pseudomonas aeruginosa.* If no source of fever is identified and the child appears well, observation in the hospital without antibiotic therapy may be considered.

Decisions regarding admission to the hospital and treatment are often more difficult in children with more benign neutropenic states. Although infections are usually mild and localized in these patients, severe infections rarely may occur. A white cell count and differential may be valuable because, in some children, the white count will rise to normal or near-normal levels during acute infection. Further laboratory investigation and treatment once again depend on the physical examination of the child as and the history of infection. In most instances, antibiotic therapy can be reserved for children with a specific source of bacterial infection. However, careful follow-up is required for untreated children in whom fever is unexplained or attributed to probable viral infection.

A particularly perplexing problem arises when a child is found to be neutropenic during an evaluation of fever. In most instances, both the fever and neutropenia are results of a viral illness. Under these circumstances, serious secondary bacterial infections are unlikely to occur, and admission to the hospital and antibiotic therapy are probably unnecessary. However, because the neutropenia usually cannot be attributed with certainty to a viral illness, other causes of neutropenia should be carefully sought. The patient or parents should be questioned about the use of drugs associated with neutropenia (e.g., penicillins, phenothiazines, phenytoin). The family history should be explored for recurrent infections or deaths in early childhood that might suggest a congenital neutropenia. Underlying disorders such as malignancies or nutritional disturbances should be considered. If the child appears even moderately ill, admission to the hospital for further evaluation is appropriate.

Disorders of Neutrophil Function

Numerous disorders of neutrophil function have been described. These disorders are associated with serious infections to a variable extent. Therefore, the evaluation and treatment of the patient with abnormal neutrophil function should be based on the specific cause and the history of serious infection. Particular attention should be paid to disorders such as chronic granulomatous disease in which the site of infection (liver, bones, GI tract) and causative organisms (*Aspergillus* species, *Pseudomonas cepacia, Serratia marcescens*) are distinctly unusual.

DISORDERS OF PLATELETS

The clinical course and management of patients with platelet abnormalities are determined primarily by the cause of the underlying disorder. For example, at the same level of thrombocytopenia, bleeding is more common in disorders of platelet production than in immune-mediated disorders of platelet survival. Consequently, the treatment of suspected bleeding after trauma should be more aggressive in the former disorder. The numerous causes of thrombocytopenia and abnormal platelet function are discussed in Chapter 65 and form an important background to the section that follows in which emphasis is placed on the management of bleeding emergencies in accordance with the underlying causes. The approach to a child with purpura or bleeding and no history of a bleeding disorder can also be found in Chapter 65. Finally, a discussion of the management of the many problems of lesser urgency that are associated with chronic platelet disorders is provided by textbooks of pediatric hematology.

Idiopathic Thrombocytopenic Purpura

Background

Idiopathic thrombocytopenic purpura (ITP) is the most commonly encountered platelet disorder in children. Serious bleeding is rare, occurring in only 2 to 4% of cases. This low incidence is particularly remarkable because the disease is most common between the ages of 1 and 4 years, when children are particularly prone to trauma as they learn to walk, run, and climb. The risk of serious bleeding decreases sharply after the first week of illness, reflecting the presence of newly formed platelets with greater hemostatic capability.

Clinical Manifestations

The diagnosis of ITP is made readily in the child with newly acquired petechiae and ecchymoses, thrombocytopenia, normal or increased megakaryocytes in the bone marrow, and the absence of any underlying disease. Epistaxis, gum bleeding, and hematuria occur less commonly than simple bruising and petechiae, but when persistent, these hemorrhagic manifestations can lead to moderate or even severe anemia. In teenage girls with ITP, heavy and prolonged menstrual bleeding can also cause a severe fall in the hemoglobin level. Fortunately, the development of anemia in children with ITP is gradual; acute, massive blood loss is extremely rare.

The major life-threatening complication of ITP is intracranial hemorrhage. This catastrophic problem may occur within a few days of diagnosis of the platelet disorder or months later. Although a history of head trauma in a child with ITP should alert the physician to the possibility of intracranial bleeding, the absence of any recognized injury is surprisingly common in patients with this complication. The symptoms of intracranial hemorrhage may be subtle, such as persistent mild headache, or they may be dramatic, such as severe headache, vomiting, and generalized or localized weakness. Intracranial bleeding in ITP is unusual once the platelet count has risen above 50,000/mm ^{3} unless a significant injury has occurred or platelet function is also impaired (e.g., as in the patient who has received aspirin).

Management

Controversy continues to surround the management of the patient with newly diagnosed ITP who has no serious bleeding. In many centers, such patients are treated with g-globulin at a dosage of 0.8 to 1.0 g/kg by IV infusion, with a second dose 24 hours later if the platelet count remains below 40,000 to 50,000/mm ^{3}. This therapy is effective in raising the platelet count in approximately 85% of patients with acute ITP. However, the low incidence of serious hemorrhagic complications of ITP, such as intracranial bleeding, has made it impossible to ascertain the overall benefits of this therapy. IV g-globulin exerts its major therapeutic effect by blocking the uptake of antibody-coated platelets by macrophages in the spleen. Unfortunately, one of the more common side effects of IV g-globulin is headache, and when this symptom persists despite slowing the rate of infusion, imaging studies of the brain may be necessary to investigate possible intracranial bleeding.

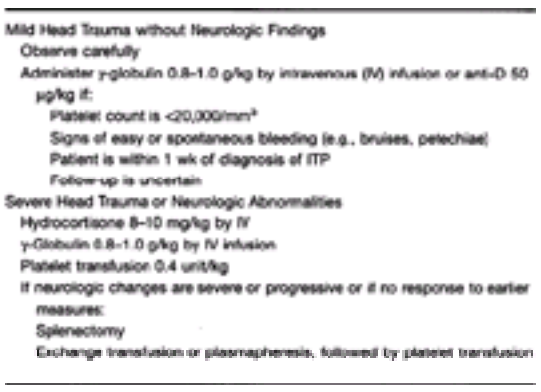
An alternative approach to the treatment of the stable patient with ITP is a 4- to 8-week course of prednisone, beginning with 2 mg/kg per day. Like g-globulin, steroids block the Fc receptors of splenic macrophages. Neither drug shortens the time until spontaneous recovery occurs. Some physicians have argued that a bone marrow aspirate to confirm the diagnosis of ITP is unnecessary in the patient with typical findings of the disorder and an absence of neutropenia or anemia. However, if steroids are used, a bone marrow aspirate should always be obtained before beginning therapy to be certain the diagnosis is absolutely correct. If a patient with acute leukemia is mistakenly diagnosed as having ITP and treated with steroids, the correct diagnosis may be delayed and long-term outcome may be affected adversely.

A more recent option for the treatment of acute ITP is the administration of antibody directed against the D-antigen of red cells. The antibody-coated erythrocytes are sacrificed to the reticuloendothelial system so that the antibody-coated platelets can continue to circulate. The effect of anti- D, usually given at a dosage of 50 µg/kg by IV infusion, is slightly delayed compared with g-globulin, and the peak platelet count may be somewhat lower. However, anti- D has the advantage of being administered over minutes rather than hours, and it rarely causes severe headache. Mild to moderate hemolysis may follow the administration of anti- D with a fall in hemoglobin level of 0.5 to 2.0 g/dL. This therapy is effective only in Rh-positive patients.

Not every physician considers it necessary to treat all patients newly diagnosed with ITP. The usually benign course of this disease must be weighed against the side effects of corticosteroids, g-globulin, and anti- D, and especially the high cost of the latter two drugs. Therefore, specific therapy with steroids, g-globulin, or anti- D may be reserved for patients with sufficient bleeding to cause moderate or severe anemia, for patients who remain severely thrombocytopenic (platelet count less than 10,000/mm^3) several weeks after diagnosis, or for patients whose physical activities cannot be effectively restricted.

For the patient with ITP and active bleeding, local therapeutic measures may be helpful until corticosteroids or infusions of g-globulin or anti- D raise the platelet count to a hemostatic level or when these drugs become ineffective. Nasal packing and topical phenylephrine are useful for persistent epistaxis. Excessive menstrual bleeding may require hormonal therapy. If bleeding does not stop despite these measures, plasmapheresis should be undertaken and, if necessary, followed by a transfusion of 0.2 unit/kg (maximum 10 to 12 units) of platelets (Table 87.2). The removal of antiplatelet antibody by plasmapheresis may increase the survival of transfused platelets sufficiently to stop or retard active bleeding.

Intracranial hemorrhage, the major cause of death in ITP, requires immediate recognition and therapy. The child with ITP who has sustained head trauma or developed signs of increased intracranial pressure (e.g., headache, vomiting, lethargy) or focal neurologic deficits should be evaluated immediately. The platelet count should be measured because intracranial hemorrhage is unusual in the child with ITP whose platelet count is above 50,000/mm ^{3}, unless trauma is exceptionally severe. The child with mild head trauma and no symptoms or signs of intracranial bleeding should be observed carefully. Whether to treat the asymptomatic child is a common and perplexing problem. Although no firm rules exist, management should be based on the duration of ITP, tendency to bleed as demonstrated by petechiae or ecchymoses, platelet count, and likelihood of careful follow-up (Table 87.13). If the head trauma has occurred within 1 week of diagnosis, the patient is still having spontaneous bleeding, the platelet count is less than 20,000/mm ^{3}, or follow-up is uncertain, one or two IV infusions of g-globulin (0.8 to 1.0 g/kg) or anti- D (50 µg/kg) may be given.



Mild Head Trauma without Neurologic Findings
- Observe carefully
- Administer γ-globulin 0.8–1.0 g/kg by intravenous (IV) infusion or anti-D 50 μg/kg if:
  - Platelet count is <20,000/mm³
  - Signs of easy or spontaneous bleeding (e.g., bruises, petechiae)
  - Patient is within 1 wk of diagnosis of ITP
  - Follow-up is uncertain

Severe Head Trauma or Neurologic Abnormalities
- Hydrocortisone 8–10 mg/kg by IV
- γ-Globulin 0.8–1.0 g/kg by IV infusion
- Platelet transfusion 0.4 unit/kg
- If neurologic changes are severe or progressive or if no response to earlier measures:
  - Splenectomy
  - Exchange transfusion or plasmapheresis, followed by platelet transfusion

Table 87.13. Management of Head Trauma in Idiopathic Thrombocytopenic Purpura (ITP)

If severe head trauma has occurred or if neurologic abnormalities are present, hydrocortisone (8 to 10 mg/kg) and g-globulin (0.8 to 1.0 g/kg) should be administered by IV and random donor platelets (0.4 unit/kg, maximum 20 units) should be infused immediately thereafter. If necessary, the volume of plasma in the platelet preparation can be reduced by centrifuging the platelets, removing a portion of the plasma, and resuspending the platelets.

If the platelet count of a patient with ITP and intracranial bleeding does not increase after steroids, g-globulin, and platelet transfusions, or if the initial neurologic changes are severe, the patient should undergo splenectomy and, if appropriate, neurosurgical exploration. If the spleen has been removed previously and if steroids, g-globulin, and platelet transfusions have failed to raise the platelet count, an exchange transfusion or plasmapheresis with subsequent platelet transfusion should be performed. However, this is a desperate situation and full recovery is unlikely.

Immune-Mediated Neonatal Purpura

Serious bleeding may occur in the newborn or young infant with isoimmune thrombocytopenia or in the infant born to a mother with ITP. A discussion of the pathophysiology and diagnosis of these disorders is beyond the scope of this section. However, because mortality may be high in these disorders, particularly in isoimmune thrombocytopenia, the management of actual or potential bleeding deserves special emphasis. The brain is the major site of serious bleeding, perhaps because of trauma sustained during vaginal delivery. IV administration of g-globulin raises the platelet count in infants with isoimmune thrombocytopenia or alloimmune thrombocytopenia caused by maternal ITP. Steroid therapy may also be effective. In isoimmune thrombocytopenia, transfusion of random donor platelets is usually ineffective; the platelets are destroyed rapidly because of the presence of the same offending antigen as that found on the infant's platelets. However, the blood center may be able to provide platelets free of the offending antigen. Maternal platelets will survive normally and may be used for the affected infant after removal of the maternal plasma, which may contain the offending antibody. Transfusion of the thrombocytopenic infant born to a mother with ITP is more difficult because survival of all donor platelets is significantly shortened. If signs of generalized bleeding are present or if vital organs are impaired by local hemorrhage, and if IV g-globulin fails to raise the platelet count, a 2-volume exchange transfusion should be performed to remove a portion of the circulating antiplatelet antibody. A platelet transfusion should be administered immediately after completion of the exchange transfusion.

The recognition of immune-mediated neonatal thrombocytopenia is important for counseling of the parents and for preparation for future deliveries, as well as for the treatment of the affected child. In some instances, maternal ITP has been recognized only after delivery of a thrombocytopenic newborn. In isoimmune thrombocytopenia, accurate diagnosis allows appropriate counseling regarding the risk to infants born of subsequent pregnancies and the management of the mother and the affected fetus. If necessary, maternal platelets can be prepared just before future deliveries so that they are available for immediate transfusion if required. These factors make it imperative for the physician to obtain appropriate diagnostic studies in the thrombocytopenic infant.

Nonimmune Thrombocytopenia and Abnormalities of Platelet Function

Serious bleeding as a result of decreased platelet production or impaired platelet function usually responds rapidly to an infusion of random donor platelets (0.4 unit/kg, maximum 10 to 20 units). However, unless they are part of a program of prophylactic therapy, transfusions should be reserved for severe or persistent bleeding. Many affected patients have chronic disorders and require repeated transfusion. The excessive use of platelet transfusions, whether prepared from multiple, single, or HLA-matched donors, may contribute to the early formation of antiplatelet antibodies, making future transfusions more difficult and, in many instances, less effective. The use of leukocyte-depleted blood products, including platelets, reduces the risk of alloimmunization and is strongly preferred in this setting.

DISORDERS OF COAGULATION

Coagulation abnormalities are responsible for a large proportion of hematologic emergencies. Indeed, parents of children with hemophilia often use the ED as their primary source of acute care because bleeding episodes often occur at odd hours and require immediate treatment. This section places particular emphasis on the management of bleeding that poses a direct threat to life or to normal, long-term organ function in children with common inherited and acquired coagulopathies. The more rare inherited disorders of coagulation are not discussed in detail. Bleeding episodes are usually similar to those found in the more common disorders. Appropriate replacement products are listed in Table 87.14.

Factor Deficiency	Replacement Therapy
Fibrinogen (I) (also dysfibrinogenemias)	Cryoprecipitate Fresh-frozen plasma
Prothrombin (II)	Fresh-frozen plasma Prothrombin complex concentrate
Factor V	Fresh-frozen plasma
Factor VII	Fresh-frozen plasma Prothrombin complex concentrates Factor VIIa (recombinant) concentrate
Factor VIII	Factor VIII concentrates DDAVP
Factor IX	Prothrombin complex concentrates Factor IX concentrates
Factor X	Fresh-frozen plasma Prothrombin complex concentrates
Factor XI	Fresh-frozen plasma
Factor XIII	Cryoprecipitate Fresh-frozen plasma
von Willebrand's disease	DDAVP[a] Certain factor VIII concentrates

Table 87.14. Specific Factor Deficiencies and Replacement Therapy

Inherited Bleeding Disorders

Background

The most common inherited bleeding disorders are factor VIII deficiency (hemophilia A), factor IX deficiency (hemophilia B), and von Willebrand's disease. The severity of bleeding in the hemophilias can usually be predicted from the level of factor coagulant activity. If less than 1% of the deficient factor is present (severe hemophilia), bleeding episodes occur frequently and are often unrelated to trauma. If the factor level is between 1 and 5% (moderate hemophilia), spontaneous hemorrhage is less common but bleeding often occurs in response to minor trauma. If the factor level is greater than 5% (mild hemophilia), significant trauma is usually required to induce bleeding. Although very low levels of factor VIII coagulant activity, von Willebrand–related antigen, and ristocetin cofactor activity are associated with severe bleeding in the rare type III (autosomal-recessive) von Willebrand's disease, the relationship between laboratory findings and clinical course is less predictable in common type I von Willebrand's disease than in hemophilia. The severity of type I von Willebrand's disease in a particular child is best judged on the basis of the patient's bleeding history.

The classification of inherited bleeding disorders according to severity is important in assessing patients who have sustained trauma or who have signs of active bleeding. For example, after mild head trauma, the patient with severe hemophilia is at greater risk of developing intracranial bleeding than the patient with mild hemophilia and therefore must be managed more aggressively. When extensive hemorrhage is seen in a child with mild hemophilia, however, significant trauma has probably occurred and injury to deeper organs should be suspected.

Human immunodeficiency virus (HIV) infection has added a new dimension to the acute and long-term management of children with hemophilia. Approximately 80% of patients with factor VIII deficiency who were treated with factor VIII concentrate between 1978 and 1985 are infected with the virus. Approximately 50% of comparably treated patients with severe factor IX deficiency are also seropositive. In general, the emergency management of HIV-related problems such as pneumonia and diarrhea is similar to the management of nonhemophiliacs with HIV infection, as described in Chapter 85. However, situations in which the presence of HIV infection alters the usual management of hemophilia emergencies are noted in the sections that follow.

Clinical Manifestations and Management

Joint Bleeding

Hemarthroses are a common complication in hemophilia that often occur in the absence of known trauma in severe disease. The large joints such as the knees and elbows are most commonly affected. Initial replacement therapy should be designed to raise the factor level to 30 to 50%. Some centers treat all joint bleeds with one or two additional doses of factor replacement, whereas others reserve further treatment for patients with persistent pain or increasing swelling. When the involved joint has been the site of recurrent hemorrhages, several doses of replacement therapy are usually required. Initial immobilization of the joint is often helpful and can be easily accomplished with a splint that extends to the next joint distally. The pain associated with joint bleeding usually resolves within a few hours of treatment; therefore, analgesics are often unnecessary. If pain is severe, however, analgesic therapy should be given orally. Aspirin must not be used because its inhibitory effect on platelet function may further aggravate the clotting disorder. Acetaminophen, either alone or in combination with codeine, is usually sufficient. Repeated, prolonged outpatient therapy with narcotics should be avoided.

Bleeding in the hip is a particularly serious problem. As the joint becomes distended, blood flow to the femoral head may be impeded, resulting in aseptic necrosis. The hip is also a difficult joint to immobilize. Consequently, rebleeding is more likely in the hip than in other joints. Local tenderness is usually present, and the child prefers to lie in the frog-leg position. A radiograph of the hip shows widening of the joint space, and ultrasonography may demonstrate fluid. Because of the importance of achieving and maintaining hemostasis in this joint, initial correction to 70 to 100% is usually followed by several days of continuing replacement therapy (30 to 50% correction every 12 hours for factor VIII deficiency or every 24 hours for factor IX deficiency). Hospitalization may be required for immobilization, using either strict bed rest or traction.

The role of arthrocentesis in the management of a hemarthrosis varies from center to center. Removal of the blood from the joint has been helpful in allowing early mobilization and maintaining normal range of motion. Other indications for arthrocentesis include relief of pain if the joint is severely distended and prevention of further synovial damage in the chronically affected joint. Although the physical examination and laboratory findings may be similar in septic arthritis and hemarthrosis, the history and the patient's description of the pain (“just like my other bleeds”) are usually sufficient to

distinguish these two disorders, making arthrocentesis for diagnostic purposes unnecessary. Furthermore, even though joint bleeding is common in hemophilia, septic arthritis is extremely rare. A possible exception is the HIV-positive hemophiliac who may have an increased risk of septic arthritis. If a decision is made to tap a joint, correction to 70 to 100% should be achieved beforehand. How prominent a role arthrocentesis plays in the emergency management of joint bleeds may ultimately depend on the availability of consistent experienced orthopedic care. As in most procedures, the risk:benefit ratio of a joint tap is greatly reduced if the physician has extensive experience with the procedure and its role in the care of the particular disease.

Muscle Bleeding

Most muscle bleeding is superficial and easily controlled with a single dose of replacement therapy to achieve 30 to 50% correction. However, emergencies may arise when substantial blood loss occurs or when nerve function is impaired. Extensive hemorrhage is most commonly found in retroperitoneal bleeds (e.g., ilealpsoas) or thigh bleeds. Retroperitoneal bleeds are often accompanied by lower abdominal pain. A mass is sometimes palpable deep in the abdomen, and sensation in the distribution of the femoral nerve may be diminished. Loss of the psoas shadow may be seen on an abdominal radiograph, and a hematoma may be demonstrated by ultrasonography. The hemoglobin level should be measured initially and, if bleeding persists, at regular intervals thereafter. Treatment consists of hospitalization, bed rest, initial correction to 70 to 100%, and maintenance of a 30 to 50% factor level until pain has resolved and ambulation has been successfully achieved.

Nerve paralysis and contracture are associated with bleeding into the volar compartment of the forearm or the lower leg. Consequently, hemorrhage in these areas should be treated with an initial correction of 70 to 100% and, if abnormal muscle or nerve function is present or if swelling increases, maintenance of factor levels above 30 to 50% until resolution of symptoms. Orthopedic consultation should be obtained to help assess the pressure in the soft-tissue compartment and to determine the possible role of surgical decompression. Patchy sensory loss is often associated with compression of superficial nerves and may persist for several months before normal sensation reappears.

Subcutaneous Bleeding

Hemorrhage under the skin may cause extensive discoloration but is rarely dangerous and usually requires no therapy unless compression of critical organs occurs. However, pressure on the airway from a subcutaneous bleed of the neck may be life-threatening, requiring steps to ensure airway patency, such as placement of an endotracheal tube, in addition to correction of the factor level to 100%. Careful observation of children with bleeding in the muscles of the neck is mandatory because airway obstruction may be sudden.

Infants with undiagnosed hemophilia may present to the ED with prolonged bleeding from the site of circumcision. A partial thromboplastin time (PTT) should be measured and interpreted carefully because of the wide normal range in healthy newborns. Additional blood should be saved for assays of specific factors. If bleeding is not severe, correction can await determination of the type of hemophilia so that a treatment product specific for the identified deficiency can be used. If immediate replacement therapy is necessary, fresh-frozen plasma should be used because it will correct both factor VIII and factor IX deficiency as well as less common inherited clotting factor deficiencies.

Oral Bleeding

Mouth bleeds are particularly common in young children with hemophilia. The presence of fibrinolysins in saliva may lead to persistent oozing in the absence of aggressive management. The site of bleeding should be identified. If a weak clot is present, it should be removed and dry topical thrombin placed on the site. Initial correction should be 70 to 100%. Often, one or more additional treatments are necessary to achieve adequate clot formation and to prevent rebleeding when the clot falls off. The antifibrinolytic agents, e-aminocaproic acid (EACA) and tranexamic acid, are useful adjuncts in the treatment of oral bleeding. EACA should be administered orally for 5 days at a dosage of 100 mg/kg every 6 hours, with a maximum of 24 g/day. Tranexamic acid is administered orally at a dosage of 25 mg/kg three or four times daily. Because children may swallow a substantial amount of blood, acute, actual blood loss may be underestimated by the patient or family, and measurement of the hemoglobin level is helpful, particularly if bleeding has persisted for more than 24 hours. As in bleeds of the neck muscles, careful evaluation of airway patency is essential. Complete airway obstruction may result from extensive bleeding in the tongue.

Gastrointestinal Bleeding

Hemorrhage from the GI tract is rarely severe in hemophilia unless an anatomic lesion such as a duodenal ulcer or diverticulum is present. Maintenance of the factor level above 30 to 50% for 2 or 3 days after initial correction to 70 to 100% is usually sufficient. If bleeding persists, appropriate diagnostic studies are necessary. A careful search for infectious causes of GI bleeding is important in HIV-positive hemophiliacs.

Urinary Tract Bleeding

Atraumatic, painless hematuria is the most common manifestation of renal bleeding in children with hemophilia. Specific lesions are rarely identified, and IV pyelography can often be reserved for patients who fail to respond to initial replacement therapy. Ultrasonography carries no risk, however, and may be helpful in identifying the occasional patient with a subcapsular or intrarenal hemorrhage. If bleeding is persistent, moderate to severe anemia may develop and the hemoglobin level should be carefully monitored. In the absence of trauma or a demonstrable lesion, several approaches to ensuring hemostasis seem equally effective. Bed rest without replacement therapy is often successful. In some centers, one or more doses of factor replacement (70 to 100%) are used in combination with bed rest for at least 24 hours after gross hematuria has ceased. A brief course of orally administered prednisone has also been effective.

Although disagreement regarding the optimal method of treating painless hematuria still persists, there is no difference of

opinion regarding the potential hazard of EACA in affected patients. The strong, antifibrinolytic activity of this drug may cause formation of ureteral clots and outflow obstruction. Although EACA is an effective and seemingly safe agent in the treatment of hematuria associated with sickle cell anemia or sickle cell trait, its use in children with hemophilia has been accompanied by obstructive uropathy and should therefore be avoided.

When the child with hemophilia develops hematuria or flank tenderness after trauma, a more aggressive approach to diagnosis and treatment is required. Ultrasonography or IV pyelography should be performed as soon as possible to look for subcapsular or intrarenal bleeding or an obstructive clot at the pelvic–ureteral junction. To prevent parenchymal damage and deterioration of renal function, replacement therapy to achieve a level of 70 to 100% should be administered immediately. If a lesion is demonstrated using the techniques noted already, replacement therapy should be continued for 5 to 10 days. If no lesion has been identified, a shorter course of therapy is usually sufficient, using resolution of pain and hematuria as an end-point. The hemoglobin level and renal function tests should be followed carefully.

Intracranial Hemorrhage

Bleeding within the cranial vault is a complication of hemophilia that fully justifies the concern, anxiety, and urgency attached to it. In practical terms, however, head trauma in children with hemophilia is common, whereas intracranial hemorrhage is comparatively rare. Thus, the physician must be able to recognize as well as treat the child at risk without exposing other patients to unnecessary hospitalization, diagnostic studies, or therapy.

The management of the hemophiliac child with head trauma but no neurologic signs requires careful attention to the severity of the bleeding disorder, type of trauma, history of intracranial bleeding, and likelihood of close follow-up. Even with consideration of these factors, good fortune remains an important ingredient. Children with seemingly insignificant trauma may develop the first obvious signs of intracranial bleeding several days later when concern has diminished. To prevent such occurrences, every child with severe hemophilia and reported head trauma is treated with at least one dose of replacement therapy in some centers. However, this approach carries the risk and expense of frequent therapy. Moreover, in an effort to prevent yet another visit to the ED, the child or parent may fail to report a serious episode of trauma. Consequently, other centers use an approach that is still conservative although slightly less rigid. If the trauma is mild (e.g., a light bump on the forehead), the child is observed at home for the usual signs of intracranial hemorrhage or increased intracranial pressure. When the trauma is somewhat more substantial (e.g., falling down two or three carpeted stairs), the child with severe hemophilia is evaluated by the physician, given replacement therapy to achieve a level of 70 to 100%, observed for several hours in the office or ED, and, if well, is discharged. The child with mild hemophilia usually does not need replacement therapy under these circumstances, whereas the child with moderate hemophilia needs particularly careful attention to the type of trauma and bleeding history for the physician to decide whether to use replacement therapy. A computed tomography (CT) scan may be useful in identifying intracranial bleeding that requires more intensive and prolonged treatment. However, the imaging study should not be used to decide on administration of an initial dose of replacement factor that should, in fact, always be given before the study is performed to avoid unnecessary delays.

If more severe trauma (e.g., hitting the head on the dashboard, falling off a changing table onto a hard floor) has occurred in any hemophiliac child, hospital admission and repeated doses of replacement therapy are essential. The initial dose of replacement should be administered as soon as it is available. A CT scan should be performed after initial correction to search for intracranial bleeding and help determine the duration of treatment.

Unfortunately, the severity of trauma usually defies quantitative analysis, and the physician is left with substantial uncertainty. When the child with head trauma is not hospitalized, parents should be well informed about signs of intracranial bleeding. In particular, parents (and physicians) must remember that bleeding may be slow, initial imaging studies may be normal, and neurologic symptoms and signs may be delayed. Repeated visits may be necessary to monitor the child's neurologic examination accurately to detect intracranial hemorrhage as early as possible. At the first suggestion of the complication (e.g., headache, vomiting), hospitalization and treatment are mandatory.

The management of the patient with hemophilia who has neurologic findings in the presence or absence of head trauma begins with replacement therapy and those measures required for life support and treatment of increased intracranial pressure. Levels of the appropriate factor should be raised to 100%. The indications for surgery are similar to those for children without coagulation disorders, provided an appropriate correction of clotting abnormalities has been achieved.

Acute neurologic changes, such as decreasing level of consciousness or seizures, may result from direct HIV infection of the brain or from associated infections that affect the immunocompromised host (see Chapter 85). In the HIV-positive hemophiliac patient, these changes may mimic the signs and symptoms of intracranial hemorrhage. Similarly, changes in vision that result from retinitis caused by CMV may be similar to visual changes caused by intraocular bleeding. The physician evaluating an HIV-positive hemophiliac patient with new neurologic findings should consider both infectious and hemorrhagic causes, and the absence of demonstrable hemorrhage on imaging studies should prompt careful attention to HIV-related disorders.

Preparation of the Hemophiliac for Emergency Surgery

The child with hemophilia is subject to the surgical emergencies that affect children with normal hemostasis (e.g., appendicitis, compound fractures) as well as those hemorrhagic complications that require immediate operative intervention. In some instances, a bleeding episode may be confused with an acute abdomen. For example, retroperitoneal hemorrhage may mimic acute appendicitis. If time allows, a trial of replacement therapy may be helpful in distinguishing the two disorders. If, however, the child's clinical condition worsens or the need for surgery has been definitely established, correction up to 100% should be given, and the PTT should be measured to ensure its normalization. Because the PTT may be normal when the factor VIII or IX level is as low as 20 to 30%, measurement of factor coagulant level is needed to assess the adequacy of response to treatment when levels above 30% are desired. This test cannot always be performed before surgery but is essential in the postoperative management of the patient with

hemophilia. Therefore, surgery in a child with hemophilia should rarely, if ever, be undertaken in a hospital without appropriate laboratory facilities.

Bleeding in von Willebrand's Disease

The sites of bleeding in mild von Willebrand's disease resemble those found in patients with platelet disorders. Epistaxis, oral bleeding, and menorrhagia are common while joint bleeding is very unusual. Children affected with more severe forms, in which the factor VIII level is very low, may have bleeding problems that resemble those found in both hemophilia and von Willebrand's disease.

Replacement Products

The four products commonly used for the treatment of children with hemophilia and other bleeding disorders are lyophilized factor concentrates, 1-deamino-8- D-arginine vasopressin (DDAVP), fresh-frozen plasma, and cryoprecipitate. The correct use of these products is important not only to ensure adequate hemostasis but also to minimize risks associated with treatment and to reduce costs when possible.

Factor Concentrates

In recent years, the development of new factor concentrates has dramatically improved the care of children with hemophilia. Recombinant factor VIII and factor IX products are presently used for almost all newly diagnosed children and for most older children as well. The recombinant factor VIII products currently available in the United States continue to have a small amount of human albumin while the recombinant factor IX product is free of all human blood components. The higher cost of these products is balanced by their extremely low likelihood of transmitting infectious diseases. Plasma-derived factor VIII and factor IX concentrates also remain available. These products are made by immunoaffinity column or related techniques that remove many contaminating plasma proteins. A variety of additional steps are used to reduce or eliminate viral contamination, a critical process since most of the plasma-derived concentrates are made from plasma collected from thousands of donors. The amount of factor in each vial of recombinant or plasma-derived factor is printed on the label so the physician can determine the exact amount administered.

Some of the factor VIII concentrates also have a relatively high concentration of von Willebrand protein, making them particularly useful for the treatment of bleeding in patient's with severe, Type 3 von Willebrand's disease or in patients with Type 2 von Willebrand's disease in which other therapies are unsafe or unavailable (see below).

Prothrombin complex concentrates are used primarily for the treatment of patients with factor VIII deficiency and inhibitors (see below) and for the treatment of the relatively rare disorders of factor II, VII, or X deficiency. The use of this product for factor IX deficiency has diminished since the pure factor IX concentrates became available. Activated prothrombin complex concentrates are used exclusively in the management of patients with inhibitors.

Fresh-Frozen Plasma

Fresh-frozen plasma contains all plasma clotting factors and is therefore particularly useful when a child with a previously undiagnosed bleeding disorder presents to the ED with a hemorrhage that requires therapy before the specific factor deficiency can be ascertained. The recent availability of fresh frozen plasma that has been treated with a solvent-detergent solution to inactivate viruses with lipid envelopes (e.g., HIV, hepatitis C) has increased the safety of this product. Nonetheless, its use is generally restricted to the treatment of an unknown inherited factor deficiency or a deficiency of a factor such as factor XI for which there is no available factor concentrate. Although the average concentration of factor in fresh-frozen plasma is 1 unit/ml, the actual concentration in a particular unit may vary widely (0.5–1.5 unit/ml) and is rarely measured. If only one or two units are being transfused, the total amount of the desired factor that is being administered is unpredictable because the units may contain unusually high or low activity. Therefore, if one chooses to use fresh-frozen plasma to treat an infant with bleeding and a suspected but uncharacterized inherited coagulopathy, one must be aware that the actual amount of transfused factor may be as little as 50% of the calculated replacement. Extra plasma may address this problem if volume is not a limiting factor.

Cryoprecipitaet

When plasma is frozen and then slowly thawed, the precipitate contains enriched factor VIII coagulant activity, von Willebrand protein, fibrinogen, and factor XIII. However, like fresh-frozen plasma, the actual amount of these clotting factors in a single unit depends on the level in the donor and may be considerably less than average. If multiple units are administered, the total factor content is more likely to reflect the average activity for units of cryoprecipitate prepared in that particular blood center.

The availability of recombinant factor VIII and, to a lesser degree, plasma-derived concentrates with an excellent safety record, makes cryoprecipitate, which does not undergo virucidal treatment, an unsuitable choice for the treatment of factor VIII deficiency and a very unlikely choice for the treatment of von Willebrand's disease. In the latter instance, cryoprecipitate may be considered in a small child for whom the limited donor exposures might appear attractive in comparison with a plasma-derived concentrate. However, the limited number of donors also carries with it the concern about underestimating the total amount of replacement factor.

Comment

Most patients seen in the ED are accustomed to receiving a particular product in their regular hemophilia care. The treating physician should pay careful attention to the patient's treatment plan as developed by the hemophilia center. The ED is rarely the place to alter the long-term treatment program for a patient with hemophilia. A particularly important situation is the treatment of a child with a previously undiagnosed bleeding disorder. Whenever possible, the diagnosis

should be established before treatment is begun. For example, the diagnosis of factor VIII or IX deficiency might save a child a one-time exposure to a plasma product. In addition, trials of treatment products are frequently available to a newly diagnosed, previously untreated child with hemophilia. A single exposure to plasma or another product may make him or her ineligible for a study that could save his or her family hundreds of thousands of dollars in drug costs. However, an unavoidable delay in making a diagnosis of a specific factor deficiency or concern about eligibility for a study should not take precedence over the timely management of a serious bleeding episode.

Calculation of Dosage

Two formulas are commonly used for determining the number of units of factor VIII or factor IX necessary to achieve a specific level:

Factor VIII:

1. Weight of patient (kg) × desired level of correction (%) × 0.5 (for recombinant) or 0.5–1.0 (for plasma-derived concentrate) = number of units
2. One unit of factor VIII per kg raises the measured factor level by 2% for recombinant product ([desired level/2] × weight of patient [kg] = number of units) or by 1–2% for plasma-derived concentrate ([desired level/1–2] × weight of patient [kg] = number of units)

Factor IX:

1. Weight of patient (kg) × desired level of correction (%) × 1.2 (recombinant factor IX) or 1.0 (plasma-derived factor IX) = number of units
2. One unit of factor IX per kg raises the measured factor IX level by 0.83% for recombinant factor IX or by 1% for plasma-derived factor IX ([desired level/1.0 × weight of patient [kg] = number of units):([desired level/0.83] × weight of patient [kg] = number of units)

In the treatment of major hemorrhages, the achieved level of factor activity should be measured directly because the recovery *in vivo* varies widely among patients and because inadequate hemostasis may lead to severe morbidity or death. In addition, the dose of plasma-derived factor VIII required in children to achieve a particular factor VIII level may be 50 to 100% greater than in adults. If a minor bleed fails to respond to conventional dosing, the post-treatment factor level should be measured to be certain that the desired level is being achieved.

When children with von Willebrand's disease are treated with concentrate or rarely with cryoprecipitate, doses of 15 to 25 units of factor VIII activity/kg and 30 to 50 units of factor VIII activity/kg are commonly used for the treatment of minor and major hemorrhages, respectively. However, the response to treatment is not as predictable in this disorder as it is in factor VIII deficiency. The clinical course is the most useful measurement of the response to therapy and the need for further treatment in von Willebrand's disease. The choice of laboratory tests with which to monitor the treatment of von Willebrand's disease is controversial. Some physicians use the bleeding time, others use the factor VIII activity, and still others use the ristocetin cofactor level.

1-Deamino-8-D-Arginine Vasopressin

After the administration of DDAVP, levels of factor VIII coagulant activity, von Willebrand antigen, and ristocetin cofactor activity increase by about threefold in most people. This activity makes DDAVP an excellent alternative to blood products for the treatment of minor bleeding episodes in patients with mild hemophilia and for the treatment of most bleeding episodes in patients with the common form (type I) of von Willebrand's disease. Because some patients with these disorders do not respond to DDAVP, it is strongly recommended that children with mild hemophilia or type I von Willebrand's disease receive a trial dose of DDAVP when well to assess individual responses to this therapy before needing treatment for a bleeding episode. The dose of DDAVP is 0.3 µg/kg, administered by IV over 30 minutes. Side effects include facial flushing, headache, and rarely, hypertension, hypotension, and water retention. Hyponatremic seizures have occurred, and the patient should avoid excessive water intake. Subsequent doses may be less effective because the drug acts by releasing factor VIII and von Willebrand protein rather than by increasing their synthesis. In patients with severe factor VIII deficiency, DDAVP is ineffective. This is also true for many patients with severe (type III) von Willebrand's disease because the baseline factor VIII level is so low. In the rare Type IIB von Willebrand's disease, DDAVP may cause or aggravate thrombocytopenia and therefore should be used with caution.

DDAVP is also available as a concentrated nasal spray that should be distinguished from the more diluted formed used for enuresis or diabetes insipidus. For patients who have been previously shown to respond adequately to this form of therapy, the nasal spray is an alternative to an IV infusion. However, the nasal spray is designed primarily for use at home. DDAVP-responsive patients who come to the ED may have already tried the intranasal spray unsuccessfully or may have bleeding that requires other therapy.

Management of Patients with Inhibitors

The treatment of bleeding episodes in the child with hemophilia and antibodies against the missing or diminished factor is difficult, controversial, and often unsatisfying. Extensive resources, experience, and ingenuity are necessary to achieve hemostasis. For serious bleeding in patients with factor VIII deficiency and relatively low inhibitor titers (less than 10 Bethesda units), large doses of factor VIII may be sufficient to overwhelm the antibody and raise the factor VIII level to hemostatic levels. Doses as high as 100 to 200 units/kg may be necessary. However, those patients who are “high responders” to the factor VIII antigen will develop rising titers of antibody within 3 to 5 days, reducing or eliminating the usefulness of further therapy with factor VIII. Despite the subsequent rise in inhibitor titer, this brief period of initial factor VIII therapy may be sufficient to stop bleeding in critical organs such as the brain. On the other hand, factor VIII replacement should not be used as initial therapy for a minor bleed in a patient with a high responding inhibitor since the

anamnestic antibody response may impair the management of a later, more serious bleed.

If the initial inhibitor titer in a patient with critical bleeding is too high to warrant a trial of factor VIII therapy, or if no response to factor VIII is obtained, alternative approaches should be initiated. Porcine factor VIII can achieve hemostatic levels in patients whose inhibitor does not cross-react with this animal protein. As with human factor VIII, treatment with porcine factor VIII can be monitored by improvement in the PTT and an increase in the factor VIII level. Another advantage of porcine factor VIII is its excellent record of viral safety. Ideally, patients with factor VIII antibodies should be tested electively to determine whether the antibody cross-reacts with porcine factor VIII. In this way, emergency treatment can be reserved for those patients who are likely to respond favorably. Unfortunately, even if there are not cross-reactive antibodies at the outset, antibodies to porcine factor VIII may develop early in the course of therapy.

Another approach to the treatment of serious bleeding in the patient with factor VIII deficiency and inhibitors is the administration of prothrombin complex concentrate or activated prothrombin complex concentrate. These products presumably are effective because of their ability to bypass factor VIII through the presence of factors II, VII, and X. The activated prothrombin complex concentrate is somewhat more effective than the nonactivated product and is generally preferred for serious bleeding. However, neither product is uniformly effective, and treatment cannot be monitored by the PTT or factor levels but only by clinical response. A new option for the treatment of bleeding in children with high-titer inhibitors is recombinant factor VIIa. This product is not derived from plasma and therefore should have little or no risk of viral transmission. However, factor VIIa has a very short half-life and must be administered every two hours. As with the prothrombin complex concentrates, success of treatment must be judged clinically rather than by changes in laboratory values.

Occasionally the condition of the patient with inhibitors will worsen despite factor therapy. For example, the child with an intracranial hemorrhage may have continued bleeding and neurologic deterioration despite high-dose factor VIII or activated prothrombin complex concentrate. In such instances, plasmapheresis may remove sufficient antibody to allow a response to factor VIII administration. However, because 50% of the IgG inhibitor is tissue-bound and will rapidly return to the plasma, further infusions of factor VIII will be unsuccessful unless preceded by additional plasmaphereses.

Minor bleeds in the patient with factor VIII deficiency and inhibitors can be treated with any of the products described above for use with major bleeds. Factor VIII is the product of choice for patients with low titer inhibitors who can achieve hemostatic factor VIII levels without anamnesis. Patients with low titer inhibitors who are unresponsive to factor VIII and patients with high titer inhibitors, joint bleeds, and other minor hemorrhages should be treated with prothrombin complex concentrate, activated prothrombin complex concentrate, porcine factor VIII, or recombinant factor VIIa. Since the clinical response to these products may be unsatisfactory or, at best, unpredictable, good local care, including splinting for joint bleeds and topical thrombin for accessible oral bleeding, remains a cornerstone of therapy.

The treatment of the rare patients with antibodies to factor IX has not been clearly defined. Nonactivated and activated prothrombin complex concentrates have been beneficial in some cases. Exchange transfusion followed by infusion of factor IX concentrate may also be useful if the hemorrhage is severe or life-threatening. However, some patients with factor IX inhibitors have developed anaphylaxis when treated with these products, even if they received them without difficulty before the inhibitor appeared. Fortunately, the incidence of factor IX deficiency is only 15% of the incidence of factor VIII deficiency, and the proportion of patients who develop inhibitors is smaller in factor IX deficiency.

The recognition of a newly developed inhibitor may be equally important as the treatment of a patient with a known inhibitor. With the development of an inhibitor, patients with severe hemophilia often experience no change in their already difficult clinical course. However, the response to therapy is usually noted to be less satisfactory. Patients with moderate hemophilia who develop inhibitors have more bleeding and an inadequate response to treatment. If an inhibitor is suspected at the time of emergency therapy, a PTT should be measured after therapy because, in the presence of a strong inhibitor, a level of factor activity adequate to normalize the PTT (20 to 30% factor VIII) is rarely achieved. An inhibitor screen can also be performed by mixing the patient's plasma with normal plasma and by demonstrating a prolonged PTT compared with normal plasma mixed with saline. The level of inhibitor can be measured using more sophisticated techniques. Although the PTT need not be measured after routine treatment of minor hemorrhages, it should always be used to demonstrate an appropriate response in vitro to the treatment of major hemorrhage because a failure to respond to initial therapy may compromise organ function or life itself.

Inherited Hypercoagulable Conditions

Background

Hemostasis is a balance between the activities of proteins that promote and inhibit clotting. Deficiencies of the factor that promote clotting, such as factor VIII and factor IX, lead to abnormal bleeding, and the recognition and emergency treatment of the hemophilias and related disorders are familiar to many physicians. Deficiencies of the factors that inhibit coagulation lead to local and sometimes disseminated thrombosis. In contrast to bleeding disorders, the identification and emergency management of these unusual hypercoagulable disorders is likely to be unfamiliar to many clinicians. In fact, the role of some of these inhibitory proteins is only now being characterized.

Clinical Manifestations

The three major proteins that serve as brakes on the coagulation pathway are antithrombin III, protein C, and protein S. Antithrombin III inactivates the serine proteases that normally promote hemostasis. Patients heterozygous for antithrombin III deficiency have an increased risk of deep vein thrombosis and pulmonary embolus. Protein C, with its cofactor protein S, inactivates factors V and VIII. Persons heterozygous for protein C or protein S deficiency have an increased risk of venous thrombosis and, to a lesser degree, arterial thrombosis. Clinical findings in heterozygotes usually appear in adolescence or adulthood. In contrast, homozygous protein C deficiency causes widespread thrombosis in the neonatal period; homozygous protein S deficiency is rarer but may have similarly severe and early

clinical manifestations. This syndrome of purpura fulminans, sometimes accompanied by cerebral thrombosis, is a dire emergency that requires immediate intervention to preserve any chance of a good outcome.

The most common inherited cause of thrombosis, factor V Leiden, is a single gene mutation that alters the amino acid composition of factor V and makes this coagulation protein resistant to the anti-thrombotic activity of protein C. Factor V Leiden is found in nearly one-half of young men and women with a venous thrombosis. It may also be a risk factor for both venous and arterial thrombosis in ill neonates. Other inherited conditions that may increase the risk of thrombosis include dysfibrinogenemias, a prothrombin variant, abnormalities in the fibrinolytic pathway, and homocystinuria.

Management

Patients who are heterozygous for antithrombin III, protein C, protein S deficiency, or factor V Leiden and who develop a venous thrombosis should be treated with heparin by IV infusion. Treatment is usually initiated with a bolus injection of 50 to 100 units/kg body weight followed by a constant infusion of 25 units/kg per hour. The heparin dose should be adjusted to maintain the PTT between 1.5 and 2.0 times normal. Once adequate anticoagulation with heparin has been achieved, warfarin should be given orally. Long-term therapy with warfarin is effective because the drug inhibits synthesis of the vitamin K–dependent clotting factors. However, in the first few days of therapy, warfarin may inhibit disproportionately the synthesis of protein C, which is also vitamin K dependent and short-lived, leading to the paradoxical effect of increased hypercoagulability. Thus, the use of warfarin as initial therapy (i.e., without heparin) should be avoided because it may cause increased clotting, which manifests as skin necrosis.

Newborns with homozygous protein C deficiency and purpura fulminans should receive fresh-frozen plasma 8 to 12 mL/kg body weight every 12 hours. Long-term therapy includes regular infusions of fresh-frozen plasma or oral anticoagulation. Cryoprecipitate plays an analogous role in the acute and chronic therapy of homozygous protein S deficiency.

In many instances, a venous thrombosis is diagnosed in a patient without a previous diagnosis of an inherited hypercoagulable condition. The family history should be carefully reviewed for the occurrence of venous thrombosis or pulmonary embolus in otherwise healthy adults. Pretreatment plasma should be obtained for measurement of antithrombin III, protein C, and protein S. Molecular analysis for the factor V Leiden mutation can be performed before or after treatment. Because levels of these proteins may be transiently decreased after a large thrombosis, the diagnosis of a specific deficiency may be difficult. Measurement of antithrombin III, protein C, and protein S levels in both parents may help clarify the diagnosis in the child with a thrombosis and a possible inherited hypercoagulable condition.

Disseminated Intravascular Coagulation

Disseminated intravascular coagulation (DIC) is an acquired disorder of hemostasis that may be a result of numerous causes, but in children, it most commonly accompanies septic shock. The clinical and laboratory findings of DIC are described in Chapter 65. The treatment of this disorder should be directed primarily toward correction of the underlying disorder. Although correction of the hemostatic abnormality may temporarily decrease bleeding or prevent formation or extension of thrombosis, mortality remains extremely high when shock is not reversed in the first several hours.

Abnormal coagulation studies are often found in the absence of actual bleeding in DIC. Attempts to correct these abnormalities are of little or no value in preventing later bleeding or in altering the outcome of the underlying illness. If persistent or severe bleeding occurs, replacement of the consumed blood products may be helpful. Platelet transfusions (0.2 to 0.4 unit/kg) and fresh-frozen plasma (15 mL/kg) should be used to correct severe thrombocytopenia or substantially prolonged tests of clotting function. Although the administration of platelets and clotting factors may theoretically provide the necessary ingredients for further pathologic clotting, there is little evidence to suggest that such therapy is, in practice, responsible for worsening organ damage. However, replacement therapy should be stopped if bleeding does not improve after one or two infusions of the appropriate product. Preliminary studies suggest a possible role for AT-III concentrates in reducing intravascular clotting or in facilitating heparin therapy (see below).

The role of therapy with heparin in DIC remains controversial. Although anticoagulation may slow the progression of disseminated thrombosis and resulting ischemia and hemorrhage, such therapy itself may lead to fatal bleeding complications. Furthermore, as noted earlier, anticoagulation does not appear to affect patient survival and therefore should not interfere with the primary goal of reversing shock. Nevertheless, administration of heparin is commonly recommended for patients with DIC and purpura fulminans (see Chapter 65) or severely compromised renal function caused by thrombosis and ischemia. Heparin may be given by intermittent IV injection (50 to 100 units/kg every 4 hours) or continuous IV infusion (12.5 to 25 units/kg per hour after an initial bolus injection of 50 to 100 units/kg). The dosage should be adjusted to maintain the PTT at 1.5 to 2 times the normal value. Once further consumption of coagulation factors has been slowed or halted, administration of plasma and platelets may restore normal components of clotting. However, the actual benefit of the seemingly paradoxical use of anticoagulants and coagulation factors is unproved.

OTHER HEMATOLOGIC EMERGENCIES

Postsplenectomy Sepsis

Splenectomy may cure or ameliorate several hematologic disorders. However, loss of the spleen is associated with a greatly increased risk of sepsis caused by *S. pneumoniae, Neisseria meningitidis, E. coli, H. influenzae,* and other bacteria, especially in young children. The frequency of pneumococcal sepsis is particularly high; this organism accounts for 50% of the episodes of postsplenectomy sepsis. If the hematologic disorder is immunologic in origin (autoimmune hemolytic anemia) or accompanied by other gaps in host defense (Wiskott-Aldrich syndrome), the incidence of sepsis is especially high. More important, the mortality from sepsis in asplenic patients is significantly increased, averaging higher than 50% and rising to more than 80% in the presence of some immunologic abnormalities.

Although pneumococcal, *H. influenzae,* and meningococcal immunization and prophylactic antibiotics may reduce the occurrence of postsplenectomy sepsis, the most important facet of management is early detection and treatment. The presence of fever in an asplenic patient demands an immediate and careful evaluation to identify a source of infection. If the fever cannot be definitely attributed to a benign process such as an upper respiratory infection or if the patient appears unusually ill, the institution of parenteral antibiotic therapy pending results of cultures is usually indicated. The rapidity with which patients develop irreversible shock makes even a brief period of observation very risky and underscores the need for aggressive management of the symptomatic child. Antibiotic therapy is similar to that described above for children with sickle cell disease and fever.

Transfusion Reaction

Background

Acute hemolytic transfusion reactions that result from blood group incompatibility constitute a major hematologic emergency and may result in massive hemorrhage, renal failure, and death. The uncommon occurrence of this problem is, in large part, a tribute to careful blood banking practices and close attention to the administration of the red cell product. Unfortunately, the rarity of acute hemolytic reactions may lead to a sense of complacency regarding transfusion and a loss of familiarity with the signs and symptoms of massive red cell destruction.

Clinical Manifestations and Management

The characteristic findings of an acute hemolytic transfusion reaction include apprehension, fever, chills, abdominal or flank pain, chest tightness, and hypotension. If one or more of these findings develops, the transfusion should be stopped immediately because the severity of symptoms is related directly to the amount of hemolysis. Saline should be administered at 1.5 to 2 times the maintenance rate (see Chapter 18). A spun hematocrit should be examined for the presence of hemoglobin, which imparts a pink color to the plasma. The urine should also be examined for hemoglobin, which causes a positive dipstick reaction for blood in the absence of red cells on microscopic analysis. The name, identification number, and blood type of the patient should be compared with those on the unit of blood to ensure that the blood was given to the patient for whom it was intended. Finally, an aliquot of the unit should be returned to the blood bank for confirmation of the original compatibility testing and labeling. A newly positive direct Coombs test confirms the diagnosis.

Further management of an acute hemolytic transfusion reaction is directed toward maintenance of normal blood pressure and urine output and treatment of intravascular coagulation. Rapid IV hydration is mandatory to prevent renal shutdown. Diuretics, including mannitol (1 g/kg), may also be helpful. Intravascular coagulation should be treated with heparin, using doses similar to those described earlier for DIC from other causes.

Delayed hemolytic transfusion reactions can occur 3 to 14 days after administration of red cells. These reactions may be due to late formation of an antibody in response to a newly encountered red cell antigen or, alternatively, to an anamnestic response of an antibody that originally developed in response to a previous transfusion but was undetectable at the time of the most recent cross-match. The rate of red cell destruction is usually slower with a delayed hemolytic transfusion reaction than with an acute hemolytic reaction. Therefore, the most prominent signs and symptoms are those of anemia and hyperbilirubinemia rather than shock and renal shutdown. Laboratory testing demonstrates a positive direct Coombs test. Antibody in the serum or in the red cell eluate is specific for the offending red cell antigen. Transfusion with compatible red cells will relieve severe or symptomatic anemia.

Nonhemolytic transfusion reactions are more common and, in most instances, less severe than hemolytic reactions. Sensitization to plasma proteins may cause urticaria. Fever, chills, and headache commonly occur in repeatedly transfused patients who have become sensitized to white cell and platelet antigens. Although these reactions pose little danger to the patient, they may be difficult to distinguish from the more urgent hemolytic reaction. Before continuing the transfusion, urine and plasma should be checked for the presence of hemoglobin. If these studies are unrewarding, the physician must decide whether the clinical condition of the child warrants discarding the remainder of the unit or finishing the transfusion with supportive therapy such as antipyretics or antihistamines. The use of filters to remove white cells has drastically reduced the incidence of nonhemolytic reactions in chronically transfused patients. As a corollary, a hemolytic reaction should be given particularly strong consideration when fever and chills occur in a child receiving filtered red cells.

Suggested Readings

Andrew M, Michelson AD, Bovill E, et al. Guidelines for antithrombotic therapy in pediatric patients. J Pediatr 1998;132:575–588.

Bernini JC. Diagnosis and management of chronic neutropenia during childhood. Pediatr Clin North Am 1996;43:773–792.

Beutler E. Glucose-6-phosphate dehydrogenase deficiency. N Engl J Med 1991;324:169–174.

Blanchette V, Imbach P, Andrew M, et al. Randomised trial of intravenous immunoglobulin G, intravenous anti-D, and oral prednisone in childhood acute immune thrombocytopenic purpura. Lancet 1994;344:703–707.

Bonilla MA, Gillio AP, Ruggeiro M, et al. Effect of recombinant human granulocyte-colony-stimulating factor on neutropenia in patients with congenital agranulocytosis. N Engl J Med 1989;320:1574–1590.

Bray G. Recent advances in the preparation of plasma-derived and recombinant coagulation factor VIII. J Pediatr 1990;117:503–507.

Chesney PJ, Wilimas JA, Presbury G, et al. Penicillin- and cephalosporin-resistant strains of streptococcus pneumoniae causing sepsis and meningitis in children with sickle cell disease. J Pediatr 1995;127:526–532.

Furie B, Limentani SA, Rosenfield CG. A practical guide to the evaluation and treatment of hemophilia. Blood 1994;84:3–9.

Gill FM, Sleeper LA, Weiner SJ, et al. Clinical events in the first decade in a cohort of infants with sickle cell disease. Blood 1995;86:776–783.

Hathaway WE. The bleeding newborn. Semin Hematol 1975;12:175–188.

Heene DL. Disseminated intravascular coagulation: evaluation of therapeutic approaches. Semin Thromb Hemostas 1977;3:291–295.

Jonsson OG, Buchanan GR. Chronic neutropenia during childhood: A 13-year experience in a single institution. Am J Dis Child 1991;145:232–235.

Lane PA. Sickle cell disease. Pediatr Clin North Am 1996;43:639–664.

Levine AB, Berkowitz RL. Neonatal alloimmune thrombocytopenia. Semin Perinatol 1991;15:35–40.

Lusher J. Approach to the bleeding patient. In: Nathan DG, Orkin SH, eds: Hematology of Infancy and Childhood. Philadelphia: WB Saunders, 1998:1574–1584.

Lux SE, Wolfe LC. Inherited disorders of the red cell membrane skeleton. Pediatr Clin North Am 1980;27:463–486.

McClure PD. Idiopathic thrombocytopenia purpura in children: diagnosis and management. Pediatrics 1975;55:68–74.

Medeiros D, Buchanan GR. Current controversies in the management of idiopathic thrombocytopenic purpura during childhood. Pediatr Clin North Am 1996;43:757–772.

Moake J. Hypercoagulable states: new knowledge about old problems. Hosp Pract 1991;26:31–42.

Montgomery RR, Hathaway WE. Acute bleeding emergencies. Pediatr Clin North Am 1980;27:327–344.

Pearson HA, Diamond LK. The critically ill child: Sickle cell disease crises and their management. Pediatrics 1971;48:629–635.

Poncz M, Kane E, Gill FM. Acute chest syndrome in sickle cell disease: etiology and clinical correlates. J Pediatr 1985;107:861–866.

Rogers ZR, Morrison RA, Vedro DA, Buchanan GR. Outpatient management of febrile illness in infants and young children with sickle cell anemia. J Pediatr 1990;117:736–739.

Vichinsky EP, Styles LA, Colangelo LH, et al. Acute chest syndrome in sickle cell disease: clinical presentation and course. Cooperative Study of Sickle Cell Disease. Blood 1997;89:1787–1792.

Wilimas JA, Flynn PM, Harris S, et al. A randomized study of outpatient treatment with ceftriaxone for selected febrile children with sickle cell disease. N Engl J Med 1993;329:472–476.

Young NS, Alter BP. Aplastic Anemia: Acquired and Inherited. Philadelphia: WB Saunders, 1994.

Zarkowsky HS, Gallagher D, Gill FM, et al. Bacteremia in sickle hemoglobinopathies. J Pediatr 1986;109:579–585.

CHAPTER 88

Toxicologic Emergencies

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POISONED CHILD

Poisoning represents one of the most common medical emergencies encountered by young children and accounts for a significant fraction of emergency department (ED) visits in the adolescent population.

Estimates of poisoning episodes annually in the United States range in the millions. Poisonings may be unintentional or intentional. Unintentional poisonings make up some 80 to 85% or more of all poisoning exposures, whereas intentional poisonings comprise the other 10 to 15%. Persons in this latter group have much higher rates of treatment in the ED, hospitalization, and intensive care. Among children aged 5 years and younger, most poisonings are unintentional events related to exploratory behavior, or they result from willful child abuse. Although less common, the physician must also consider the possibility of environmental exposures, suicide attempts in children, and neonates exposed to toxins in utero.

The unintentional ingestion of a toxin by a toddler represents a complex interplay of host, agent, and environmental factors and may be considered a subset of the modern traumatic injury model. In this model, each factor contributes, more or less, in a given context to the probability of the injury occurring. Some children are more at risk because of peak age of 1 to 4 years, male gender, temperament that leans toward hyperactivity, and increased finger–mouth activity and/or pica. Some agents are more culpable because of ease of access, attractiveness/palatability, and toxic potential. Two classic examples are iron tablets—they look like candy, are widely available over the counter (OTC), and are toxic in significant overdose—and mouthwash—it has a bright color, pleasant taste, and smell and is packaged in large volumes without safety caps and has a surprisingly high ethanol content (15 to 25%). Typical environmental factors include an acute stressor, such as a recent move or new baby in the household, and chronic issues, such as parental illness/disability. The concordance of child, agent, and environmental factors may lead predictably to the statistical

likelihood of a toddler ingestion. Pediatricians have led the way in poison prevention strategies by modifying these risk factors with traditional anticipatory guidance and by spearheading the lobby for child-safety caps on particularly dangerous medications and household products. These efforts have resulted in a dramatic decrease in childhood poisoning morbidity and mortality over the past three decades, but such poisonings continue to occur and demand the emergency physician's attention.

The scope of toxic substances involved in poisonings is broad, requiring a wide range of knowledge. Table 88.1 presents the categories of substances most commonly reported in human exposures in the United States for the year 1997. Table 88.2 presents the 10 most common toxic exposures involved in human deaths for the year 1997. The former listing much more closely approximates the profile of pediatric poisonings, whereas the latter is more typical of intentional adult exposures. The most important difference between the pediatric and the adult profile by type of agent is in the higher percentage of cases in which psychopharmacologic drugs (sedatives, tranquilizers, and antidepressants) cause poisoning in adults and the much higher frequency of exposures to household and personal care products and plants in children.

Substance	Percentage of Total Exposures
Cleaning products	10.4
Analgesics	10.3
Cosmetics	9.0
Plants	5.6
Cough and cold preparations	5.1
Bites/envenomations	4.4
Foreign bodies	4.3
Insecticides/pesticides	4.0
Topicals	3.7
Food products/food poisoning	3.6

Modified with permission from Litovitz TL, et al. 1997 Annual Report of the American Association of Poison Control Centers Toxic Exposure Surveillance System. Am J Emerg Med 1998;16:443–457.

Table 88.1. Substances Most Often Reported in Human Exposures

Analgesics	Antidepressants
Sedatives-hypnotics, antipsychotics	Stimulants and street drugs
Alcohols	Cardiovascular drugs
Gases and fumes	Chemicals
Anticonvulsants	Antihistamines
Cleaning substances	

Modified with permission from Litovitz TL, et al. 1997 Annual Report of the American Association of Poison Control Centers Toxic Exposure Surveillance System. Am J Emerg Med 1998;16:443–457.

Table 88.2. Toxic Exposures Associated with the Most Deaths

There are six basic modes of exposure to poisoning: ingestion, ocular exposure, topical exposure, envenomation, inhalation, and transplacental exposure. Poisonings may be the result of acute or chronic exposures. Most poisonings are acute, and the victims are typified by the child who surreptitiously invades the medicine cabinet or the storage area for household cleaners or the adolescent or adult who takes a massive number of pills in a fit of despair. *Chronic poisoning* refers to toxicity produced over time in which a substance accumulates in the body, producing toxic results; it is best exemplified by environmental exposure to lead or other heavy metals. In the drug category, chronic toxicity can also exist. Acetaminophen hepatotoxicity that occurs in infants and small children or aspirin poisoning in older adults as a result of salicylate accumulation after administration of too much drug for too long is typical of chronic toxicity. Chronic toxicity is a special problem for the clinician because the source is not always apparent, the toxicity is not always clear, and the toxic process is not often obvious until serious clinical derangements occur.

GENERAL APPROACH TO THE POISONED CHILD

Following the analogy between unintentional accidental poisoning and traumatic injury, a similar model may be used in formulating a management approach. The poisoned patient often represents an acute-onset emergency with a broad spectrum of multiorgan system pathophysiology that shares many features with the multiple trauma victim. In essence, poisoning might be viewed as a multiple chemical trauma. The concept of a brief window of opportunity to make critical diagnostic and management decisions is likewise analogous. Thus, with a nod toward the widely acclaimed Advanced Trauma Life Support model of the American College of Surgeons, one may conceptualize a management approach that attempts to prioritize critical assessment and, at times, simultaneous management interventions (Table 88.3). The initial phase (or “primary survey”) addresses traditional ABCs of airway securement and cardiorespiratory support, with a slight additional emphasis on emergent toxicologic considerations. The more specific evaluation and detoxification phase (or “secondary survey”) is aimed at simultaneously initiating generic treatment while assessing the actual extent of intoxication (in cases of known or presumed exposures) and/or identifying the actual toxins involved (in unknown, but highly suspected intoxications).

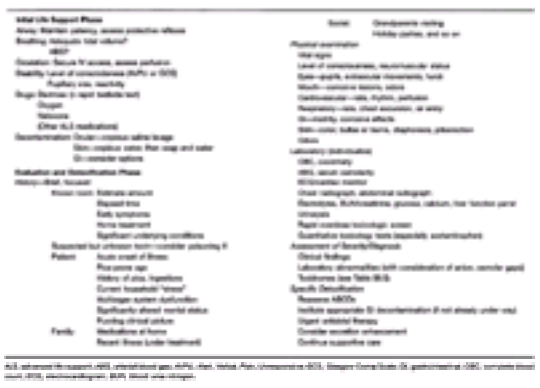

Table 88.3. General Approach to the Known or Suspected Intoxication

Initial Life Support Phase

The general approach to recognition and support of vital airway and cardiorespiratory functions (or “ABCDs”) is well known to most readers and is covered in detail in Chapter 1. In the context of the poisoned child, a few points deserve special emphasis. In addition to the usual signs of airway obstruction, the physician must pay special attention to evidence of disturbed airway protective reflexes. Many poisoned patients will vomit or undergo procedures such as gastric lavage or administration of charcoal, which pose an aspiration risk. Elective endotracheal intubation (see Chapter 5) may thus be indicated at a slightly lower threshold in this context than in another child with comparable central nervous system (CNS) depression.

It is also particularly important to anticipate imminent respiratory failure in the deeply comatose poisoned child. Cyanosis and overt apnea are late findings with progressive drug-induced medullary depression. Thus, clinical assessment of early ventilatory insufficiency and/or measurement of P_{CO_2} on arterial blood gas analysis is critical in such patients, to avoid the chaos of a precipitous respiratory arrest. Likewise, it is far easier to establish intravenous (IV) access in a child with normal circulatory status than in a child in shock; early efforts to obtain a secure IV line in symptomatic overdose patients are thus well worth the time and effort.

Having reached the “D” in our mnemonic, the patient is evaluated for “ *d*isability” (e.g., neurologic status), empiric “ *d*rug” treatment, and emergent “*d*econtamination.” Level of consciousness may be assessed rapidly with a semiquantitative scale such as the Glasgow Coma Scale or the AVPU scale (spontaneously *a*lert, response to *v*erbal stimulation or *p*ain, or *u*nresponsive). Pupillary size and reactivity may be quickly noted. Rapid changes in mental status are common in serious intoxications and may herald precipitous cardiorespiratory failure.

Empiric drug treatment is warranted for most symptomatic poisoned children with altered mental status. All such patients may initially be given humidified *oxygen* and their oxyhemoglobin saturation monitored, if possible, by pulse oximetry. If available, rapid bedside blood glucose testing may be used; if low, or not readily available, a trial dosage of 0.25 to 1.0 g/kg glucose as 10 to 25% solution should be infused. It should be noted that drug- or toxin-induced hypoglycemia does not present uniformly with coma or seizures. Almost any neuropsychiatric picture may predominate, including aphasia; slurred, dysarthric speech; and focal neurologic signs. Adrenergic signs, such as diaphoresis and tachycardia, are not uniformly present. Hypoglycemia is a complication seen in ingestions of ethanol, oral hypoglycemics, b-blockers, salicylates, and of course, insulin injection. As basic as this intervention seems, in our experience, it is still one of the most often missed (or more accurately, *delayed*) critical treatments in the management of the poisoned patient.

Thiamine (100 mg IV), although routinely administered to adult overdose patients who receive hypertonic glucose to obviate precipitating Wernicke's encephalopathy, is not generally warranted in the pediatric population. Perhaps it should be considered in adolescent patients who may be thiamine deficient secondary to eating disorders, chronic disease (e.g., inflammatory bowel disease), or alcoholism. Last, empiric naloxone therapy is just as important in potentially poisoned toddlers with altered mental status as it is in adults. Although substance abuse is admittedly uncommon in the average 2-year-old child, it is amazing how many narcotics find their way into the curious child's mouth. Many households contain a variety of oral opioid analgesic agents, as well as cough medicines (codeine, dextromethorphan), antidiarrheal agents (paregoric, diphenoxylate), and partially naloxone-responsive antihypertensive agents such as clonidine. In addition, the possibility of unintentional ingestion of a “stash” of illicit opioids does exist. Thus, naloxone should be used as a therapeutic/diagnostic trial when there is a reasonable possibility that altered mental status is drug induced. Previous recommendations have based dosing on weight (e.g., 0.01 to 0.1 mg/kg); however, many authorities now prefer a unified dose of 1 to 2 mg for acute overdose patients of all ages (outside the neonatal period). Such an approach conceptualizes naloxone dosing as based on total narcotic load and number of opioid receptors that require competition for binding sites. In general, this latter approach is easier to remember and has not been associated with complication in the ED. Adolescent patients with a strong clinical picture for opioid intoxication (without habituation) may receive 2-mg bolus doses every 2 minutes, up to a total dose of 8 to 10 mg before abandoning hope of benefit because several congeners of morphine (e.g., propoxyphene, illicit fentanyl derivatives, pentazocine) may require such large doses. If chronic abuse is suspected, lower initial doses (0.2–0.11 mg) are warranted. Administration of flumazenil to adolescents exhibiting depressed consciousness after an unknown drug overdose is contraindicated (see also discussion in Sedative-Hypnotic section).

The rationale for decontamination of the poisoned child is discussed in the following section, Evaluation and Detoxification. This treatment phase may begin urgently, after, or in concert with attention to the ABCDs. At times, a decision to perform gastric decontamination through the preferred technique can be made almost immediately upon presentation and, if so, should be instituted as soon as possible in light of the patient's clinical status and the number of hands available to assist in management. For example, a toddler with coma, shock, and massive hematochezia who is

rushed into the ED by the rescue squad—and for whom there is witnessed or strong circumstantial evidence of massive iron overdose—requires a concerted team effort directed toward resuscitation, stabilization, and urgent gastric decontamination—in this case, probably initiated with gastric lavage after endotracheal intubation. However, an apparently asymptomatic adolescent who admits to ingesting 10 g of acetaminophen 2 hours before arrival at the ED may be more fully evaluated in a timely but orderly manner (as outlined in the next section) and within short order can be considered for less emergent gastric decontamination—in this case, likely an oral dose of activated charcoal. Significant dermal or ocular exposures require immediate copious lavage, and precautions from exposure should be taken to protect the health care providers tending to the patient.

At the completion of this initial life support phase, the poisoned patient should have been assessed for compromise of vital airway and cardiorespiratory function and for global neurologic status and should have had resuscitative measures instituted. Patients with significant altered mental status have been critically evaluated for respiratory status, have had IV access secured, and have had their therapeutic trials of oxygen, glucose, and naloxone. Other advanced life support interventions such as anticonvulsants or antiarrhythmics have been instituted as necessary. Consideration of decontamination options has begun.

Evaluation and Detoxification Phase

History

A brief and focused *historical evaluation* should be addressed as soon as the life support phase has been accomplished. The primary goal is to determine the potential severity of the exposure. This assessment requires toxin and patient-related data alike.

For a known or highly suspected toxin, an attempt is made to estimate the total amount ingested (number of pills missing, ounces left in the bottle, dosage of pills, concentration of alcohol, and so forth). The best estimate of time elapsed since ingestion is also sought. Parents should be questioned regarding early symptoms noted at home or on route to the ED, and any treatments administered before arrival. Certain underlying medical conditions may be relevant (e.g., glucose-6-phosphate dehydrogenase [G6PD] deficiency for mothball ingestions); thus, any significant past medical history ought to be noted.

Often, children who are poisoned do not come to the ED with a clear history of exposure followed by onset of symptoms. Often, they develop signs and symptoms that mimic other diseases and give no history of toxic exposure. Thus, the ED staff must always consider the possibility of an ingestion when treating young children.

General historical features that suggest the possibility of poisoning include 1) acute onset; 2) age range of 1 to 5 years; 3) history of pica or known, unintentional ingestion; 4) substantial environmental stress, either acute (e.g., arrival of a new baby, serious illness in a parent) or chronic (marital conflict, parental disability); 5) multiple organ system involvement; 6) significant alteration in level of consciousness; and 7) a clinical picture that seems especially puzzling.

Certain family and social history variables are also important. Medications used by other household members, and particularly new medications introduced into the home environment by virtue of recent illnesses or visits from grandparents and other relatives, are a common source of ingested drugs. Changes in routine and large family gatherings (e.g., holiday parties, moving to a new home) are particularly risky occasions for decreased parental supervision and new (or less carefully guarded) potentially toxic medications or household products.

Physical Examination

The focused physical examination should begin with a reassessment of vital functions and complete recording of vital signs, including core temperature. With secure airway and cardiorespiratory function confirmed, the examination should then focus on the central and autonomic nervous systems, eye findings, changes in the skin and/or oral and gastrointestinal (GI) mucous membranes, and odors (see Chapter 47) on the breath or clothing of the patient. These features represent those areas most likely affected in toxic syndromes and, when taken together, often form a constellation of signs and symptoms referred to as toxidromes (Table 88.4 and Table 88.5). Such toxidromes may be so characteristic as to provide guidance for early therapeutic trials before precise historical or laboratory confirmation of a specific exposure is available.

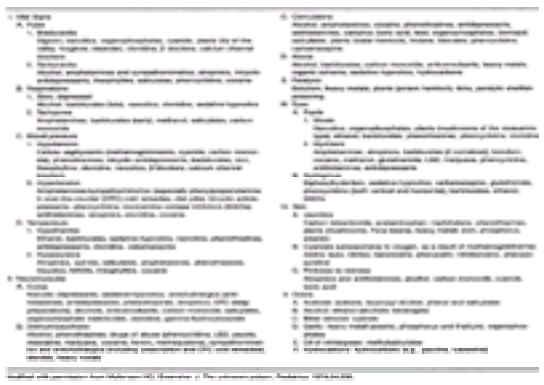

Table 88.4. Clinical Manifestations of Poisoning

	Sympathomimetics (Antipsychotics, Cocaine)	Anticholinergics (Antihistamines, Many Others)	Organophosphates (Insecticides, Nerve Gases)	Opiates/ Clonidine	Barbiturates/ Sedative-Hypnotics	Salicylates	Theophylline
Mental status/ CNS	Agitation, delirium, psychosis, convulsions	Delirium, psychosis, coma, convulsions	Confusion, fasciculations, coma	Euphoria, somnolence, coma	Somnolence, coma	Lethargy, convulsions	Agitation, tremor, convulsions
Heart rate	Increased	Increased	Decreased or increased	Decreased	–	–	Increased
Blood pressure	Increased	Increased	–	Decreased	Decreased	–	Decreased
Temperature	Increased	Increased	–	Decreased	Decreased	Increased	–
Respirations	–	–	Increased	Decreased	Decreased	Increased	Increased
Pupils	Large, reactive	Large, sluggish	Small	Pinpoint	–	–	–
Bowel sounds	Present	Diminished	Hyperactive	–	–	–	–
Skin	Diaphoresis	Flushed, dry	Diaphoresis	–	–	–	–
Miscellaneous	–	–	"SLUDGE"*	–	–	Vomiting	Vomiting

CNS, Central nervous system.
*"SLUDGE" is a mnemonic representing salivation, lacrimation, urination, defecation, gastric cramping, and emesis.

Table 88.5. Toxidromes

Laboratory Evaluation

The laboratory may be helpful in confirming diagnostic impressions or in demonstrating toxin-induced metabolic aberrations. However, there is no “tox panel” that is uniformly helpful or necessary. Most poisonings can be managed appropriately without extensive laboratory studies, and in particular, the reflex ordering of rapid overdose toxicology “screens” has been found to be rarely helpful in acute patient management. They have important, nonemergent roles (e.g., in resolving medicolegal issues or considering drug-induced causes of behavioral changes in a psychiatric patient). In toddlers with a known or strongly suspected specific ingestion, rapid drug screens are rarely indicated. In the adolescent intentional overdose patient who is not critically ill or who does not have a particularly puzzling clinical picture, the drug screen again is rarely helpful, although the finding of an unexpected toxic level of acetaminophen (which may have been omitted in the history) may impact on management, and some authors recommend that quantitative acetaminophen levels (in lieu of “tox screens”) be sought in such patients.

The comprehensive drug screen that requires blood and urine samples may be useful for patients who are seriously ill with an occult ingestion or for the occasional intentional overdose adolescent patient whose clinical picture does not fit with the stated history. Often of greater help is the critical interpretation of routine measurements of serum chemistries and osmolarity in patients with altered mental status. The presence of hypoglycemia or aberrations of serum electrolytes may be crucial information about the poisoned patient. In certain circumstances, tests of liver or renal function, urinalysis, creatine phosphokinase levels, and other select tests may be useful. Metabolic acidosis with a high anion gap is found in many clinical syndromes and toxidromes, reflected by the often-cited mnemonic *MUDPILES*, for *m*ethanol and *m*etformin; *u*remia; *d*iabetic and other ketoacidoses; *p*araldehyde; *i*soniazid, *i*ron, and *i*nborn errors of metabolism; *l*actic acidosis (seen with hypoxia, shock, carbon monoxide, cyanide, and many drugs that cause compromised cardiorespiratory status or prolonged seizures); *e*thanol and *e*thylene glycol; and *s*alicylates. Differences between calculated and measured serum osmolarity (Calculated = 2 × [Serum Na] + BUN/2.8 + glucose/18 with normal osmolarity = 290 mOsm/kg) may suggest intoxication with ethanol, isopropanol, or more rarely in pediatric patients, methanol or ethylene glycol.

An immediate determination of quantitative levels is helpful in making management decisions for some drugs, and these are outlined in Table 88.6. Furthermore, many important causes of coma and altered vital signs are not detected on even the most sophisticated “comprehensive” toxicology panels (which are usually biased toward psychoactive medications and illicit drugs). An overview of such agents is presented in Table 88.7. An electrocardiogram (ECG) should be performed in all seriously ill patients in whom poisoning is being considered. Detectable conduction delays may precede life-threatening cardiac rhythm disturbances.

Drug/Toxin	Optimal Time After Ingestion
Acetaminophen	4 hr
Carbamazepine	2–4 hr
Carboxyhemoglobin	Immediate
Digoxin	4–6 hr
Ethanol	½–1 hr
Ethylene glycol	½–1 hr
Iron	4 hr
Lithium	2–4 hr*
Methanol	½–1 hr
Methemoglobin	Immediate
Phenobarbital	1–2 hr
Phenytoin	1–2 hr
Salicylates	2–4 hr*
Theophylline	1–2 hr*

Modified with permission from Weisman, RS, Howland MA, Flomenbaum, NE. The Toxicology Laboratory. In: Goldfrank LR, Flomenbaum NE, Lewin NA, Weisman RS, Howland MA, eds. Toxicologic Emergencies. Norwalk, CT: Appleton & Lange, 1990.
*Repeat levels over 8–12 hours may be necessary with sustained-release preparations.

Table 88.6. Frequently Useful Quantitative Toxicology Tests in Pediatric Patients

Coma-Causing	Hypotension-Causing
Bromide	β-Blockers[a]
Carbon monoxide	Calcium channel blockers[a]
Chloral hydrate	Clonidine[a]
Clonidine	Colchicine
Cyanide	Cyanide
Organophosphates	Digitalis[a]
Tetrahydrozoline (in over-the-counter eye drops)	Iron

Modified with permission from Wiley JF II. Difficult diagnoses in toxicology: poisons not detected by the comprehensive drug screen. Pediatr Clin North Am 1991;38:725–737.
[a]Hypotension often is seen with bradycardia.

Table 88.7. Important Drugs and Toxins Not Detected by Most Drug Screens

Assessment of Severity and Diagnosis

At this juncture, most intoxicated patients may be readily stratified by specific toxin or category of drug(s) ingested and some judgment made as to the potential or current severity of the exposure. For some children, clinical features of a complex illness of acute onset may suggest an intoxication without a specific history of such ingestion. In a few cases, some laboratory confirmation of clinical suspicion will be available on an immediate basis. Using all the clinical clues available and with some familiarity of the “toxidrome” approach to differential diagnosis as detailed previously (Table 88.4 and Table 88.5) and, at times, with help from the laboratory, the emergency physician must now establish a working diagnosis and proceed with consideration of options for specific detoxification.

Specific Detoxification

Again, the proviso that the patient be continually reassessed and managed for impaired vital function is addressed. All decisions about further decontamination and/or specific antidotal therapy involve a complex interplay of toxin-related and patient's condition-related factors.

Gastrointestinal Decontamination

The effort to “get the poison out” has long been a mainstay of the traditional discussion of toxicologic management. Recently, considerable controversy has arisen over the optimal method of gastric emptying and its overall value in the management of poisoned patients. Many authorities now eschew gastric emptying in all but a few select cases, preferring an enlarged role for the early use of activated charcoal. Many studies that support this trend have been published over the past 10 years. It is emphasized here that many of the relevant human studies used relatively nontoxic “overdoses” in adult volunteers or excluded (at the attending physician's discretion) most critically ill patients. Few young children were included in any of these studies. It is likely that as further research is conducted, particularly as directed toward the pediatric population, current dogma regarding optimal GI decontamination will evolve. For the sections that follow, several appropriate techniques for gastric decontamination are reviewed, all of which may be useful under certain circumstances. An approach to the overall decision process in a given patient is then offered.

Simple Dilution. Dilution may be indicated only when the toxin produces local irritation or corrosion. Water or milk is an acceptable diluent. For drug ingestion, however, dilution alone should not be used because it may increase absorption by increasing dissolution rates of the tablets or capsules, or it may promote more rapid transit into the lower GI tract. However, administration of fluids during the induction of emesis is appropriate.

Gastric Emptying. The goal of gastric emptying is to rid the stomach of remaining poison to prevent further local effect or systemic absorption. The utility of gastric emptying diminishes with time and is most effective if done within the first hour. In certain circumstances, such as the delayed gastric emptying accompanying intoxication with anticholinergic drugs, benefit may be noted longer after ingestion. *Emesis* was once a favored means of gastric emptying, but currently induction of emesis is typically limited to first aid in the home setting. The drug of choice for inducing emesis is syrup of ipecac. A dose of 30 mL for adolescents, 15 mL for children, and 10 mL for infants 6 to 12 months of age, given with 8 oz of water or other liquid, reliably produces vomiting. If vomiting does not occur in 20 minutes, the dose should be repeated.

The efficacy of ipecac-induced emesis in reducing bioavailability of ingested drug has recently been called into question. Study results have varied widely with ranges from 0 to 70% decrease in absorption. Despite a long history of use, it has been difficult to prove improved clinical outcome in poisoned patients given ipecac compared with patients treated with activated charcoal alone. Ipecac has a long record of safe pediatric experience, and despite rare case reports of complications secondary to particularly prolonged or forceful emesis, its administration is generally well tolerated by patients and parents. It may be the most useful decontamination procedure for young children who ingest plant parts, seeds, mushrooms, and the like, in whom lavage is less apt to be successful because of particle size and for whom charcoal alone may not be as effective. Ipecac has been found to delay the time to administration of activated charcoal and some specific antidotes. It is contraindicated when patients have ingested caustics or hydrocarbons, when patients have uncontrolled hypertension or increased intracranial pressure, and when the presence or possibility of compromised airway protective reflexes exists.

An alternative to ipecac-induced emesis for emptying the stomach is *gastric lavage.* This procedure is usually reserved for patients who have ingested a potentially life-threatening amount of a poison, in cases where the procedure can be performed safely within 60 minutes of ingestion and charcoal alone is not believed to be adequate. To carry out a satisfactory lavage, the patient should be on his or her left side, head slightly lower than feet, and the largest orogastric

lavage tube that can reasonably be passed should be used (e.g., 24-Fr orogastric tube for a toddler, 36-Fr orogastric tube for an adolescent). A smaller caliber nasogastric (NG) tube is sufficient for only some liquid toxins. Gastric contents should be aspirated initially before any lavage fluid is introduced. Normal saline aliquots of 50 to 100 mL in young children and 150 to 200 mL in adolescents can be lavaged repeatedly until the return is clear. Like induced emesis, gastric lavage's efficacy in reducing drug absorption has been reviewed critically in recent studies. Again, the efficacy has been highly variable and lavage has not been demonstrated to improve outcome in poisoned patients. Several important risks are associated with gastric lavage, including oxygen desaturation, aspiration, and mechanical trauma to the oropharynx and esophagus.

Activated Charcoal. Activated charcoal minimizes absorption of drugs by adsorbing them onto its surface. Charcoal administration has become the decontamination strategy of choice for most pediatric poisonings and is most effective when used in the first few hours after ingestion. A number of notable compounds, such as iron and lithium, do not adsorb well to activated charcoal (Table 88.8). The usual dose of activated charcoal is 1 g/kg; adolescents and adults should receive 50 to 100 g. Most activated charcoal is now available premixed with water to make a slurry that can be taken orally or administered by NG tube. Considerable pharmaceutical research is being done toward the goal of making charcoal easier to administer and more palatable. “Superactivated” charcoals are available with increased adsorptive surface area. It should be noted that the once advocated “universal antidote” that consisted of activated charcoal, magnesium oxide, and tannic acid is not recommended. Similarly, burnt toast is not effective.

Substances poorly (or not) adsorbed
Common electrolytes
Iron
Mineral acids or bases
Alcohols
Cyanide
Most solvents
Most water-insoluble compounds (e.g., hydrocarbons)

Table 88.8. Activated Charcoal

Activated charcoal was “rediscovered” by the toxicology community during the 1980s, with several studies finding its use to be superior to gastric emptying alone, and at least equivalent to the combination of gastric emptying plus charcoal administration. The use of charcoal alone is less invasive and intuitively is probably less likely to be associated with complications in the clinical setting than gastric emptying. Aspiration of charcoal can be a serious concern in patients with poor airway protective reflexes, and vomiting remains the most common difficulty associated with its use. Some preparations of activated charcoal are premixed with a sorbitol cathartic in an effort to speed fecal toxin elimination and reduce constipation. Charcoal might be contraindicated in patients with an unprotected airway or a disrupted GI tract (e.g., after severe caustic ingestion) or in patients in whom charcoal therapy may increase the risk and severity of aspiration (e.g., hydrocarbons). The use of multiple doses of activated charcoal is addressed later in this chapter.

Catharsis. Two types of osmotic cathartics have been used to treat poisoned patients: the saccharide cathartics (e.g., sorbitol) and the saline cathartics (e.g., magnesium citrate, magnesium sulfate). A special type of catharsis, *whole bowel irrigation (WBI),* is discussed in the next section. Unfortunately, little evidence exists to suggest that standard cathartics accomplish their goal of reducing drug absorption by decreasing GI transit time. It is still unclear whether cathartics administered with activated charcoal reduce subsequent constipation, and some believe cathartics increase the incidence of vomiting. Repetitive doses of osmotic cathartics are associated with considerable diarrhea and cramping, and hypernatremic dehydration has been reported in young infants. A single dose of premixed charcoal/sorbitol is safe for most pediatric ingestions, but this preparation should be used with caution in young infants. Mineral oil or stimulant cathartics such as castor oil are discouraged because they may increase absorption of some poisons or unnecessarily extend the cathartic effect.

Whole Bowel Irrigation. An additional technique of GI decontamination that has been developed over the past several years is that of intestinal irrigation with large volumes and flow rates of a polyethylene glycol-balanced electrolyte solution such as Colyte or GoLYTELY. These solutions are not significantly absorbed nor do they exert an osmotic effect, so the patient's net fluid/electrolyte status is unchanged. They have a long track record of safe use, including in infants, in the surgical application of preoperative bowel preparation. WBI has been found to be particularly useful in pediatric iron overdoses, in which gastric lavage may be limited by tube size and the substance is not bound to charcoal. It has been used for other metal ingestions (e.g., lead), for overdoses of sustained-release medications (e.g., lithium, theophylline) with increasing levels after initial decontamination, and for ingestions of crack vials or cocaine packets. It might also be useful in particularly massive and/or late-presenting overdoses for which the efficacy of gastric emptying and/or charcoal is expected to be suboptimal. The technique may be used by mouth in cooperative patients or by NG tube; the usual recommended dosing is 500 mL/hour in toddlers and 2 L/hour in adolescents and adults.

Gastrointestinal Decontamination Strategies

It should be apparent that no unique approach to GI decontamination of all poisoned patients is optimal in every case. Factors to be considered include the expected degree of toxicity from the drug, the physical nature of the drug, the current location of the drug within the body, and the presence of contraindications or alternatives. A risk-benefit decision

must be made before the institution of any decontamination strategy.

Syrup of ipecac is used primarily as first-aid treatment of recent potentially toxic ingestions in the home. Today, induced emesis is rarely favored in the ED. Its use may be considered after ingestions of particulate poisons, like plant leaves, in toddlers. The use of gastric lavage as a decontamination strategy has become more limited in recent years. It is still considered to have a potentially important role in patients with recent ingestions of extremely toxic substances that put them at risk for a lethal course, especially when those substances do not bind well to charcoal. Likewise, patients with truly massive overdoses may benefit from gastric lavage because standard charcoal preparations have diminished effectiveness when the charcoal to drug ratio is less than 10:1. The correct technique for gastric lavage requires that careful attention be given to prevent aspiration and anatomic trauma.

Some of the patients in question will have undergone endotracheal intubation during the initial life support phase of management, as detailed previously, or they may be strong candidates for such airway protection because of borderline mental status and in anticipation of their ensuing critical course. Others may be awake, alert, and cooperative and may have normal airway protective reflexes, and thus be managed by elective, voluntary lavage without prior endotracheal intubation. The combative, agitated patient poses a dilemma and must be carefully managed on an individualized basis—the benefit of a rapid sequence endotracheal intubation performed primarily to allow gastric lavage, per se, would only occasionally outweigh the risks involved. A severely depressed, uncooperative adolescent or a frightened, combative toddler who had ingested a full bottle of colchicine or lithium within 1 hour of presentation might be such an example. A similar overdose of only benzodiazepine, or only acetaminophen, would probably not.

Overall, the mortality from acute poisoning is less than 1%. Suicidal overdoses in adolescents typically have more inherent lethality than unintentional overdoses in toddlers. When gastric decontamination is warranted, the administration of activated charcoal, without gastric emptying, is most often the appropriate choice. WBI is of theoretic benefit to body packers and stuffers, and patients who have ingested toxic amounts of sustained-release preparations or agents not adsorbed well by activated charcoal. An attempt to summarize these considerations is diagrammed in Figure 88.1, although it should be reiterated that all decisions regarding gastric decontamination involve multiple patient and toxin-related factors and should not be made with a “cookbook” approach.

FIGURE 88.1. Approach to gastrointestinal decontamination. [a]For patients in whom the toxin is no longer believed to be in the stomach, activated charcoal administration and/or whole bowel irrigation might still be valid considerations.

Antidotal Therapy

The overall number of ingestions for which a specific antidote is necessary or available is small. When a specific antidote can be used, it is vital that it be administered as early as possible and in an appropriately monitored dose. Those antidotes that should be available for immediate administration include sodium bicarbonate (cyclic antidepressants), sodium nitrite/sodium thiosulfate (cyanide), atropine and pralidoxime (cholinesterase inhibitors), ethanol or 4-methylpyrazole (ethylene glycol and methanol), deferoxamine (iron), dextrose (ethanol, salicylates, oral hypoglycemics), methylene blue (methemoglobinemic agents), oxygen (carbon monoxide), flumazenil (benzodiazepines), and naloxone (opioids). Other antidotes usually do not require such urgent administration and may be given subsequent to initiation of other management modalities. Even when available, antidotes do not diminish the need for meticulous supportive care or other therapy. Indiscriminant use of antidotes without other forms of management should be discouraged. Table 88.9 summarizes a list of commonly used antidotes, suggested doses, and their indications for use. Because of its frequent use, naloxone is discussed further here.

Table 88.9. Summary of Antidotes

Naloxone. Naloxone, a pure narcotic antagonist, is one of the broadest acting, safest, and most effective of any true antidotes now available. It is effective against all opioids. Naloxone is a synthetic congener of oxymorphone but is devoid of morphine agonist or depressant effects. It has no significant side effects in the treatment of acute overdose except narcotic withdrawal symptoms in the addicted patient. These symptoms include GI upset, tachycardia, hyperpnea, mydriasis, rhinorrhea, diaphoresis, sialorrhea, increased blood pressure, anxiety, restlessness, discomfort, and hyperalgesia. These symptoms are not life-threatening to teenagers and adults but can be fatal to an infant born to an addicted mother. Withdrawal symptoms, if observed during acute overdose treatment, would be expected to last no more than 30 minutes and should generally be treated with supportive care. The serum half-life of naloxone is 1 hour; its duration of action 1 to 4 hours. Initial reversal of narcosis may then revert to coma, requiring ongoing reassessment and readministration of naloxone. There are a few case reports of other adverse effects, including hypertension, pulmonary edema, ventricular irritability, and seizures after naloxone-induced reversal of narcosis in the perioperative setting, typically in patients with underlying cardiopulmonary disease and in the presence of additional medications or anesthetic agent use.

The mechanism of action of naloxone is by competitive displacement of narcotic analgesics at central narcotic receptor sites. It can be used as a diagnostic test when faced with a questionable history. Current dosage recommendations reflect the proven safety of naloxone in large doses and the necessity of such doses to reverse effects of synthetic opioids such as propoxyphene and pentazocine. If respiratory depression is present, the initial dose should be 2 mg IV in any patient. Repeat doses may be given every 2 minutes until 10 mg have been administered for adolescent patients with suspected opioid overdose who fail to respond to the lower dosages. Of course, concomitant airway management is vital. In patients without respiratory depression, an initial dose of 1.0 mg can be used. In adolescents suspected of chronic opiate abuse, smaller initial doses (e.g., 0.2–0.4 mg) are warranted. Again, if there is no response but a strong clinical suspicion, 2-mg doses can be repeated up to a total of 10 mg before concluding that further dosing will be of no benefit. Naloxone can also be given intramuscularly, sublingually, or by endotracheal tube if no IV access is available.

If a lightening response occurs, naloxone will have to be repeated at the effective total dose every 20 to 60 minutes. An alternative approach is to provide a continuous IV infusion; generally about two-thirds of the total reversal dose will need to be infused per hour initially, with subsequent adjustments as necessary.

Nalmefene and naltrexone are longer-acting opioid antagonists that may have use in some clinical situations in which a longer duration of action (4 to 6 hours for nalmefene, 24 hours for naltrexone) is deemed beneficial, such as in reversal of postoperative opioid depression or as aids in opioid detoxification programs. However, as antidotes for acute opioid overdose in the adolescent or pediatric population, their longer duration may be problematic in assessing the actual time course for resolution of clinical toxicity and/or in precipitating prolonged withdrawal symptoms in habituated patients. Nalmefene may be a useful substitute for prolonged naloxone infusions in cases for which such opioid antagonism is necessary, but little pediatric experience and few dosing guidelines for its use are currently available.

Enhancing Excretion

The procedures available for enhancing the elimination of an absorbed poison that have the greatest value are multiple-dose activated charcoal, ionized diuresis, dialysis, and hemoperfusion. Because some risk is involved, these measures are indicated only in those cases in which the patient's recovery would be otherwise unlikely or in which a specific significant benefit is expected.

Diuresis

Diuresis has historically been advocated in cases of poisoning with agents that are excreted primarily by the renal route. Although it is important to maintain high glomerular filtration rates in the presence of rhabdomyolysis or when chelating with agents such as ethylenediaminetetraacetic acid (EDTA), forced diuresis has limited value in the treatment of acute poisoning. Similarly, diuretic use has fallen out of favor with the possible exception of mannitol therapy for ciguatera poisoning.

Ionized diuresis takes advantage of the principle that excretion is favored when a drug is in its ionized state. Urinary alkalinization promotes excretion of salicylate (a weak acid) and may also enhance clearance of phenobarbital, chlorpropamide, and chlorophenoxy herbicides. Urine alkalinization can be initiated with sodium bicarbonate at a dose of 1 to 2 mEq/kg by IV over a 1- to 2-hour period. Careful attention should be given to total fluid and sodium load administered, especially in patients at risk for congestive heart failure or pulmonary edema. Hypokalemia can interfere with the ability to alkalinize the urine and should be corrected. The rate of bicarbonate infusion can be adjusted to maintain a urinary pH of 7.5 to 8.5. Urinary acidification is almost never indicated because it may lead to serious side effects such as systemic acidosis and exacerbation of renal impairment in the context of myoglobinuria.

Dialysis

Dialysis is indicated for selected cases of severe poisoning or when renal failure is present. Indications for dialysis depend on patient-related and drug-related criteria. Patient-related criteria include 1) anticipated prolonged coma with the high likelihood of attendant complications, 2) development of renal failure or impairment of normal excretory pathways, and 3) progressive clinical deterioration despite careful medical supervision. Drug-related criteria are 1) satisfactory membrane permeability, 2) a correlation between plasma drug concentration and drug toxicity of the agent, 3) plasma levels in the potentially fatal range or the presence of a significant quantity of an agent that is normally metabolized to a toxic substance, and 4) significant enhancement of clearance. Hemodialysis is the most effective means of dialysis. Because it requires highly technical skills, as well as a physician and a technician, it is not always available,

but is an essential consultative service for units that manage severe poisoning cases.

Hemoperfusion

Hemoperfusion, the process of passing blood through an extracorporeal circuit and a cartridge containing an adsorbent after which the detoxified blood is returned to the patient, is also effective in drug removal. Although there are some reservations regarding the extent to which hemoperfusion can be used, it appears to be at least as effective as and possibly more effective than hemodialysis for a number of agents. Indications for use are similar to those for hemodialysis. Table 88.10 summarizes the generally accepted common drugs and drug concentrations for which hemodialysis and hemoperfusion should be considered, in light of the previous discussion regarding clinical criteria.

Hemodialysis	Hemperfusion
Lithium, 4.0 mEq/L	Phenobarbital, 100 mg/L
Ethylene glycol, 50 mg/dL	Theophylline, 6–10 mg/dL (60–100 mg/L)
Methanol, 50 mg/dL	Paraquat, 0.1 mg/dL
Salicylates, 100 mg/dL	Glutethimide, 4 mg/dL
	Methaqualone, 4 mg/dL
	Ethchlorvynol, 15 mg/dL
	Meprobamate, 10 mg/dL

Modified with permission from Winchester JF. Active methods for detoxification. In: Haddad LM, Winchester JF (eds.). *Clinical Management of Poisoning and Drug Overdose*. Philadelphia: WB Saunders, 1983:162–166.

Table 88.10. Drugs and Their Plasma Concentrations for Which Hemodialysis or Hemoperfusion Should Be Considered

Multiple-Dose Activated Charcoal (Gastrointestinal Dialysis)

Several studies have shown significant increase in clearance for a number of drugs when repeated doses of 0.5 to 1.0 g/kg of activated charcoal are given every 4 to 6 hours. By using a nearly continuous stream of fresh charcoal that descends through the intestinal tract, a constant concentration gradient is maintained that favors the back diffusion of free drug from periluminal capillary blood into the intestinal lumen, where it may be bound immediately to the newer charcoal, so that the free drug concentration in the intestinal lumen remains low. In addition, enterohepatic recirculation of some drugs may be interrupted as reabsorption from bile is prevented. To be safe and effective, this technique requires active peristalsis and an intact gag reflex or a protected airway. Common pediatric poisonings for which repetitive charcoal dosing may be indicated include phenobarbital, carbamazepine, phenytoin, digoxin, salicylates, and theophylline. Cathartics, such as sorbitol, should be administered no more frequently than every third dose.

Supportive Care

The final step in optimizing treatment for the poisoned child is the direction of scrupulous attention to supportive care, including continued close monitoring of ABCDs, fluid and electrolyte status, urine output, and level of consciousness. The value of these efforts usually far outweighs that which may be ascribed to any specific toxicologic interventions in most cases. Severely symptomatic patients are most properly cared for in specialized facilities that have skilled pediatric critical care staff and access to toxicology consultation.

NONTOXIC INGESTION

Often, the emergency physician will be asked about a childhood ingestion of some common household products, many of which are nontoxic, unless taken in huge amounts. The availability of a list of such nontoxic products often leads to immediate relief of parental anxiety and avoids the institution of unnecessary noxious interventions. Before using such a list, however, several precautions need to be borne in mind. The fact that an ingestion is nontoxic does not necessarily mean that it has no medical significance. Ingestions often occur in the context of a suboptimal environment. There may be poor supervision or unusual family stresses surrounding the accident, or the ingestion may not have been accidental. Several criteria have been suggested by Mofenson and Greensher to qualify an ingestion as “nontoxic.” These include the assurance that only one identifiable product is ingested in a well-approximated amount, that the product label includes no cautionary signal word, that the child is symptom-free and less than 5 years old, and that an appropriate mechanism is available for telephone follow-up. When used with these criteria, Table 88.11 provides an updated list of nontoxic ingestions. In certain cases, consultation with a regional poison control center is often helpful.

Table 88.11. Products That Are Nontoxic When Ingested In Small Amounts

PEDIATRIC OVERDOSES

The following section highlights selected agents that are ingested by children. They have been chosen because of their common occurrence, because they represent the potential for serious or life-threatening toxicity, and because timely recognition and appropriate treatment may be lifesaving.

Acetaminophen

Background

Acetaminophen, *N*-acetyl-*p*-aminophenol (APAP), is the most popular pediatric analgesic-antipyretic and has now become one of the most common pharmaceutical preparations ingested by young children. It is also one of the 10 most common drugs used by adolescents and adults in intentional self-poisoning. Acetaminophen also occasionally turns up as an unreported coingestant in intentional overdoses. Fortunately, accidental ingestion in young children has been associated with little morbidity, although occasional cases of hepatotoxicity occur, particularly in the context of inadvertent repetitive overdosing.

Pathophysiology

The major toxicity of APAP is severe hepatic damage. Acetaminophen is metabolized in three ways by the liver: 1) glucuronidation, 2) sulfation, and 3) metabolism through the cytochrome P-450 pathway to form a potentially toxic intermediate, which conjugates with glutathione. In a massive overdose, glutathione becomes depleted, thus allowing the undetoxified intermediate to bind to hepatocytes, leading to cellular necrosis. This damage is reflected by rising liver enzymes, hepatic dysfunction, and in severe poisonings, hepatic failure and death. The use of *N*-acetylcysteine as an antidote relates in part to this molecule's ability to act as a glutathione precursor.

Clinical Findings

Initially, the signs and symptoms of APAP ingestion are vague and nonspecific but include nausea and vomiting, anorexia, pallor, and diaphoresis. These manifestations usually resolve within 12 to 24 hours, and the patient appears well for 1 to 4 days. During this latent period, liver enzymes may rise, and jaundice with liver tenderness may ensue. Most patients have a gradual resolution of their hepatic dysfunction, although without antidotal treatment, about 2 to 4% of intoxications that develop toxic plasma levels will go on to hepatic failure and death. Such patients with severe toxicity develop further clinical evidence of hepatic disease at 3 to 5 days after ingestion and some develop renal damage. Anorexia, malaise, and abdominal pain may progress to signs of liver failure with hepatic coma.

The potential severity of an acute intoxication may be predicted by the amount ingested, if accurately known, and the plasma level of APAP. APAP in single doses of less than 150 mg/kg in children is likely to be harmless. Severe toxicity in adolescents or adults usually occurs with overdoses of 10 to 15 g. Initial GI symptoms, although vague, are generally more pronounced when the overdose is large. However, the only reliable indication of the potential severity of the hepatic damage is the plasma APAP level, taken at least 4 hours after ingestion. A nomogram (Fig. 88.2) is available for using this value in the prediction of likely toxicity. We recommend use of the lower line of the nomogram, plotted 25% below the possible toxicity line, to err on the safe side in making therapeutic decisions.

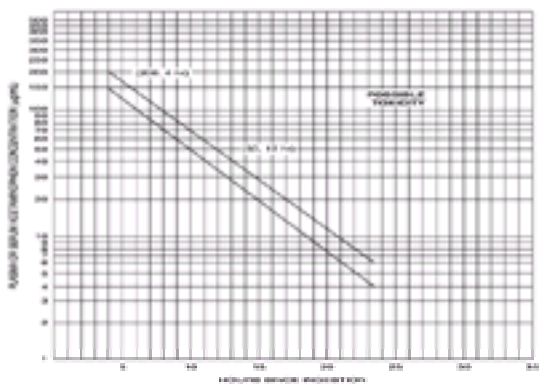

FIGURE 88.2. Nomogram for estimating severity of acute acetaminophen poisoning. (Modified with permission from Rumack BH, Matthew H. Pediatrics 1975;55:871–876. Copyright, American Academy of Pediatrics, 1975.)

Management

The basic toxicologic principles of preventing absorption apply to APAP overdoses, with a few points deserving special emphasis. Immediate-release acetaminophen is rapidly absorbed, and it would generally be unlikely to expect significant return from a gastric emptying procedure done much after 1 hour after ingestion. An extended-release preparation of acetaminophen has recently been marketed, and this preparation may be more amenable to gastric emptying.

Furthermore, many patients will require the use of the oral antidote, *N*-acetylcysteine (NAC), and patients given ipecac often have prolonged vomiting when attempting to drink the unpleasant tasting NAC. Although some have speculated that activated charcoal would decrease the bioavailability of the NAC and thus should be avoided, recent studies have demonstrated clinically insignificant decreases in NAC absorption, even when using large doses of charcoal. In general, acetaminophen ingestions presenting within less than 1 hour may rarely benefit from gastric emptying, particularly for massive overdoses or in the context of mixed ingestions when the coingestant is particularly toxic. However, for most cases of acetaminophen overdose per se, and particularly for those typically seen 2 to 4 hours after ingestion, charcoal alone is probably effective and should not significantly alter the ability to use NAC several hours later. In cases that present after 4 hours have elapsed, gastric decontamination is usually not warranted.

NAC, given orally, essentially prevents the occurrence of hepatotoxicity when instituted within 8 hours of ingestion. It also lessens the severity of hepatic damage if used within 16 hours. This antidote can be mixed with fruit juice or soda to disguise its foul smell, or it can be administered by lavage tube. Only mild GI side effects result from its use. Persistent vomiting is an occasional obstacle to completing the 72-hour course of therapy. This may be obviated by giving the dose slowly or by NG or duodenal tube infusion. Antiemetic therapy with metoclopramide or ondansetron may also be helpful. An IV preparation is currently undergoing clinical trials in the United States and has been used in Canada and Europe for some time with good effects, with the advantage of utility in the face of protracted vomiting or other GI complications from coingestants or coincident medical problems; however, its use has been associated with occasional anaphylactoid reactions.

The protocol for NAC therapy may be summarized as follows:

1. Consider GI decontamination options as already noted.
2. If patient presents less than 4 hours after ingestion, wait to draw 4-hour level and base therapeutic decision on nomogram (assumes rapid turnaround time so that level will be available by 8 hours after ingestion); if necessary, initiate treatment as described next. For extended-release preparations, McNeil Consumer Products Co., a manufacturer of acetaminophen, suggests a second APAP level drawn 4 hours after the first; antidotal therapy is to be instituted if either level suggests possible toxicity.
3. If patient presents more than 6 to 8 hours after ingestion, give a loading dose of NAC, 140 mg/kg orally (regardless of time since ingestion, consider increasing dose by 40%, or repeating the loading dose, if charcoal had been given within the preceding 1 to 2 hours). Obtain level and base subsequent course of therapy on nomogram.
4. If level plots out above the lower line on the nomogram, admit the patient to hospital and continue NAC at 70 mg/kg orally every 4 hours for a total of 17 doses. Monitor complete blood count (CBC), renal, and liver function tests.
5. Treatment for patients who present more than 24 hours after ingestion is controversial, as is treatment for patients with subacute, repetitive overdosing over several days. We generally advise treatment for children who receive more than 150 mg/kg per day for 1 to 2 days or more and/or patients with levels above 10 µg/mL who present more than 24 hours after ingestion.
6. In rare cases, oral administration of NAC may be impossible. Protocols exist for the IV administration of the oral NAC preparation. A regional poison control center may be able to assist in this situation.

Alcohols and Glycols

The alcohols and glycols are some of the most commonly found organic compounds in the environment. Ethanol is a commonly encountered solvent and is used as a topical antiseptic, chemical intermediate, beverage, and in some instances, a rubbing alcohol. It is also an ingredient in most perfumes, colognes, and toilet waters. Methanol, or methyl alcohol, functions as an antifreeze (in windshield washers/de-icers and gasoline antifreeze) and as a solvent in many industrial and home products. Isopropyl alcohol serves as a rubefacient. Ethylene glycol is used primarily as a de-icer or antifreeze. A related class of compounds, the glycol ethers, are widely used in rug shampoos and other cleaning compounds. The toxicity of these agents is thought to be comparable to (if not more severe than) ethylene glycol.

Ethanol

The most commonly ingested alcohol is ethanol. After ingesting ethanol, children may develop nausea, vomiting, stupor, and ataxia. Coma and death from apnea may occur if significant quantities are consumed. In adolescents, blood concentrations of less than 50 mg/dL rarely result in sensory or motor impairment. Values of 100 to 150 mg/dL are consistent with intoxication and cause mild neurologic findings. Lethal blood alcohol concentrations are generally greater than 500 mg/dL. Infants and toddlers who ingest ethanol have a clinical course that is significantly different from that in adolescents and adults; a triad of coma, hypothermia, and hypoglycemia appears once ethanol levels exceed 50 to 100 mg/dL. This triad may be accompanied by metabolic acidosis.

The blood ethanol level may be estimated by calculating the osmolol gap (measured serum osmolality - [2 × Serum Na + BUN/2.8 + Serum glucose/18]). An osmolal gap of 22 to 25 mOsm/kg H_2O exists for every 100 mg/dL of ethanol in the serum.

The amount of an ethanol-containing liquid that is of concern when ingested by a child depends on the alcohol concentration. However, a rough rule is that ingestion of 1 g/kg of ethanol is sufficient to raise blood alcohol to 100 mg/dL. Therefore, for a beverage such as beer (5% alcohol), approximately 10 to 15 mL/kg must be ingested before serious toxicity results. Similar estimates are 4 to 6 mL/kg for wine (14% alcohol) and 1 to 2 mL/kg for an 80-proof liquor (40% alcohol).

The management of ethanol ingestion in children begins with prompt recognition and evaluation of blood glucose. Airway or ventilatory compromise should be treated with endotracheal intubation. If seizures result from hypoglycemia, they should be promptly treated with 10 to 50% (0.25 to 1.0 g/kg) IV glucose and anticonvulsants if necessary. Warming techniques should be instituted to increase core temperature. Because ethanol is rapidly absorbed from the gut and is not adsorbed by activated charcoal, there is rarely a role for GI decontamination if the patient presents more than 1 to 2

hours after ingestion. However, if presentation is earlier, gastric lavage should be performed. Because of the high risk of drug coingestion, activated charcoal should be administered to adolescents.

Alcohol is metabolized by the hepatic enzyme alcohol dehydrogenase; its elimination rate is dose dependent. This means that the higher the blood alcohol concentrations, the longer the elimination process because the capacity of the body to produce alcohol dehydrogenase is limited. The rate of reduction in blood alcohol concentration varies from 10 to 25 mg/dL per hour. Although hemodialysis effectively enhances elimination (threefold to fourfold), it is rarely necessary. The institution of hemodialysis may be useful in those patients who have impaired liver function or a blood alcohol concentration greater than 450 to 500 mg/dL.

Isopropyl Alcohol

Poisoning with isopropyl alcohol may be particularly insidious because oral ingestion is not the only route of exposure. Children may develop severe intoxication, including coma, after topical application of isopropyl alcohol for the relief of fever (although such exposure may represent inhalational exposure rather than direct, dermal absorption). Because isopropyl alcohol is usually available in a 70% concentration by volume, ingestion of 2 to 2.5 mL/kg of this solution may lead to symptoms. Ingestion of this compound causes many of the same features as ethanol ingestion, with the additional complication of severe gastritis. Unlike the other toxic alcohols, isopropyl does not lead to metabolic acidosis. This is because its metabolite, acetone, is not an acid. However, it is approximately twice as intoxicating as ethanol, leading to greater mental status impairment at comparable serum levels. The life-threatening toxicity of isopropyl alcohol is cardiac; at high serum concentrations, direct myocardial depression occurs, leading to hypotension and shock.

In any patient with coma and an unexplained osmolar gap (the difference between calculated and observed osmolarity), isopropyl alcohol should be strongly considered. The presence of ketonuria in conjunction with the absence of metabolic acidosis effectively makes the diagnosis of isopropyl intoxication.

Isopropyl alcohol is easily removed by hemodialysis. However, hemodialysis is rarely necessary because life-threatening toxicity does not occur until serum levels exceed 400 to 500 mg/dL. Therefore, the sole indication for hemodialysis is considered hemodynamic instability, regardless of serum concentration. Treatment is otherwise supportive.

Methanol

Although methanol is used primarily as a solvent for industrial purposes, it is found in other household products, including fuels for stoves, jelled fuels for heating small dishes (e.g., Sterno), paint removers, and antifreezes.

Methanol is a model for the few drugs that, rather than being detoxified, become more toxic as they are metabolized. Thus, although methanol has little or no inherent toxicity, its metabolism by alcohol dehydrogenase to form formaldehyde and formic acid creates highly toxic compounds. Formic acid is a potent organic acid that results in severe metabolic acidosis and ocular symptoms. Fortunately, because methanol is metabolized slowly, toxicity appears after some delay, permitting time for intervention. Ingestions approaching 100 mg/kg should be considered dangerous.

The clinical effects of a methanol ingestion usually occur after a latent period of 8 to 24 hours. This delay occurs as the result of the metabolic conversion of methanol to its toxic byproducts. In large ingestions, acute methanol poisoning may cause severe CNS depression, metabolic acidosis, and a number of reversible or irreversible optic changes. In the early stages of intoxication, funduscopic examination may be remarkable for hyperemia. However, left untreated, methanol intoxication results in blindness with the appearance of a pale, avascular retina. In subacute ingestions, the nonspecific neurologic symptoms of methanol intoxication resemble those of ethanol with a “hangover,” malaise, headache, and dizziness. During recovery from a mild ingestion, occasional paresthesias of the extremities may develop.

The most significant clinical concern from a methanol ingestion is severe metabolic acidosis. This acidosis is primarily the result of formic acid production. The metabolic acidosis may be intractable and results in multiorgan dysfunction, which includes cardiac arrhythmias, seizures, and pancreatitis. The ophthalmologic abnormalities that develop during methanol intoxication may be temporary or permanent. These include blurred or double vision, changes in color perception, and sharply reduced visual acuity. Permanent abnormalities may include diminished pupillary light reaction or frank blindness. The occurrence of permanent visual defects correlates directly with the degree of metabolic acidosis, the duration of the acidosis, and the quantity of methanol ingested.

Management

The treatment of methanol ingestion consists of supportive care, administration of specific therapies, and enhancement of elimination. As with other alcohol ingestions, emesis or gastric lavage should be performed only if the ingestion occurs within 2 hours of ED presentation. Activated charcoal does not adsorb methanol effectively and is unnecessary.

Laboratory assessment includes serial arterial blood gases, electrolytes, BUN, creatinine, glucose, toxin screen, serum osmolality, and methanol level. Serum methanol concentration in milligrams per deciliter can be estimated by the formula (osmolal gap × 3).

There are three specific treatments for methanol intoxication: sodium bicarbonate, folic acid, and ethanol (or 4-methylpyrazole—see the following). Sodium bicarbonate should be administered aggressively to correct metabolic acidosis. Folate is provided because of its role in formic acid disposition within the tetrahydrofolate cycle. Customary doses are 1 mg/kg IV every 6 hours.

Because serum methanol levels of 20 mg/dL or greater are associated with toxicity if untreated, higher levels require treatment to prevent its metabolism and/or interventions to enhance its elimination. Ethanol, which has a higher affinity for alcohol dehydrogenase than methanol, is provided to “block” further production of toxic metabolites. 4-Methylpyrazole,

another alcohol dehydrogenase antagonist, shows promise as an antidote for methanol intoxication. Currently in the United States, it has received indication for use only in adult ethylene glycol poisoning (see next section). Its use in childhood methanol poisoning is still experimental at this time.

Ethanol should be instituted if the calculated or measured methanol concentration is 20 mg/dL or greater. It is administered with the goal of maintaining serum ethanol concentrations at 100 mg/dL or greater. Ethanol may be given by continuous IV infusion (600 mg/kg bolus followed by a 110 mg/kg per hour) or by oral administration. IV ethanol is preferred but has the problems of being often unavailable, hyperosmolar (preventing its administration in small veins), and requiring large fluid volumes. Therefore, the oral route must often be used in children. Given orally, any beverage that contains ethanol may be used. It must be remembered that proof designation of a beverage is twice the alcohol concentration expressed as a percentage (e.g., 80 proof equals 40% alcohol). Children must be closely monitored for the complications of ethanol administration, including mental status depression, hypoglycemia, and hypothermia.

Patients who have a blood methanol concentration of 50 mg/dL or greater require hemodialysis in addition to ethanol administration. During hemodialysis, the infusion rate of the ethanol must be doubled because hemodialysis will also remove ethanol.

Ethylene Glycol

The ingestion of ethylene glycol, although uncommon, causes significant morbidity and occasional mortality in adolescents and young adults. The toxicity of ethylene glycol, like that of methanol, is the result of drug toxification; ethylene glycol has virtually no toxicity in its parent state. However, metabolism by alcohol dehydrogenase produces several toxic intermediates, including glycolaldehyde, glycolic acid, and oxalate. These metabolites result in severe metabolic acidosis and deposition of calcium oxalate crystals in all vital organs. Therefore, ethylene glycol intoxication is associated with more systemic toxicity than methanol poisoning. Also, because ethylene glycol is metabolized more rapidly than methanol (elimination half-life approximately 3 hours), toxicity appears rapidly after ingestion.

The clinical syndrome of ethylene glycol intoxication appears in three different stages. The first stage consists predominantly of CNS manifestations and is accompanied by a profound metabolic acidosis. In this early stage, mild hypertension, tachycardia, and a leukocytosis are often present. Nausea and vomiting commonly occur, and with larger doses, coma and convulsions may appear within a few hours. Another common finding is the presence of hypocalcemia. This is believed to result from the widespread formation of calcium oxalate. Hypocalcemia may be severe enough to cause tetany and cardiac conduction disturbances. Urinalysis usually reveals a low specific gravity, proteinuria, microscopic hematuria, and crystalluria. The second distinct state is ushered in by coma and cardiopulmonary failure; it is usually the result of acidosis and hypocalcemia. The third stage usually occurs after 24 to 72 hours. Here, renal failure emerges as the dominant problem. Usually, a picture of acute tubular necrosis develops with either polyuria or anuria. Urine sediment contains blood, protein, and casts. Victims often require dialysis for extended periods and may be left with permanent renal insufficiency.

Consideration of ethylene glycol poisoning should be based either on the history or, in the absence of diabetic ketoacidosis, the presence of any of the following criteria: 1) alcohol-like intoxication without the odor of alcohol, 2) large anion-gap metabolic acidosis, 3) an elevated osmolar gap in the absence of ethanol or methanol ingestion, or 4) a urinalysis that demonstrates oxalate crystals. Another diagnostic tool is to perform a Woods' lamp examination of urine. If the ingested substance is radiator antifreeze, the fluorescein dye that it contains will be excreted in urine and fluoresce under Woods' lamp. Arterial blood gases should be obtained frequently because of the rapid evolution of metabolic acidosis. The availability of ethylene glycol levels varies by institution.

Gastric emptying is the only decontamination measure that is effective after ethylene glycol ingestion and should be performed if the patient arrives within 1 hour of ingestion. Activated charcoal negligibly adsorbs ethylene glycol and is unnecessary.

As with methanol intoxication, treatment of ethylene glycol poisoning falls into three areas: supportive care, administration of pharmacologic agents, and enhancement of elimination. Supportive care includes close monitoring of vital signs and anticipation of life-threatening events, particularly cardiac arrhythmias secondary to hypocalcemia. An ECG should be obtained, and the patient should be placed on a cardiac monitor. Intubation and mechanical ventilation should be provided as needed for control of acid-base balance.

Pharmacologic therapy is subdivided into four areas: administration of sodium bicarbonate, calcium, pyridoxine with thiamine, and ethanol or 4-methylpyrazole. Correction of acidosis should begin immediately with the administration of sodium bicarbonate and appropriate ventilation. Hypocalcemia may present as skeletal muscle disturbances (tetany) or cardiac dysfunction (prolonged Q-T interval). These may be alleviated by the prompt institution of calcium (e.g., 10% calcium gluconate, 0.3–0.6 ml/kg). Thiamine and pyridoxine are vitamins that act as cofactors in the nontoxic metabolic pathways of ethylene glycol and, theoretically, divert its metabolism toward formation of nontoxic metabolites. Therefore, thiamine (0.25 to 0.5 mg/kg) and pyridoxine (1 to 2 mg/kg) are recommended for the first 24 hours of treatment.

Ethanol administration is one option to inhibit ethylene glycol metabolism by alcohol dehydrogenase (previously discussed under Methanol). It should be initiated as soon as possible to interrupt further formation of organic acids. As with methanol, ethanol is indicated for ethylene glycol concentrations of 20 mg/dL or greater. If a serum ethylene glycol cannot be obtained in a timely fashion, it can be estimated by the formula (osmolar gap × 6), assuming that no other alcohols are contributing to the osmolar gap. For serum ethylene glycol concentrations of 50 mg/dL or greater, both ethanol and hemodialysis are recommended. Hemodialysis is also indicated if there is renal failure or severe electrolyte disturbances, regardless of the serum ethylene glycol concentration.

4-Methylpyrazole is a competitive inhibitor of alcohol dehydrogenase recently approved for the treatment of ethylene glycol poisoning in adults. Currently, use of ethanol is recommended in children; but if available, 4-methylpyrazole could

be considered therapeutically for adolescent patients. 4-Methylpyrazole is an oral or IV agent with quick onset of action, and unlike ethanol, its use is not associated with CNS depression. The loading dose is 15 mg/kg IV. The maintenance dose is 10 mg/kg every 12 hours for four doses, then 15 mg/kg every 12 hours thereafter. During hemodialysis, additional doses may be required.

Antihistamines

Antihistamines are used to treat children with allergic diseases, as sedatives and antinauseants, and to prevent motion sickness. They are present in many cough syrups, available both OTC and by prescription. Antihistamines may also be found in combination with analgesics, sympathomimetic amines, and caffeine for the symptomatic relief of the common cold. They are combined with analgesics, such as salicylamide, and an anticholinergic drug, such as scopolamine, for use as a nonprescription sleep medication. Finally, they are included in some liquid cough and cold preparations that also may contain ethanol as the solvent.

Antihistamines may depress or stimulate the CNS. Used therapeutically, CNS depression is most commonly seen as drowsiness or dizziness. With increasing doses, stimulation results in insomnia, nervousness, and restlessness. In antihistamine overdose, the CNS stimulatory effects of the drug predominate. In children, CNS stimulation causes excitement, tremors, hyperactivity, hallucinations, and with higher dosages, tonic-clonic convulsions. Children are also more likely to have signs and symptoms of anticholinergic poisoning: flushed skin, fever, tachycardia, and fixed dilated pupils. The nonsedating antihistamines terfenadine (no longer available in the United States) and astemizole have caused cardiac arrhythmia after overdose and as a result of drug–drug interactions. Cetirizine, loratadine, and fexophenadine have not produced this complication. Death from antihistamine ingestion in children usually is the result of uncontrolled seizures that progress to coma and cardiorespiratory arrest.

The treatment of antihistamine poisoning requires an accurate history of the time of ingestion and the type and quantity of drug consumed. Of particular importance is the type of drug ingested because numerous sustained-release antihistamine products are on the market. Options for GI decontamination include gastric emptying and the use of activated charcoal, as described in general previously. Overdoses with the sustained-release preparations may benefit from WBI.

Patients with seizures (see Chapter 70 and Chapter 83) require anticonvulsant therapy immediately. Preferably, short-term control may be gained using diazepam, in a dose of 0.1 to 0.2 mg/kg IV. Severely agitated patients with a clear anticholinergic toxidrome may have improved sensorium after administration of physostigmine. This is usually administered in an initial dose of 0.5 mg IV slowly over 3 minutes. The 0.5-mg dose may be repeated every 10 to 15 minutes (maximum 2 mg) to establish the effective total dose. This minimal effective dose may be repeated in several hours if necessary. It should be noted that when administered too rapidly or in too large a dose, physostigmine may precipitate seizures or asystole. Physostigmine would be particularly dangerous to use in the context of any coingestants that might affect intracardiac conduction, such as tricyclic antidepressants. A 12-lead ECG should be examined for conduction delays before physostigmine is given. Cardiac rhythm should be monitored closely during antidote infusion, and atropine should be available to reverse severe cholinergic effects that may also occur with physostigmine use. The potential risks encountered with physostigmine may favor use of a benzodiazepine for treatment of anticholinergic delirium.

Meticulous attention to supportive care is critical. Some patients may develop extreme hyperthermia and thus require aggressive measures to reduce core body temperature, including ice water baths and fans. There is little evidence for therapeutic efficacy of dialysis or hemoperfusion because of the high-plasma protein binding and large volumes of distribution for most of these agents.

Aspirin

Background

Aspirin continues to be a common cause of poisoning in children and adolescents. Salicylism is the result of acute ingestion in about 60% of cases and chronic ingestion in the remaining 40%. Clinical features of acute versus chronic salicylate intoxication often require a different management approach, depending on the manner of intoxication.

Several factors work in concert to make chronic salicylate intoxication so common. The primary factor is aspirin's elimination pattern. As serum salicylate concentrations increase, the ability of the liver to metabolize the drug diminishes until predictable, first-order elimination kinetics are replaced by unpredictable, dose-dependent, zero-order elimination. Thereafter, increments in dose are associated with disproportionate increases in serum salicylate concentration. Also, much of aspirin elimination is through urinary excretion of unchanged drug. Therefore, in the face of dehydration and decreased glomerular filtration, drug clearance is impaired even more. Finally, because aspirin is often prescribed for illnesses that may be associated with hepatic dysfunction, reduced biotransformation initiates the spiraling increase in serum concentration. Unfortunately, because chronic salicylism is associated with nonspecific symptoms (e.g., fever, vomiting, tachypnea), diagnosis may be delayed until more striking signs of intoxication appear.

Pathophysiology

The direct effects of aspirin on metabolism are multiple. Aspirin stimulates the medullary respiratory center, which leads to tachypnea and respiratory alkalosis—its hallmark. Metabolic disturbances are widespread and include hyperglycemia (or, with chronic salicylism, hypoglycemia), as well as abnormalities in lipid and amino acid metabolism. Inhibition of Krebs cycle enzymes and uncoupling of oxidative phosphorylation in conjunction with lipid disturbances create the combined lactic and ketoacidosis responsible for metabolic acidosis (which leads to the mixed respiratory alkalosis and metabolic acidosis found on the arterial blood gas). In addition to inhibiting platelet function, aspirin intoxication is associated with disturbances in vitamin K–dependent and vitamin K–independent clotting factors, resulting in a

significant coagulopathy. Mild elevations in liver enzymes are also common. Other features of aspirin intoxication include leukocytosis and electrolyte disturbances, particularly hypokalemia. Physical manifestations include fever, tachypnea, nausea, vomiting, lethargy, slurred speech, and seizures. Children with chronic salicylism are more likely to present with severe metabolic acidosis and seizures than those with acute intoxication.

The combination of respiratory alkalosis with metabolic acidosis produces an arterial blood gas that is almost pathognomonic for salicylism. Serum pH typically ranges from 7.41 to 7.55, except in severe cases in which metabolic acidosis combined with respiratory acidosis from severe CNS depression leads to pH less than 7.35, and P_{CO_2} is generally less than 30 mm Hg. Serum bicarbonate is mildly depressed, often ranging from 15 to 20 mEq/L. However, although adults may continue to hyperventilate for extended periods when poisoned with salicylates, children with mild to moderate poisoning quickly lose this respiratory drive and are more likely to present with metabolic and respiratory acidosis.

As mentioned, glucose homeostasis is seriously altered in acute aspirin poisoning. Early in the course, hyperglycemia usually occurs because of glycogenolysis and decreased peripheral use. Later, hypoglycemia may supervene as glucose stores are depleted.

Fluid and electrolyte disturbances are multifactorial, resulting in dehydration, hyponatremia or hypernatremia, and hypokalemia. Among contributing factors are increased insensible water losses through both skin and lungs, emesis, and increased renal water and potassium loss. The patient with severe salicylate poisoning may lose 4 to 6 L of water/m^2.

Clinical Findings

The initial clinical signs and symptoms, the estimate of dose ingested, and the measurement of salicylate levels all serve to gauge the severity of a given acute aspirin poisoning. However, in cases of chronic therapeutic salicylism, the clinical picture is the most useful guideline. Because of the nonspecific nature of symptoms with salicylism, the initial differential diagnosis is broad and may include diabetic ketoacidosis, iron intoxication, and ethylene glycol ingestion.

Signs and symptoms of salicylism depend on the method and severity of intoxication. Acute ingestion amounts of 150 to 300 mg/kg are associated with mild symptoms, 300 to 500 mg/kg are associated with moderate toxicity, and more than 500 mg/kg are associated with death. With mild toxicity (serum concentrations 30 to 50 mg/dL), manifestations may be confined to GI upset, tinnitus, and mild tachypnea. With moderate salicylate poisoning (serum level 50 to 100 mg/dL), more visible signs of toxicity—fever, diaphoresis, and agitation—appear. After severe salicylate poisoning (serum concentrations greater than 100 mg/dL), signs and symptoms are primarily neurologic and consist of dysarthria, coma, and seizures. Pulmonary manifestations, particularly pulmonary edema, may appear in severe cases. In victims of chronic salicylism, these same conditions appear at significantly lower serum salicylate concentrations. Death from salicylism results from severe CNS toxicity with complete loss of function in cardiorespiratory centers, leading to respiratory and/or cardiac arrest. A nomogram that strives to correlate clinical toxicity with serum salicylate levels and the time of ingestion exists. This nomogram has many shortcomings that diminish its clinical utility. Severity of salicylate intoxication is best assessed by physical examination, electrolytes, and blood gas analysis.

Management

Assessment of the victim of salicylate intoxication begins with an accurate history that identifies the patient as having acute or chronic poisoning. Laboratory assessment is extensive and includes serum salicylate concentration, electrolytes, arterial blood gas, liver function tests, CBCs, prothrombin and partial thromboplastin times, urinalysis, and an ECG. In the case of intentional ingestions by adolescents, a complete toxin screen should be obtained with particular attention to a serum acetaminophen measurement (because many OTC analgesics contain aspirin and acetaminophen in combination).

Supportive care includes assessment of ventilatory function, cardiac monitoring, and vascular access. Because aspirin overdose is associated with delayed gastric emptying and drug coalescence to form bezoars, GI decontamination should receive careful consideration in those patients who present within 4 to 6 hours of ingestion. In patients who present more than 6 hours after ingestion or in those with chronic salicylism, activated charcoal should still be administered because it may enhance postabsorptive elimination of salicylates (through GI dialysis).

Specific therapeutic goals in salicylate intoxication include correction of fluid and electrolyte disturbances and the enhancement of salicylate excretion.

Fluid therapy should be aimed at restoring hydration and electrolyte balance and at promoting renal salicylate excretion. If dehydration is significant, the fluid infusion rate should be 10 to 15 mL/kg per hour to initiate fluid resuscitation. However, fluids should be given prudently to prevent precipitation of pulmonary edema, particularly in patients with severe intoxication. For patients with symptomatic salicylate intoxication, urine alkalinization should be combined with fluid resuscitation. The administration of sodium bicarbonate, by increasing urinary pH, ionizes filtered aspirin, increasing tubular secretion and inhibiting its tubular reabsorption (ion trapping). The initial fluid is therefore designed to replace both sodium and bicarbonate losses as well as promote urine alkalinization. It should contain 5% dextrose with 50 to 100 mEq/L of sodium bicarbonate. In cases of severe acidosis, additional bicarbonate may be necessary. Because hypokalemia impairs the ability of the urine to create an alkaline urine and is exacerbated by administration of sodium bicarbonate, potassium must be added to IV fluids. Forced diuresis does not appear to enhance salicylate excretion more than the clearance accomplished by alkalinization alone. Therefore, fluids are only given as needed to restore normal hydration and to produce 1 to 2 mL/kg per hour of urine. Calcium homeostasis should also be monitored during therapy with exogenous bicarbonate. Both urine alkalinization and repetitive oral charcoal should be continued until salicylate concentration falls below 30 to 40 mg/dL and symptoms resolve.

Salicylate elimination can be also be enhanced by hemodialysis or hemoperfusion. Although hemoperfusion results in

superior clearance technique, hemodialysis is usually preferred because it permits correction of fluid and electrolyte imbalances. Hemodialysis should be reserved for seriously ill patients. Specific indications include 1) serum salicylate levels greater than 100 mg/dL after acute ingestion, 2) a serum salicylate level of 60 to 70 mg/dL or greater after chronic salicylism, 3) severe acidosis or other electrolyte disturbance, 4) renal failure, 5) persistent neurologic dysfunction, and/or 6) progressive clinical deterioration despite standard treatment.

Cardiac Drugs

b-***Adrenergic Blockers and Calcium Channel Blockers***

The approach to overdoses of these two categories of cardiovascular agents are discussed together because of similarities of clinical presentation and management approach. They are both commonly prescribed to adult patients with a variety of cardiovascular disorders, including angina and status postmyocardial infarction, hypertension, and arrhythmias. As such, experience with pediatric overdoses has been increasing in recent years.

b-Blockers (BBs) vary considerably in terms of receptor specificity and pharmacokinetics, but most overdose experience is with propranolol. Similarly, the three calcium channel blockers (CCBs) most commonly used in the United States (verapamil, nifedipine, and diltiazem) are chemically dissimilar and have varied degrees of effect on vasodilation, myocardial contractility, and sinoatrial (SA)–atrioventricular (AV) node function. Most of the clinical overdose experience is with verapamil.

Both BBs and CCBs may present with fulminant cardiovascular and neurologic findings after a large overdose. Typical presentations of both agents include marked bradycardia and hypotension; particularly with the CCBs, common additional findings are those of abnormal AV node conduction, with AV block or accelerated junctional rhythm. The CNS may also be affected, with coma and/or convulsions that occur in either category of overdoses. Metabolic disturbances include hypoglycemia with BBs and hyperglycemia and metabolic acidosis with CCBs. Bronchospasm may complicate BB toxicity further in patients with underlying reactive airway disease.

Management begins with aggressive gastric decontamination for both types of agents. Gastric lavage may be considered for the child who presents early after overdose. Activated charcoal/cathartic should be administered. Sustained-release preparations may cause prolonged effects, and WBI may be considered in this context. Bradycardia and hypotension may improve with standard treatment such as atropine, fluid boluses, and pressors; however, many cases prove resistant to these measures.

Additional therapy includes calcium infusion for the CCBs, with the recommended adult initial dose being 10 mL of 10% calcium chloride, or 30 mL of 10% calcium gluconate, which may be repeated two or three times as necessary (e.g., an initial pediatric dose of approximately 0.2 mL/kg calcium chloride or 0.6 mL/kg of calcium gluconate). Serum calcium should be monitored if several doses are used to avoid severe hypercalcemia. Glucagon increases intracellular cyclic adenosine monophosphate (cAMP) by a mechanism independent of b receptors or sites of calcium antagonist action and has been used with success to improve heart rate and blood pressure in overdoses of both types of agents. The usual adult dosing regimen is 3 to 5 mg by IV bolus, which may be repeated to a total dose of 10 mg, followed by infusion at 2 to 5 mg/hour. Such dosing translates to 50 to 150 µg/kg boluses and similar amounts per hour for pediatric patients.

Currently the role of euglycemic insulin/glucose administration is being investigated. Another experimental antidote, 4-aminopyridine, has shown promise in Eastern European studies for CCB overdose. Severe cases may also benefit from pacemaker insertion and consideration of aortic balloon pump and/or cardiopulmonary bypass. It is unlikely that hemodialysis or hemoperfusion would benefit most of these cases.

Clonidine

Clonidine is an antihypertensive that appears to have growing popularity, part of which comes from its efficacy in illnesses other than hypertension, including nicotine withdrawal and attention deficit disorder. Also, the advent of clonidine in transdermal patches has become a convenient and somewhat unique vehicle for drug administration.

Clonidine exerts its antihypertensive effect through stimulation of CNS a $_2$-adrenergic receptors. These receptors are located on presynaptic neurons in cardiorespiratory centers of the midbrain. Their stimulation results in decreased secretion of catecholamines into the synaptic cleft, resulting in decreased pulse and blood pressure. In addition, clonidine appears to interact with or modulate CNS opiate receptors; this interaction has been used to explain clonidine's efficacy in opiate withdrawal and the picture of coma and miosis that accompanies clonidine intoxication. An imidazoline compound, clonidine is related to other medications, including tetrahydrozoline and oxymetazoline—common vasoconstrictors found in nasal decongestants and ophthalmic agents.

Clonidine is an extremely potent drug with typical doses of 100 to 200 µg in adults. Therefore, ingestions of small amounts can potentially lead to significant toxicity in children. Initial toxic manifestations include altered mental status that may range from lethargy to coma. Victims also may develop significant hypothermia. In severe intoxications, coma, miosis, and respiratory depression may appear. The cardiovascular changes that accompany clonidine intoxications may range from profound hypotension and bradycardia to hypertension. Clonidine-induced hypertension occurs uncommonly and is thought to result from a-adrenergic effects at peripheral vascular receptors that override the central, antihypertensive effect. The clinical picture of clonidine intoxication typically lasts 8 to 24 hours.

Management

The treatment of clonidine intoxication requires immediate assessment of the ABCs. Because patients with severe intoxication often have coma and respiratory depression, emergency endotracheal intubation may be necessary. Also, because of blood pressure instability, vascular access should be achieved immediately for better hemodynamic control.

Hypotension should be treated with fluids and vasopressors as needed. Hypertension is generally uncommon, very transient, and would rarely require specific treatment.

GI decontamination may be ineffective more than 2 hours after ingestion because clonidine is rapidly absorbed from the GI tract. However, the potential benefits of activated charcoal administration far outweigh any of its risks.

In addition to supportive care measures, other pharmacologic interventions may be effective. Naloxone has been suggested as a specific antidotal agent after clonidine intoxication, based on case reports of improved mental status and cardiorespiratory function after its administration. However, in reported case series, there have not been consistent improvements after naloxone administration.

Because naloxone is a benign agent and may potentially improve mental status to the extent that intubation becomes unnecessary, a trial dose of 1 to 2 mg should be administered. Large amounts of naloxone (up to 8 mg) must be provided before it can be concluded that the intoxication is not responsive to this therapy. If effective, repeat doses or a continuous infusion of naloxone may become necessary. Other pharmacologic agents that have been used include yohimbine, tolazoline, and phentolamine. Specific efficacy from these agents has not been demonstrated, and they are not considered important in the treatment of clonidine intoxication.

Digoxin

Digoxin, although less commonly used, remains an important cardiac inotrope. It is still widely used in young infants with congenital heart disease as well as elderly patients with congestive heart failure. This continued popularity, its narrow therapeutic index, and the appealing color of digoxin elixir make it a source of many childhood poisoning episodes annually. Also, related agents, particularly the foxglove and oleander plants, are occasionally ingested by children, leading to a clinical picture identical to that of digoxin.

Digoxin's primary pharmacologic action is to inhibit activity of sodium-potassium adenosine triphosphatase (ATPase), which is responsible for maintaining the electrical potential of excitable tissues through transmembrane concentration of electrolytes. Therefore, the effects of digoxin are largely related to disturbances in this action.

In all victims of digoxin poisoning, two distinct pictures of toxicity exist: acute and chronic. These pictures have several differences: The victim of acute digoxin ingestion is typically a toddler who ingests a relative's medication. The toddler is generally healthy with no underlying cardiac disease. The child with chronic digoxin poisoning, however, by definition has preexisting heart disease and is likely to be taking other medications known to modulate the effects of digoxin poisoning (e.g., diuretics). Therefore, it is the latter victim who is more likely to have severe toxic manifestations after digoxin intoxication.

Digoxin pharmacokinetics are complex. After ingestion, absorption is complete within 2 to 4 hours. However, after peak serum levels are achieved, the drug is rapidly redistributed, resulting in dramatic falls in serum concentration. This has particular importance with the victim of acute digoxin intoxication who may have an initial serum digoxin concentration in the highly toxic range that falls to the therapeutic range within a matter of hours. After redistribution, digoxin elimination occurs primarily through renal excretion of unchanged drug. Therefore, any condition associated with decreased renal function may be associated with the insidious development of intoxication.

The therapeutic serum digoxin concentration (SDC) is less than 2 ng/mL. A concentration in the slightly higher range often does not correlate with clinical manifestations and may be of limited value. However, when SDC exceeds 4 ng/mL, some evidence of intoxication usually appears. This toxicity is influenced by many host factors, including patient age, underlying illness, and disturbances in serum potassium, magnesium, and calcium.

With significant intoxication, the symptoms of digoxin poisoning include nausea, vomiting, and visual disturbances. With more severe intoxication, additional symptoms, including lethargy, disorientation, electrolyte disturbances, and cardiac disturbances, appear. The hallmark of severe acute digoxin toxicity is hyperkalemia, the result of profound inhibition of sodium-potassium ATPase activity. The typical pattern of cardiac toxicity with digoxin overdose initially is prolonged atrioventricular dissociation that appears as heart block that ranges from first to third degree. These conduction disturbances can lead to the development of ventricular or supraventricular escape rhythms. In patients with chronic digoxin intoxication, these symptoms may be more striking than in those with acute, single digoxin overdoses. In fact, children with acute digoxin intoxication rarely develop life-threatening illness if their peak SDC remains below 10 ng/mL.

Management

The management of the patient with digoxin intoxication begins with evaluation of the vital signs, particularly hemodynamic status. Patients should have an ECG followed by continuous cardiac monitoring. If significant cardiac arrhythmias are already present, they are treated initially according to advanced cardiac life support protocols.

GI decontamination should include administration of activated charcoal with a cathartic. Clinical assessment requires an ECG, electrolytes (including magnesium and calcium), urinalysis, and SDC. If coingestants are suspected, a complete toxin screen should also be performed. Electrolyte disturbances should be treated aggressively because they will aggravate any digoxin-induced arrhythmias.

Some digoxin-specific antibody fragments have become specific antidotal therapy for reversing the toxic manifestations. These fragments are the result of sheep-derived immunoglobulin that is cleaved to extract only the Fab fragment. This small-molecular-weight antibody fragment is capable of avidly binding free digoxin so that a gradient results that favors digoxin removal from receptor sites into interstitial water. The effect of this gradient is that sodium-potassium ATPase function is immediately restored. The digoxin-antibody complex is then rapidly excreted in the urine. Of note, after digoxin antibody fragments are administered, SDC increases astronomically, reflecting bound, inactive digoxin that has diffused

into the vascular compartment.

These antibody fragments are indicated in the following circumstances after digoxin poisoning: 1) progressive signs and symptoms of intoxication, 2) life-threatening cardiac arrhythmias, or 3) severe hyperkalemia (defined as a serum potassium of 5.5 mEq/L or greater). The dose of antibody fragments is calculated on the basis of ingested digoxin dose (in the case of acute intoxication) or on the basis of SDC (in the case of chronic intoxication). Each 40-mg vial of digoxin-Fab will bind 0.6 mg of digoxin. The total dose of Fab needed (in vials) may be estimated by dividing a known ingested dose by 0.6, or calculated for the steady state context as body load of digoxin:

$$\text{No. of vials} = \frac{\text{SDC (ng/mL)} \times \text{wt (in kg)}}{100}$$

Complications from the administration of antibody fragments are low and consist of an allergic reaction (in approximately 0.6% of patients), precipitation of congestive heart failure (secondary to the abrupt loss of digoxin's inotropic action), and rebound hypokalemia. These complications should be anticipated and treated accordingly.

Disc Batteries

The development and widespread use of disc batteries in home toys and appliances has led to a burgeoning increase in the rate of disc battery ingestions in young children. A somewhat unusual feature of these ingestions is the frequency among children 4 to 8 years of age who often ingest them accidentally or out of curiosity. Children with hearing aids form another group at particular risk.

Disc batteries contain a number of potentially toxic substances, including mercury, lithium, and potassium hydroxide. However, their toxic potential is primarily confined to their corrosive action when they are in contact with a mucosal surface for an extended time. Thus, disc batteries that are placed in nasal or aural cavities should be removed immediately.

With the history of a disc battery ingestion, all patients should receive an immediate chest radiograph. This is because disc batteries that are retained in the esophagus act as local corrosives, leading to esophageal injury or perforation. If the disc battery is found in the esophagus, it must be removed immediately. If the battery is beyond the esophagus, the patient may be discharged.

The natural history of disc battery ingestion is that the object is usually expelled within 72 hours of ingestion without inducing symptoms. Therefore, the treatment of these ingestions involves no intervention. Rather, parents are asked to monitor stools for 3 days to document passage of the battery. In the event the battery is not passed within that time, an abdominal radiograph should be obtained to confirm that the battery has not been incarcerated in a bowel loop. If the battery is still in the gut, there is continued observation. Surgical removal of these objects is almost never necessary.

Foods/Fish

In addition to drugs and medications and household products and plants, toxic ingestions may occur through normal diet when the ingested product contains a toxin that is preformed by microorganisms. The largest class of such toxins are the enterotoxins produced by organisms that include *Shigella, Salmonella, Yersinia, Escherichia coli, Staphylococcus, Bacillus cereus, Clostridium, Vibrio, and Clostridium botulinum*. After this large group of toxins, the next most common cause of foodborne intoxications results from the ingestion of contaminated marine life.

The general approach to the patient with diarrhea and infectious causes of the gastroenteritis syndrome is discussed in Chapter 19 and Chapter 84, respectively. The association of hemolytic uremic syndrome with GI infection by *E. coli* O157:H7 is discussed in Chapter 84 and Chapter 86. Plant toxicity is discussed later in this chapter under its own heading. Here, the common causes of acute bacterial toxin-induced food poisoning are outlined, followed by a discussion of marine-related illness.

When similar GI symptoms occur in a group of persons who share the same meal or the same food on separate occasions, the emergency physician may consider the possibility of foodborne disease. Detailed epidemiologic investigations are usually beyond the capacity of the ED setting, but the hospital infection control officer and/or local health department can often be helpful.

Staphylococcal food poisoning is probably the most common cause of such cases in the United States. The heat-stable toxins typically produce acute abdominal pain, nausea, vomiting, and diarrhea within 1 to 6 hours of eating the contaminated meal. The illness is usually self-limiting, although occasional patients develop severe symptoms and dehydration.

Other bacterial toxin-induced diarrheal food poisonings include those secondary to *Bacillus cereus, Clostridium perfringens*, and *Vibrio* species. The onset of clinical illness and usual food sources of these and staphylococcal disease are outlined in Table 88.12. All these illnesses are generally self-limiting, and treatment is supportive, with careful attention given to fluid and electrolyte status in unusually severe cases (e.g., the rare occurrence of cholera in the United States).

Table 88.12. Common Causes of Diarrheal Food Poisoning in the United States

Infant botulism shares many pathophysiologic and clinical features with foodborne botulism (it is discussed in detail in Chapter 83). The etiology of the foodborne disease differs, of course, in that preformed toxin is ingested at the time of consuming contaminated food, typically improperly home-canned, low-acidity vegetables (e.g., potatoes, onions, beans) or poorly refrigerated pot pies or meats. The incubation period is usually 12 to 36 hours, with initial GI symptoms soon followed by weakness, malaise, and then cranial nerve symptoms, particularly diplopia, dysphagia, and dysarthria. The neurologic examination is notable for normal mental status and symmetric ocular findings, such as ptosis, lateral rectus weakness, and pupillary abnormalities.

Diagnosis should be suspected clinically and may be buttressed with positive serum or stool analyses for botulinum toxin and suggestive electromyelograph (EMG) findings. The management of foodborne botulism shares with infant botulism the requirement for meticulous, intensive supportive care, with special attention to airway and ventilatory status. In addition, unlike the case in infant botulism, administration of trivalent antitoxin is recommended for all symptomatic patients. This antitoxin and details regarding its optimal use are available from the Centers for Disease Control and Prevention (404-639-2206 during workdays; 404-639-2888 after hours).

Scombroid Poisoning

Scombroid poisoning is an intoxication that occurs shortly after ingestion of spoiled fish from the Scombroidea family (e.g., tuna, bonita, skipjack), as well as ingestion of non-Scombroidea fish (e.g., bluefish, mahi-mahi). The ingested toxin(s) has not been completely characterized, but large quantities of histamine are invariably found in fish that produce scombroid.

The clinical picture of scombroid poisoning consists of sudden-onset headache, facial flushing, a peppery taste in the mouth, dizziness, nausea, and vomiting. An urticarial eruption with pruritus may develop. In its extreme, victims may develop tachycardia, bronchospasm, respiratory distress, and hypotension.

In patients with severe symptoms, treatment is directed toward ensuring adequate ventilation and hemodynamic stability. Fluids and vasopressor support may be needed to treat hypotension. Pharmacologic treatment of scombroid poisoning includes administration of antihistamines, corticosteroids, and if necessary, adrenergic agents. Both diphenhydramine and cimetidine have been used successfully to treat the symptoms of scombroid poisoning. In the event of severe bronchospasm, other bronchodilators, including inhaled b_2 agonists may be necessary adjuncts.

Ciguatera

Ciguatera is an illness endemic to the South Pacific but is considerably less common in the continental United States, where it is largely confined to the lower Atlantic states. However, because it does occasionally appear in the United States or may occur in recent visitors from endemic areas, its clinical manifestations should be recognized.

Ciguatera results from ingestion of a toxin elaborated by the dinoflagellate, *Gambierdiscus toxicus.* This parasite is ingested by small fish that begin to concentrate the toxin. As predators ingest those small fish, the toxin ascends the food chain until ingested by humans. The fish that most commonly harbor ciguatoxin include barracuda, grouper, red snapper, and parrot fish. The physiologic actions of ciguatoxin are primarily neurologic. The toxin decreases CNS concentrations of gamma-aminobutyric acid (GABA) and dopamine. This action occurs in conjunction with sodium channels being "locked-open," permitting unrestricted sodium ingress.

The clinical picture of ciguatera poisoning begins 4 to 36 hours after ingestion of contaminated fish. After a brief period of nausea and vomiting, victims develop paresthesias, particularly perioral, or weakness. A hallmark of ciguatera toxin is the reversal of hot–cold sensation. In severe cases, CNS dysfunction, including coma, may appear. Toxic manifestations may persist for days to months after significant exposure.

The diagnosis of ciguatera intoxication is clinical, based on the history of ingestion of a fish known to carry this toxin. Because symptoms appear many hours after ingestion of contaminated fish, there is no clear role for GI decontamination.

Management of ciguatera is supportive. Primary attention should be paid to CNS status and its effects on airway and ventilation. IV mannitol has shown great promise in reversing many of the neurologic manifestations, particularly coma. It is administered in a dose of 0.5 to 1.0 g/kg.

Paralytic Shellfish Poisoning

The dinoflagellate *Gonyaulax* is responsible for elaborating the toxin (saxitoxin) that causes paralytic shellfish poisoning (PSP). The name red tide is based on the characteristic red pigment of the *Gonyaulax.* Paralytic shellfish poisoning appears in large bloom between the months of May and October and is found primarily along the eastern seaboard (although blooms have increased across the world in recent years and may be found on either U.S. coast). The animals that ingest and concentrate this toxin are primarily bivalve shellfish, including mussels, clams, oysters, and uncommonly, scallops. The toxin, saxitoxin, is capable of reversibly binding neuronal sodium channels, resulting in depolarization disturbances. The toxin is heat stable.

After ingestion of contaminated shellfish, victims quickly develop GI distress with nausea and vomiting. This is followed

by generalized paresthesias, cranial nerve disturbances, and weakness. In severe intoxications, cardiorespiratory failure may ensue.

Treatment of PSP is supportive. Victims may require ventilatory support until the intoxication resolves over hours to days.

Household Cleaning Products and Caustics

Household Cleaning Products

Background

Until the early 1950s, cleaning products used for home laundering, household maintenance, and personal hygiene were usually some form of soap. However, soap has the disadvantage of forming an insoluble precipitate that clings to surfaces such as skin, bathtubs, clothes, and dishes. Most products today use synthetic detergents that do not form such precipitates. Soap is one type of surface active agent ("surfactant"). A "detergent" is any cleansing product. However, in common use, the word *detergent* has come to mean a household cleaning product that is based on nonsoap surfactants, used mainly for laundering and dishwashing. Other cleaning products include disinfectant cleaners; cleaners for drains, ovens, and toilet bowls; bleaches; and ammonia. These agents are of concern because their accessibility to children makes them commonly involved in human ingestions. Furthermore, animal studies and clinical observations have shown some of these products to be injurious after topical applications.

Each year, about 6% of reported unintentional ingestions involve soap, detergents, or cleaners; 2 to 3% involve household bleaches; and 1 to 2% involve corrosive acids and alkalis (e.g., ammonia, drain cleaners). Most of these cases involve children less than 5 years of age, of whom only 1 to 2% of those ingesting noncorrosive products require hospitalization. Most such exposures occur inside the home, while the product is in use. In almost half of these cases, the product had been transferred out of its original container, often unfortunately, to an empty glass or soda bottle.

Caustics

Background and Pathophysiology

Many agents possess corrosive potential when they are placed in direct contact with biologic tissues. These agents, collectively referred to as corrosives, may be acidic, alkaline, or rarely, have neutral pH (e.g., silver nitrate, concentrated hydrogen peroxide). Essentially all corrosives found in the home are acids or alkalis. Strong alkalis and acids cause direct destruction of tissue but with differing histopathologic patterns. Acids produce coagulation necrosis that usually causes superficial damage, rather than deep, penetrating burns. Alkalis, in contrast, cause a deep and penetrating liquefaction necrosis, that often has severe consequences, such as esophageal perforation. Such deep burns are often associated with severe scarring and, ultimately, with stricture formation. Acid corrosives include the mineral acids, such as hydrochloric, sulfuric, nitric, and hydrofluoric acids. Common household products that contain acid corrosives include toilet bowl and drain cleaners. However, many home accidents involve acids brought home (often in unmarked food containers) by parents from the workplace.

Alkali caustics are found in several household products. Sodium hydroxide (lye), which is available in crystalline and liquid forms, is used primarily as an oven cleaner or drain pipe cleaner (e.g., Drano, Liquid Plumr, Easy-Off). Sodium hydroxide is also an ingredient in the tablets used to test for glycosuria (Clinitest). Other products may contain alkaline corrosives, including powdered laundry and dishwasher detergents.

Clinical Findings

Ingestions of acid and alkali corrosives cause immediate severe burning of exposed surfaces, usually with intense dysphagia. Associated glottic edema may cause airway obstruction and asphyxia. Severe acid ingestions most often cause gastric necrosis and may be complicated by gastric perforation and peritonitis. With alkalis, severe damage is more commonly found in the esophagus; deep-tissue injury may quickly lead to esophageal perforation, mediastinitis, and death. As already noted, alkalis also produce severe esophageal strictures in survivors.

Management

The initial step in the management of a caustic ingestion is to determine whether the agent is, in fact, corrosive and, if so, whether it is an alkaline or acid corrosive. Many products that are thought to have corrosive potential (e.g., household bleach) are simple irritants and do not require intervention. Identification of ingredients and their corrosive potential can be found in texts on household products or through consultation with a regional poison control center.

The approach to management of cleaning products and caustic ingestions, as outlined in Figure 88.3 and Figure 88.4, begins with rapid clinical assessment of cardiorespiratory function, neurologic status, and evidence of GI hemorrhage. Life support measures may be needed emergently to secure the airway and to treat shock or metabolic acidosis. As noted previously, most patients with significant exposures develop symptoms early and may appear critically ill. However, even patients with minimal symptoms and the absence of oral lesions may have significant esophageal injury; thus, all patients with a convincing history of exposure to a caustic substance need esophagoscopy to be evaluated fully for the presence of esophageal burns.

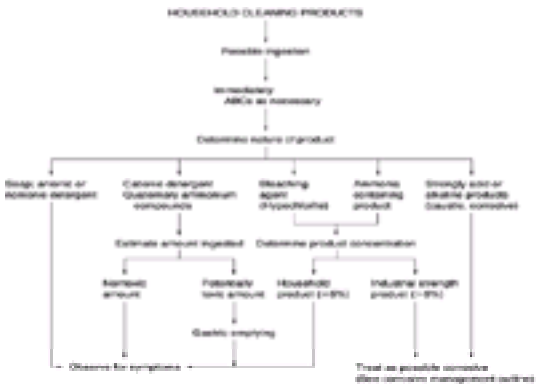

FIGURE 88.3. Algorithm for management of household cleaning product ingestion. (Modified with permission from Temple AR, Lovejoy FH Jr. Cleaning Products and Their Accidental Ingestion. New York: Soap and Detergent Association, 1980.)

FIGURE 88.4. Algorithm for management of corrosive ingestion. (Modified with permission from Temple AR, Lovejoy FH Jr. Cleaning Products and Their Accidental Ingestion. New York: Soap and Detergent Association, 1980.)

Simple dilution has been suggested as being safe and potentially diagnostic. However, there are several reasons why this should not be attempted. First, in the event esophageal injury or perforation has occurred, fluids may extravasate, inducing severe mediastinitis. Also, because esophagoscopy is the diagnostic procedure of choice in establishing the extent of injuries, an empty stomach is necessary for minimizing the risks of anesthesia. Finally, if administered fluids are alkaline or acidic, an exothermic reaction may occur that also can worsen esophageal injury. No GI decontamination is conducted after the ingestion of corrosive agents.

If the eyes are involved (something that should always be considered if a caustic has splashed on the face), copious irrigation should be provided and carried out for at least 15 minutes, with longer periods for crystalline caustics. The physician should perform pH testing of fluids in the ocular cul-de-sac after irrigation to confirm that corrosives have been neutralized; the normal pH of tears is 7. Alkali eye injuries require urgent ophthalmologic consultation. Skin contamination also deserves prolonged rinsing with water and removal of contaminated clothing. Irrigation should continue until the skin is free of alkali, as determined by disappearance of the soapy sensation.

The next phase of management calls for further evaluation. All exposed surfaces, especially the oropharynx, should be examined scrupulously. A CBC and chest radiograph should be obtained, the latter particularly if any respiratory signs or symptoms are noted. Referral to an endoscopist must be done on an immediate basis.

Analgesic therapy may be necessary for severe pain. An IV line should be established if not previously done for basic life support. Conflicting data regarding the role of corticosteroids in the treatment of corrosive esophageal injury exist. First-degree burns typically heal without long-term sequelae. Circumferential second-degree burns may be less likely to stricture after steroid administration; therefore, corticosteroids with empiric antibiotics may be considered in this scenario. Third-degree burns are likely to scar despite treatment, and administration of steroids in this situation may provide more risk than benefit. The consulting otolaryngologist may elect to administer steroids in select patients based on endoscopic findings. All patients are admitted for supportive care and monitoring for acute complications such as mediastinitis, pneumonitis, and peritonitis.

The long-term management of survivors with severe caustic esophageal burns and stricture formation is complex, involving many surgical, medical, and psychologic stresses to the patient. Years of repeated bougienage may be necessary, and some patients will require esophagectomy with colonic interposition in an effort to replace the destroyed esophagus. The patient may be incapable of tolerating solid foods for prolonged periods.

Hydrocarbons

Hydrocarbons are carbon compounds that become liquid at room temperature. The term *hydrocarbons* is somewhat confusing and is often used interchangeably with the term *petroleum distillates.* However, whereas all petroleum distillates are hydrocarbons, not all hydrocarbons are petroleum distillates (e.g., pine oil). Hydrocarbons can be found in solvents, fuels, household cleaners, and polishes.

Hydrocarbons are typically divided into three categories: the aliphatic hydrocarbons, the aromatics, and the “toxic”

hydrocarbons. The aliphatic hydrocarbons are petroleum distillates and are found in such household products as furniture polish, lamp oils, and lighter fluids. The aromatics are cyclic structures and include toluene, xylene, and benzene. These agents are found in solvents, glues, nail polish, paints, and paint removers. The “toxic” hydrocarbons consist of a broad class of substances that possess no specific profile of toxicity. These agents include halogenated hydrocarbons and hydrocarbons that serve as vehicles for toxic substances such as pesticides.

The major toxicity of hydrocarbons varies from class to class. However, the feature that these agents have in common is a low viscosity that permits them to spread freely over large surface areas, such as the lungs, when aspirated. This property (plus their solvent actions) leads to a necrotizing, often fatal chemical pneumonitis when these compounds are aspirated. The high volatility of these substances is responsible for alterations in mental status, including narcosis, inebriation, and frank coma. In addition to these toxicities, the solvents possess additional toxicities (see Inhalants), including the risk of bone marrow injury (in the case of benzene). Finally, with the toxic hydrocarbons, additional toxicities may occur as a result of actions such as cardiotoxicity or as a result of the pharmacologic properties of the other agents contained within these compounds. The major toxicity of hydrocarbons is classified in Table 88.13.

Nontoxic (unless complicated by gross aspiration)
Asphalt, tars
Mineral oil
Liquid petrolatum
Motor oil, axle grease
Baby oils, suntan oils
Systemic Toxicity
Halogenated (carbon tetrachloride, trichloroethane)
Aromatic (benzene, toluene, xylene)
Additives (camphor, organophosphates, heavy metals)
Aspiration Hazard (without significant systemic toxicity unless ingested in massive quantity)
Turpentine
Gasoline
Kerosene
Mineral seal oil (furniture polish)
Charcoal lighter fluid
Cigarette lighter fluid
Mineral spirits

Table 88.13. Classification of Hydrocarbons

The amount of a hydrocarbon that has been ingested by a pediatric patient is often difficult to quantify. However, any degree of aspiration results in signs, including coughing, gagging, or tachypnea. Less than 1 mL of some compounds, when aspirated directly into the trachea, may produce severe pneumonitis and eventual death. When ingested, these compounds are poorly absorbed from the GI tract. In a retrospective study of hydrocarbon ingestions in children, most children (880 of 950) developed no symptoms.

The major aspiration hazard associated with hydrocarbons can be quantified by their viscosity. Products with a viscosity of 150 to 250 Saybolt Seconds Units (SSU), such as oils, pose a small risk of chemical pneumonitis; those with a viscosity under 60 SSU, such as furniture oils or polishes, have a high aspiration hazard.

Clinical manifestations of hydrocarbon ingestion depend largely on the specific profile of toxicity of the ingested substances. All these agents cause significant GI irritation that may be associated with nausea and bloody emesis. CNS effects may range from inebriation to coma. Hemolysis with hemoglobinuria has been reported after significant ingestions. Finally, hydrocarbon ingestion may be associated with the development of fever and leukocytosis in up to 15% of patients in the absence of clinically evident pneumonitis.

Because most hydrocarbons cause clinical toxicity only when aspirated, the mainstay of treatment is to leave ingested compounds in the gut (when possible) and to prevent emesis or reflux. Gastric emptying is generally reserved only for those compounds with the potential for systemic toxic effects (Table 88.13). These compounds include the halogenated hydrocarbons (e.g., trichloroethane, carbon tetrachloride) and aromatic hydrocarbons (e.g., toluene, xylene, benzene). In addition, all petroleum distillates, regardless of their viscosity, should be evacuated from the GI tract if they contain dangerous additives, such as heavy metals or insecticides. Finally, gastric emptying should be considered when a huge amount of hydrocarbon has been ingested, as in a suicide attempt.

Patients who have aspirated may exhibit immediate choking, coughing, and gagging as the product is swallowed and then vomited after ingestion. Aspiration of the product may also occur at the time of the initial swallowing. ED management of these patients is outlined in Figure 88.5. If the patient has any cough or respiratory symptoms upon arrival to the ED, a chest radiograph should be obtained immediately. Because there is a gradual evolution of abnormal radiographs, an initially negative chest radiograph should be repeated at 4 to 6 hours after ingestion. All patients with abnormal chest radiographs or persistent respiratory symptoms after 4 to 6 hours of ED observation should be admitted for observation. Patients who are asymptomatic after this period of observation may be discharged. Because pneumonitis occasionally appears 12 to 24 hours after exposure, detailed instructions should be provided for warning signs of respiratory dysfunction.

FIGURE 88.5. Management of petroleum distillate ingestion. (Modified with permission from Shannon M. Petroleum Distillate Poisoning. Harwood-Nuss A, ed. Philadelphia: JB Lippincott, 1991.)

Treatment of hydrocarbon pneumonitis consists of airway control if there is mental status depression and intubation if ventilation is impaired. Adult respiratory distress syndrome may ensue, and heroic measures such as extracorporeal membrane oxygenation has been successfully employed. Antibiotics should not be used prophylactically but should be reserved for specific infections, should they develop. The use of corticosteroids in the treatment of aspiration from hydrocarbons has been associated with increased morbidity and is not recommended. In the event of hypotension or bronchospasm, epinephrine is contraindicated because hydrocarbons are known to cause ventricular irritability and predispose to fibrillation, an effect that is exacerbated by catecholamines.

Iron

Background

Iron poisoning is one of the most common, potentially fatal intoxications in children. Most serious childhood poisonings result from ingestion of prenatal vitamins or ferrous sulfate tablets (which unfortunately look much like candy) that were intended for adults. A common scenario is that the victim is a toddler whose mother has just had a new baby; the increased demands on the mother's attention and almost universal prescription of iron to postpartum women combine to set the stage for this ingestion. In addition, numerous exposures result from ingestion of iron-fortified children's vitamins, but these tend to be far less toxic.

Sufficient data to define a safe lower limit for toxic iron ingestions are not available. As little as 20 mg/kg of elemental iron has caused toxicity, whereas ingestions of more than 50 mg/kg often produce toxic effects. Of course, it is often impossible to know the exact number of tablets ingested. As few as ten 300-mg $FeSO_4$ tablets have been fatal to a young child. Furthermore, the elemental iron content of whole bottles of chewable vitamins is usually about 1200 mg. Legislation now demands child-proof caps for vitamin bottles that contain more than 250 mg of elemental iron.

Pathophysiology

Iron toxicity results from the direct caustic effect on the GI mucosa and the presence of free iron in the circulation. Pathologic changes include hemorrhagic necrosis of stomach and intestinal mucosa and lesions in the liver that range from cloudy swelling to areas of complete necrosis. Occasionally, pulmonary congestion and hemorrhage are noted. Excess free iron is believed to act as a mitochondrial poison, particularly in the liver, with resulting changes in cellular energy metabolism and the production of metabolic acidosis.

Clinical Manifestations

The clinical effects of iron poisoning are classically divided into four phases. Phase I represents the effects of direct mucosal injury and usually lasts 6 hours. Vomiting, diarrhea, and GI blood loss are the prominent early signs; when severe, the patient may lapse into early coma and shock caused by volume loss and metabolic acidosis.

Phase II, which lasts from 6 to 24 hours after ingestion, is marked by diminution of the GI symptoms. With appropriate therapy to replace fluid and/or blood losses, the child may seem relatively well and often goes on to full recovery without any subsequent symptoms. However, this remission may be transient and may be followed by phase III, characterized by cyanosis and profound metabolic acidosis. The child may develop coma, seizures, and intractable shock. This phase is believed to represent hepatocellular injury with consequent disturbed energy metabolism; elevated levels of lactic and citric acids are noted in experimental iron poisoning before cardiac or respiratory failure occurs. Jaundice and elevated transaminases are noted in this phase. A phase IV has been described in survivors of severe iron poisoning, marked by pyloric stenosis that results from scarring and consequent obstruction.

Laboratory abnormalities often associated with severe iron intoxication include metabolic acidosis, leukocytosis, hyperglycemia, hyperbilirubinemia and increased liver enzymes, and a prolonged prothrombin time. If fluid loss is significant, there will be hemoconcentration and elevated BUN. Abdominal films may show radiopaque material in the stomach, but the absence of this finding does not indicate a trivial ingestion.

Management

All children alleged to have ingested iron are potentially at significant risk for life-threatening illness. However, severe iron poisoning is uncommon compared with the number of children who develop only mild symptoms or remain entirely

asymptomatic. Thus, the emergency physician needs an approach that encompasses the response to the severely poisoned child and to most who will remain well.

As noted earlier, the amount of iron ingested is often hard to quantify, and minimal “safe” amounts are not well established. Serum iron levels have been shown to correlate with the likelihood of developing symptoms (usually a reflection of the serum iron that exceeds the iron-binding capacity and results in free-circulating iron). Usually, iron levels below 350 µg/dL, when drawn 3 to 5 hours after ingestion, predict an asymptomatic course. Patients with levels in the 350 to 500 µg/dL range often show mild phase I symptoms but rarely develop serious complications. Levels higher than 500 µg/dL suggest significant risk for phase III manifestations. However, the serum iron determination is not always available on a stat basis.

Although serum iron levels are useful, toxicity from iron overdose remains a clinical diagnosis. Ill patients require vigorous hydration and support. Children who are completely asymptomatic 6 hours after ingestion are unlikely to develop systemic illness. Among laboratory studies, the presence of metabolic acidosis or acidemia probably best correlates with toxicity. A white blood cell (WBC) count higher than 15,000/mm ^{3} paired with a serum glucose higher than 150 mg/dL is fairly predictive of elevated serum iron levels (but are not sensitive). Radiopaque material on abdominal radiograph also suggests significant absorption of iron. Measurement of the total iron-binding capacity is no longer believed to be useful in acute management. With these observations in mind, it is possible to construct a protocol for the triage and initial management of the patient who has ingested a possibly toxic amount of iron (Fig. 88.6).

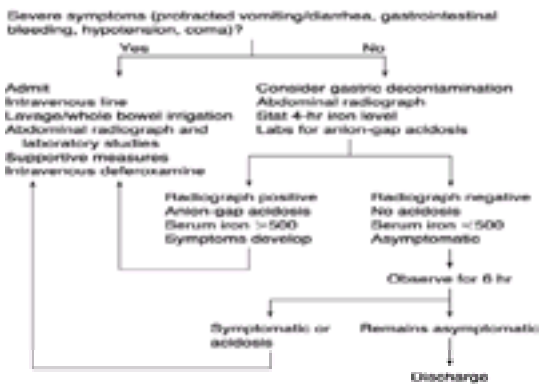

FIGURE 88.6. The initial approach to the patient ingesting a possibly toxic dose of iron.

Categorization

Patients who arrive with severe early symptoms, including vomiting, diarrhea, GI bleeding, depressed sensorium, or circulatory compromise require urgent, intensive treatment in the ED. The first priority is to obtain venous access. Simultaneously, blood is drawn for CBC, blood glucose, electrolytes, BUN, liver function tests, serum iron, and type and crossmatch. GI decontamination is begun as detailed in the following. Blood pressure should be supported with normal saline or Ringer's lactate (see Chapter 3). Specific chelation therapy with IV deferoxamine is begun immediately in all severely poisoned patients. An abdominal radiograph should be obtained as soon as possible after GI decontamination to determine its efficacy and to investigate for the presence of iron-pill concretions.

Patients with only mild vomiting and diarrhea in the early postingestion period still need urgent treatment but usually do well. Again, GI decontamination strategies should be promptly addressed. Blood studies, as previously noted, are drawn, and parenteral deferoxamine therapy is begun.

The asymptomatic patient who has presumably ingested iron in toxic amounts and who presents within 30 minutes of ingestion may benefit from ipecac-induced emesis if spontaneous vomiting has not occurred. If serum iron levels are available, blood should be sent for this study, an abdominal radiograph should be obtained, and the patient should be observed for 6 hours. An iron level of less than 350 µg/dL taken 3 to 5 hours after ingestion in an asymptomatic patient with a normal radiograph suggests that the patient is at minimal risk and may be discharged. Iron levels greater than 500 µg/dL, the development of any symptoms, or a positive radiograph should lead to admission and management as previously described for the mild to moderately ill patient.

When serum iron levels are not available on an emergency basis, the WBC count and serum glucose may be used as screening laboratory tests, in addition to the electrolytes and abdominal radiograph. Patients are observed for 6 hours in the ED. Those who have normal screening tests and remain asymptomatic may be discharged. Patients with abnormal screening laboratory tests should have an iron level sent for later reference. Acidotic or symptomatic patients should be admitted and treated with deferoxamine. Although rarely used today, the deferoxamine challenge test (50 mg/kg IM up to a maximum of 1 g) may be useful when iron levels are unavailable and the patient's screening laboratory studies and clinical status are borderline. The appearance of a pinkish-orange (vin rose) color to the urine indicates the presence of iron-deferoxamine complex and correlates well with a significantly elevated serum iron level. A positive deferoxamine challenge also mandates admission and further treatment. Patients asymptomatic after 6 hours with a negative challenge may be discharged.

Treatment

The treatment for acute iron poisoning includes efforts to decrease absorption and hasten excretion, as well as appropriate supportive care.

Ipecac-induced emesis may be helpful in preventing absorption, if the patient presents within 30 minutes, has a normal level of consciousness, and has an intact gag reflex. However, most children with toxic iron exposures will exhibit spontaneous vomiting. Furthermore, the empiric early use of ipecac may interfere with the clinical interpretation of early signs of iron toxicity, as detailed previously. Activated charcoal is not effective in binding iron salts. For serious poisonings, gastric lavage with normal saline can be considered in patients who present early in the hope of minimizing any direct mucosal injury caused by residual particulate matter and possibly contributing to the dissolution of pill concretions.

The mainstay of GI decontamination currently for iron-poisoned patients is the early and aggressive use of WBI (p. 894). This approach is believed effective in decreasing iron absorption and additionally in breaking up pill concretions that might be a risk for direct mucosal injury. As noted previously, an abdominal radiograph should be obtained early in the evaluation of symptomatic patients, but after gastric emptying procedures if these have been used. If this study demonstrates significant radiopaque material and the patient's condition allows it, WBI should be instituted for at least 4 to 6 hours. A few hours of WBI (until rectal effluent is clear) may be indicated in symptomatic cases even without definite radiographic findings to hasten elimination of residual iron pill particles or “sludge,” as long as there is no evidence of peritonitis or perforation. In patients with considerable initial radiographic findings, particularly pill concretions, a follow-up radiograph should be obtained to assess the adequacy of bowel cleansing. Further options of gastroscopy or even gastrotomy are reserved as last resorts to effect iron pill removal. Large clumps of coalesced iron tablets in the stomach or duodenum have led to severe hemorrhagic infarction of these viscera with subsequent perforation, peritonitis, and death. As previously noted, even in such patients who survive the acute phase, there is considerable risk of subsequent pyloric or bowel stenosis with obstruction, usually 4 to 6 weeks after ingestion. In this regard, we also urge early pediatric surgical consultation for patients in the first few days after ingestion who show any evidence of peritoneal irritation.

Chelation therapy with parenteral deferoxamine enhances the excretion of iron. The most efficacious route is a continuous IV infusion, and the maximum recommended dose is 15 mg/kg per hour (maximum daily dose 360 mg/kg, up to 6 g total). A higher infusion rate has been associated with hypotension but may be necessary (in conjunction with blood pressure support) for severe ingestions. Chelation is continued until the serum iron level returns to normal, metabolic acidosis has resolved, the patient is clinically improved, and the urine color returns to normal. The dose of deferoxamine may be titrated down in concert with the patient's clinical response and fall in iron levels.

Once the patient has been stabilized initially, further problems may include hypotension, profound metabolic acidosis, hypoglycemia or hyperglycemia, anemia and colloid loss caused by GI hemorrhage (after equilibration), renal shutdown resulting from shock, and hepatic failure with an associated bleeding diathesis. The maintenance of an adequate urine output is critical to prevent renal failure and to foster excretion of the iron-deferoxamine complex. If renal failure supervenes, chelation may be continued with concurrent dialysis because the complex is dialyzable.

Isoniazid

The recent increase in the incidence of tuberculosis has escalated the use of isoniazid (INH), the primary treatment for this infection. Thus, cases of INH poisoning in children continue to increase. Because this poisoning can be recognized promptly and specific antidotal therapy is available, a review of its toxicity is warranted.

The greatest increases in tuberculosis have been found in select groups, including new immigrants, the homeless, and people with acquired immunodeficiency syndrome (AIDS). Therefore, these people and their children form the primary at-risk group for such toxic ingestions.

Even when taken appropriately INH has many actions that can lead to clinical toxicity. These include hepatic dysfunction and interactions with foods such as those containing tyramine. However, its greatest toxicity appears after acute single ingestions of more than 20 mg/kg in children or more than 1.5 g in an adult.

INH's mechanism of toxicity is incompletely understood but seems to involve its potent effect at reversing the biologic activity of vitamin B_6 (pyridoxine). This action, as well as other effects on the synthesis of catecholamines and the neurotransmitter GABA, provides an explanation for the epileptogenic toxicity of the drug. INH also prevents hepatic conversion of lactate to pyruvate.

In overdose, the hallmark of INH poisoning is the triad of seizures, metabolic acidosis, and coma. Seizures induced by INH are typically generalized and appear to have a rhythmic recurrence. They are generally difficult to treat; patients usually remain comatose between seizures. The metabolic acidosis of INH can be severe; pH values of as low as 6.4 have been reported. This places INH on the list of substances associated with the development of high-anion gap metabolic acidosis (see MUDPILES mnemonic on p. 892). Of all these drugs, only INH possesses seizures as a prominent characteristic. Interestingly, in animal models of INH poisoning, metabolic acidosis does not occur if seizures are prevented through paralysis. Finally, the coma of INH intoxication can be severe and prolonged.

Because of the striking clinical picture of INH poisoning, diagnosis is often easily made on the basis of demographic characteristics and clinical manifestations. INH is not usually detected on routine toxin screens, and serum concentrations are of little value in acute management. Laboratory tests that are important in initial assessment include arterial blood gas, electrolytes, liver function tests, creatine kinase, and urinalysis.

Management

Management of INH intoxication begins with advanced life support. Because of seizures and coma, airway protection and ventilation are typically necessary. Cardiac monitoring should be initiated to monitor for the development of cardiac

arrhythmias (resulting from severe metabolic acidosis).

Aggressive GI decontamination is warranted if ingestion occurs within 2 to 3 hours of ED arrival. Syrup of ipecac is contraindicated after INH overdose because of the risk of rapid-onset seizures. Therefore, gastric emptying, if performed, is best accomplished by orogastric lavage. Activated charcoal with a cathartic should be administered. Theoretically, giving multiple doses of activated charcoal to enhance postabsorptive elimination of INH is advantageous.

Pharmacologic treatment for INH intoxication includes sodium bicarbonate, anticonvulsants, and pyridoxine. Sodium bicarbonate is provided as needed to restore serum pH to normal. In treating seizures, effective anticonvulsants include the benzodiazepines or phenobarbital (both of which are GABA agonists). Either diazepam (0.1 to 0.3 mg/kg) or lorazepam (0.1 mg/kg) should be administered IV to terminate seizures.

Administration of pyridoxine has been shown to provide specific antidotal therapy for INH poisoning. After administration of vitamin B_6, seizures and metabolic acidosis promptly resolve. Pyridoxine is given by IV in a dose that equals the estimated dose of INH in milligrams. In cases in which the ingested amount is unknown, a single dose of 5 g (70 mg/kg in children) of pyridoxine is administered. Rarely, repeat administration is necessary.

Although INH clearance can be enhanced by hemodialysis or hemoperfusion, these techniques are rarely necessary if pyridoxine, activated charcoal, and aggressive supportive care are provided.

Lead

Background

Although lead poisoning is usually the result of chronic ingestion by pica-prone children or of occupational exposure in adults, patients with lead poisoning may come to the ED with varied complaints of recent onset that often mimic diverse acute illnesses. Fortunately, severe lead encephalopathy is now rare, attributable in large part to widespread screening programs and early treatment of asymptomatic or mildly ill children. However, the risk of lead intoxication still exists, and the emergency physician and pediatrician in every community must maintain an index of suspicion.

Sources of Lead

The major source of excess lead absorption in children is lead-based paint, widely used in home interiors through the 1950s. In addition to the ingestion of macroscopic-sized chips of paint, inner-city children are often exposed to house dust, with a high lead content that results from finely crumbled paint particles, which gets on their hands and toys. Repetitive mouthing can lead to increased lead exposure even in the absence of observable pica. Although classically a disease of poor inner city residents, the recent phenomenon of young, middle socioeconomic level families moving into older sections of large cities and renovating townhouses has led to an expanded population at risk. This is because the sanding, stripping, and burning of lead-based paint from woodwork in such houses has also been associated with lead intoxication in the occupants. Other unusual sources of lead exposure include the burning of battery casings for heat, soft well water carried by outdated lead pipes, improperly home-glazed ceramics, drinking glass glazed decals, and dust or dirt alongside heavily traveled roads (resulting from auto emissions in communities still using leaded gasoline). Infants have also been demonstrated to develop elevated lead levels when parents prepare formula with first-draw tap water or when they boil the water before mixing.

Pathophysiology

Absorption of lead occurs through GI and pulmonary routes, although the former predominates in pediatric intoxications. Lead is then compartmentalized into three main areas: bone, soft tissues, and blood. Excretion occurs slowly through urine, feces, and sweat. Children are probably at double jeopardy compared with adults in that there is experimental evidence that younger animals have increased absorption and also a heavier distribution into soft tissues (including the brain). Concomitant nutritional deficiency, especially low dietary iron and calcium, may enhance intestinal lead absorption. Unfortunately, the same children at greatest risk for lead poisoning by virtue of age and residence are also likely to be at risk for dietary deficiency, especially iron.

Lead exerts its toxic effect principally by two mechanisms: by interference with calcium function at the cellular level and by enzyme inhibition, particularly on enzymes rich in sulfhydryl groups. In humans, the most obvious effects are on neurologic function and on the heme synthesis pathway, which is interrupted at several points, resulting in abnormally high levels of porphyrins and their precursors.

Clinical Findings

Early signs and symptoms of plumbism are notably vague and nonspecific. Abdominal complaints, including colicky pain, constipation, anorexia, and intermittent vomiting, are common; of course, these same symptoms are often ascribed to relatively normal 2 year olds by their parents. The child with early plumbism may also show listlessness and irritability. When encephalopathy begins, the child develops persistent vomiting and becomes drowsy, clumsy, or frankly ataxic. As encephalopathy worsens, the level of consciousness deteriorates further, and seizures commonly occur. Pathologic examination of brains of children who have died of lead encephalopathy shows severe cerebral edema with vascular damage; intracranial pressure is often, although not invariably, increased during the encephalopathy. When spinal fluid is examined, it often reveals a picture similar to that of aseptic meningitis with a mononuclear pleocytosis and elevated protein; however, lumbar puncture should be avoided if possible because of the risk of subsequent herniation. Peripheral neuropathy often occurs in adults with lead poisoning but is rare in children, although it is seen occasionally in those with an underlying hemoglobinopathy. Other organs may be damaged by lead. The kidneys may develop disturbances that range from slight aminoaciduria to a full Fanconi's syndrome with glycosuria and phosphaturia (in addition to aminoaciduria). High levels are also associated with a microcytic anemia that results from a defect in hemoglobin

synthesis. However, most anemia seen in children with excess lead levels is actually caused by concurrent iron deficiency. A moderately sensitive laboratory measure of lead effect on heme synthesis is the evaluation of erythrocyte protoporphyrin (EP), a heme precursor. Moderately elevated EP levels are seen in iron deficiency, but levels above 250 to 300 µg/dL are almost always the result of chronic lead poisoning.

Management

The asymptomatic child discovered to have a lead level in the 20 to 44 µg/dL range, particularly if the EP is greater than 250 µg/dL, deserves immediate referral to a pediatrician or toxicologist. Such children require environmental investigation, further evaluation, and possibly, chelation therapy. All symptomatic children and those with lead levels higher than 44 µg/dL need urgent treatment as outlined next, as well as pediatric consultation to ensure adequate postchelation follow-up.

The remainder of this discussion is addressed primarily to the early recognition and treatment of plumbism, including acute lead encephalopathy. This single aspect of chronic childhood lead poisoning is focused on because it represents a true medical emergency.

Recognition

As stated previously, to recognize mildly symptomatic patients with lead poisoning (or asymptomatic children with high lead levels at great risk to soon become symptomatic) requires a high index of suspicion. All children between 1 and 5 years of age are suspect if they have 1) persistent vomiting, listlessness or irritability, clumsiness, or loss of recently acquired developmental skills; 2) afebrile convulsions; 3) a strong tendency to pica, including a history of acute accidental ingestions or aural or nasal foreign body; 4) a deteriorating pre–World War II house or a parent with industrial exposures; 5) a family history of lead poisoning; 6) iron deficiency anemia; or 7) evidence of child abuse or neglect.

The child aged 1 to 5 years who comes to the ED with an acute encephalopathy presents the physician with a dilemma: lead intoxication requires urgent diagnosis, but confirmation with a blood level is usually not available on an immediate basis. A constellation of historical features of lead poisoning increases the likelihood of the diagnosis. These features include 1) a prodromal illness of several days' to weeks' duration (suggestive of mild symptomatic plumbism); 2) a history of pica; and 3) a source of exposure to lead. Several nonspecific laboratory findings make lead poisoning likely enough to warrant presumptive chelation therapy until confirmation by lead levels is available. These findings include 1) microcytic anemia; 2) elevated EP level, especially if greater than 250 µg/dL (conversely, a normal or minimally elevated EP level, less than 50 µg/dL, would make lead encephalopathy caused by chronic lead paint exposure unlikely); 3)basophilic stippling of peripheral erythrocytes or, if feasible, of red cell precursors on bone marrow examination; 4) elevated urinary coproporphyrins; 5) glycosuria; 6) aminoaciduria; 7) radiopaque flecks on abdominal radiographs; and 8) dense metaphyseal bands on radiographs of knees and wrists (lead lines).

Abnormalities on examination of cerebrospinal fluid (CSF) are also indicative of lead encephalopathy, including a lymphocytic pleocytosis, elevated protein, and increased pressure. However, a lumbar puncture should not be performed if lead encephalopathy is strongly suspected because the risk of herniation is considerable. If CSF must be examined to rule out bacterial meningitis, the minimal amount (less than 1 mL) necessary should be obtained. Alternatively, one might institute treatment for presumed meningitis, perform a determination of the lead level, and consider a delayed lumbar puncture after several days if the lead level is normal.

Treatment

The treatment of lead poisoning involves relocation of the child to a lead-free environment, chelation therapy, and appropriate supportive care. Symptomatic patients are at risk of developing encephalopathy with subsequent death or neurologic sequelae. In addition, asymptomatic patients with high lead levels (especially greater than 100 µg/dL) are also at significant risk for developing CNS involvement and might require urgent treatment.

The specific chelating drugs commonly used for symptomatic lead intoxication are edathamil calcium disodium (CaEDTA) and 2,4-dimercaptopropanol (British Anti-Lewisite or BAL) (Table 88.14). Side effects of CaEDTA include local reactions at injection sites, fever, hypercalcemia, and renal dysfunction manifested by rising BUN and abnormal urine sediment with proteinuria, hematuria, and/or epithelial cells. The major side effects of BAL include nausea and vomiting, so for the first day or two of BAL therapy, it is prudent to maintain the patient on IV fluids and clear liquids or nothing by mouth. BAL also induces hemolysis in patients with glucose-6-phosphate dehydrogenase (G6PD) deficiency. Its use is hazardous if the patient has severe hepatic dysfunction, and it forms a toxic complex if given concurrently with iron. Succimer (dimercaptosuccinic acid, or DMSA) has been approved for pediatric use in cases in which lead levels exceed 45 µg/dL (see Table 88.14). This water-soluble analog of BAL may be taken orally, and several studies have found such use to be as effective as CaEDTA given parenterally. However, experience with its use in symptomatic patients is limited.

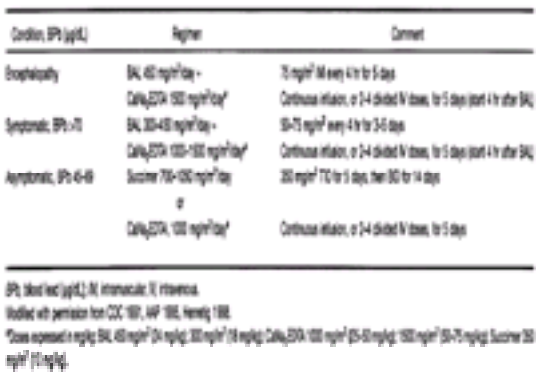

Table 88.14. Guidelines for Chelation Therapy of Lead Poisoning

Asymptomatic children found to have lead levels of 45 to 69 µg/dL should have urgent referral and treatment for 5 days with CaEDTA or DMSA alone. If the lead level is above 69 µg/dL, BAL and EDTA are used for at least the first 2 days (Table 88.14). Supportive care includes adequate hydration to promote good urine output. Symptomatic children without frank encephalopathy should receive chelation therapy with a combination of CaEDTA and BAL for 5 days. Supportive care includes close monitoring for signs of encephalopathy and, again, maintenance of urine flow.

Patients with encephalopathy require combination chelation therapy with CaEDTA and BAL, as well as intensive supportive care. Fluid therapy is critical and must be individualized. Adequate urine flow is needed to excrete the lead-chelate complexes; however, fluid overload must be avoided so that cerebral edema is not exacerbated. A reasonable goal is to supply basal water requirements, maintaining urine production at 0.35 to 0.5 mL/kcal per 24 hours. Basal water needs in children average 1 mL/kcal and may be calculated as 100 kcal/kg for 0 to 10 kg, plus 50 kcal/kg for 10 to 20 kg, plus 20 kcal/kg for each kilogram above 20 kg.

Seizures commonly occur in acute encephalopathy and should be controlled with anticonvulsant drugs (see Chapter 83). Hypothetical precautions have been made about the use of phenobarbital in lead encephalopathy (i.e., synergistic disturbances in porphyrin metabolism), but its clinical use has not been associated with any noticeable deleterious effect.

Recent advances in the management of cerebral edema and increased intracranial pressure (see Chapter 125) have not been evaluated in a controlled fashion in the context of lead encephalopathy. However, it seems reasonable to expect that such measures as forced hyperventilation, mannitol or glycerol osmotic therapy, and high-dose steroids would have a salutary effect. Whether more aggressive measures such as continuous intracranial pressure monitoring, induced hypothermia, and barbiturate coma would decrease mortality or morbidity any further is unknown.

Oral Hypoglycemics

Although almost all juvenile diabetics require insulin therapy for control, the frequent prescription of oral hypoglycemic agents for patients with non–insulin-dependent, adult-onset diabetes has made the availability and, consequently, the ingestion of these medications commonplace among toddlers. The scenario typically involves visits to a grandparent's home (or conversely, a visit by the grandparent to the child's home). The sulfonylureas (chlorpropamide, glipizide, glyburide) are capable of inducing significant hypoglycemia in a toddler after the ingestion of a single tablet. In addition, the onset of hypoglycemia may be delayed up to 16 to 24 hours after ingestion. Thus, prudent management of such exposures generally implies 24-hour hospitalization with close observation and frequent blood glucose testing. All such patients should receive charcoal/cathartic. There may be theoretic benefit to the use of repeat charcoal dosing for ingestions of glipizide, which has an enterohepatic circulation. Excretion of chlorpropamide may be enhanced by urinary alkalinization. The biguanides (e.g., metformin) are unlikely to create hypoglycemia but may promote metabolic acidosis.

Maintenance of euglycemia is usually accomplished in symptomatic patients with the infusion of hypertonic glucose (e.g., 10 to 20%) solutions, supplemented as necessary by bolus doses. Occasionally, patients may still exhibit hypoglycemia, requiring additional treatment. Diazoxide has been a useful adjunct to glucose infusion, in similar doses to its more familiar role as an antihypertensive (e.g., 3 to 5 mg/kg), although given slowly over 30 minutes, to limit any hypotensive effect. This dose may be repeated every 4 to 6 hours. An oral form is also available. Recently, octreotide (1–10 mcg/kg), a somatostatin analog, has been used effectively in cases of refractory hypoglycemia. Occasionally, patients require glucose infusion for several days; as their condition improves, the glucose load may be tapered gradually with frequent monitoring.

Organophosphates

Background

Organophosphates are lipid-soluble insecticides that are commonly applied in sprayed dust or emulsion formulations. These compounds are found in agricultural and home use, and they form the basis of “nerve gases” in chemical warfare agents (see Chapter 132). Organophosphates are readily degraded in the environment and metabolized in mammals by hydrolic cleavage. Some of these chemicals are “systemic” insecticides, meaning that they are taken up by the roots of the plants and translocated into foliage, flowers, and/or fruit.

Pathophysiology

Compounds of this class can be absorbed by inhalation, ingestion, and skin penetration. They irreversibly phosphorylate the enzyme acetylcholinesterase in tissues, allowing acetylcholine accumulation at cholinergic junctions in autonomic effector sites (causing muscarinic effects), in skeletal muscle or autonomic ganglia (causing nicotinic effects), and in the CNS.

Clinical Findings

The symptoms of acute poisoning usually develop during the first 12 hours of contact. These include findings related to the CNS (dizziness, headache, ataxia, convulsions, and coma); nicotinic signs, including sweating, muscle twitching, tremors, weakness, and paralysis; and muscarinic signs characterized by the SLUDGE mnemonic, including *s*alivation, *l*acrimation, *u*rination, *d*efecation, *g*astrointestinal cramping, and *e*mesis. In addition there may be miosis, bradycardia,

bronchorrhea, and wheezing; in severe cases, pulmonary edema develops. Severe intoxications may also cause a toxic psychosis that resembles alcoholism.

A history of exposure to organophosphates and the clinical manifestations already discussed are the best clues to an organophosphate poisoning. A depression of plasma or red blood cell cholinesterase activity provides the best laboratory marker of excessive absorption of organophosphates, although it is rarely available on a stat basis. A decrease in the cholinesterase activity of the red blood cells is more specific for organophosphate inhibition than is the plasma assay. Although plasma cholinesterase is depressed by liver injury from various causes, and a small percentage of the population has a genetically determined deficiency of plasma cholinesterase activity, a depression of 25% or more is strong evidence of excessive organophosphate absorption. However, it is important that treatment not be delayed until confirmation of plasma cholinesterase is obtained.

Management

The management of a patient who has ingested organophosphates must always include safeguards against exposure for the persons who treat the patient. If the compound is ingested orally, gastric lavage should be considered; care should be taken that the gastric aspirate or vomitus not be splashed on the ED staff because the organophosphates are readily absorbed through the skin and mucous membranes. Patients who have been poisoned by the topical application of organophosphates should receive a thorough scrubbing with a soap solution on admission to prevent further absorption of organophosphates. In addition, all contaminated clothing must be removed and stored in a plastic bag to protect the institutional personnel. Activated charcoal should be instilled for those who have been poisoned by the oral route.

After decontamination, antidotal therapy begins with the administration of atropine sulfate given in a dose of 0.05 to 0.1 mg/kg to children and 2 to 5 mg for adolescents and adults. This dose should be repeated every 10 to 30 minutes or as needed to obtain and maintain full atropinization, as indicated by clearing of bronchial secretions and pulmonary rales. Therapy is continued until all absorbed organophosphate has been metabolized and may require 2 mg to more than 2000 mg of atropine over the course of a few hours to several days. After atropinization has been instituted, severe poisonings should be treated with the addition of pralidoxime. This drug is particularly useful in poisonings characterized by profound weakness and muscle twitching. A dose of 25 to 50 mg/kg should be administered in 250 mL of saline by infusion over approximately 30 minutes; adults may receive 1 to 2 g by IV. This may be repeated at 1 hour intervals if muscle weakness is not relieved and then at intervals of 6 to 8 hours for 24 to 48 hours. In patients with severe poisoning, a 2.5% concentration may be infused continuously at the rate of 500 mg/hour in adolescents and adults, or approximately 20 mg/kg per hour in children. Occasionally, patients may require more than 48 hours of therapy; the end-point should be persistent relief of neurologic and cholinergic signs.

Organophosphates are usually dissolved in hydrocarbon bases; thus, the clinician should be prepared to treat hydrocarbon pneumonitis if it develops. Also, bronchopneumonia that complicates the pulmonary edema has been observed in acute poisonings.

Because the organophosphates cause elevated levels of acetylcholine in the plasma, compounds that effect the uptake of acetylcholine and/or its release should be avoided in the management of these patients. Specifically, aminophylline and phenothiazines are contraindicated. In situations in which identification of the ingested insecticides is difficult, consultation may be obtained from the National Insecticide/Pesticide Hotline (800-858-7378). This hotline provides an around-the-clock consultation service for advice on pesticides.

Phenothiazines

Background and Pathophysiologys

The phenothiazines are commonly prescribed major tranquilizers. Phenothiazines are also often used to treat nausea and vomiting in young children. The toxic effects of phenothiazines primarily involve the three components of the nervous system: central, autonomic, and extrapyramidal.

The three subgroups of phenothiazines—aliphatic, piperazine, and piperidine—vary in their effects on the different components of the CNS. In general, the aliphatic group (e.g., chlorpromazine) may cause sedation and hypotension in overdose. The piperazine group (e.g., prochlorperazine) are more likely to create extrapyramidal side effects.

Clinical Findings

The manifestations of phenothiazine toxicity may be dose dependent or dose independent (idiosyncratic). These have significantly different features.

With dose-dependent effects, the manifestations of intoxication after acute ingestion vary from mild to severe. In mild intoxication, CNS signs such as sedation, ataxia, and slurred speech occur. The anticholinergic effects of these drugs may cause constipation, urinary retention, and blurred vision. Because phenothiazines have potent actions on the temperature-regulating center of the hypothalamus, temperature disturbances occur in up to 30% of patients and may consist of hypothermia or hyperthermia. Orthostatic hypotension, the probable result of peripheral vasodilation, may also be noted with mild intoxication.

In moderate intoxications, the patients may have significant depression in level of consciousness. Extrapyramidal effects become notable at this level of intoxication with muscle stiffness or “cogwheel” rigidity seen on passive movement of the neck, biceps, or quadriceps. Anticholinergic manifestations are severe and include acute urinary retention and paralytic ileus; hypotension may be profound. Cardiac conduction disturbances may make their appearance and are often heralded by a prolonged Q-T interval.

In severe overdoses, patients are unarousable. Deep tendon reflexes may be hyperactive. Dystonic reactions may occur, involving the head and neck and the cranial nerves (torticollis and opisthotonos). Arrhythmias and shock may result in death.

The dose-independent effect of the phenothiazines is the dystonic reaction. This striking clinical occurrence consists of episodic spasm of voluntary muscles, particularly those of the head and neck. Patients may develop torticollis, bruxism, tongue protrusion, or oculogyric crisis. Dystonic reactions are unrelated to the amount of ingested phenothiazine. They may or may not occur after the first dose. Their onset is 8 to 40 hours after ingestion of a single dose of phenothiazine. This marked delay between ingestion and manifestations often interferes with obtaining an accurate history of ingestion. Fortunately, although painful and distressing, dystonic reactions are rarely life-threatening and usually resolve quickly after administration of anticholinergics.

Management

Treatment of acute phenothiazine intoxication hinges on the severity of ingestion. In patients with mild overdose, hospitalization is usually unnecessary. The autonomic signs and symptoms are most often transient and require no treatment. In patients with moderate or severe overdoses, the potential for life-threatening manifestation requires prompt evaluation of vital signs, GI decontamination (if ingestion was within 4 to 6 hours of ED arrival), vascular access, and cardiac monitoring. Pressors such as norepinephrine (see Chapter 3) may be used to correct the hypotension. In those rare instances of hypertension, the use of nitroprusside (see Chapter 35) may be indicated. Severe arrhythmias should be treated aggressively as detailed later under Tricyclic Antidepressants (p. 926) and in Chapter 82. Attention should be directed to the treatment of temperature instability and other autonomic disturbances.

Dystonic reactions are effectively controlled by the IM or IV administration of diphenhydramine in a dose 1 to 2 mg/kg. This dose may be repeated within 15 to 20 minutes if no effect is noted. An alternative agent is benztropine mesylate (1 to 2 mg for an older child, adolescent). This agent reportedly causes less sedation than diphenhydramine. After resolution of the dystonic reaction, oral treatment should be continued for an additional 24 to 48 hours to prevent recurrences.

Plants/Mushrooms

Plant Toxicity

Plants are among the more commonly reported accidental ingestions in children. Most such ingestions involve common house and garden plants. Fortunately, of the many varieties of such plants, only a small fraction pose a serious toxic hazard (Table 88.15 and Table 88.16).

Table 88.15. Common Nontoxic Plants

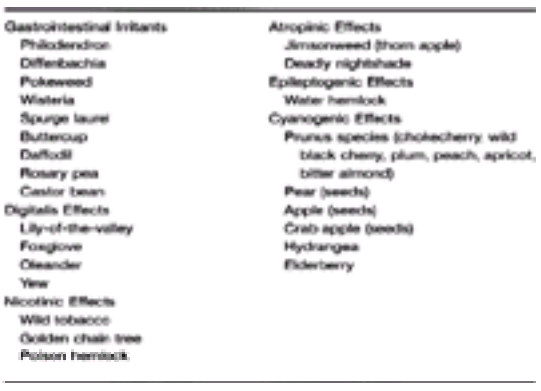

Table 88.16. Common Plant Toxidromes

When a child visits the ED after a plant ingestion, a general evaluation should be performed. Unless the plant can be identified absolutely and is known to be nontoxic, ipecac may be considered. Activated charcoal may also be extremely useful in adsorbing plant toxins. If the plant is subsequently identified and not believed to have serious toxic potential, or if the plant is not available (or recognizable) for identification, the child should be observed for a short period in the ED.

The child who remains asymptomatic may then be discharged and observed at home. Children who develop symptoms or for whom there is strong suspicion or confirmation that the ingested plant poses a potentially serious intoxication should be admitted for further observation and specific or supportive treatment.

Specific Categories of Plant Toxidromes

Plants with Gastrointestinal Irritation

Plants that cause GI irritation account for most plant poisonings in the United States. The range of symptoms extends from mild oral burning to a severe gastroenteritis syndrome. Representative species include *Philodendron* and *Dieffenbachia* species (leaves), which cause minor mouth and throat burning; pokeweed (roots, stem), Wisteria (seeds), spurge laurel (berries), buttercup (leaves) and daffodil (bulbs, accidentally substituted for onions), which cause severe vomiting, colicky abdominal pain, and diarrhea; and the toxalbumin-containing plants such as rosary pea and castor bean (seeds), which can cause a violent hemorrhagic gastroenteritis that leads to profound dehydration and circulatory collapse when the seeds are chewed up. The management of this group of ingestions consists essentially of fluid and electrolyte therapy.

Plants with Digitalis Effects

Several common garden or wildflowers contain digitalis, and they have been responsible for fatal ingestions. Instances of chewing on leaves or flowers or swallowing the berries of lily-of-the-valley, foxglove, squill, and oleander all have led to such poisonings. Intoxication has even occurred when water from a vase that contained these flowers was ingested. Early after ingestion, the child may complain of intestinal symptoms such as mouth irritation, vomiting, and diarrhea. As the digitalis is absorbed, typical digitalis effects may ensue, with conduction defects and, at times, serious arrhythmias. Treatment may include administration of digoxin-specific antibody fragments as was previously discussed (p. 908).

Plants with Nicotinic Effects

Several species of plants contain nicotine or closely related alkaloids. Ingestion of wild tobacco (leaves), golden chain tree (seeds), and poison hemlock (leaves, seeds) usually leads to spontaneous vomiting within 1 hour. Salivation, headache, fever, mental confusion, and muscular weakness may follow, and the child may deteriorate to convulsions, coma, and death from respiratory failure. Charcoal is especially useful in adsorbing these nicotinic alkaloids. Further treatment consists of intensive supportive care, with anticonvulsants and ventilatory assistance.

Plants with Atropinic Effects

The most common atropine-containing plant in the United States is jimsonweed, which is widely distributed. Cases most commonly occur in rural areas but have been seen in inner-city children who managed to find this weed growing in their neighborhoods, where flora in general is scarce.

Symptoms and signs are those of atropinization (Table 88.7) and include visual blurring, dilated pupils, dryness of the mouth, hot, dry skin, fever, delirium, and psychosis. Convulsions and coma may follow. Treatment consists of supportive care and, in severe cases, physiologic antagonism with physostigmine (Table 88.4).

Plants That Cause Convulsions

Convulsions represent the principal toxic effect of some plants. Water hemlock, with its potent cicutoxin, is the main species to cause convulsions in the United States. Within 1 hour after ingestion, nausea, vomiting, and profuse salivation occur. These initial symptoms are followed by tremors, muscle rigidity, and multiple major motor seizures. Treatment is with anticonvulsants, as for status epilepticus (see Chapter 70 and Chapter 83).

Plants That Contain Cyanogenic Glycosides

Many plants and particularly fruit seeds (pits) contain the cyanogenic glycoside amygdalin (Table 88.16). Symptoms and signs after ingestion are those of cyanide poisoning, with resultant cellular hypoxia. Initially, there is CNS stimulation and headache, with tachypnea, hypertension, and reflex bradycardia. Anxiety and excitation may progress to opisthotonus and seizures. Respiratory depression, with cyanosis, tachycardia, and hypotension, follows. An odor of bitter almonds may be detected. Treatment is initiated with 100% oxygen and cardiopulmonary resuscitation (CPR) as necessary. Antidotal therapy with amyl nitrite, sodium nitrite, and sodium thiosulfate is administered as detailed in Table 88.9 and in Chapter 132.

Mushrooms

Mushrooms cause an estimated 50% of all deaths from plant poisoning in the United States. The difficulty in accurate identification of mushrooms makes reliance on such identification for appropriate management of ingestions extremely hazardous in the ED. Rather, the approach advocated by Lampe is reviewed here.

Two main groups of mushrooms can be characterized on the basis of the time interval between ingestion and symptom onset: those with the immediate onset of symptoms and those with delayed onset. Regardless of the mushroom, the initial management for all suspected poisonings includes activated charcoal and catharsis.

Onset of symptoms within 6 hours of ingestion usually confers a benign prognosis, although careful attention to fluid and electrolyte management is critical. Most mushrooms have GI effects. There are five general classes of mushrooms in this group, each possessing a unique toxicologic feature. Some “early-onset” mushrooms cause muscarinic effects, usually

within 15 minutes, such as sweating, salivation, colic, and pulmonary edema. This syndrome responds to atropine therapy. Other early-onset mushrooms cause anticholinergic effects, including drowsiness, followed by mania and hallucinations. Another subgroup of early onset mushrooms produces a severe gastroenteritis syndrome. Hallucinogenic mushrooms such as psilocybin make up another class of mushrooms with early-onset symptoms. Finally, some mushrooms precipitate an Antabuse reaction if they are coingested with alcohol. Management for all these agents consists of supportive care and careful monitoring of fluid status.

The second, more important, category of mushrooms that are responsible for 90% of mushroom-related deaths are those associated with onset of symptoms that occur more than 6 hours after ingestion. The most important members of this group are those mushrooms that belong to the *Amanita phalloides* species. With these mushrooms, after a latent period of many hours, GI upset appears. Approximately 24 hours after ingestion, hepatic dysfunction appears, which results in fulminant hepatic failure. Without liver transplantation, such victims generally die.

Two compounds are known to produce the toxic effects of *A. phalloides.* Phallotoxin acts first, causing GI symptoms, including nausea, vomiting, abdominal pain, and diarrhea. Fever, tachycardia, and hyperglycemia may also occur during this stage. The other toxin, amatoxin, causes renal tubular and hepatic necrosis.

Treatment of the gastroenteric phase includes fluid and electrolyte replacement. If renal failure develops, dialysis may be necessary. Hepatic damage after *A. phalloides* ingestion may be attenuated by early use of repetitive activated charcoal, which appears to interrupt enterohepatic recirculation of amatoxin.

Additional therapies have shown mixed results in the treatment of *A. phalloides* poisoning. High-dose penicillin, cimetidine, thioctic acid, prophylactic charcoal hemoperfusion, and other modalities await further investigation. A regional poison control center may offer guidance with experimental therapies, but multiple-dose activated charcoal and vigorous attention to supportive care remain the standard.

In management of mushroom ingestions in which the specific mushroom cannot be identified, GI decontamination, including activated charcoal, should always be provided. Identification of the agent may be possible through consultation with a local mycologist, although mushroom cohabitation makes identification uncertain even if a fragment of mushroom is brought for direct inspection.

Theophylline

Theophylline was once the primary agent in the treatment of reactive airway diseases, including asthma. Since the advent of specific b-adrenergic agonists, however, it has become less widely used. Nonetheless, theophylline continues to be commonly prescribed for asthma, as well as for neonatal apnea. Because of its narrow therapeutic index, theophylline also continues to be a common cause of intoxication. In overdose, it may lead to clinical toxicity, including nausea, vomiting, muscle tremor, electrolyte disturbances, hypotension, cardiac arrhythmias, seizures, and death.

Theophylline has many physiologic actions, including bronchodilation, respiratory stimulation, diuresis, and augmented cardiac output. However, its exact mechanism of action remains uncertain. Formerly thought to act through inhibition of the enzyme phosphodiesterase, thereby potentiating intracellular activity of cAMP, current evidence indicates that this is not the case. More recent theories suggest that theophylline acts as an antagonist at adenosine receptors. These receptors are found on bronchial smooth muscle, in the CNS and on the myocardium.

The pharmacokinetics of theophylline are complex and responsible for many of the drug-disease or disease-disease interactions that so often result in accidental intoxication. Metabolism of theophylline is by the hepatic mixed function oxidases (cytochrome P-450), with the production of inactive metabolites. The metabolic rate is subject to considerable variability based on factors such as age (with neonates having significantly longer elimination half-lives compared with children and adolescents), viral illness (which may decrease P-450 activity), concomitant medications (including erythromycin and cimetidine, both potent inhibitors of P-450 activity), and drug dose. Because theophylline has a Michaelis-Menten elimination pattern, increases in dose are associated with a reduction in elimination rate and greater risk of intoxication.

To a limited extent, clinical manifestations of theophylline toxicity correlate with serum theophylline concentration. The therapeutic theophylline level is 10 to 20 µg/mL, although, even in this range, signs of toxicity such as nausea and vomiting may be present. With serum levels of 20 to 50 µg/mL, toxicity generally consists of nausea, vomiting, and muscle tremor. When theophylline exceeds 60 to 70 µg/mL, severe toxicity appears with the development of seizures and arrhythmias. However, this correlation only exists for those patients with acute, single overdoses of theophylline rather than those with chronic intoxication. Because differences in toxic manifestations appear between acute and chronic theophylline intoxication, it is necessary to determine the method of intoxication as soon as this poisoning is recognized.

In addition to the clinical manifestations, theophylline may cause a number of metabolic disturbances, including hyperglycemia, hypokalemia (with serum potassium as low as 2.2 mEq/L), metabolic acidosis, hypercalcemia, and hypophosphatemia.

Management

Emergency management of theophylline intoxication begins with rapid assessment of vital signs. If seizures are present, airway protection may be necessary, followed by the administration of anticonvulsants (see Chapter 83). Preferred anticonvulsants with theophylline poisoning are benzodiazepines and barbiturates. Blood pressure is typically low with theophylline poisoning. If there is significant hemodynamic compromise, fluid administration followed by vasopressor support may be necessary (see Chapter 3). Cardiac monitoring should be provided immediately; arrhythmias are treated according to standard protocols (see Chapter 1 and Chapter 82).

GI decontamination is important after acute theophylline overdose because sustained-release preparations often lead to delayed peak absorption. Activated charcoal has an important role in the treatment of theophylline poisoning, and antiemetics should be administered aggressively to permit charcoal retention. Antiemetics may include metoclopramide or ondansetron. In addition, ranitidine, which reduces gastric secretions, may reduce vomiting. Phenothiazines are relatively contraindicated because they may lower seizure threshold. For the patient with significant overdose of sustained-release preparations or increasing serum theophylline levels despite adequate initial GI decontamination, use of WBI may be valuable (p. 894).

Laboratory assessment should include electrolytes, blood glucose, urinalysis, and theophylline level. A complete toxin screen should be obtained if coingestants are suspected. Because of the unpredictable absorption pattern that follows overdoses of sustained-release preparations, serial theophylline concentrations (every 2 to 4 hours) should be obtained until a plateau has been identified.

There are several methods of enhancing theophylline elimination. Activated charcoal, through GI dialysis, is capable of enhancing fecal elimination of theophylline; it is highly effective even if theophylline intoxication is the result of IV administration. Therefore, the foundation of treatment is repeated doses of oral charcoal (0.5 to 1.0 g/kg), every 2 to 4 hours. This regimen generally requires coadministration of antiemetics. In addition to multiple-dose activated charcoal, both hemodialysis and hemoperfusion are highly effective at removing theophylline. Hemoperfusion is the preferred procedure because it increases theophylline elimination fourfold to sixfold. However, because it is not widely available, hemodialysis is an acceptable alternative, capable of doubling theophylline clearance. These procedures appear to be effective only as prophylactic measures; if they are performed before the appearance of seizures or arrhythmias, they can prevent their occurrence. However, if they are initiated after seizures or arrhythmias develop, they do not clearly alter clinical course. The indications for hemoperfusion or hemodialysis in otherwise healthy children are generally considered to be 1) serum theophylline level greater than 80 to 100 µg/mL in the patient with acute theophylline poisoning; 2) serum theophylline level greater than 40 µg/mL in the patient with chronic theophylline poisoning; or 3) severe toxicity unresponsive to supportive measures, regardless of serum concentration. All patients who meet these criteria should be admitted to the intensive care unit or referred to a tertiary care medical center for intervention and close observation.

Tricyclic Antidepressants

Background

The ingestion of tricyclic antidepressant compounds is a significant problem in pediatric patients. The availability of these compounds in the household may be the result of therapy for depression for a parent or a grandparent or of treatment for enuresis in the patient or a sibling.

Clinical Findings

The ingestion of 10 to 20 mg/kg of most tricyclic antidepressants represents a moderate to serious exposure, with coma and cardiovascular symptoms expected. The ingestion of 35 to 50 mg/kg may result in death. Children have been reported to be more sensitive than adults to tricyclic antidepressants and often have symptoms at lower dosages.

Cyclic antidepressants have many pharmacologic effects. Anticholinergic activity causes altered sensorium and sinus tachycardia. a-Adrenergic blockade may lead to hypotension. However, the more severe cardiovascular effects are primarily caused by the membrane-depressant or quinidinelike effects that depress myocardial conduction and may lead to multiple focal premature ventricular contractions and ventricular tachycardia. It has been shown that a QRS interval over 0.1 second is associated with a significant morbidity and mortality in these patients; this delay in conduction may progress to complete heart block and cardiac standstill and/or the previously mentioned ventricular arrhythmias. Another typical electrocardiographic finding suggestive of cyclic antidepressant poisoning is the finding of an R wave of greater than 3-mm amplitude in the QRS complex in lead aVR.

Neurologic findings include lethargy, disorientation, ataxia, hallucinations, and with severe overdoses, coma, and seizures. Fever is commonly present initially, but hypothermia may occur later. Additional anticholinergic symptomatology includes decreased GI motility, which delays gastric emptying time, and urinary retention. Muscle twitching has been observed and may be associated with increased deep tendon reflexes. Although the pupils may be dilated, they usually respond to light.

Management

Severe tricyclic antidepressant overdoses require gastric decontamination. Lavage can be considered even for those patients who present as late as 4 to 12 hours after ingestion. Because tricyclic antidepressants decrease GI motility, unabsorbed drug may be left in the stomach for prolonged periods. Certainly, the administration of activated charcoal should be performed. Significant conduction delays or arrhythmias resulting from tricyclic antidepressants may benefit from alkalinization of the blood. A sodium bicarbonate bolus of 1 to 2 mEq/kg can be given during continuous ECG monitoring. Bicarbonate infusion can then be used to keep the serum pH at 7.45 to 7.55. These therapeutic maneuvers likely serve to decrease drug binding to the myocardium. Additional bolus doses of sodium bicarbonate may be required if the QRS interval is noted to widen. An additional benefit of the sodium cation may be to partially overcome the sodium channel blockade that is believed to represent the biomolecular substrate of the membrane depressant effect of these agents. If arrhythmias persist, appropriate antiarrhythmic therapy should be instituted, perhaps using lidocaine (see Chapter 1 and Chapter 82). Quinidine or procainamide should be avoided because each may increase heart block in this situation. Physostigmine, although previously recommended for its antidotal effects on the anticholinergic aspects of these poisonings, has the potential to worsen ventricular conduction defects and to lower the seizure threshold. Its use is currently considered to be contraindicated in cyclic antidepressant overdoses. In the presence of hypotension, many clinicians have advocated the use of norepinephrine infusions (0.1 to 0.3 µg/kg per minute). This is based on the

observation that the hypotension is the result of norepinephrine depletion secondary to the block of catecholamine uptake caused by tricyclic antidepressants. Other clinicians have reported that dopamine is as effective; however, the occurrence of ventricular arrhythmias has been reported with dopamine. During the recovery period, serum electrolytes should also be monitored because the infusion of bicarbonate may cause hypokalemia, which may aggravate tricyclic antidepressant-induced cardiac arrhythmias. It must be remembered in the treatment of such antidepressants that these compounds have long half-lives and slow elimination rates; therefore, the therapy for these ingestions is often protracted and intensive.

Other Antidepressants

Besides the trycyclic antidepressants, numerous agents designed to elevate mood are prescribed. The chemical structure of these agents and their profile of toxicity are diverse. Major groupings of nontricyclic antidepressants include 1) the selective serotonin reuptake inhibitors (SSRIs) (e.g., fluoxetine, sertraline), 2) the monoamine oxidase inhibitors (e.g., phenelzine, tranylcypromine), and 3) other atypical antidepressants (e.g., amoxapine, venlafaxine, bupropion).

SSRIs most commonly produce CNS depression in overdose. Seizures may occur after large ingestions. Life-threatening events from acute overdose of these compounds rarely occurs. The serotonin syndrome, manifested by muscular rigidity, myoclonus, flushing, and autonomic instability, more typically occurs as the result of drug interaction and is potentially lethal. Amoxapine has anticholinergic activity but is best known for its convulsant properties and the tendency for victims to present with status epilepticus. Bupropion, prescribed perhaps most commonly in smoking cessation programs, prevents reuptake of biogenic amines and is also seizurogenic. The a-adrenergic antagonism of trazodone may lead to hypotension.

The monoamine oxidase inhibitors (MAOIs), although pharmacologically effective and therapeutically important, are some of the most toxic medications known. Acute single overdoses of as little as 6 mg/kg have been associated with a fatal outcome. In addition, because of their irreversible inhibition of the enzyme monoamine oxidase, which is responsible for the degradation of most biogenic amines, MAOIs possess several important interactions with foods and other medications that can lead to severe toxicity, even in the patient who takes them in appropriate doses. There are three important clinical pictures of MAOI toxicity. First, because GI tract activity of monoamine oxidase is also inhibited by these drugs, patients who take them appropriately and then ingest foods or drugs that contain biogenic amines (e.g., tyramine in wines, cheese, or soy sauce and decongestants) may develop severe hypertension with headache, seizures, or stroke. The second picture of MAOI toxicity appears when those who take the drug therapeutically are given certain sympathomimetic or serotonergic agents causing the serotonin syndrome. Important examples of such drugs include common agents in OTC cough and cold preparations such as dextromethorphan, analgesics such as meperidine, and psychotropic medications such as clomipramine and fluoxetine or other SSRIs. In these patients, this drug combination may quickly lead to hyperpyrexia, skeletal muscle rigidity, cardiac arrhythmias, and death. This is one of the few fatal drug interactions known. Finally, those with acute MAOI overdoses develop a clinical syndrome that includes blood pressure instability, hyperpyrexia, skeletal muscle rigidity, seizures, and death.

Because of the toxicity of these agents and the frequent delay in their onset of activity (up to 24 hours), all patients with a history of MAOI ingestion, regardless of symptoms, should be admitted to the hospital for 24 hours. Management of the patient with MAOI toxicity is largely dictated by the specific toxic manifestations. In those with hypertensive reactions, treatment consists of the immediate administration of an antihypertensive. The ideal agent may be nitroprusside because its brief duration of action permits titration of effect. In the treatment of hyperpyrexia, cooling measures are promptly instituted. Because hyperpyrexia is often accompanied by skeletal muscle rigidity and rhabdomyolysis, serum creatine kinase should be measured and close attention should be paid to the urine for any signs of myoglobinuria. Benzodiazepines are often helpful in this situation and neuromuscular blockade may be beneficial in patients who have severe muscle rigidity with hyperthermia. In the patient with acute overdose, treatment is directed to hemodynamic stability. Because blood pressure changes occur quickly and consist of hypotension and hypertension, hypertension should be treated with short-acting agents (see Chapter 35) and hypotension with fluid and vasopressor support (see Chapter 3). Intensive care unit admission is mandatory for these patients because of their clinical instability.

Drugs Dangerous in Small Doses

Toddlers often are brought to EDs for evaluation after possibly having ingested one or two doses of a medication. This can be a particularly vexing problem. Most often these children will be fine with little treatment beyond reassurance. There are circumstances, however, when this situation can be life-threatening and proper intervention can be lifesaving. A large list of chemicals and poisons can be extremely toxic in small amounts, and that is beyond the scope of this discussion. However, it is wise to be familiar with a modest list of pharmaceuticals that may cause dangerous toxicity to young children with just one or two doses (Table 88.17). Many of these agents have been discussed earlier in this chapter. The actual incidence of life-threatening toxicity of each of these drugs, when just one or two doses has been ingested, is as yet undefined.

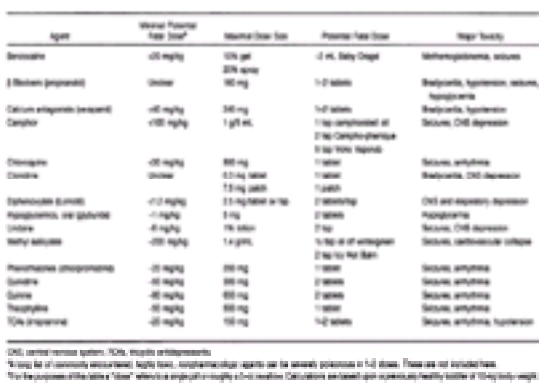

Table 88.17. Medications Dangerous to Toddlers in 1–2 Doses [a]

A systematic approach to these patients includes a careful history, an examination with attention to the presence of toxidromes (Table 88.5), and a guided laboratory assessment. This approach may allow narrowing of the differential diagnosis and may allow a determination of the possible severity of the ingestion. If the differential diagnosis includes any of the drugs listed in Table 88.17, it may be prudent to provide decontamination and prolonged observation. An algorithmic approach to this situation is provided in Figure 88.7.

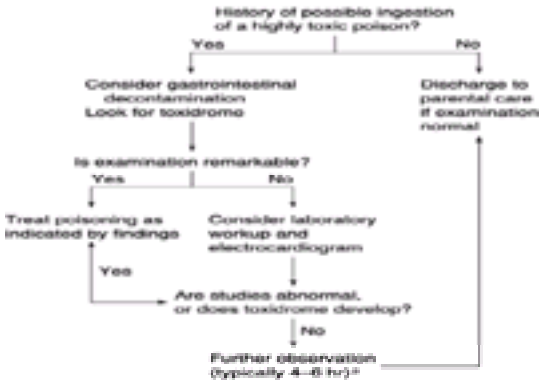

FIGURE 88.7. Algorithmic approach to toddler having ingested 1 to 2 doses (1–2 pills or 5–10 cc swallow) of a drug. [a]Medications notorious for their ability to have delayed onset in toxicity, beyond 4–6 hours, include oral hypoglycemic agents, sustained-release preparations, monoamine oxidase inhibitors, drugs taken concomitantly with anticholinergic agents, and acetaminophen.

SUBSTANCE ABUSE

As a special category of pediatric toxicology, exposures to psychoactive drugs is outlined in this section. In a discussion of substance abuse, three distinct age populations in the pediatric group may be placed at risk from such exposures: 1) the adolescent or preadolescent who abuses drugs for their mind-altering effects (and, in the case of females, may do so when pregnant); 2) the neonate who is exposed to substances of abuse during gestation and manifests signs of intoxication or abstinence after birth; and 3) the infant or toddler who becomes exposed to drugs of abuse either through active administration by a caretaker (“chemical child abuse”), the ingestion of a drug left in an accessible place (e.g., the coffee table), or passive exposure created by being in an environment where drugs of abuse are used (e.g., marijuana, cocaine, PCP, methamphetamine). In any of these circumstances, the exposure can be sufficient to produce severe intoxication. Thus, knowledge of the epidemiology and manifestations of substance abuse become important in the management of children of all ages.

Further discussion regarding the general phenomenon of chronic adolescent drug abuse and addiction, including alcoholism, is found in Chapter 130.

Clinical Manifestations

The drug-abusing child or adolescent may present to the ED after an accidental overdose, a suicidal gesture, suicide attempt, sudden bizarre behavior, or after multiple trauma (e.g., assault, motor vehicle accident). Often, the history of drug exposure is undeclared and may not be diagnosed unless there is a high index of suspicion and appropriate diagnostic tests are performed. In such cases, the patient's mental status can range from fully awake and responsive to comatose; physical examination can be without any signs of drug exposure or with overt signs of toxicity (e.g., seizures). Table 88.18 provides a summary of the common drugs of abuse, their typical routes of administration, associated symptoms, toxic levels, and duration of action.

Table 88.18. Drug Abuse: Summary of Toxicity

Initial history from patient, family, or friends must specify drugs taken and estimate quantities when a history of exposure is given. In the absence of a history of exposure, it is important to inquire whether the patient has a history of any psychoactive drug abuse. In many cases, the patient may admit to using a drug but may identify it by a street name. Although drug terminology tables are often available in pharmacology or toxicology texts or on the Internet, temporal and regional changes in street drug terminology generally make such tables of limited value.

Management

Primary attention is paid toward assessment of vital signs and life support as needed to provide a patent, secure airway; to ensure adequate respiratory function; and to treat seizures, shock, or cardiorespiratory arrest. A key but often overlooked feature in the assessment of such patients is an accurate temperature because many drug intoxications are associated with hyperpyrexia. If there is any suspicion of hyperthermia, a rectal temperature must be obtained. In the agitated patient, physical and/or chemical restraint may be necessary to obtain vital signs. Chemical restraint should be used liberally to prevent the patient from harming himself or others. The preferred agents in such cases are diazepam (0.1 to 0.3 mg/kg IV) or midazolam (0.05 to 0.1 mg/kg IV). Haloperidol (0.05 mg/kg) given intramuscularly is also effective but reduces heat-dissipating capability and lowers the seizure threshold.

Management must also include consideration of the need for GI decontamination. It is common with psychoactive drug use that several distinct routes of exposure are possible (e.g., ingestion, inhalation, injection, nasal insufflation). Therefore, GI decontamination is not always necessary or appropriate. However, because those who abuse drugs almost invariably use more than one drug, decontamination must be performed if there is any possibility of an ingestion. In the event that decontamination seems appropriate, assessment of the patient's mental status and gag reflex must be performed; in the presence of obtundation or a diminished gag reflex, airway protection by endotracheal intubation should be accomplished before decontamination measures.

Disposition

In the case of the adolescent who presents with intentional drug abuse, after initial assessment and medical stabilization, an evaluation must be made of the severity of the drug use problem. This is accomplished through discussion with the parents or caretakers. Although issues of patient confidentiality may require the physician to provide limited information to parents, obtaining a thorough psychosocial evaluation is necessary for complete management of the acute event. Such discussions may require or may be facilitated by an interview with a social services or psychiatry consultant. Once the severity of the drug problem has been established, referral to a treatment program should be discussed. Primary care physicians may be comfortable managing those patients who have no long-standing histories of drug abuse. Compulsive users or anyone who presents with a drug abstinence syndrome must be referred for intensive rehabilitation. Family therapy often is a component of this rehabilitation.

SPECIFIC DRUGS

The major categories of drugs of abuse that require the physician's familiarity with the whole spectrum of their physiologic effects are 1) hallucinogens (phencyclidine, lysergic acid diethylamide [LSD], marijuana), 2) stimulants (amphetamines, cocaine), 3) central anticholinergics, 4) sedatives (benzodiazepines, barbiturates), 5) opioids (morphine, codeine, heroin, methadone), 6) inhalants, and 7) alcohol. (Acute alcohol overdose is discussed in the previous section; chronic alcoholism in teenagers is discussed in Chapter 130.)

Hallucinogens (Psychedelics)

No single characteristic distinguishes psychedelics from other classes of centrally active drugs such as anticholinergics, cocaine, and amphetamines. All of these drugs can produce a number of mental status changes, including illusions, hallucinations, delusions, and paranoid ideation. However, the psychedelic state is described as consisting of vivid and unusual visual experiences with diminished control over what is experienced. Images and sensations take on profound meaning, and the ability to differentiate oneself from the environment is decreased. Most drugs in this category are related to the indolealkylamines (e.g., LSD, psilocybin, dimethyltryptamine, and diethyltryptamine), to the phenylethylamines (mescaline), or to phenylethylamines (methylenedioxymethamphetamine MDMA, Ecstasy]).

Phencyclidine (Angel Dust)

Identification

PCP was developed in the 1950s as a general anesthetic. It rapidly fell into disuse because of disturbing emergence syndromes that developed in postoperative patients. Sporadic abuse occurred in the 1960s, but its popularity peaked in the 1970s. The drug remains common in several metropolitan areas and a similar agent, ketamine, is finding increased popularity. PCP is easily synthesized and is often sold on the streets misrepresented as LSD, mescaline, or marijuana. It is well absorbed across all mucous membranes and is most popularly used by inhalation (often mixed into cigarettes or marijuana “joints”), but it can be ingested, injected, or insufflated.

Chemically, PCP is an arylcyclohexylamine. This group of drugs has a range of CNS actions that range from hallucinations with smaller doses, to stimulation with moderate doses (occasionally associated with seizures), to profound CNS depression with respiratory arrest with large doses.

Pharmacodynamics

There is great variability in the metabolism of PCP. In general, 0.1 µg/mL is considered a toxic serum level. One cigarette

may contain 1 to 100 mg. A dose of 5 to 10 mg may produce stupor and coma; with doses exceeding 10 mg, respiratory depression and convulsions occur. A fatal dose is in the range of 1 mg/kg. Because PCP has a long elimination half-life (18 hours), clinical symptoms may last for more than 12 hours; also, patients may have cyclic symptoms because the drug is enterogastrically recirculated.

Pharmacologically, PCP acts as a dissociative anesthetic, meaning that it interferes potently with association pathways that link the cerebral cortex with deeper structures in the brain, thus diminishing the ability to integrate sensory input into meaningful behavior. Its anesthetic actions also lead to a marked diminution of pain sensation. In conjunction with bizarre behavior, this often leads victims to have feelings of invulnerability and to attempt life-threatening actions (e.g., stepping into automobile traffic).

Clinical Symptoms

Small doses of PCP produce signs and symptoms of inebriation with staggering gait, slurred speech, and nystagmus (vertical or rotatory). Users may also be diaphoretic and have catatonic muscular rigidity with a blank stare. Having sympathomimetic actions, it is often associated with hypertension and tachycardia. Moderate doses cause other signs of intoxication, including hypersalivation, pyrexia, repetitive movements, and muscle rigidity. Larger doses can cause seizures, coma, or respiratory arrest. The typical “high” from a single dose lasts 4 to 6 hours and is followed by an extended “coming-down”; PCP-induced psychotic states may be long lasting and may recur (“flashbacks”). Tolerance develops to the behavioral and toxic effects of the drug. Chronic users report persistent difficulties with recent memory, speech, and thinking that last from 6 months to 1 year after the last dose; they also may be left with personality changes such as withdrawal, isolation, anxiety, nervousness, and depression.

Management

PCP is easily detected through a qualitative analysis of urine. Serum levels are rarely available and do not correlate with clinical manifestations. Therefore, management must often be based solely on a history of exposure or index of suspicion. Initial treatment is directed at stabilizing vital signs and treating life-threatening events such as seizures. If exposure is the result of ingestion, GI decontamination should be performed by administration of activated charcoal. A quiet room may be helpful, although the ability to monitor the patient cannot be compromised. Physical restraints should be avoided if possible because they may lead to significant rhabdomyolysis with resulting myoglobinuria and renal injury. For chemical restraint diazepam (0.1 to 0.3 mg/kg IV) or lorazepam (0.1 mg/kg IV) may be effective, although a major tranquilizer (e.g., haloperidol) is often necessary.

Although urine acidification (pH below 5.0) enhances the urinary excretion of PCP, it should never be performed in these patients because it exacerbates metabolic acidosis and may promote deposition of myoglobin in renal tubules. In a review of 27 confirmed cases of PCP poisoning, 3 patients developed rhabdomyolysis and 2 progressed to acute renal failure. Both patients had received acidification measures before diagnosis. If tests for muscle enzymes and/or renal function are abnormal and the urine has a positive test for hemoglobin without red blood cells, the patient should be assumed to have rhabdomyolysis and should be treated accordingly (see Chapter 86).

LSD (Blotter, Acid)

Pathophysiology

LSD and related psychedelic drugs such as psilocybin, mescaline, and dimethyltryptamine (DMT) have actions at multiple sites in the CNS (from the cortex to the spinal cord). In addition, dozens of congeners of these agents exist in mushrooms or have been synthesized, and they also cause signs and symptoms similar to LSD. The pharmacologic action that these drugs seem to have in common is as agonists of presynaptic serotonin-2 receptors (which modulate serotonin release into the synaptic cleft). These agents all have structural similarities to serotonin (5-hydroxytryptamine).

Pharmacodynamics

In humans, the somatic symptoms of dizziness, weakness, drowsiness, nausea, and paresthesias may be observed after one oral dose of 0.5 to 2 µg/kg. Between the dose range of 1 to 16 µg/kg, the intensity of LSD's psychoactive effects is proportional to the dose. A typical LSD “hit” is 200 to 400 µg. A high degree of tolerance to the behavioral effects develops after three to four daily doses, with sensitivity returning after a drug-free interval. Deaths directly attributable to LSD are virtually unknown, although fatal accidents and suicides have occurred during states of intoxication.

Clinical Symptoms

In general, the somatic effects of hallucinogens are sympathomimetic and include pupillary dilation, hypertension, tachycardia, hyperreflexia, and hyperpyrexia. Doses as low as 20 to 25 µg can produce CNS effects such as euphoria, visual perceptual distortions, alteration of subjective time so that time passes slowly, lability of mood, or even an acute panic episode. Hallucinations and psychosis with hyperalertness are commonly seen. The clinical duration of action of LSD is somewhat dose dependent but averages 6 to 12 hours. The psychedelic state includes a heightened awareness of sensory input, often accompanied by an enhanced sense of clarity but a diminished control over what is experienced. There is often a feeling that one part of the self is a passive observer while another part receives vivid sensory input. The ability to separate one object from another or to separate self from the environment is diminished. There is an enhanced sense of oneness with mankind or the cosmos.

Management

LSD intoxication is rarely associated with life-threatening events. However, vital signs should be assessed to ensure that

the patient is stable and in the event there has been drug coingestion.

Because LSD is ingested in minuscule doses and onset of symptoms occurs hours after ingestion, GI decontamination is unnecessary, unless coingestion is suspected.

Clinical management involves placing the patient in a quiet room. Someone who knows the patient may be able to quietly “talk down” and reassure the patient. The patient's loss of boundaries and fear of fragmentation or self-disintegration create a need for a structuring or a supportive environment. Both benzodiazepines (e.g., diazepam 0.1 to 0.3 mg/kg IV or midazolam 0.05 to 0.1 mg/kg IV) and haloperidol 0.05 mg/kg IM are effective tranquilizers in the event that anxiety or agitation persists.

Marijuana (Pot, Reefer, Smoke, Grass, Hemp)

Pathophysiology

With the exception of ethanol, marijuana remains the most popular psychoactive drug of abuse. It is typically sold in “nickel” bags that produce two to three joints. Marijuana is occasionally laced with other psychoactive substances, including PCP and cocaine. Hashish is the concentrated resin of marijuana.

The flowering tops of the female marijuana plant contain the highest concentration of the active constituent, tetrahydrocannabinol (THC). In the 1970s, most marijuana contained approximately 1 to 2% THC by weight. Recently, cultivated seedless varieties of marijuana (“sensimilla”) have become popular, and they contain 5 to 8% THC by weight. Therefore, a joint is now likely to lead to a greater degree of altered mental status than previously.

Within minutes of smoking this material, perceptual, behavioral, and emotional states become altered for several hours. Patients often have the appearance of inebriation with dysarthria and ataxia. However, violence, hallucinations, and agitation are uncommon after marijuana use.

Pharmacodynamics

It is estimated that no more than 50% of THC in a marijuana cigarette is actually absorbed. Pharmacologic effects begin immediately. In contrast, the onset of effects after ingestion occurs in 30 minutes to 1 hour, and peak effects may not occur until the second and third hours after ingestion; THC is three times more potent when smoked than when taken by mouth.

Clinical Symptoms

The most prominent effects in humans are on the CNS and cardiovascular system. In doses of up to 20 mg, THC produces effects on mood, memory, motor coordination, cognitive ability, sensorium, time sense, and self-perception. There is an increased sense of well-being or euphoria accompanied by feelings of relaxation or sleepiness when subjects are alone. With greater intake of THC, short-term memory is impaired, and the capacity to carry out tasks that require multiple mental steps to reach a specific goal deteriorates. This effect on memory-dependent, goal-directed behavior has been called temporal disintegration and is correlated with a tendency to confuse past, present, and future and with depersonalization, a sense of strangeness and unreality about one's self. Marijuana smokers often report a voracious appetite (“the munchies”), dry mouth and throat, more vivid visual imagery, and a keener sense of hearing. Altered time perception is a consistent effect of cannabinoids, so minutes seem like hours. Larger doses of THC can produce frank hallucinations, delusions, and paranoid feelings. Thinking becomes confused and disorganized. Anxiety that reaches panic proportions may replace euphoria, often as a feeling that the drug-induced state will never end. Because of the rapid onset of effects when marijuana is smoked, most users can regulate their intake to avoid the excessive doses that produce these unpleasant effects. Marijuana may cause an acute exacerbation of symptoms in stabilized schizophrenics. Cardiovascular effects include tachycardia, hypertension, and marked conjunctival injection. Chronic smoking of marijuana and hashish is associated with bronchitis and asthma, even though THC is a mild bronchodilator.

Infants and toddlers passively exposed to marijuana may develop profound lethargy or coma, occasionally with tachycardia.

Management

In general, the only treatment required is discontinuation of the drug. In the adolescent patient with a psychotic reaction or acute toxic delirium, a sedative such as diazepam, 5 to 10 mg by mouth or 0.1 mg/kg by IV, may be necessary. These acute symptoms should improve with drug abstinence over 4 to 6 hours.

Stimulants

Amphetamines (Crank, Speed)

Pathophysiology

Amphetamines have powerful CNS stimulant actions, in addition to peripheral adrenergic actions. Unlike epinephrine, amphetamines are effective after oral administration. However, they are often taken by injection and nasal insufflation. Amphetamines have been used medically to treat narcolepsy, obesity, fatigue, and nasal congestion. Several decongestant nasal inhalers continue to add amphetamine agents that may be extracted and ingested by drug-seeking adolescents. The pharmacologic effects of amphetamines include increased blood pressure, occasionally with a reflex slowing of heart rate, contraction of bladder sphincter, and dramatic CNS stimulation. Like other indirect

sympathomimetics, amphetamines act by releasing endogenous biogenic amines from the presynaptic neurons.

Methamphetamine is the most commonly abused of these drugs, reportedly because its greater lipid solubility is associated with more potent CNS effects.

Abuse patterns of amphetamines have changed in recent years because these drugs have begun to approach cocaine as widely abused stimulants. This increase has paralleled government interdiction efforts that have reduced the illegal entry of cocaine. Many drug users prefer amphetamines over cocaine because the clinical duration of action is considerably longer than that of cocaine. Also, a smokable form of methamphetamine (“ice”) has appeared, associated with more striking and prolonged alterations in CNS function. Finally, amphetamines have become a popular substance of abuse in pregnant women, leading to increases in neonatal intoxication or abstinence syndromes.

Pharmacodynamics

The therapeutic dose of dextroamphetamine in adolescents is typically 5 mg three times daily. The toxic dose is variable but is rarely less than 15 mg. Severe reactions have been reported at 30 mg, yet doses up to 400 to 500 mg may cause only mild symptoms. Tolerance is striking, with chronic users taking 10 to 15 g daily without ill effects. The elimination half-life of the amphetamines is about 3 hours, with much of the drug being excreted in the urine unchanged.

Clinical Symptoms

The psychic effect of amphetamines depends on the dose, mental state, and personality of the drug user. In general, 10 to 30 mg cause wakefulness, alertness, decreased sense of fatigue, and elevation of mood, with increased initiative, self-confidence, ability to concentrate, elation, euphoria, and increase in motor and speech activity. Physical performance in athletes may be improved. Prolonged use of large doses is followed by depression and fatigue. Amphetamines have an appetite-suppressant effect through an action on the lateral hypothalamic feeding center. However, tolerance to this effect also develops; thereafter, the effect is insufficient to reduce weight for a sustained period.

The acute toxic effects of amphetamine are usually extensions of its therapeutic actions. The central effects induce euphoria, restlessness, dizziness, tremor, hyperactive reflexes, talkativeness, irritability, weakness, insomnia, and fever. In addition, confusion, assaultiveness, anxiety, delirium, paranoid hallucinations, panic states, and suicidal or homicidal tendencies can occur, especially in patients who have underlying mental illnesses. However, these psychotic effects may occur in anyone who chronically abuses amphetamines. Cardiotoxic effects include palpitations, anginal pain, and rarely, hypertensive crisis or circulatory collapse. GI effects include anorexia, nausea, vomiting, diarrhea, and abdominal cramps. Severe overdoses may cause convulsions, coma, and cerebrovascular accidents. Both psychological and physical dependence occurs with chronic use. Chronic amphetamine abuse causes symptoms similar to many of those seen after acute overdose. The most common serious effect is a psychotic reaction with vivid hallucinations and paranoid delusions, often mistaken for schizophrenia. Recovery may or may not occur after withdrawal of the drug. In patients with persistent psychotic symptoms, it has been theorized that the amphetamine has hastened the onset of incipient schizophrenia. Chronic amphetamine abuse is also associated with the development of cerebral vasculitis.

An amphetamine derivative, methylenedioxymethamphetamine (MDMA or Ecstasy), has become popular among drug-abusing college students and may find its way into the high school scene. It is a drug with mildly hallucinogenic effects. In larger doses, it causes perceptual distortions, hallucinations, and agitation.

Management

Treatment of intoxication after ingestion of these agents should include GI decontamination. For severe agitation, specific treatment consists of administration of a benzodiazepine (e.g., diazepam 0.1 to 0.2 mg/kg IV) or haloperidol (0.01 to 0.05 mg/kg IM). Severe hypertension unresponsive to benzodiazepines may be treated with such agents as phentolamine, hydralazine, or IV sodium nitroprusside. Because up to 45% of amphetamines are excreted in the urine unchanged, ample fluids are beneficial.

Cocaine

Pathophysiology

Cocaine occurs in the leaves of *Erythroxylon coca* and other species of *Erythroxylon* trees indigenous to Peru and Bolivia, where the leaves have been used for centuries by the natives to increase endurance and to promote a sense of well-being. Chemically, cocaine is benzoylmethylecgonine. Ecgonine is an amino alcohol base closely related to tropine, the amino alcohol in atropine. Cocaine may be used by injection, inhalation (in the form of cocaine alkaloid or “crack”), nasal insufflation, and rarely, ingestion. Although ingestion is uncommon, there are two circumstances under which cocaine may be ingested in toxic quantities: the “body packer” and the “body stuffer.” In the body packer, large quantities of cocaine are enclosed in plastic and ingested in an attempt to smuggle the drug, usually across international boundaries. In the case of the body stuffer, the person in fear of being found with the substance suddenly ingests cocaine. Body stuffers are typically at greater risk of cocaine intoxication because they do not take sufficient care to guarantee that the cocaine does not leech from the bag.

Cocaine is reportedly used in up to 15% of women during pregnancy. This has led to epidemic increases in the number of “cocaine babies” who are often preterm, small for age, irritable, and show neurodevelopmental delay. Beyond the postnatal age, passive cocaine exposure in infants and toddlers has been associated with severe intoxication, including the development of convulsions.

Pharmacodynamics

The relief from fatigue that occurs with cocaine use results from central stimulation that masks the sensation of fatigue. Cocaine potentiates the excitatory and inhibitory responses of sympathetically innervated organs to norepinephrine and epinephrine by blocking the reuptake of catecholamines at adrenergic nerve endings. This explains why cocaine, unlike other local anesthetics, produces vasoconstriction and mydriasis. The most important pharmacologic action is its ability to block the initiation or conduction of the nerve impulse after local application. Cocaine is still widely used as a local anesthetic for ophthalmologic or otorhinolaryngologic procedures. It is also used as a topical anesthetic for laceration repair in the form of TAC (tetracaine, adrenaline, cocaine).

Although fatalities have been associated with cocaine doses as low as 30 mg, 1 to 2 g is generally the lethal dose in adults. Ingested cocaine is less toxic than that taken by other routes because of its prolonged absorption by this route. The elimination half-life of cocaine is approximately 1 hour. Cocaine metabolism is complex and consists of nonenzymatic degradation to form benzoylecgonine and metabolism by plasma cholinesterases to form ecgonine methyl ester. A small fraction of cocaine is also metabolized through the cytochrome P-450 enzymes to form norcocaine. People with congenital deficiencies in plasma cholinesterase are thought to have exaggerated responses to cocaine; cocaine abusers have been known to ingest inhibitors of cholinesterases or P-450 enzymes (e.g., organophosphate insecticides, cimetidine) to enhance the effect of the cocaine. Cocaine metabolites are readily detected in urine for approximately 3 days after exposure.

Cocaine is absorbed from all sites of application, including GI mucosa. Body-packing may lead to severe toxicity (seizures and cardiorespiratory collapse) if the container ruptures. If smuggling is suspected, a flat plate may show opaque densities within the bowel highlighted by a gas halo.

In addition to nasal application, cocaine can be used by injection or inhalation. The latter is called freebase (crack) and is a dangerous practice. In making crack, street cocaine (which is in the form of cocaine hydrochloride) is converted to cocaine alkaloid by removal of the salt moiety. This is accomplished by mixing the cocaine with water and sodium bicarbonate. The crack is then separated from the water by filtration and drying. The paste hardens and is cut into chips that resemble soap. It is then smoked in a pipe or sprinkled onto a cigarette or joint. A small piece, called a quarter rock, produces a 20- to 30-minute high when smoked in a water pipe. Probably because of its enhanced lipid solubility, crack crosses the blood–brain barrier rapidly, causing an intense rush of pleasure. This habit is highly addictive. Presently crack is primarily used by older teenagers and persons in their early twenties, in part because it is relatively inexpensive (approximately $5 to $10 per rock).

Clinical Symptoms

Cocaine's most dramatic clinical effect is CNS stimulation. In humans, this manifests in a feeling of well-being and euphoria, often accompanied by garrulousness, restlessness, excitement, and a sense of clarity. However, as the dose is increased, tremors, forced speech, agitation, and even tonic-clonic convulsions may result from excessive stimulation.

Initially, small doses (1 to 1.5 mg/kg) may slow the heart rate through central vagal stimulation. After moderate doses, pulse increases, the result of both central and peripheral adrenergic effects. Hypertension may appear abruptly and lead to cerebrovascular accidents. Fortunately, hypertension is generally short lived. Larger doses of cocaine may cause hypertension that may be followed quickly by cardiovascular collapse, often the result of myocardial ischemia and infarction. Myocardial injury that ranges from angina pectoris to massive infarction can be seen in young adults after acute cocaine exposure. With chronic cocaine use, a cardiomyopathy may develop that results in depressed cardiac function and death.

Rhythm disturbances are also characteristic of acute cocaine intoxication. These may consist of ventricular or supraventricular tachyarrhythmias and may be intractable. Arrhythmias are the most common cause of death after severe cocaine exposure.

Use of crack has been associated with a number of pulmonary disturbances, including bronchospasm, hemoptysis, pneumothorax, and pneumomediastinum. These are thought to result from the barotrauma associated with inhalation of hot, particulate matter, followed by a Valsalva maneuver.

Cocaine has been associated with other syndromes of organ dysfunction, including hyperpyrexia and renal failure. “Coke fever” (or pyrexia) is a common occurrence after acute cocaine use. It is often associated with muscle rigidity (resembling neuroleptic malignant syndrome) or rhabdomyolysis (the result of agitation and/or physical restraint). Rhabdomyolysis may go on to induce myoglobinuric renal failure if not promptly recognized and treated.

Infants exposed to cocaine may also exhibit CNS excitation that includes hyperactivity, dystonic posturing, altered mental status, or frank seizures.

Management

Among substances of abuse, cocaine is most likely to create the unstable patient with life-threatening manifestations. Therefore, this intoxication requires rapid, thorough assessment and management. Immediate attention should be paid to the vital signs, including temperature (which should be obtained rectally). The patient who develops seizures requires immediate airway control as well as anticonvulsant therapy. Benzodiazepines (e.g., diazepam 0.1 to 0.3 mg/kg) are considered the anticonvulsants of choice because of their rapid onset of action and because animal data have associated their use with decreased mortality from cocaine intoxication. Benzodiazepines should also be administered liberally to the patient with mild to moderate toxicity (agitation, hypertension, tachycardia) because of their efficacy in

reversing many of these clinical manifestations.

Because circulatory function can range from hypertensive crisis to cardiovascular collapse, early vascular access is important. Blood pressure instability should be anticipated and treated accordingly. For treatment of hypertensive crises, liberal benzodiazepine use may be combined with a short-acting antihypertensive (e.g., nitroprusside). Immediate treatment of hypertension is recommended because it may lead to cerebrovascular or myocardial injury. Cardiac arrhythmias are treated according to advanced cardiac life support protocols (see Chapter 1 and Chapter 82).

Hyperthermia must be recognized and treated promptly to prevent its complications. Management is discussed in Chapter 89. Diuresis should be induced if urinalysis is suggestive of myoglobinuria. Patients with CNS depression or a lateralizing neurologic examination should receive cranial tomography to rule out an intracranial vascular event.

Because cocaine is rarely ingested, the need for GI decontamination is confined to body packers/stuffers or when drug coingestion is suspected. With body stuffers, because bag leakage can lead to abrupt onset of severe intoxication and possibly death, activated charcoal should be administered immediately. Gastric emptying maneuvers and endoscopic removal of cocaine bags are relatively contraindicated because of the risk of bag rupture. Instead, decontamination is confined to administration of activated charcoal and WBI. Multiple-dose activated charcoal is recommended to maximize the opportunity for cocaine to be adsorbed by the charcoal. Because cocaine bags and crack vials are radiopaque in up to 50% of cases, an abdominal radiograph is recommended to determine the location and extent of retained packets after decontamination has been initiated. A contrast study may be considered to improve detection.

In the event of severe intoxication or ingestion of more than 1 to 2 g of cocaine, transfer to the intensive care unit is essential for appropriate monitoring.

Central Anticholinergics

Pathophysiology

Increasingly, drugs, plants, and mushrooms with anticholinergic properties are ingested for their psychoactive effects. Because compounds such as antidepressants, antihistamines, antispasmodics, and belladonna alkaloids are in widespread use, these are more readily available than illicit psychoactive substances. Also, many OTC drugs having anticholinergic activity are available without prescription and are ingested to get “high.” All of these agents are competitive antagonists with acetylcholine at the neuroreceptor site (Table 88.19). The major effects of these drugs are on the myocardium, CNS, smooth muscle, and exocrine glands. These effects include tachycardia, mydriasis, facial flushing, hyperpyrexia, cardiac arrhythmias, urinary retention, dry mucous membranes, decreased sweating, and decreased or absent bowel sounds. CNS effects include delirium, anxiety, hyperactivity, visual hallucinations, illusions, and disorientation. These signs and symptoms lead to the common mnemonic, “Mad as a hatter, red as a beet, dry as a bone, blind as a bat, and hot as Hades.” In excess, anticholinergics may lead to severe toxicity that includes cardiac arrhythmias, seizures, and death.

Antidepressants: amitriptyline (Elavil), imipramine (Tofranil), doxepin (Sinequan, Adapin)
Antihistamines: chlorpheniramine (Ornade, Teldrin), diphenhydramine (Benadryl), orphenadrine (Norflex)
Ophthalmologic Preparations: cyclopentolate (Cyclogel), tropicamide (Mydriacyl)
Antispasmodic Agents: propantheline (Probanthine), clidinium bromide (Librax)
Antiparkinson Agents: trihexyphenidyl (Artane), benztropine (Cogentin), procyclidine (Kemadrin)
Proprietary Drugs: Sleep-Ezs (scopolamine, methapyrilene), Dominex (scopolamine, methapyrilene), Asthma-Cor (belladonna alkaloids), Excedrin-PM (methapyrilene)
Belladonna Alkaloids: Atropine, homatropine, hyoscine, hyoscyamus, scopolamine
Toxic Plants: Mushroom (*Amanita muscaria*), bitter-sweet (*Solanum dulcamara*), Jimson weed (*Datura stramonium*), potato leaves and sprouts (*Solanum tuberosum*), deadly nightshade (*Atropa belladonna*)

Table 88.19. Drugs and Chemicals That May Produce the Central Anticholinergic Syndrome

Pharmacodynamics

The effects of anticholinergics vary according to the specific drug ingested, particularly because the many classes of drugs lead to secondary actions that are independent of anticholinergic actions. An important, universal anticholinergic effect, however, is decreased GI motility. This is associated with delayed absorption of drug and, if GI decontamination is not performed, the appearance of severe toxicity may be delayed 12 to 24 hours after ingestion.

Management

The management of a patient with a known central anticholinergic syndrome is a challenge, particularly because one must also be prepared for the other distinct toxicities of the ingested drug or plant. Also, most plants and many drugs are not detected on toxin screen, so the diagnosis must rely on history and clinical suspicion. Along similar lines, serum drug levels do not predict the degree of anticholinergic symptoms.

GI decontamination is essential after anticholinergic ingestion. Unlike most toxic ingestions, there is clear efficacy to decontamination 12 to 24 hours after ingestion because of the likelihood of drug persistence in the gut lumen for an extended time. Even gastric emptying may be effective for many hours after ingestion and should be considered for

patients who present within 4 to 8 hours of ingestion. Activated charcoal with a cathartic is added to enhance fecal expulsion of the drug-charcoal complex in the face of diminished gut motility.

Based on presenting signs and symptoms, the patient may require sedation, and monitoring in an intensive care unit setting to provide ventilatory support for coma, anticonvulsants for seizures, and antiarrhythmic drugs for cardiac arrhythmias. Adequate sedation may be achieved with titrated doses of benzodiazepines. Physostigmine, a potent anticholinesterase, is a recognized antidote for anticholinergic-induced mental status alterations; however, its use is controversial. Physostigmine can produce bronchospasm, bradycardia, hypotension, and seizures. It is therefore reserved for those who have normal ECGs and mental status dysfunction confined to hallucinations or severe agitation. The adult dose is 1 to 2 mg via slow IV infusion over 5 minutes. The trial dose can be repeated in 10 to 15 minutes up to a maximum of 4 mg. The pediatric dose is 0.5 mg administered slowly intravenously, with repeat every 10 minutes up to a maximum of 2 mg. The smallest effective dose may be repeated every 30 to 60 minutes if symptoms recur over 6 to 8 hours. The muscarinic toxicity of physostigmine may be treated with IV atropine at one-half the physostigmine dose given; physostigmine-related seizures may be treated with benzodiazepines.

Central Nervous System Sedative-Hypnotics

Pathophysiology

The sedative-hypnotics reversibly depress the activity of all excitable tissues. For most of these agents, CNS effects occur with little action on skeletal, cardiac, or smooth muscle. Uncommonly, serious depression in cardiovascular and other functions may occur. The prevalence of abuse of these agents was formerly exceeded by opioid abuse. However, with the increasing popularity of cocaine, these agents have become the preferred choice in treating cocaine-induced tension and anxiety. Many of these agents, including glutethimide, meprobamate, methaqualone, and barbiturates, are uncommonly available and have been replaced by the benzodiazepines. Because they have retained some popularity and still make periodic appearances on the streets, however, they should be included in discussions of such drugs.

Pharmacodynamics

The sedative-hypnotics have tranquilizing, euphoriant effects that may be similar to morphine. With all these agents—prescribed for this tranquilizing action—it is difficult to draw the line between appropriate use, abuse, habituation, and addiction. However, for all, tolerance is common and physical dependence quickly develops. Therefore, their abuse potential is considered high.

The pharmacologic characteristics of each drug are largely determined by their specific chemical nature. For example, all barbiturates are bound by plasma proteins. These characteristics have important implications in affecting their renal elimination and the effectiveness of extracorporeal drug removal techniques (hemodialysis, hemoperfusion).

For all sedative-hypnotics, patterns of abuse vary, ranging from infrequent sprees of intoxication to compulsive daily use. Introduction to these drugs may be through street use or drug trade (which is most common in adolescents), but, commonly, exposure is initiated through a physician's prescription to a parent for insomnia or anxiety. Because tolerance develops to most of the actions of these drugs, no signs of chronic use may be apparent.

Clinical Symptoms

After sedative-hypnotic use the adolescent may exhibit sluggishness, difficulty in thinking, dysarthria, poor memory, faulty judgment, emotional lability, and short attention span. Irritability and lability are common. With chronic use, these drugs also lead to dependence so that a picture of abstinence may appear after their disuse, with clinical manifestations of apathy, weakness, tremulousness, agitation, or frank convulsions. In its mildest form, the abstinence syndrome may consist only of rebound increases of rapid eye movement sleep, insomnia, or anxiety.

Management

With victims of acute sedative-hypnotic ingestion, attention should be directed to ensuring a patent airway and an intact gag reflex. Cardiovascular disturbances are rare after sedative use, but because of the possibility of drug coingestion, thorough hemodynamic assessment is necessary. Most sedative-hypnotics are detectable on comprehensive toxin screens so that specimens of serum and urine should be sent for analysis. GI decontamination should be considered and can typically be confined to administration of activated charcoal with a cathartic. Repeated doses of charcoal with cathartic have been shown to enhance clearance of certain barbiturates and benzodiazepines. Urinary alkalinization (p. 898) aids in the excretion of phenobarbital. In extreme cases, charcoal hemoperfusion should be considered.

Optional treatment of sedative overdose includes continuous monitoring in an intensive care unit with intubation and ventilator support as indicated. Flumazenil, a recently released benzodiazepine antagonist, can be administered in cases of suspected benzodiazepine ingestion. Its pediatric dose is 0.01 to 0.02 mg/kg IV. Indications for flumazenil administration may be 1) to reverse a witnessed, unintentional benzodiazepine overdose in a young child, or 2) to prevent airway intubation after an iatrogenic overdose. Flumazenil must not be given empirically in unknown or intentional overdoses for which induction of seizures may be life-threatening.

Opioids (Morphine, Codeine, Heroin, Methadone, Propoxyphene)

Pathophysiology

In the United States, three distinct groups who abuse opioids have been described: 1) those who are prescribed an opioid as medical treatment and then go on to become dependent and develop drug-seeking behaviors (this group

constitutes a minority of opioid abusers); 2) those who begin with recreational drug use and quickly progress to regular use; and 3) women who abuse opioids when pregnant. Such women and their offspring are at risk for a number of adverse pregnancy outcomes. For the patient who abuses opiates through injection (the most common route of abuse), other consequences include the risk of hepatitis, endocarditis, AIDS, and vasculitis.

The opioids produce their major effects by combining with receptors in the brain and other tissues. Effects include analgesia, drowsiness, change in mood, respiratory depression, decreased GI motility, nausea, and vomiting. The opiate receptors appear to be the normal sites of action of several endogenous opioidlike substances (e.g., the endorphins).

Pharmacodynamics

Generally, the toxic opioid dose for a person who is not addicted depends on the particular drug. For example, with morphine, clinical toxicity (excessive sedation) may appear with doses that exceed 5 mg in the adolescent. Tolerance rapidly develops to many CNS effects. However, death may occur as a result of marked respiratory depression and consequent anoxia. Other toxicities of opiates include (neurogenic) pulmonary edema, mast cell degranulation (which leads to histamine release and an “anaphylactoid” reaction), cardiac disturbances (with propoxyphene intoxication), and neurotoxicity with seizures (with meperidine intoxication).

Clinical Symptoms

Opioids invariably cause miosis, even after development of tolerance. Respiration may be depressed because of decreased responsiveness of brain stem respiratory centers to increases in carbon dioxide tension. Therapeutic doses of morphine have no effect on blood pressure or cardiac rate or rhythm. When blood pressure changes occur, they result from histamine release. Because histamine dilates capacitance blood vessels and decreases the ability of the cardiovascular system to respond to gravitational shifts, sitting or standing may produce orthostatic hypotension.

Many opioids have extensive effects on the GI tract. They decrease secretion of hydrochloric acid, GI motility, and pancreatic secretions while increasing colonic tone to the point of spasm. In addition, the tone of the anal sphincter is augmented. Therapeutic doses of morphine and codeine can also increase biliary tract pressure, producing epigastric distress and biliary colic.

Management

The presence of coma, pinpoint pupils, and depressed respiration should suggest opioid poisoning in the absence of history. The finding of needle marks on the body further suggests this diagnosis. To confirm the diagnosis, toxicologic analysis of urine and serum should be conducted.

The first management step with opioid intoxication is to ensure adequate ventilation of the patient. Endotracheal intubation may be necessary if there is severe respiratory depression or pulmonary edema. If appropriate, GI decontamination should be performed. The narcotic antagonist naloxone (1 to 2 mg) should be given by IV. If there is no response despite the suspicion of opiate intoxication, the naloxone dose should be repeated (up to 8 to 10 mg), depending on effect and level of suspicion. Naloxone can precipitate an abstinence syndrome in those who have developed physical dependence; in such patients, smaller initial doses of 0.2 to 0.4 µg, with upward titration as needed, are preferable.

When patients who are addicted to opiates are hospitalized, small doses of an opiate may be necessary to prevent severe withdrawal. Methadone substitution is the preferred agent, because in small doses, it is less euphorigenic and its long elimination half-life permits once- or twice-daily dosing. With the patient under observation, 10 to 20 mg of methadone are given, ideally before the appearance of withdrawal symptoms (insomnia, irritability, agitation, piloerection).

Gamma Hydroxybutyrate and Gamma Hydroxybutyrolactone

The related agents, gamma hydroxybutyrate (GHB) and gamma hydroxybutyrolactone (GBL), have become popular substances of abuse among teenagers and young adults. GHB is an endogenous compound with neurotransmitter and/or neuromodulator function, and interacts with dopamine, serotonin, GABA, and endogenous opioid-based neural systems. GHB sales and transportation across state lines is currently restricted by federal law, and possession is illegal in some states. However, it is widely available through the purchase of kits (e.g., by mail order, via the Internet) that allow its home synthesis. GBL is actually a precursor to GHB and is the primary ingredient of such kits, and continues to be legally sold in health food stores. However, GBL is rapidly metabolized in vivo to GHB, and the clinical effects of ingesting either agent are nearly indistinguishable. These agents are used for a variety of reasons, but primarily as euphoriants and aphrodisiacs at parties or all-night dance clubs (“raves”). GHB has gained a particular notoriety as a date-rape agent. They also have a reputation in the body-builder community as growth hormone stimulants and thus enhancers of muscle development and fat loss.

GHB and GBL are CNS depressants that cause rapid onset of deep sleep that can progress to coma and respiratory depression. Patients who have overdosed may have transient seizure activity, and are often hypothermic and bradycardic. The coma is usually relatively short in duration, on the order of 1 to 2 hours. During emergence, transient delirium and vomiting are often observed. Depressed respiratory effort and airway-protective reflexes are common in the more severe cases, although aspiration pneumonia as a complication has been rare. Many patients are surprisingly responsive to stimulus, and attempts at laryngoscopy to effect endotracheal intubation in a seemingly deeply comatose patient may result in an angry, combative patient who sits up and swears at the endoscopist.

Most patients with acute overdose can be managed with the provision of ambient oxygen, suctioning, and airway and antiaspiration positioning. A nasal trumpet is helpful in some cases, and endotracheal intubation may be required

occasionally, although it may necessitate rapid sequence induction for the reasons previously noted. Atropine has been used for severe bradycardia with success. Blood pressure support is rarely necessary.

Inhalants

The High School Senior Survey, conducted by the National Institute on Drug Abuse, has suggested that the abuse of inhalants is relatively common in adolescents with a lifetime prevalence rate of up to 20%. Additional data suggest that these psychoactive agents are even more common in school-aged children and preadolescents. The prevalence of inhalant abuse among young children has been related to the ready availability of these products. Patterns of abuse are also strikingly region specific, with the highest rates of abuse in the southwestern and southeastern United States.

The psychoactive inhalants can be placed into three broad categories: 1) hydrocarbons, 2) nitrous oxide, and 3) nitrites. The hydrocarbons can be subdivided further into the aliphatic hydrocarbons, the halogenated hydrocarbons, and solvents. Regardless of class, all inhalants possess the pharmacologic property of narcosis, leading to euphoria and light-headedness after inhalation. Typically, the agents are abused by “huffing” or “bagging.” In huffing, the agent is placed into a rag or handkerchief, held under the nose, and then deeply inhaled. With bagging, a common method of abuse at parties, the compound is placed into a large bag (e.g., garbage bag) with the drug user placing his or her head into the bag.

Several distinct profiles of toxicity have been described after inhalant abuse. The inebriation that these agents produce may be associated with mental status changes that include coma with respiratory arrest or aspiration. The halogenated hydrocarbons all possess potent cardiotoxicity, leading to myocardial irritability and cardiac arrhythmias. This action has been associated with many reports of spontaneous ventricular fibrillation in adolescents during a binge. Finally, the act of bagging is associated with the risk of simple asphyxia. A syndrome known as sudden sniffing death has been described in adolescents who abuse inhalants. This syndrome may be the result of any of these previously described toxicities. Finally, acute exposure to inhalants may lead to methemoglobinemia, often severe.

Other toxicities are associated with chronic inhalant abuse. The solvents, particularly toluene, may lead to a syndrome that includes abdominal pain, muscle wasting, electrolyte disturbances, and renal tubular acidosis. Victims of chronic solvent abuse also may develop a leukoencephalomalacia with cerebral atrophy.

Management

Because inhalant abuse may lead to the development of life-threatening symptoms, close attention should be directed to the vital signs and their stability. Patients with depressed levels of consciousness may require airway support and ventilation. Because of the risk of cardiac arrhythmias, vascular access should be established early. Arrhythmias should be treated according to standard protocol (see Chapter 1 and Chapter 82); however, the use of epinephrine is relatively contraindicated because it has been associated with worsening of rhythm disturbances. As a part of the evaluation, a complete metabolic panel that includes electrolytes with calcium, phosphate and magnesium, amylase, liver function tests, creatine phosphokinase, and urinalysis should be obtained.

Treatment of methemoglobinemia is discussed in Chapter 87.

Suggested Readings

REFERENCE TOXICOLOGY TEXTBOOKS

Haddad LM, Shannon MW, Winchester JF, eds. Clinical Management of Poisoning and Drug Overdose. 3rd ed. Philadelphia: WB Saunders, 1998.

Goldfrank LR, Flomenbaum NE, Lewin NA, et al, eds. Goldfrank's Toxicologic Emergencies. 6th ed. Stamford, CT: Appleton & Lange, 1998.

GENERAL APPROACH AND MANAGEMENT

American Academy of Clinical Toxicology, European Association of Poisons Centres and Clinical Toxicologists. Position statement: cathartics. J Toxicol Clin Toxicol 1997;35:743–752.

American Academy of Clinical Toxicology, European Association of Poisons Centres and Clinical Toxicologists. Position statement: gastric lavage. J Toxicol Clin Toxicol 1997;35:711–719.

American Academy of Clinical Toxicology, European Association of Poisons Centres and Clinical Toxicologists. Position statement: ipecac syrup. J Toxicol Clin Toxicol 1997;35:699–709.

American Academy of Clinical Toxicology, European Association of Poisons Centres and Clinical Toxicologists. Position statement: single-dose activated charcoal. J Toxicol Clin Toxicol 1997;35:721–741.

American Academy of Clinical Toxicology, European Association of Poisons Centres and Clinical Toxicologists. Position statement: whole bowel irrigation. J Toxicol Clin Toxicol 1997;35:753–762.

Henretig FM. Special considerations in the poisoned pediatric patient. Emerg Med Clin North Am 1994;12:549–567.

Litovitz TL, Klein-Schwartz W, Dyer KS, et al. 1997 annual report of the American Association of Poison Control Centers Toxic Exposure Surveillance System. Am J Emerg Med 1998;16:443–497.

Litovitz TL, Manoguerra A. Comparison of pediatric poisoning hazards: an analysis of 3.8 million exposure incidents (a report from the American Association of Poison Control Centers). Pediatrics 1992;89:999–1006.

Osterloh JD, Snyder JW. Laboratory principles and techniques to evaluate the poisoned or overdosed patient. In: Goldfrank LR, Flomenbaum NE, Lewin NA, eds. Goldfrank's Toxicologic Emergencies. Stamford, CT: Appleton & Lange, 1998.

Perry H, Shannon M. Emergency department gastrointestinal decontamination. Pediatr Ann 1996;25:19–26.

Shannon MW, Haddad LM. The emergency management of poisoning. In: Haddad LM, Shannon MW, Winchester JF, eds. Clinical Management of Poisoning and Drug Overdose. 3rd ed. Philadelphia: WB Saunders, 1998:2–331.

Walton WW. An evaluation of the poison prevention packaging act. Pediatrics 1982;69:363–370.

Wiley JF II. Difficult diagnoses in toxicology: poisons not detected by the comprehensive drug screen. Pediatr Clin North Am 1991;38:725–737.

Woolf AD, Shannon MW. Clinical toxicology for the pediatrician. Pediatr Clin North Am 1995;42:317–333.

ACETAMINOPHEN

Anker AL, Smilkstein MJ. Acetaminophen: concepts and controversies. Emerg Med Clin North Am 1994;12:335–349.

Harrison PM, Keays R, Alexander GJ, et al. Improved outcome of paracetamol-induced fulminant hepatic failure by late administration of acetylcysteine. Lancet 1990;335:1572–1573.

Henretig FM, Selbst SM, Forrest C, et al. Repeated acetaminophen overdosing causing hepatotoxicity in children. Clin Pediatr 1989;28:525–528.

Perry H, Shannon MW. Acetaminophen. In: Haddad LM, Shannon MW, Winchester JF, eds. Clinical Management of Poisoning and Drug Overdose. 3rd ed. Philadelphia: WB Saunders, 1998:664–674.

Rumack BH. Acetaminophen overdose in children and adolescents. Pediatr Clin North Am 1986;33:691–701.

Rumack BH, Peterson RG. Acetaminophen overdose, incidence, diagnosis and management in 416 patients. Pediatrics 1978;62(Suppl):898.

Smilkstein MJ, Knapp GL, Kulig KW, et al. Efficacy of oral *N*-acetylcysteine in the treatment of acetaminophen overdose—analysis of the national multicenter study (1976 to 1985). N Engl J Med 1988;319:1557–1562.

Yip L, Dart RC, Hurlbut KM. Intravenous administration of oral *N*-acetylcysteine. Crit Care Med 1998;26:40–43.

ALCOHOLS AND GLYCOLS

Arditi M, Killner MS. Coma following use of rubbing alcohol for fever control. Am J Dis Child 1987;141:237–238.

Bates BA, Shannon MW, Woolf AD. Ethanol-related visits by adolescents to a pediatric emergency department. Pediatr Emerg Care 1995;11:89–92.

Church AS, Witting MD. Laboratory testing in ethanol, methanol, ethylene glycol, and isopropanol toxicities. J Emerg Med 1997;15:687–692.

Hylander B, Kjellstrand CM. Prognostic factors and treatment of severe ethylene glycol intoxication. Intens Care Med 1996;22:546–552.

Liu JJ, Daya MR, Carrasquillo O, et al. Prognostic factors in patients with methanol poisoning. J Toxicol Clin Toxicol 1998;36:175–181.

Scherger DL, Wruk KM, Kulig KW, et al. Ethyl alcohol (ethanol)-containing cologne, perfume, and after-shave ingestions in children. Am J Dis Child 1988;142:630–632.

Shannon M. Toxicology reviews: fomepizole—a new antidote. Pediatr Emerg Care 1998;14:170–172.

Trummel J, Ford M, Austin P. Ingestion of an unknown alcohol. Ann Emerg Med 1996;27:368–374.

ANTIHISTAMINES

Freedberg RS, Friedman GR, Palu RN. Cardiogenic shock due to antihistamine overdose. JAMA 1987;257:660.

Hestand HE, Teske DW. Diphenhydramine hydrochloride intoxications. J Pediatr 1977;90:1017.

Shannon M. Toxicology reviews: physostigmine. Pediatr Emerg Care 1998;14:224–226.

Wiley JF II, Gelber ML, Henretig FM, et al. Cardiotoxicity following astemizole overdose in children. J Pediatr 1992;120:799–802.

Woodward GA, Baldassano RN. Topical diphenhydramine toxicity in a five year old with varicella. Pediatr Emerg Care 1988;4:18–20.

ASPIRIN POISONING

Dugandzic RM, Tierney MG, Dickinson GE, et al. Evaluation of the validity of the Done nomogram in the management of acute salicylate intoxication. Ann Emerg Med 1989;18:1186–1190.

Vertrees JE, McWilliams BC, Kelly HW. Repeated oral administration of activated charcoal for treating aspirin overdose in young children. Pediatrics 1990;85:594–597.

Yip L, Dart RC, Gabow PA. Concepts and controversies in salicylate toxicity. Emerg Med Clin North Am 1994;12:351–364.

b-ADRENERGIC BLOCKERS

Kenyon CJ, Aldinger GE, Joshipura P, et al. Successful resuscitation using external cardiac pacing in beta adrenergic antagonist-induced bradyasystolic arrest. Ann Emerg Med 1988;17:711–713.

Kerns W II, Kline J, Ford MD. Beta-blocker and calcium channel blocker toxicity. Emerg Med Clin North Am 1994;12:365–390.

Love JN, Litovitz TL, Howell JM, et al. Characterization of fatal beta blocker ingestions: a review of the American Association of Poison Control Centers data from 1985 to 1995. J Toxicol Clin Toxicol 1997;35:353–359.

Peterson CD, Leeder JS, Sterner S. Glucagon therapy for beta-blocker overdose. DICP 1984;18:394–398.

CALCIUM CHANNEL BLOCKERS

Kerns W II, Kline J, Ford MD. Beta-blocker and calcium channel blocker toxicity. Emerg Med Clin North Am 1994;12:365–390.

Passal DB, Crespin FH Jr. Verapamil poisoning in an infant. Pediatrics 1984;73:543.

Ramoska EA, Spiller HA, Winter M, et al. A one-year evaluation of calcium channel blocker overdoses: toxicity and treatment. Ann Emerg Med 1993;22:196–200.

CLONIDINE

Fiser DH, Moss M, Walker W. Critical care for clonidine poisoning in toddlers. Crit Care Med 1990;18:1124–1128.

Heidemann SM, Sarnaik AP. Clonidine poisoning in children. Crit Care Med 1990;18:618–620.

Wiley JF, Wiley CC, Torrey SB, et al. Clonidine poisoning in young children. J Pediatr 1990;116:654–658.

DIGOXIN

Lewander WJ, Gaudreault P, Einhorn A, et al. Acute pediatric digoxin ingestion—a ten-year experience. Am J Dis Child 1986;140:770–773.

Woolf AD, Wenger T, Smith TW, et al. The use of digoxin-specific Fab fragments for severe digitalis intoxication in children. N Engl J Med 1992;326:1739–1744.

DISC BATTERIES

Litovitz T, Schmitz BF. Ingestion of cylindrical and button batteries: an analysis of 2382 cases. Pediatrics 1992;89:747–757.

Sheikh A. Button battery ingestions in children. Pediatr Emerg Care 1993;9:224–229.

FOODBORNE INTOXICATIONS

Mines D, Stahmer S, Shepherd SM. Poisonings: food, fish, shellfish. Emerg Med Clin North Am 1997;15:157–177.

Morrow JD, Margolies GR, Rowland J, et al. Evidence that histamine is the causative toxin of scombroid-fish poisoning. N Engl J Med 1991;324:716–720.

Shewmake RA, Dillon B. Food poisoning: causes, remedies, and prevention. Postgrad Med 1998;103:125–136.

Swift AE, Swift TR. Ciguatera. J Toxicol Clin Toxicol 1993;31:1–29.

HOUSEHOLD CLEANING PRODUCTS AND CAUSTICS

Anderson KD, Rousetin, Randolph JG. A controlled trial of corticosteroids in children with corrosive injury of the esophagus. N Engl J Med 1990;323:637–640.

Christesen HB. Prediction of complications following unintentional caustic ingestion in children: is endoscopy always necessary? Acta Paediatr 1995;84:1177–1182.

Crain EF, Gershel JC, Mezey AP. Caustic ingestions—symptoms as predictors of esophageal injury. Am J Dis Child 1984;138:863–865.

Gaudreault P, Parent M, McGuigan MA, et al. Predictability of esophageal injury from signs and symptoms: a study of caustic ingestion in 378 children. Pediatrics 1983;71:767–770.

Howell JM, Dalsey WC, Hartsell FW, et al. Steroids for the treatment of corrosive esophageal injury: a statistical analysis of past studies. Am J Emerg Med 1992;10:421–425.

Previtera C, Giusti F, Guglielmi M. Predictive value of visible lesions (cheeks, lips, oropharynx) in suspected caustic ingestion: may endoscopy reasonably be omitted in completely negative pediatric patients? Pediatr Emerg Care 1990;6:176–178.

Temple AR, Lovejoy FH Jr. Cleaning products and their accidental ingestion. New York. Soap and Detergents Association, 1980.

HYDROCARBONS

Chyka PA. Benefits of extracorporeal membrane oxygenation for hydrocarbon pneumonitis. J Toxicol Clin Toxicol 1996;34:357–363.

Truemper E, De La Rocha SR, Atkinson SD. Clinical characteristics, pathophysiology, and management of hydrocarbon ingestion: case report and review of the literature. Pediatr Emerg Care 1987;3:187–193.

Victoria MS, Nangia BS. Hydrocarbon poisoning: a review. Pediatr Emerg Care 1987;3:184–186.

IRON

Henretig FM, Karl SR, Weintraub WH. Management of severe iron poisoning with enteral and intravenous deferoxamine. Ann Emerg Med 1983;12:306–309.

Howland MA. Risks of parenteral deferoxamine for acute iron poisoning. J Toxicol Clin Toxicol 1996;34:491–497.

McGuigan MA. Acute iron poisoning. Pediatr Ann 1996;25:33–38.

Palatnick W, Tenenbein M. Leukocytosis, hyperglycemia, vomiting, and positive X-rays are not indicators of severity of iron overdose in adults. Am J Emerg Med 1996;14:454–455.

Tenenbein M. Whole bowel irrigation in iron poisoning. J Pediatr 1987;111:142–145.

Tenenbein M. Benefits of parenteral deferoxamine for acute iron poisoning. J Toxicol Clin Toxicol 1996;34:485–489.

ISONIAZID

Orlowski FP, Paganini EP, Pippenger CE. Treatment of a potentially lethal dose isoniazid ingestion. Ann Emerg Med 1988;17:73–76.

Shah BR, Snatucci K, Sinert R, et al. Acute isoniazid neurotoxicity in an urban hospital. Pediatrics 1995;95:700–704.

Shannon MW. Isoniazid. In: Haddad LM, Shannon MW, Winchester JF, eds. Clinical Management of Poisoning and Drug Overdose. 3rd ed. Philadelphia: WB Saunders, 1998;721–726.

Wason S, Lacouture PG, Lovejoy FH Jr. Single high-dose pyridoxine treatment for isoniazid overdose. JAMA 1981;246:1102–1104.

LEAD

American Academy of Pediatrics, Committee on Drugs. Treatment guidelines for lead exposure in children. Pediatrics 1995;96:155–160.

Angle CR. Childhood lead poisoning and its treatment. Annu Rev Pharmacol Toxicol 1993;33:409–434.

Centers for Disease Control and Prevention. Preventing lead poisoning in young children. US Department of Health and Human Services, 1991.

Glotzer DE. Management of childhood lead poisoning: strategies for chelation. Pediatr Ann 1994;23:606–612.

Glotzer DE, Weitzman M. Commonly asked questions about childhood lead poisoning. Pediatr Ann 1995;24:630–632.

Henretig FM. Lead. In: Goldfrank LR, Flomenbaum NE, Lewin NA, et al, eds. Goldfrank's Toxicologic Emergencies. 6th ed. Stamford, CT: Appleton & Lange, 1998:1277–1318.

Liebelt EL, Shannon MW. Oral chelators for childhood lead poisoning. Pediatr Ann 1994;23:616–626.

Shannon MW. Lead. In: Haddad LM, Shannon MW, Winchester JF, eds. Clinical Management of Poisoning and Drug Overdose. 3rd ed. Philadelphia: WB Saunders, 1998:767–783.

Shannon M, Graef J. Lead intoxication in infancy. Pediatrics 1992;89:87–90.

ORAL HYPOGLYCEMICS

Quadrani DA, Spiller HA, Widder P. Five year retrospective evaluation of sulfonylurea ingestion in children. J Toxicol Clin Toxicol 1996;34:267–270.

Szlatenyi CS, Capes KF, Wang RY. Delayed hypoglycemia in a child after ingestion of a single glipizide tablet. Ann Emerg Med 1998;31:773–776.

PHENOTHIAZINES

Buckley NA, Whyte IM, Dawson AH. Cardiotoxicity more common in thioridazine overdose than with other neuroleptics. J Toxicol Clin Toxicol 1995;33:199–204.

Gupta J, Lovejoy FH Jr. Acute phenothiazine toxicity in childhood: a five-year study. Pediatrics 1967;39:771–774.

Knight Me, Roberts RJ. Phenothiazine and butyrophenone intoxication in children. Pediatr Clin North Am 1986;33:299–309.

ORGANOPHOSPHATES

Farrar HC, Wells TG, Kearns GL. Use of continuous infusion of pralidoxime for treatment of organophosphate poisoning in children. J Pediatr 1990;116:658–661.

O'Malley M. Clinical evaluation of pesticide exposure and poisonings. Lancet 1997;349:1161–1166.

Zweiner RJ, Ginsburg CM. Organophosphate and carbamate poisoning in infants and children. Pediatrics 1988;81:121–126.

PLANTS/MUSHROOMS

Centers for Disease Control and Prevention. Jimson weed poisoning—Texas, New York, and California, 1994. MMWR 1995;44:41–44.

Challoner KR, McCarron MM. Castor bean intoxication. Ann Emerg Med 1990;19:1177–1183.

Furbee B, Wermuth M. Life-threatening plant poisoning. Crit Care Clin 1997;13:849–888.

Goldfrank LR. Mushrooms: toxic and hallucinogenic. In: Goldfrank LR, Flomenbaum NE, Lewin NA, et al, eds. Goldfrank's Toxicologic Emergencies. Stamford, CT: Appleton & Lange, 1998.

Hall AH, Spoerke DG, Rumack BH. Mushroom poisoning: identification, diagnosis, and treatment. Pediatr Rev 1987;8:291–298.

Horn S, Horina JH, Krejs GJ, et al. End-stage renal failure from mushroom poisoning with Cortinarius orellanus: report of four cases and a review of the literature. Am J Kidney Dis 1997;30:282–286.

Krenzolak EP, Jacobsen TD. Plant exposures—a national profile of the most common plant genera. Vet Hum Toxicol 1997;39:248–249.

O'Brien BL. A fatal Sunday brunch: amanita mushroom poisoning in a Gulf Coast family. Am J Gastroent 1996;91:581–583.

Ridker PM. Toxic effects of herbal teas. Arch Environ Health 1987;42: 133–136.

Safadi R, Levy I, Amitai Y, et al. Beneficial effect of digoxin-specific Fab antibody fragments in oleander intoxication. Arch Int Med 1995;155:2121–2125.

Schneider SM, Borochovitz D, Krenzelok EP. Cimetidine protection against alpha-amanitin hepatotoxicity in mice: a potential model for the treatment of amanita phalloides poisoning. Ann Emerg Med 1987;16: 1136–1140.

THEOPHYLLINE

Heath A, Knudsen K. Role of extracorporeal drug removal in acute theophylline poisoning. Med Toxicol Adv Drug Exp 1987;2:294–308.

Minton NA, Henry JA. Treatment of theophylline overdose. Am J Emerg Med 1996;14:606–612.

Shannon M. Predictors of major toxicity after theophylline overdose. Ann Intern Med 1993;119:1161–1167.

Shannon MW. Theophylline and caffeine. In: Haddad LM, Shannon MW, Winchester JF, eds. Clinical Management of Poisoning and Drug Overdose. 3rd ed. Philadelphia: WB Saunders, 1998:1093–1106.

Shannon M, Lovejoy FH Jr. Effect of acute versus chronic intoxication on clinical features of theophylline poisoning in children. J Pediatr 1992;121:125–130.

TRICYCLIC ANTIDEPRESSANTS/OTHER ANTIDEPRESSANTS

Hayes PE, Kristoff CA. Adverse reactions to five new antidepressants. Clin Pharm 1986;5:471–480.

Henry JA. Epidemiology and relative toxicity of antidepressant drugs in overdose. Drug Safety 1997;16:374–390.

Liebelt EL. Toxicology reviews: targeted management strategies for cardiovascular toxicity from tricyclic antidepressant overdose: the pivotal role for alkalinization and sodium loading. Pediatr Emerg Care 1998; 14:293–298.

Liebelt EL, Francis PD, Woolf AD. ECG lead aVR versus QRS interval in predicting seizures and arrhythmias in acute tricyclic antidepressant toxicity. Ann Emerg Med 1995;26:195–201.

Linden CH, Rumack BH, Strehike C. Monoamine oxidase inhibitor overdose. Ann Emerg Med 1984;13:1137–1140.

Mills KC. Serotonin syndrome: a clinical update. Crit Care Clin 1997; 13:763–783.

Spiller HA, Morse S, Muir C. Fluoxetine ingestion: a one year retrospective study. Vet Human Toxicol 1990;32:153–156.

Wedin GP, Oderda GM, Klein-Schwartz W, et al. Relative toxicity of cyclic antidepressants. Ann Emerg Med 1986;15:797–804.

DRUGS DANGEROUS IN SMALL DOSES

Koren G. Medications which can kill a toddler with one tablet or teaspoonful. Clin Toxicol 1993;31:407–413.

Liebelt EL, Shannon MW. Small doses, big problems. Pediatr Emerg Care 1993:292–297.

SUBSTANCE ABUSE

Aaron CK. Sympathomimetics. Emerg Med Clin North Am 1990;8:513–526.

Baldridge EB, Bessen HA. Phencyclidine. Emerg Clin North Am 1990; 8:541–550.

Bateman KA, Heagarty MC. Passive freebase cocaine (“crack”) inhalation by infants and toddlers. Am J Dis Child 1989;143:25–27.

Blum RW. Adolescent substance use and abuse. Arch Pediatr Adol Med 1997;151:805–808.

Ford M, Hoffman RS, Goldfrank LR. Opioids and designer drugs. Emerg Med Clin North Am 1990;8:495–512.

Henretig F. Inhalant abuse in children and adolescents. Pediatr Ann 1996;25:47–52.

Henretig FM, Slap GB. A guide to acute medical management of intoxication in adolescents. American Academy of Pediatrics: Adol Health Update 1994;6:1–7.

Kulig K. LSD. Emerg Med Clin North Am 1990;8:551–565.

Li J, Stokes SA, Woeckener A. A tale of novel intoxication: a review of the effects of gamma-hydroxybutyric acid with recommendations for management. Ann Emerg Med 1998;31(6):729–736.

LoVecchio F, Curry SC, Bagnasco T. Butyrolactone-induced central nervous system depression after ingestion of RenewTrient, a “dietary supplement” (Letter). N Engl J Med 1998;339:847–848.

Mueller PD, Benowitz NL, Olson KR. Cocaine. Emerg Med Clin North Am 1990;8:481–494.

Perry HE, Shannon MW. Diagnosis and management of opioid- and benzodiazepine-induced comatose overdose in children. Curr Opin Pediatr 1996;8:243–247.

CHAPTER 89

Environmental Emergencies

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DROWNING AND NEAR DROWNING

Background

Drowning is defined as water submersion with resultant asphyxiation and death within 24 hours; *near drowning* implies that resuscitation has extended survival beyond 24 hours. Each term is further classified according to whether aspiration has occurred.

Drowning is responsible for approximately 4000 deaths per year in the United States, affecting all age groups and affecting males two to three times more often than females. Drowning ranked fourth among unintentional injury deaths in recent years.

Older infants and toddlers are disproportionately represented in these accidents, and their survival rates are lower. They are vulnerable to immersion in household buckets, bath and hot tubs, swimming pools, and other bodies of water near their homes. Young teenagers are also at greater risk because adult supervision decreases and bravado increases. However, coexisting trauma, drug or alcohol use, and suicidal intent must be considered in each case. The importance of careful surveillance and public safety measures, such as fences around pools, cannot be overemphasized.

Pathophysiology

When a child is submerged, either breath-holding or laryngospasm occurs. If hypoxemia follows, loss of consciousness and cardiovascular collapse may occur without aspiration of fluid. Alternatively, submersion and frantic struggling may result in gasping, with subsequent aspiration. If antecedent head trauma, drug ingestion, seizure activity, or cardiac arrhythmia impair consciousness and protective airway reflexes, aspiration is more likely to occur. Although most organs may become involved, the major morbidity occurs in the pulmonary and central nervous systems.

Fresh water aspirated into the lungs is rapidly taken up into the circulation, resulting in a transient rise in circulating blood volume that is quickly redistributed through the body. In a canine study of cold water drowning, aspiration of fresh water caused body weight to increase an average of 16.5% with concomitant hemodilution. Aspiration of salt water caused body weight to increase only 6% with hemoconcentration and diminished intravascular volume. In humans, changes in the hematocrit are not predictable, and those that occur are more closely related to coexisting trauma than to effects of hypertonic or hypotonic fluids. Occasionally, however, massive hemolysis may occur. Electrolyte abnormalities that occur after massive aspiration in laboratory animals rarely achieve clinical significance in either adult or child victims.

However, even small (1 to 3 mL/kg) quantities of fresh water cause disruption of surfactant, a rise in surface tension in the lungs, and alveolar instability. Capillary and alveolar membrane damage allow fluid to leak into the alveoli with subsequent pulmonary edema.

Aspiration of salt water (osmolality greater than normal saline) does not denature surfactant but creates an osmotic gradient for fluid to accumulate in the lungs. This accumulated fluid greatly exceeds the volume that was aspirated and effectively removes surfactant from the alveolar–gas interface.

Both fresh and salt water aspiration decrease pulmonary compliance, increase airway resistance and pulmonary artery pressure, and diminish pulmonary flow. As nonventilated alveoli are perfused, an intrapulmonary shunt develops, leading to a drastic fall in partial pressure of arterial oxygen (Pa O_2).

In other animal studies, aspiration of as little as 2.2 mL/kg of fresh water led to a fall in Pa O_2 to about 60 mm Hg in 3 minutes, whereas a similar amount of sea water precipitated an even greater drop (to about 40 mm Hg). In humans, even lower levels of PaO_2 are seen; tissue hypoxia then leads to severe metabolic acidosis. The victim is usually able to correct a rise in partial pressure of arterial carbon dioxide (Pa CO_2). Aspiration of bacteria, gastric contents, and foreign materials may cause additional trauma to the lungs.

Hypoxemia, whether secondary to upper airway obstruction (here, laryngospasm) or to impaired gas exchange after aspiration, results in loss of consciousness. If anoxia ensues, irreversible central nervous system (CNS) damage begins after 4 to 6 minutes. Fear or cold may trigger the diving reflex (commonly encountered in infancy), which shunts blood to the brain and heart primarily and affords several minutes of additional perfusion. Experience with drowning victims and in cardiovascular surgery indicates that cold water is relatively protective of the CNS, but probably only if immersion hypothermia develops very rapidly or before compromise of oxygenation. Onset of hypothermia is more rapid in the victim who is younger (greater surface:volume ratio) or is struggling in or swallowing icy water. If, however, laryngospasm or aspiration occurs before a fall in core body temperature and cerebral metabolic rate, protection is probably minimal.

Cardiovascular effects are primarily those expected with myocardial ischemia, severe systemic acidosis, hypothermia, and intravascular volume changes. After aspiration of fresh water, the transient rise in intravascular volume later contributes to problems of cerebral edema and pulmonary function.

Clinical Manifestations

In the first moments after rescue, the appearance of the child who has nearly drowned may range from apparently normal to apparently dead. Body temperature is often low, even in temperate, warm-water environments. Respiratory efforts may be absent, irregular, or labored, with pallor or cyanosis, retractions, grunting, and cough productive of pink, frothy material. The lungs may be clear, or there may be rales, rhonchi, and wheezing. Infection may develop as a consequence of aspirated mouth flora or organisms in stagnant water, but this is not usually important in the first 24 hours.

Respiratory function may improve spontaneously or deteriorate rapidly as pulmonary edema and small airway dysfunction worsen. Alternatively, deterioration may ensue slowly over 12 to 24 hours.

Intense peripheral vasoconstriction and myocardial depression may produce apparent or actual pulselessness.

Neurologic assessment may show an alert, normal child or any level of CNS compromise. A child may display agitation and combative behavior, blunted responsiveness to the environment, or profound coma with stereotypic posturing or flaccid extremities. Superficial evidence of head trauma may be noted in a few children whose submersion episode was a secondary event.

Pulmonary and neurologic damage need not occur together. Although extensive pulmonary destruction and resultant hypoxemia may cause neurologic damage, all combinations of mild and severe lung and brain damage are possible.

Management

The ultimate outcome of serious immersion accidents depends on the duration of submersion, the degree of pulmonary damage by aspiration, and in some cases, effectiveness of initial resuscitative measures. When all children who experience immersion accidents are considered as a group, most are salvageable, and all should receive the benefit of excellent cardiopulmonary resuscitation without delay at the scene, according to the principles elaborated in Chapter 1. In particular, children should be given the maximum concentration of supplemental oxygen possible (100%) in transport to an emergency facility. Even those rescued with spontaneous ventilation and minimal or no neurologic dysfunction should receive the benefit of supplemental oxygen to minimize the risk of progressive hypoxemia and acidosis with secondary myocardial and cerebral damage. Physical examination is notoriously insensitive to hypoxemia; a seriously hypoxemic child may be alert and talking. Once the child has arrived at an emergency facility (and cardiovascular stability is achieved), pulmonary and neurologic assessment should guide further treatment.

Several recent pediatric studies have attempted to predict outcome in submersion accidents. One prospective investigation devised a prediction rule for children submerged in nonicy water who presented to the emergency department (ED) in a comatose state: lack of pupillary light reflex, male sex, and initial blood glucose were variables used to predict unfavorable outcome (vegetative state or death). A retrospective study of children presenting to the ED after warm-water submersion suggested that hemodynamic, rather than neurologic, status was more highly predictive of poor neurologic outcome.

Effective therapy of near drowning depends on the reversal of hypoxemia and metabolic acidosis. The pulmonary status is assessed initially with a chest radiograph (Fig. 89.1) and with measurement of arterial oxygen saturation ($Sa{O_2}$) and arterial blood gas (ABG), as in Table 89.1. If oxygenation is normal on breathing room air, the child can be assumed to have suffered near drowning without aspiration. Observation for 12 to 24 hours with repeat ($Sa\ O_2$) or ABG determination should be sufficient to assess the possibility of late deterioration in gas exchange.

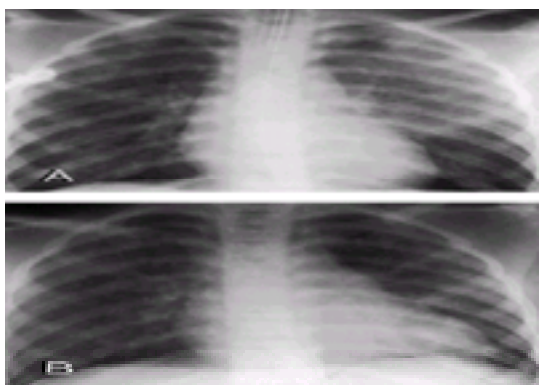


FIGURE 89.1. Near drowning in a 4-month-old girl. **A.** There is bilateral disseminated alveolar pattern, more on the left than on the right, consistent with the pulmonary edema of near drowning. This change may be the result of neurologic pulmonary edema rather than aspirated water. **B.** Two days later, the patient has been extubated and there is marked improvement in appearance of pulmonary edema secondary to near drowning. (Courtesy of Soroosh Mahboubi, MD).

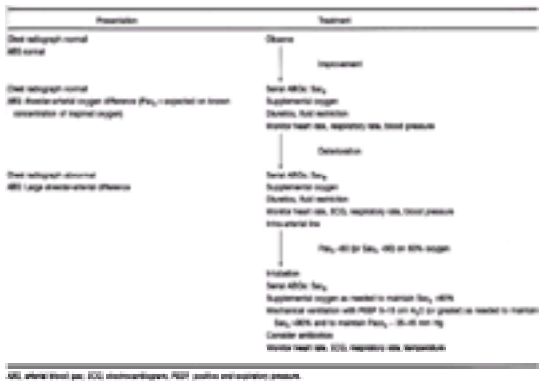


Table 89.1. Pulmonary Assessment after Near Drowning

Other initial laboratory evaluation should include complete blood count (CBC), electrolytes, and urinalysis.

Patients with abnormalities of gas exchange and acid-base status (but with normal chest radiographs) can usually be managed with supplemental oxygen, pulmonary physiotherapy, and bicarbonate therapy. Additional blood gas analysis should be done to document adequate oxygenation and reversal of metabolic acidosis. Any change in mental status or increase in respiratory distress may reflect arterial hypoxemia and should also prompt a repeat ABG determination. Continuous Sa_{O_2} or serial Pa_{O_2} measurements will guide the physician to continue conservative treatment or to intensify ventilatory support.

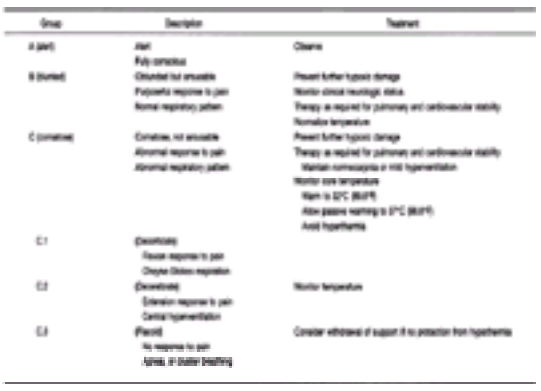
Patients with obvious respiratory distress, hypoxemia (Sa_{O_2} less than 90% or Pa_{O_2} less than 60 on 60% inspired oxygen) and extensive pulmonary edema or infiltration generally require more vigorous treatment and more extensive monitoring. All should be monitored for heart rate, cardiac rhythm, respiratory rate, and blood pressure. Most will require frequent blood gas analysis and may be more easily monitored through arterial cannulation. Intubation, supplemental oxygen, and mechanical ventilation with positive end-expiratory pressure (PEEP, 5 to 15 cm H_2O) should be provided.

Once blood pressure is stabilized, fluid restriction (to approximately one-half the maintenance rate) and diuretic therapy (e.g., furosemide 0.5 to 1 mg/kg intravenously) may improve gas exchange. In the setting of extensive pulmonary damage, pulmonary and cardiovascular components of the disease are intimately entwined. Optimum management requires monitoring of blood gases and systemic arterial pressure.

The risk of pulmonary infection is always present, but retrospective studies have not demonstrated benefit from prophylactic antibiotics, which should be reserved for proven bacterial infection. Exceptions may be made when grossly contaminated water is aspirated and, in the worst cases, when maximal ventilatory support is required to provide any margin for survival. Most studies of bronchoalveolar lavage show no improvement. Steroids have no demonstrated benefit.

Renal function must be maintained. If significant hemoglobinuria exists, forced diuresis is required. Maintenance of an adequate hemoglobin level (more than 10 g/100 mL) and normal electrolytes is obviously necessary. Specific problems vary from patient to patient in an unpredictable way, and an understanding of the principles outlined in Chapter 86 and Chapter 87 is essential.

The patient's clinical condition in the ED dictates further management and may provide prognostic clues. Patients may be assigned to one of three groups (Table 89.2). Those who are awake, alert, and fully responsive have survived the episode, presumably without CNS damage. Conservative observation for only 12 to 24 hours is warranted. The second group includes those who are obtunded but arousable and exhibit a normal respiratory pattern and purposeful responses to pain. These patients have suffered certain but reversible CNS hypoxia; the goal is to prevent further hypoxic damage through intensive management of cardiopulmonary disease. Repeated neurologic evaluation is essential, and fluid restriction and diuretic therapy within the limits of cardiovascular stability may decrease the risk of cerebral edema. There is no demonstrated value in the use of steroids in this setting (see Chapter 125). In both groups, temperature normalization should be prompt. If reversal of coexisting pulmonary damage is effective, neurologic recovery should be complete.



Group	Description	Treatment
A (alert)	Alert Fully conscious	Observe
B (stunned)	Obtunded but arousable Purposeful response to pain Normal respiratory pattern	Prevent further hypoxic damage Monitor clinical neurologic status Therapy as required for pulmonary and cardiovascular stability Normalize temperature
C (unresponsive)	Comatose, not arousable Abnormal response to pain Abnormal respiratory pattern	Prevent further hypoxic damage Therapy as required for pulmonary and cardiovascular stability Maintain normocapnia or mild hyperventilation Monitor core temperature Warm to 32°C (89.6°F) Allow passive warming to 37°C (98.6°F) Avoid hyperthermia
C.1	(Decorticate) Flexion response to pain Cheyne-Stokes respiration	
C.2	(Decerebrate) Extension response to pain Central hyperventilation	Monitor temperature
C.3	(Flaccid) No response to pain Apneustic or fixed breathing	Consider withdrawal of support if no protection from hypothermia

Table 89.2. Neurologic Assessment after Near Drowning

There is controversy over patients in a third group—those who have experienced severe CNS asphyxia. These children are not arousable and can be further divided into three subcategories according to neurologic findings: 1) those with decorticate response to pain and Cheyne-Stokes breathing; 2) those with decerebrate response to pain and central hyperventilation; and 3) those who are flaccid with fixed, dilated pupils and apneustic breathing or apnea. Again, reversal of hypoxemia and acidosis is critical. Fluid resuscitation should be designed to prevent hyperglycemia. Avoiding hypercapnia and resultant cerebral hyperemia is generally accepted, but hyperventilation, barbiturate coma, and other measures initially thought to provide cerebral protection and prevent or treat elevated intracranial pressure have not been helpful in these patients.

Hypothermia does appear to have some protective value. Extreme hypothermia should be corrected to at least 32°C (89.6°F) to achieve hemodynamic stability and to minimize the risk of infection. The child should then be allowed to rewarm passively. Although data in humans are limited, animal studies suggest that maintenance of mild brain hypothermia may minimize reperfusion injury. Hyperthermia, a common result of active rewarming, should be avoided.

The prognosis for this group is certainly more grim, with a much greater risk of death or severe anoxic/ischemic encephalopathy. Risk increases with depth of coma on presentation. Patients in the third subdivision (flaccid with fixed, dilated pupils) rarely survive intact regardless of treatment, although coexistent hypothermia has provided some remarkable exceptions.

Recent studies indicate that patients with asystole on arrival in the ED have uniformly poor neurologic outcomes. In each case, consideration should be given to the possibility that continued resuscitation will salvage only the cardiovascular system; in these cases, the physician may reasonably discontinue resuscitative efforts.

SMOKE INHALATION

Background

Among unintentional injuries, fires rank fifth, killing nearly 4000 people annually in the United States. Although fire has been the cause of much death and misery throughout human history, the importance of smoke inhalation has been recognized only in the last 50 years. Respiratory complications of smoke inhalation rank with carbon monoxide poisoning (see next section) as a major cause of early death from fire. Although serious pulmonary disease may occur in the absence of cutaneous injury, inhalation injury dramatically increases the morbidity and mortality associated with any given percent body surface area burn.

The severity of carbon monoxide inhalation and respiratory problems is related to the duration of exposure, the occurrence in a closed space (more likely in very young or elderly victims), the nature of materials involved, and the presence of products of incomplete combustion. Severe hypovolemic shock, massive tissue destruction, extensive fluid resuscitation, and infection further complicate direct inhalation trauma.

Pathophysiology

The relatively low heat capacity of dry air and the excellent heat exchange properties of the nasopharynx usually limit direct thermal injury to the upper airway. Dry air above 160°C (320°F) has little effect on the lower airway. The greater heat capacity of steam increases the risk of lower airway damage. In addition, continuing combustion of soot particles carried deeply into the lung may exacerbate thermal injury.

Chemical injury may occur at any level of the respiratory tract. Oxides of sulfur and nitrogen combine with lung water to form corrosive acids. Incomplete combustion of any carbon-containing material, such as wood, may produce carbon monoxide. Combustion of cotton or plastic generates aldehydes that cause protein denaturation and cellular damage. One example is acrolein, known to cause upper airway irritation at concentrations of 5.5 parts per million (ppm) and pulmonary edema within seconds at 10 ppm. Polyvinylchloride releases chlorine and hydrochloric acid, whereas polyethylene produces hydrocarbons, ketones, and other acids. One of the hydrocarbons, benzene, may anesthetize the upper airway to facilitate passage of toxins and particles. Burning polyurethane may produce cyanide gas. Fire retardants that contain phosphorus may actually produce phosgene gas. The upper airway filters most soot particles, but those carried into the lung may adsorb various substances and cause reflex bronchospasm to further extend chemical damage.

Upper airway lesions include actual burns of varying severity as well as severe edema of the nose, mouth, pharynx, and laryngeal structures. A murine model of wood smoke inhalation suggests that combustion produces hydroxyl radicals in

the gas phase of smoke that cause a reflex apneic response. In the investigation, a hydroxyl radical scavenger, applied to the larynx, attenuated the response. Upper airway edema increases inspiratory and expiratory resistance and causes a dramatic increase in the work of breathing. If airway narrowing is severe or complete, acute respiratory failure with hypercarbia and hypoxemia occurs and sets the stage for subsequent cardiovascular collapse.

Lower airway lesions depend on the toxin involved. A variety of features may aggravate pulmonary pathology, including circulatory, metabolic, and infectious complications, as well as therapeutic interventions such as endotracheal intubation, oxygen administration, mechanical ventilation, and fluid therapy.

Immediate effects of smoke inhalation on the lower airway include loss of ciliary action, mucosal edema, bronchiolitis, alveolar epithelial damage, and impaired gas exchange, particularly oxygenation. In addition, areas of atelectasis or air trapping worsen ventilation-perfusion mismatch and hypoxemia. Loss of surfactant activity exaggerates this phenomenon. Hours later, sloughing of tracheobronchial mucosa and mucopurulent membrane formation increase the degree of obstruction and poor gas exchange as well as the likelihood of infection. Beyond the first 24 hours, pulmonary pathology that results from smoke inhalation is largely indistinguishable from adult respiratory distress syndrome, which arises from other insults.

The Dellwood nursery fire demonstrated that infants who died sustained serious respiratory damage in the absence of cutaneous injury. Necrosis of bronchial, bronchiolar, and alveolar epithelium; vascular engorgement and edema; and formation of membranes or casts of the airway produce small and large airway obstruction. Severe cutaneous injury increases alveolar capillary permeability, leading to pulmonary hemorrhage, edema, and hyaline membrane formation.

Clinical Manifestations

A history of exposure in a closed space should heighten concern for smoke inhalation. Need for cardiopulmonary resuscitation at the site implies significant carbon monoxide poisoning and/or hypoxia secondary to decreased ambient oxygen concentration or severe respiratory disease. The physician should also consider the types of material involved to determine the risk of poisoning from carbon monoxide or other toxins.

Physical examination that reveals facial burns, singed nasal hairs, pharyngeal soot, or carbonaceous sputum justifies a presumption of smoke inhalation. Any sign of neurologic dysfunction, including irritability or depression, should be presumed related to tissue hypoxia until proven otherwise. Signs of respiratory distress may be delayed for 12 to 24 hours, but tachypnea, cough, hoarseness, stridor, decreased breath sounds, wheezing, rhonchi, or rales may be detected on presentation. Auscultatory findings often precede chest radiograph abnormalities by 12 to 24 hours. Radiographic changes may include diffuse interstitial infiltration or local areas of atelectasis and edema (Fig. 89.2).

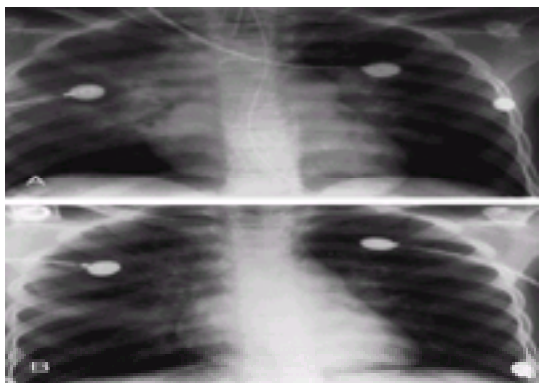


FIGURE 89.2. Smoke inhalation in a 9-year-old girl. **A.** There is bilateral central alveolar process consistent with acute smoke inhalation. **B.** A day later, the patient has been extubated and there is marked improvement in the appearance of pulmonary edema. (Courtesy of Soroosh Mahboubi, MD)

Acute respiratory failure may occur at any point. The cause may be asphyxia or carbon monoxide exposure and subsequent CNS depression initially, or airway obstruction or parenchymal dysfunction later. ABG analysis provides the ultimate assessment of effective respiratory function. Xenon lung scanning may provide evidence for smoke inhalation but does not add significantly to repeated clinical assessment and blood gas determinations in the ED. Bronchoscopy can document the extent of inhalation injury and help remove debris but may worsen airway edema. In general, it is respiratory function, not appearance of lesions, that guides supportive care, and therefore most patients can be treated effectively without bronchoscopy.

Management

Initial assessment and resuscitation at the scene of the fire should proceed according to the principles outlined in Chapter 1. Because of the likelihood of carbon monoxide exposure and the difficulty of assessing hypoxemia clinically, all victims should receive the maximum concentration of inspired oxygen possible in transport and in the ED until further evaluation is complete (Table 89.3).

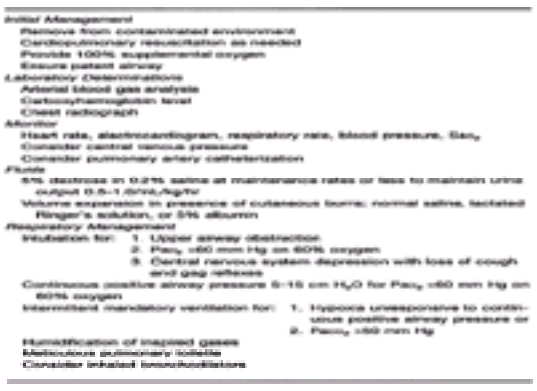


Table 89.3. Management of Smoke Inhalation

Upon the patient's arrival in the ED, assessment of the airway and respiratory functions must proceed simultaneously with cardiovascular stabilization. Thermal injury to the nose, mouth, or face, or compromise of the upper airway (stridor, hoarseness, barking cough, retractions, delayed inspiration, or difficulty handling secretions) indicates the need for direct laryngoscopy. The presence of significant pharyngeal, supraglottic, or glottic edema mandates elective endotracheal intubation. Although some clinicians may elect to observe closely, worsening edema over 24 hours may lead to respiratory arrest and difficult emergency intubation through a distorted airway. Elective tracheostomy may be considered if placing or securing the endotracheal tube will further traumatize an edematous airway or severe facial burns. However, in the presence of extensive cutaneous burns, tracheostomy dramatically increases the risk of systemic and pulmonary infection.

Cardiovascular stabilization depends on fluid replacement, which is complex when major surface burns have occurred. The details of therapy are elaborated in Chapter 114, but in general, the goals are stabilization of cardiovascular function without fluid overload and compromise of gas exchange. Pulse rate and blood pressure should guide administration of fluid volume. Maintenance of urine output of at least 0.5 mL/kg per hour should provide adequate tissue perfusion. Decreased urine output may respond to diuretics or inotropic agents. Although adequate fluid administration is essential, careful monitoring of renal and cardiovascular systems may prevent or minimize acute pulmonary edema and delayed pulmonary dysfunction secondary to late fluid mobilization and infection.

Oxygen saturation and serial blood gas determinations should be obtained to guide oxygen supplementation and to assess adequacy of ventilation. Intubation is indicated if adequate oxygenation (Sa O_2 less than 90 or PaO_2 60 mm Hg or greater) cannot be maintained with an inspired oxygen concentration of 40 to 60%, if Pa CO_2 rises above 50 mm Hg, or if the work of breathing appears unsustainable. In the presence of small airway edema and disrupted surfactant activity, continuous distending airway pressure may improve oxygenation. Spontaneous ventilation with continuous positive airway pressure (CPAP) causes less cardiovascular interference, but in the patient with severe CNS depression or severe pulmonary parenchymal damage, mechanical ventilation with PEEP will likely be necessary. Maximally humidified oxygen should be delivered by mask or artificial airway to prevent inspissation of debris and occlusion of the airway. The patient with a natural airway should also receive humidified gas mixtures and be encouraged to take deep breaths and cough frequently. If an endotracheal tube is necessary, meticulous pulmonary toilette is essential, with frequent suctioning to remove edema fluid, mucus, and sloughed epithelium that may otherwise occlude the endotracheal tube.

A recent study of lung mechanics in children with inhalation injury compared two modes of ventilation. High-frequency percussive ventilation was superior to conventional mechanical ventilation in reducing work of breathing.

After the first few hours, diuretic therapy (furosemide 0.5 to 1 mg/kg intravenously) within the limits of cardiovascular stability may also improve oxygenation and pulmonary compliance, leading to more effective ventilation. Chemical and particulate irritation of upper airway receptors may cause reflex bronchoconstriction and contribute to lower airway obstruction. Bronchodilators such as nebulized albuterol (2.5 mg in 2.5 mL, 0.9% sodium chloride) or intravenous (IV) terbutaline (load 10 µg/kg per dose intravenously or subcutaneously; drip 0.1 to 1.0 µg/kg per minute) may help reverse bronchospasm, but relief depends mostly on removal of secretions and debris from the respiratory tree.

Studies have not demonstrated a role for steroids in reducing airway edema or in decreasing the inflammatory response to smoke inhalation. When steroids are used, there is evidence that sodium and fluid retention increase, healing is delayed and bacterial clearance from the lung is decreased. Little argument remains for their routine use.

Similarly, there is no value in the use of prophylactic antibiotics. Institution of antimicrobial therapy should await specific indications, which rarely occur in the first 24 hours.

CARBON MONOXIDE POISONING

Background

Each year, unintentional carbon monoxide poisoning claims about 500 lives and is largely responsible for early deaths related to fire. However, exposure may occur in a variety of other settings unrelated to accidental fires, including incomplete combustion of any carbon-containing fuel. Poisoning may occur with exposure to improperly vented wood- or coal-burning stoves, and to automobile exhaust in garages (in most cases, garage doors and windows were actually open). Passengers may be poisoned in vehicles with open backs, or with faulty or blocked exhaust systems. During a 1996 blizzard in New York City, snow blocked exhaust pipes of idling automobiles and rendered 21 people (8 less than

16 years of age) unconscious.

Pathophysiology

In the normal person, carboxyhemoglobin levels are less than 1%. In smokers, levels of 5 to 10% are common. Inhaled carbon monoxide has two important effects that conspire to cause tissue hypoxia: 1) carbon monoxide binds to hemoglobin with an affinity 200 to 300 times greater than that of oxygen, and 2) it shifts the oxyhemoglobin dissociation curve to the left and changes the shape from sigmoidal to hyperbolic (Fig. 89.3). The first effect decreases oxygen content of the blood, whereas the second causes oxygen release at lower-than-normal tissue oxygen levels. Other endogenous (anemia) and exogenous (high altitude, consumption, or displacement of ambient oxygen during fires) factors contribute further to hypoxia. Although oxygen content of the blood is low, the Pa O_2 remains normal. Because carotid body receptors respond to Pa O_2, respiration may not be stimulated until late, when metabolic acidosis activates other centers. Tissue hypoxia increases cerebral blood flow, cerebrospinal fluid pressure, and cerebral capillary permeability, which predispose the patient to cerebral edema.

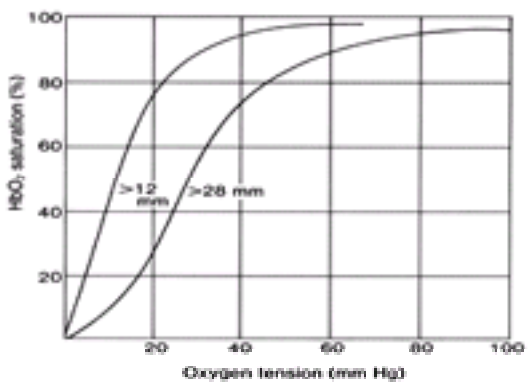


FIGURE 89.3. Carbon monoxide shifts the oxyhemoglobin dissociation curve to the left and changes its shape. This makes unloading of oxygen in the tissues more difficult and provides an inadequate diffusion gradient. (*Left curve,* P_{50} = 12 mm; *right curve,* P_{50} = 28 mm.)

Carbon monoxide interacts with several other cellular proteins, including cytochrome oxidase. It appears to interfere with oxidative energy production and may generate free radicals, which exacerbate CNS dysfunction. Carboxyhemoglobin may even act as a neurotransmitter, altering smooth muscle tone and indirectly contributing to a reperfusion injury.

Clinical Manifestations

History provides the most valuable clue to diagnosis. Carbon monoxide poisoning should be suspected in all fire victims and considered in children exposed to other hazards noted earlier. Presence or absence of the classically described “cherry red” skin color is of no diagnostic value. In fact, patients with thermal injury may appear red, whereas those with vasoconstriction may be quite pale. Both color and respiratory rate may be deceptive and may lead the physician away from recognition of severe tissue hypoxia. Pa O_2 and arterial saturation as determined by pulse oximetry (Sa O_2) are likely to be normal in carbon-monoxide intoxication; low values reflect coexistent pulmonary dysfunction.

Determination of blood levels of carboxyhemoglobin may help document the diagnosis and may aid prognosis. Spectrophotometric methods are most widely used clinically. Venous blood may be used because of the high affinity of carbon monoxide for hemoglobin, but an arterial sample provides more precise information about acid-base balance and adequacy of ventilation. The level of hemoglobin should also be determined.

Levels of carboxyhemoglobin as low as 5% in nonsmokers may impair judgment and fine motor skills. Mild intoxication (20% carboxyhemoglobin) produces headache, mild dyspnea, visual changes, and confusion. Moderate poisoning (20 to 40%) produces drowsiness, faintness, nausea and vomiting, tachycardia, dulled sensation, and decreased awareness of danger. At lower levels, these symptoms are noted only with exertion, but as the fraction approaches 40%, they are present at rest. Between 40 and 60%, weakness, incoordination, and loss of recent memory occur, and cardiovascular and neurologic collapse is imminent. Above 60%, coma, convulsions, and death are almost certain. Although carboxyhemoglobin levels and symptoms tend to follow the pattern just described, individual patients may be more or less symptomatic than predicted. An important caveat is that blood carboxyhemoglobin levels will fall rapidly with time and may not reflect cellular dysfunction, especially in high-demand tissues of the heart and CNS.

Patients with severe poisoning are peculiarly vulnerable to pressure trauma to skin, subcutaneous tissue, and muscle, especially at sites that support body weight or that are pinned under fallen objects. The history may suggest which sites are most vulnerable, and pain is an early symptom. Muscle breakdown and myoglobin deposition in renal tubular cells may precipitate acute renal failure.

A syndrome of delayed neuropsychologic sequelae (DNS) had been described in patients after exposure to carboxyhemoglobin. These patients develop neurologic symptoms acutely, appear to recover with treatment, and then exhibit a broad spectrum of neurologic and psychiatric abnormalities days to weeks after the exposure. Neuropsychiatric testing of children has obvious difficulties. Studies of DNS, many of which are methodologically flawed, have elucidated neither an exact mechanism nor a consensus on prevention and treatment.

Management

Most important and obvious is the immediate need to remove the victim from the contaminated environment (Table 89.4). Resuscitation should proceed according to general principles. As soon as possible, the patient suspected of suffering carbon monoxide poisoning should be provided 100% oxygen. If the patient is breathing spontaneously, this can be accomplished with well-fitting masks supplied with nonrebreathing valves and a reservoir bag. Entrainment of room air precludes simple masks from providing more than 40% oxygen. The half-life of carboxyhemoglobin is approximately 4 hours in a patient breathing room air at sea level and approximately 1 hour if pure oxygen is inspired. The half-life is further reduced to less than 30 minutes if the patient has access to hyperbaric oxygen (HBO) at 2 to 3 atmospheres' pressure. There is no widespread agreement on indications for HBO, and transfer to a hyperbaric chamber should not jeopardize meticulous conventional cardiopulmonary stabilization. However, HBO administration may have effects beyond the mere reduction in carboxyhemoglobin half-life. Some studies in adults suggest a role for HBO in reducing the incidence of mortality and DNS. Well-controlled studies in children would have to be undertaken to answer these questions definitively but would have obvious ethical and methodologic difficulties. In any case, early consultation with an HBO facility should be considered while the patient is receiving 100% oxygen.

Initial Management
Remove from contaminated environment
Cardiopulmonary resuscitation as needed
Provide 100% supplemental oxygen
Laboratory Determinations
Arterial blood gas analysis
Carboxyhemoglobin level
Complete blood count, electrolytes
Urinalysis for myoglobin
Monitor
Heart rate, electrocardiogram, respiratory rate, blood pressure
Treatment
Correct anemia Hgb <10 g/dL
Continue supplemental oxygen until carboxyhemoglobin ≤5%
Decrease oxygen consumption with bedrest, avoid producing anxiety
Maintain urine output >1 mL/kg/hr
Consider hyperbaric oxygen

Table 89.4. Management of Carbon Monoxide Poisoning

Use of inspired gas mixtures that contain carbon dioxide (5%) to increase minute ventilation may precipitate respiratory failure if used at the scene, as ventilation cannot be well monitored or assisted. It may be considered under closely controlled situations in the hospital but probably is of little benefit.

Metabolic acidosis, if present, should be treated with sodium bicarbonate, although adequacy of ventilation must be assessed to prevent paradoxical intracellular acidosis. The possibility of coexistent cyanide poisoning should be considered in patients involved in closed-space fires (especially where nitrogen-containing synthetic materials have burned) who have a persistent metabolic acidosis in the context of normal carboxyhemoglobin and methemoglobin. Cyanide has high mortality but a short half-life (approximately 1 hour), so empiric cyanide levels on patients who have survived the scene are not recommended generally unless confirmation is needed. If cyanide poisoning is strongly suspected in an early-presenting patient, the cyanide antidote kit (formerly known as the Lilly kit) may be considered. This two-step kit must be used with caution because the nitrite-containing first step induces methemoglobinemia. In case of doubt, the thiosulfate-containing second step, which is able to scavenge cyanide without significant additional toxicity, may be given alone. Anemia (hemoglobin less than 10 g/100 mL) must be corrected to maximize oxygen-carrying capacity. If myoglobinemia or myoglobinuria is present, vigorous hydration and forced diuresis with furosemide (1 mg/kg intravenously) and/or mannitol (0.25 to 1 g/kg intravenously) with close attention to urine output may preserve renal function. If hydration and diuresis are ineffective, renal failure should be considered and fluids restricted accordingly (see Chapter 86).

The patient should be observed for at least 24 hours to identify other sequelae of smoke inhalation.

ENVIRONMENTAL AND EXERTIONAL HEAT ILLNESS

Background

Environmental and exertional heat illness occurs with excessive heat generation and storage. These conditions arise when high ambient temperature prevents heat dissipation by radiation or convection, and not on humidity limits cooling by sweat evaporation. The spectrum of illness is broad, including heat cramps, heat exhaustion, and heat stroke; the latter is an acute medical emergency with significant associated morbidity and mortality.

Heat illness is a serious tropical health hazard, but even in the United States, summer heat is implicated in the deaths of more than 4000 people per year. Heat waves bring epidemic illness, but sporadic cases are also common. The elderly are most vulnerable (more than 80% of cases occur in people older than 50), but heat illness is significant among healthy young people as well. Military recruits, laborers, and athletes who work in a hot, humid atmosphere are notoriously vulnerable. Obesity, physical disability, heart disease, and alcohol and drug use increase risk. Pediatricians need to understand the special risk to young athletes, children with cystic fibrosis, infants left in automobiles on hot days, and the rare child with congenital absence of sweat glands. Mortality related to acute heat stroke has been estimated to range from 17 to 70%, depending on the severity of heat stress and age of the patient.

Pathophysiology

Under normal conditions, body core temperature is maintained constant within 0.6°C (1°F) when the environment varies from 9.4° to 60°C (49°–140°F) in dry air. This represents a remarkable balance between heat production and heat loss by the body. Heat is produced as 1) a byproduct of basal metabolism, 2) a consequence of muscle activity (including shivering), and 3) the effects of thyroxine and sympathetic stimulation on cellular processes. Heat is lost by 1) conduction to objects and air, 2) convection through air or liquid that surrounds tissues, 3) evaporation, and 4) radiation of infrared energy.

Conduction to objects represents a small fraction (about 3%) of heat lost. Conduction to air and convection represent another 12% in still air. As air movement increases, the proportion of heat lost by these mechanisms may increase to nearly 60%. Evaporative losses normally account for 25% of heat lost, and radiant heat losses account for about 60%. However, the body gains heat by conduction and radiation when ambient temperature exceeds skin temperature and loses heat only by evaporation. High ambient humidity and absence of convection currents decrease the rate of evaporation.

Heat-sensitive centers of the posterior hypothalamus control sympathetic tone. This tone regulates vasoconstriction of arterioles and subcutaneous arteriovenous anastomoses, which, in turn, controls heat conduction from the body core to the skin. Flow through these areas may represent 0 to 30% of total cardiac output. High flow provides efficient heat transfer from the body core to the skin, which is an effective radiator. Low flow to the skin prevents radiation and allows only inefficient diffusion through the insulating skin and subcutaneous tissues.

When body temperature rises, blood in the preoptic area of the anterior hypothalamus is warmer than optimal. Impulses from this area increase and are conducted through autonomic pathways to the spinal cord and then, through cholinergic fibers to the sweat glands, where sweat is released. Exercise and certain emotional states release circulating epinephrine and norepinephrine to increase sweat production.

The sweat gland is composed of two portions. The deep coiled portion actively elaborates a precursor secretion in response to cholinergic stimulation. At low rates of sweating, much of the sodium chloride (NaCl) contained in the precursor secretion is reabsorbed before the sweat is conducted to the skin surface. At higher rates, flow exceeds the capacity of the duct to reabsorb solute, and substantial total body NaCl depletion may occur.

In the unacclimatized adult, sweating may vary from negligible amounts at rest in a cool, dry environment to 1.5 L/hour during vigorous activity in hot weather. Long-term exposure to tropical weather results in a steady increase in sweating rate over approximately 6 weeks, to a maximum of about 4.0 L/hour. Initially, enormous salt losses may occur (15 to 20 g/day). However, aldosterone secretion rises and stimulates active reabsorption of NaCl in the ducts of sweat glands (as in the kidney), and salt losses decrease to a normal 3 to 5 g/day.

In humans, behavioral control over temperature regulation is probably as important as all other mechanisms. When body temperature changes, sensations of excessive warmth or cold prompt efforts to correct the situation. One moves out of the cold or into the shade, selects warmer or cooler clothing, initiates maneuvers that warm or cool the environment, or alters levels of activity. Choice of clothing that traps a layer of air around the body prevents significant heat loss by convection. A reflective inner surface decreases radiant heat loss by more than half. Light-colored clothing permeable to moisture but impervious to radiant heat from the environment, however, prevents the formation of an insulating layer of air. This allows for heat loss by evaporation and prevents radiant heat gain.

Clinical Findings

Three types of heat illness are recognized and represent different physiologic disturbances (Table 89.5). Heat cramps refer to the sudden onset of brief, intermittent, and excruciating cramps in muscles after they have been subjected to severe work stress. Cramps tend to occur after the work is done, on relaxing or on taking a cool shower. Occasionally, abdominal muscle cramps may simulate an acute abdomen. The usual victim is highly conditioned and acclimatized. Typically, these individuals can produce sweat in large quantities and provide themselves with adequate fluid replacement but inadequate salt replacement. Electrolyte depletion is probably the cause of cramps.

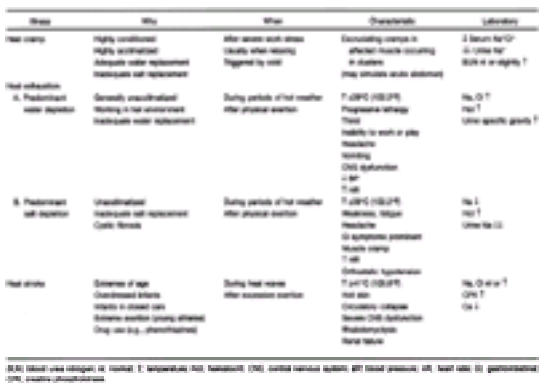


Table 89.5. Characteristics of Heat Illness

Most spasms last less than a minute, but some persist for several minutes, during which a rock-hard mass may be palpated in the affected muscle. Cramps often occur in clusters. Rapid voluntary contraction of a muscle, contact with

cold air or water, or passive extension of a flexed limb may reproduce a cramp. Laboratory investigation reveals hyponatremia and hypochloremia and virtually absent urine sodium. The blood urea nitrogen (BUN) level is usually normal but may be mildly elevated.

Heat exhaustion is less clearly demarcated from heat stroke than are heat cramps. In most cases, water depletion predominates because individuals who live and work in a hot environment do not voluntarily replace their total water deficit. Progressive lethargy, intense thirst, and inability to work or play progress to headache, vomiting, CNS dysfunction (including hyperventilation, paresthesias, agitation, incoordination, or actual psychosis), hypotension, and tachycardia. Hemoconcentration, hypernatremia, hyperchloremia, and urinary concentration are typical. Body temperature may rise but rarely over 39°C (102.2°F). If unattended, heat exhaustion may progress to frank heat stroke.

Heat exhaustion may also occur secondary to predominant salt depletion. As in heat cramp, water losses are replaced but without adequate electrolyte supplementation. Symptoms include profound weakness and fatigue, frontal headache, anorexia, nausea, vomiting, diarrhea, and severe muscle cramps. Tachycardia and orthostatic hypotension may be noted. Unlike heat cramp victims, these patients are typically unacclimatized. Hyponatremia, hemoconcentration and significantly diminished urine sodium are consistent findings. Children with cystic fibrosis, particularly those who are young and unable to meet increased salt requirements, are at risk for electrolyte depletion because salt losses in their sweat apparently do not respond to acclimatization and aldosterone stimulation of the sweat gland.

Heat stroke (Table 89.5) is a life-threatening emergency. Classic signs are hyperpyrexia (41°C [105.8°F] or higher); hot, dry skin that is pink or ashen, depending on the circulatory state; and severe CNS dysfunction. Often, but not invariably, sweating ceases before the onset of heat stroke.

The onset of the CNS disturbance may be abrupt, with sudden loss of consciousness. Often, however, premonitory signs and symptoms exist. These include a sense of impending doom, headache, dizziness, weakness, confusion, euphoria, gait disturbance, and combativeness. Posturing, incontinence, seizures, hemiparesis, and pupillary changes may occur. Any level of coma may be noted. Cerebrospinal fluid findings are usually normal. The extent of damage to the CNS is related to the time and extent of hyperpyrexia and to the adequacy of circulation. Once the body temperature is lowered, consciousness usually is restored quickly, but coma may persist for 24 hours or more.

Patients able to maintain cardiac output adequate to meet the enormously elevated circulatory demand are most likely to survive. Initially, the pulse is rapid and full, with an increased pulse pressure. Total peripheral vascular resistance falls as a result of vasodilation in the skin and muscle beds, and splanchnic flow diminishes. If hyperpyrexia is not corrected, ashen cyanosis and a thin, rapid pulse herald a falling cardiac output. The cause may be either direct thermal damage to the myocardium or significant pulmonary hypertension with secondary right ventricular failure. Even after body temperature is returned to normal, cardiac output remains elevated and peripheral vascular resistance remains low for several hours, resembling the compensatory hyperemia after ischemia noted in posttrauma, postshock, and postseptic states. Persistently circulating vasoactive substances probably account for this phenomenon.

Severe dehydration is not a necessary component of heat stroke but may play a role if prolonged sweating has occurred. Electrolyte abnormalities may occur, especially in the unacclimatized victim, if NaCl has not been replaced. In acclimatized persons, NaCl is conserved but often at the expense of a severe potassium deficit. Polyuria is sometimes noted, often vasopressin resistant and possibly related to hypokalemia. Acute tubular necrosis may be seen in as many as 35% of cases and probably reflects combined thermal, ischemic, and circulating pigment damage. Hypoglycemia may also be noted.

Nontraumatic rhabdomyolysis and acute renal failure have been described as consequences of various insults, including hyperthermia and strenuous exercise in unconditioned persons. Clinically, there may or may not be musculoskeletal pain, tenderness, swelling, or weakness. Laboratory evidence includes elevated serum creatinine phosphokinase (300 to 120,000) and urinalysis that is orthotoludin (Hematest)-positive without red blood cells (RBCs) and shows red–gold granular casts. Typically, serum potassium and creatinine levels rise rapidly relative to BUN. An initial hypocalcemia, possibly a consequence of deposition into damaged muscle, progresses to hypercalcemia during the diuretic phase a few days to two weeks later.

Management

Most cases of heat cramps are mild and do not require specific therapy. Rest and increased salt intake from liberally salted foods are sufficient. In severe cases with prolonged or frequent cramps, IV infusion of normal saline is effective. Approximately 5 to 10 mL/kg over 15 to 20 minutes should be adequate to relieve cramping. Oral intake of fluids and salted foods can then complete restoration of salt and water balance.

Heat exhaustion as a result of predominant water depletion is treated with rehydration and rest in a cooled or well-ventilated place. If the child is able to eat, he or she should be encouraged to drink cool liquids and be allowed unrestricted dietary sodium. If weakness or impaired consciousness preclude oral correction, IV fluids are given as in any hypernatremic dehydration.

Exhaustion caused by predominant salt depletion also requires rest in a cool environment. Alert, reasonably strong children can be given relatively salty drinks such as consommé or tomato juice and should be encouraged to salt solid foods. Hypotonic fluids (e.g., water, Kool-Aid) should be avoided until salt repletion has begun. Patients with CNS symptoms or gastrointestinal dysfunction may be rehydrated with IV isotonic saline or Ringer's lactate. Initial rapid administration of 20 mL/kg over 20 minutes should improve intravascular volume with return of blood pressure and pulse toward normal. Further correction of salt and water stores should be achieved over 12 to 24 hours. In especially severe cases with intractable seizures or muscle cramps, hypertonic saline solutions may be used. The initial dose of 3% saline solution is 5 mL/kg by IV. An additional 5 mL/kg should be infused over the next 4 to 6 hours.

Treatment of heat stroke centers on two priorities: 1) immediate elimination of hyperpyrexia and 2) support of the cardiovascular system (Table 89.6). Clothing should be removed and the patient should be cooled actively. Patients should be transported to an emergency facility in open or air-conditioned vehicles. Ice packs may be placed at the neck, groin, and axilla of the patient. Although immersion in ice water may be a more efficient means of lowering body temperature, it may complicate other support and monitoring. Among the most efficient but invasive methods is iced peritoneal lavage. However, a canine model of heat stroke suggested that an evaporative technique in which fans blew room air over subjects sprayed with 15°C (59°F) tap water was equally efficient. Temperature should be monitored continuously with a rectal probe, and active cooling should be discontinued when rectal temperature falls to approximately 38.5°C (101.3°F).

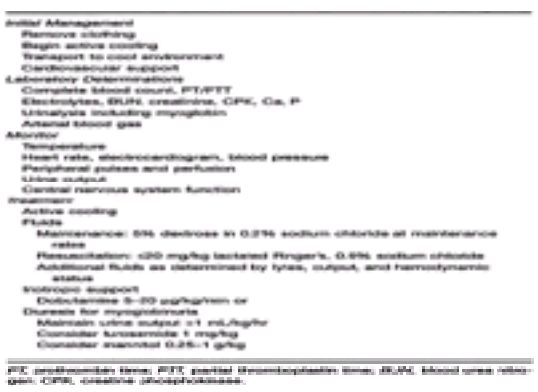


Table 89.6. Management of Heat Stroke

The severity of the patient's presentation determines the degree of cardiovascular support. If the skin is flushed and blood pressure adequate, lowering body temperature with close attention to heart rate and blood pressure may be sufficient. Although severe dehydration and electrolyte disturbances are uncommon, these should be assessed and corrected if necessary. Fluids cooled to 4°C (39.2°F) hasten temperature correction but may precipitate arrhythmias on contact with an already stressed myocardium. Adult patients rarely have required more than 20 mL/kg over the first 4 hours, but determinations of electrolytes, hematocrit, and urine output, and clinical assessment of central vascular volume should guide precise titration of fluids and electrolytes.

Patients with ashen skin, tachycardia, and hypotension demonstrate cardiac output insufficient to meet circulatory demand and are in imminent danger of death. Monitoring of the electrocardiogram (ECG) and arterial blood pressure (with an indwelling arterial line) should determine support.

Inotropic support may be required after a fluid challenge (see Chapter 3). Dobutamine is probably most appropriate: its b-agonist properties increase myocardial contractility and maintain peripheral vasodilation. Isoproterenol has been used successfully in the past but may cause myocardial oxygen consumption to exceed oxygen delivery. Additional fluid resuscitation may be necessary with the initiation of either dobutamine or isoproterenol to fill the effectively increased vascular space. Normal saline or albumin should be given to maintain the arterial blood pressure in the normal range. Dopamine may also be effective, infused at rates compatible with inotropic support without vasoconstriction. In cases of extreme hemodynamic instability, extracorporeal circulation may provide both circulatory support and a means of rapid temperature correction.

Agents with a-agonist characteristics (epinephrine and norepinephrine) are not recommended for initial management; they cause peripheral vasoconstriction, interfere with heat dissipation, and may compromise hepatic and renal flow further. Atropine and other anticholinergic drugs that inhibit sweating should be avoided.

Renal function should be monitored carefully, especially in patients who have been hypotensive or in whom vigorous exercise precipitated heat stroke. In general, BUN, creatinine, electrolytes, calcium, and urinalysis for protein and myoglobin should be obtained. Once the patient's vascular volume has been restored and arterial pressure normalized, hourly urine output should be monitored. If urine output is inadequate (less than 0.5 mL/kg per hour) in the face of normovolemia and adequate cardiac output, furosemide (1 mg/kg by IV) and/or mannitol (0.25 to 1 g/kg by IV) should be given. If the response is poor, acute renal failure should be suspected, and fluids should be restricted accordingly. Rapidly rising BUN or potassium (K^+) should prompt consideration of early dialysis.

ACCIDENTAL HYPOTHERMIA

Background

Elevated body temperature is a routine concern for most physicians, especially pediatricians. However, hypothermia, defined as core temperature at or below 35°C (95°F), is often overlooked. Reduced body temperature may be a consequence or cause of many disorders but is diagnosed only if health care providers maintain a high index of suspicion.

Populations at high risk for hypothermia are similar to those vulnerable to heat illness. Neonates and the elderly are most often affected. Physical disability, especially immobilizing conditions, and drug or alcohol ingestion increase risk at any age. Healthy young people who work or play to exhaustion in a cold environment are also at risk. The rising popularity of cold weather sports is producing more cases of accidental hypothermia. However, environmental conditions need not be

extreme, and the diagnosis should be considered even in temperate climates.

Primary CNS dysfunction, endocrinopathies, sepsis, protein caloric malnutrition, and various metabolic derangements may also depress core temperature.

Mortality rates, reported from 30 to 80%, depend more on the underlying disorder than on the degree of temperature depression.

Pathophysiology

Human core temperature is normally maintained within 0.6 °C (1°F). As described in the section on heat illness, this represents a fine balance between heat production and heat loss. When core temperature begins to fall below 37°C (98.6°F), physiologic mechanisms that produce and conserve heat are activated. Cooled blood stimulates the hypothalamus to increase muscle tone and metabolism (oxidative phosphorylation and high-energy phosphate production) and to augment heat production by 50% during nonshivering thermogenesis. When muscle tone reaches a critical level, shivering begins, and heat production increases two to four times basal levels.

Although surface temperature of the body, especially of the extremities, may drop nearly to environmental temperature, several mechanisms work to conserve heat and to protect blood and core structures from ambient air temperature, humidity, and wind. Sweating is abolished, decreasing heat loss by evaporation (unless there is external moisture), while vasoconstriction of cutaneous and subcutaneous vessels reduce losses further. Piloerection, which in many animal species traps an insulating layer of air next to the skin, occurs but is ineffective in humans.

When any component of the balance between heat production and loss is altered, the risk of hypothermia increases. Neonates, with large surface:volume ratios and small amounts of subcutaneous fat, conserve heat poorly and are unable to produce heat by shivering. The capacity for nonshivering thermogenesis—primarily metabolism of brown fat—is intact, but oxygen consumption is significantly increased. Hypoxemia may result, as well as metabolic acidosis, hypoglycemia, and hypocalcemia. Therefore, minor deviations in the thermal environment may produce hypothermia in neonates. More pronounced environmental stresses are required to overcome the greater compensatory capacity of older children.

Immersion in cold water causes the most rapid fall in body temperature. Struggling or swimming movements increase blood flow to the extremities and hasten hypothermia. Death occurs in 15 minutes in water at 0°C (32°F), but significant hypothermia occurs even in water at 21°C (69.8°F). Exposure to extreme cold is an obvious risk, taxing the body's ability to conserve and produce heat maximally. Voluntary motor activity produces heat, and physically fit, acclimatized persons may be able to increase activity to balance heat loss even in exceptionally cold environments. However, the metabolic cost of physical activity increases in the cold, and less fit persons quickly exhaust muscle glycogen supplies, are unable to maintain adequate heat production, and are likely to become hypothermic quickly. Wet, windy conditions hasten loss of body heat and may precipitate hypothermia even in temperate environments. Adolescents are psychologically less likely to conserve energy and to take preventive or corrective measures, thus increasing their risk of hypothermia.

Once homeostatic mechanisms fail and core temperature falls, predictable physiologic changes take place. If shivering does not occur, basal metabolic rate decreases steadily, reaching 50% of normal at 28°C (82.4°F). As a result, oxygen consumption and carbon dioxide production decline. The oxygen–hemoglobin dissociation curve shifts to the left.

Although respiratory depression occurs late, impaired mental status and cold-induced bronchorrhea predispose the patient to airway obstruction and aspiration. Acid-base balance follows no predictable pattern. Although respiratory acidosis occurs, tissue hypoxia, increased lactic acid production, and decreased lactate clearance by the liver produce metabolic acidosis.

Decreased heart rate contributes primarily to a fall in cardiac output. Peripheral vasoconstriction and an early increase in central vascular volume cause a transient rise in blood pressure, which later falls to become clinically significant below 25°C (77°F). A variety of cardiac conduction abnormalities arise, including decreased sinus rate, T-wave inversion, prolongation of ECG intervals, and the appearance of pathognomonic J waves (Fig. 89.4), which may provide the first clue to the diagnosis. Atrial fibrillation may occur at temperatures below 33°C (91.4°F) but is usually not hemodynamically significant. Below 28°C (82.4°F), myocardial irritability increases dramatically, and ventricular fibrillation becomes likely.

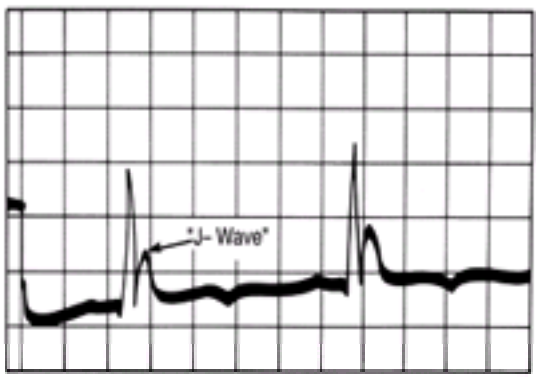


FIGURE 89.4. J wave, pathognomonic of hypothermia. Rounded contour distinguishes it from an RSR' pattern. It may also be confused with a T wave with a short Q-T interval. (Reprinted with permission from Welton D, Mattox K, Miller R, et al. Treatment of profound hypothermia. JAMA 1978;240:2291.)

Cold-induced vasoconstriction and elevated central blood volume and pressure contribute to a diuresis, which subsequently diminishes intravascular volume. At lower temperatures, tubular dysfunction allows salt and water loss. Acidosis causes potassium to shift from cells to the urine, where it is lost. Increased capillary permeability results in loss of fluid into the extracellular space.

Hematologic abnormalities may also occur. Plasma loss causes an increased hematocrit, whereas splenic sequestration may be responsible for a fall in white blood cell and platelet counts. Disseminated intravascular coagulation is sometimes seen.

CNS abnormalities are progressive. Each fall of 1°C produces a 6 to 7% decline in cerebral blood flow. Plasma loss increases blood viscosity, which further contributes to impaired cerebral microcirculation and mentation. Peripheral nerve conduction slows, and deep tendon reflexes decrease. Pupils dilate and react sluggishly, if at all, below 30°C (86°F). The electroencephalogram deteriorates progressively with falling temperature from high-voltage slow waves, to burst suppression patterns, to electrical silence at 20°C (68°F).

Gastrointestinal motility decreases below 34°C (93.2°F). The liver's capacity for detoxification or conjugation of drugs and products of metabolism is poor. Insulin release abates and serum glucose rises. Frank pancreatic necrosis may also occur, producing clinical evidence of pancreatitis.

Clinical Manifestations

The astute clinician must consider the possibility of hypothermia if the diagnosis is to be made in a timely manner. A history of sudden immersion in icy water or prolonged exposure to low environmental temperatures provides the obvious clue, but significantly low core temperatures may occur under much less suggestive circumstances. Examples include trauma victims found unconscious or immobile on a wet, windy, summer day; infants from inadequately heated homes or left exposed during prolonged medical evaluation; adolescents with anorexia nervosa; and patients with sepsis or burns. Severe hypothermia, coma, and cardiac arrest may present as the sudden infant death syndrome. Hypothermia may go undetected if the patient's temperature falls below the lower limit of the thermometer in use or if the thermometer is not shaken down adequately. Low-recording thermometers should be available in EDs and intensive care units. The diagnosis should be borne in mind for any patient with a suggestive history or coma of uncertain cause.

Physical examination reveals a pale or cyanotic patient. At mild levels of hypothermia, mental status may be normal, but CNS function is progressively impaired with falling temperature until frank coma occurs at approximately 27°C (80.6°F). Blood pressure also falls steadily below 33°C (91.4°F) and may be undetectable. Heart rate slows gradually unless atrial or ventricular fibrillation occurs. Intense peripheral vasoconstriction and bradycardia may render the pulse inapparent or absent. Below 32°C (89.6°F), shivering ceases, but muscle rigidity may mimic rigor mortis. Pupils may be dilated and may not react. Deep tendon reflexes are depressed or absent. Evidence of head trauma or other injury, drug ingestion, and frostbite should be sought (Fig. 89.5 and Fig. 89.6).

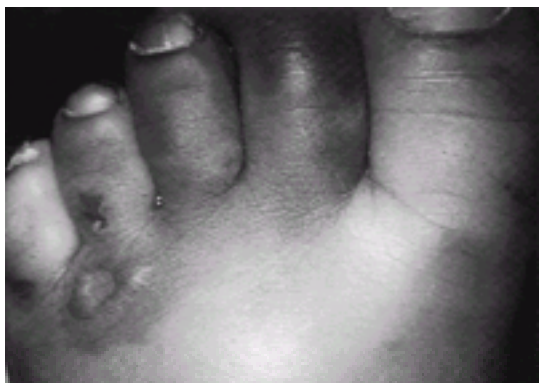


FIGURE 89.5. Frostbite of toes. Note the line of demarcation and ulcerative lesion.

FIGURE 89.6. Swollen fingers of a child with cold exposure.

Severe hypothermia mimics death. However, the significant decrease in oxygen consumption may allow life to be sustained for long periods, even after cessation of cardiac function. Signs usually associated with certain death (i.e.,

dilated pupils or rigor mortis) have little prognostic value. If the patient's history suggests that hypothermia is the primary event and not a consequence of death, resuscitation should be attempted and death redefined as failure to revive with rewarming.

Initial laboratory tests should include CBC, platelet count, clotting studies, electrolytes, BUN and creatinine, glucose, serum amylase, and ABGs corrected for temperature (Table 89.7). Urine should be sent for drug screening.

	For Each Elevation of 1°C	For Each Depression of 1°C
pH	-0.015	+0.015
$Paco_2$ (mm Hg)	+4.4%	-4.4%
Pao_2 (mm Hg)	+7.2%	-7.2%

Table 89.7. Effect of Body Temperature on Arterial Blood Gases Measured at 37° C (98.6° F)

Management

Therapy for hypothermia can be divided into two parts: general supportive measures and specific rewarming techniques (Table 89.8). Once hypothermia is diagnosed, temperature must be monitored continuously as treatment progresses.

Initial Management
Provide supplemental oxygen
Cardiopulmonary resuscitation for asystole, ventricular fibrillation
Laboratory Determinations
Arterial blood gas analysis corrected for temperature
Complete blood count, platelet count
Prothrombin time, partial thromboplastin time
Electrolytes, blood urea nitrogen, creatinine
Glucose, amylase
Urine drug screen
Monitor
Heart rate, electrocardiogram, respiratory rate, blood pressure
Temperature
Consider central venous pressure
Treatment
Correct hypoxemia, hyperkarbia
Correct hypokalemia
Correct hypoglycemia, 25% dextrose 1 g/kg IV
Tolerate hyperglycemia
Temperature: ≥32°C (89.6°F): passive rewarming or simple external rewarming
<32°C (89.6°F) (acute): external or core rewarming
<32°C (89.6°F) (chronic): core rewarming
Fluid replacement:
(acute) 5% dextrose in 0.2% saline at maintenance rates
(chronic) Normal saline, 5% albumin, fresh-frozen plasma to maintain blood pressure

Table 89.8. Management of Hypothermia

All patients should be given supplemental oxygen. Patients with profuse secretions, respiratory depression, or impaired mental status should be intubated and mechanically ventilated. Intubation should be performed as gently as possible to minimize the risk of arrhythmias.

A decreased metabolic rate produces less carbon dioxide, and usual minute ventilation would produce respiratory alkalosis, increasing the risk of dangerous arrhythmias. Therefore, ventilation should begin at approximately one-half the normal minute ventilation.

Assessment of acid-base status and ventilation in the hypothermic patient is the subject of considerable confusion. Blood gas machines heat the patient's blood sample to 37°C (98.6°F) before measuring pH and gas partial pressures (thus providing theoretical values if the patient were 37°C [98.6°F]). If the patient's actual temperature is provided with the sample, the machine can correct the values according to the nomogram of Kelman and Nunn. (Table 89.7 shows one set of guidelines for appropriate correction.) However, it is most important to understand two concepts. The first is the ectothermic principle, which relies on the following aspect of physiology: dissociation of ions and partial pressures of gases are decreased in cooled blood. In hypothermia, therefore, neutral pH is higher, whereas “normal” P CO_2 is lower than is encountered at 37°C (98.6°F). For example, hypoventilation of the hypothermic patient with a pH of 7.5 would actually induce an undesirable respiratory acidosis. A second, more practical concept is that if the patient's blood volume is restored and oxygenation maintained, acidosis will be corrected spontaneously as the patient is warmed.

Heart rate and rhythm should be monitored continuously and the patient handled gently to avoid precipitation of life-threatening arrhythmias in an exquisitely irritable myocardium. Sinus bradycardia, atrial flutter, and atrial fibrillation are common but rarely of hemodynamic significance. Spontaneous reversion to sinus rhythm is the rule when temperature is corrected. Ventricular fibrillation may occur spontaneously or with trivial stimulation, especially at temperatures below 28° to 29°C (82.4° to 84.2°F). Electrical defibrillation is warranted but frequently often is ineffective until core temperature rises. Closed chest massage should be initiated and maintained until the temperature is above 30°C (86°F), when defibrillation is more likely to be effective. Drug therapy is rarely effective and fraught with hazards associated with decreased hepatic and renal metabolism.

Fluid replacement is essential. Relatively little plasma loss occurs in acute hypothermia (as it does after cold-water immersion), but losses may be great in hypothermia of longer duration. Normal saline or lactated Ringer's solution, warmed to about 43°C (109.4°F) in a blood warming coil, is appropriate initially. Electrolyte determinations should guide further replacement. If clotting abnormalities occur, fresh-frozen plasma (10 mL/kg) is a useful choice for volume expansion (see Chapter 87). As temperature rises and peripheral vasoconstriction diminishes, hypovolemia is expected. Fluid volume should be sufficient to provide an adequate arterial blood pressure.

Hypoglycemia, if present, is treated with glucose (1 g/kg by IV). Hyperglycemia, which may result from impaired insulin release in the hypothermic pancreas, should be tolerated to avoid severe hypoglycemia with rewarming.

A number of rewarming strategies exist (Fig. 89.7). Passive rewarming implies removal of the patient from a cold environment and use of blankets to maximize the effect of basal heat production. For patients with mild hypothermia (temperature over 32°C [89.6°F]), this may be adequate.

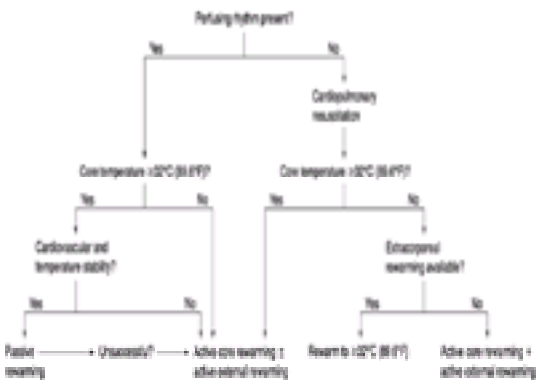


FIGURE 89.7. Algorithm for rewarming. (Adapted from data published in Danzl DF, Pozos RS. Accidental hypothermia. N Engl J Med 1994;331(26):1756–1760.)

Active rewarming is divided into external and core rewarming techniques. Electric blankets, hot water bottles, overhead warmers, and thermal mattresses are simple, easily available sources of external heat. Immersion in warm-water baths is also possible but complicates monitoring or response to arrhythmias. All of these methods, however, cause early warming of the skin and extremities with peripheral vasodilation and shunting of cold, acidemic blood to the core. The well-known “after-drop” of core temperature results. Severe hypotension may also occur in chronic cases as vasodilation increases the effective vascular space. External rewarming techniques limited to the head and trunk may minimize vasodilation and afterdrop. In acute hypothermia, active external rewarming is appropriate, but there is some evidence that in chronic cases (more than 24 hours), mortality is higher if active external rewarming is used instead of simple passive techniques.

Core rewarming techniques are almost certainly more rapid and less likely to be associated with after-drop, dangerous arrhythmias, or significant hypotension. These methods are especially valuable in the setting of severe chronic hypothermia (temperature below 32°C [89.6°F]), where fluid shifts are most likely to occur. A nonshivering human model of severe hypothermia indicated that inhalation rewarming offered no rewarming advantage, whereas forced air warming (approximately 200 W) allowed a sixfold to tenfold increase in rewarming rate over controls. A canine study of experimental hypothermia found that heated aerosol inhalation alone contributed less heat than endogenous metabolism, but that peritoneal lavage and pleural lavage had similar effect on rewarming (6°C/hour per meter squared). In humans, peritoneal dialysis, with dialysate warmed to 43°C (109.4°F), is effective and requires only equipment routinely available in most hospitals. Limited clinical experience suggests that pleural lavage is a relatively simple and useful measure. Hemodialysis, extracorporeal blood rewarming, and mediastinal irrigation are effective but require mobilization of sophisticated equipment and personnel. Gastric or colonic irrigation has also been advocated, but placement of the intragastric balloon may precipitate dysrhythmias.

Each increment in core temperature produces a “new” patient who requires reassessment and appropriate management, but most children with hypothermia have a good prognosis. In patients with mild temperature depression (above 32°C [89.6°F]), external rewarming techniques, and supportive care based on vital signs, ABGs, and metabolic parameters such as glucose and calcium levels, should result in prompt recovery. Patients with temperatures below 32°C (89.6°F), and especially those in whom hypothermia developed over 24 hours or more, require meticulous attention to continuously changing vital signs and metabolic needs. More elaborate core rewarming techniques are appropriate.

ELECTRICAL INJURIES

Background

Since the beginning of time, people have viewed lightning with fear and fascination. This natural phenomenon arises when warm, moist atmospheric air encounters cold air and is forced aloft. As warm air rises, it cools, condenses into a cloud, and loses electrons. A charge differential develops within the cloud, and lower aspects become relatively negatively charged, inducing a positive charge on the ground. Once a critical charge difference develops, electrons are discharged toward the ground, where positive charges meet the current from above. Current then flows to normalize the charge differential and superheats the air, causing the familiar flash of lightning and audible shock waves of thunder.

The last two centuries have witnessed the incorporation of controlled electricity into daily life and a better understanding

of its properties and physiologic effects. Availability of electricity has also meant increased exposure to electrical hazards and accompanying injuries. Electrical injury is responsible for approximately 700 deaths per year, of which 10% are children. No federal safety standards exist for household electrical cords, the major cause of electrocution in children 12 years of age and younger. High-tension electrical injuries dominate in older children who climb on trees, buildings, or utility structures.

Pathophysiology

The spectrum of electrical injury is enormous, ranging from low-voltage household accidents to million-volt lightning strikes (Table 89.9). Appropriate management requires an understanding of the basic physical aspects of electricity, the physiologic responses to injury, and the potential for immediate and delayed damage.

Factor	Lightning	High-Voltage
Duration	Brief	Prolonged
Energy level	100,000,000 V 200,000 amps	Much lower
Type of current	Direct	Usually alternating
Shock wave	Present	Absent
Cardiac	Asystole	Ventricular fibrillation
Burns	Superficial, minor	Deep, frequently obscured
Renal failures	Rare	Common secondary to myoglobinuria
Fasciotomy and amputation	Rare	Common, early, extensive

Table 89.9. Lightning versus High-Voltage Electrical Injury

The severity of electrical injury depends on six factors: 1) the resistance of skin, mucosa, and internal structures; 2) the type of current (alternating or direct); 3) the frequency of the current; 4) the intensity; 5) the duration of contact; and 6) the pathway taken by the current. Precise separation of the effect of these factors, which are interrelated, is impossible. Together, they produce either heat or current, and a variety of injuries result.

Resistance is a major factor determining the amount of current flow through tissue. Tissue injury is inversely related to resistance. Dry skin provides resistance of approximately 40,000 ohms, whereas thick, callused palms may provide up to 1×10^6 ohms. Thin, moist, or soiled skin lowers resistance to the 300- to 1000-ohm range. The highly vascular, moist oral mucosa has even lower resistance.

Once surface resistance is overcome, current flows between points of contact, not necessarily along anatomic structures such as nerves or blood vessels. Although the resistance of various tissues is known, the voltage difference may determine the actual path taken. Low-voltage current usually follows the path of least resistance, whereas high-voltage current follows a more direct course to ground with less regard for tissue type. Assessment of the most likely current pathway does not reliably predict injury. However, current that passes through the head or thorax may cause respiratory center or cardiac injury and increases the risk of cardiopulmonary arrest. Hand-to-hand flow carries risks of 60% mortality related to myocardial injury, spinal cord transection at C4–C8, and tetanic contraction of thoracic muscles with suffocation. Hand-to-foot current passage is associated with cardiac arrhythmias but lower mortality (20%). Foot-to-foot injuries are rarely fatal.

The type of current is another important determinant of injury. Alternating current (AC) at low voltage is able to induce tetanic muscle contraction and is therefore more dangerous than direct current (DC). These contractions prevent the victim from releasing his grip (“ locking-on”), thus extending the duration of contact. Normal household 60-Hz current changes direction 120 times per second, a frequency that induces an indefinite refractory state at neuromuscular junctions. Higher-frequency commercial currents are less likely to induce such a state and may be less harmful.

DC is used in medical settings for cardiac defibrillation, countershock, and pacing. Currents as low as 1 mA may trigger ventricular fibrillation, and high currents may damage the heart and conducting tissues directly. Lightning is another example of DC, discharged in a single, massive bolt that lasts $^1/_{10,000}$ to $^1/_{1,000}$ second. The brevity of exposure makes deep thermal injury unlikely.

In general, high-voltage injury is more dangerous than low-voltage injury. A higher voltage is more likely to cause “locking-on” and associated deep-tissue injury, although its tendency to throw victims from the source of current may mitigate this effect. The possibility of head and cervical spine injuries must be considered in these cases. The value of the current, or amperage, is of even greater importance than the voltage. Flow as low as 1 to 10 mA may be perceived as a tingling sensation. Progressively higher flows may paralyze muscles and ventilation, precipitate ventricular fibrillation, and cause deep-tissue burns.

Clinical Manifestations

Electrical injury may produce a variety of clinical pictures, ranging from local damage to widespread multisystem disturbances. Typically, deceptively small entry and exit wounds mask extensive damage to subcutaneous tissue, muscle, nerves, and blood vessels. Direct effects on the heart and nervous system are particularly common, and injury to all other symptoms can occur. Much of the injury is revealed immediately, but late complications are often encountered.

Victims of the most severe accidents are commonly pulseless, apneic, and unresponsive. Current that passes directly through the heart may induce ventricular fibrillation. Brainstem (medullary) paralysis or tetanic contractions of thoracic muscles may result in cardiopulmonary collapse. Lightning injury is capable of inducing asystole, from which the heart may recover spontaneously, but the accompanying respiratory failure is commonly prolonged. Unless ventilation is initiated promptly, hypoxia leads to secondary ventricular fibrillation and death.

Other cardiac disorders, including arrhythmias and conduction defects, are common among survivors. Supraventricular tachycardia, atrial and ventricular extrasystoles, right bundle branch block, and complete heart block are most common. Complaints of crushing or stabbing precordial pain may accompany nonspecific ST-T wave changes. Some patients sustain myocardial damage or even ventricular wall perforation. Despite evidence of important cardiac injuries, patients without secondary hypoxic-ischemic injury usually regain good myocardial function.

Nervous system injury is also extremely common and may involve the brain, spinal cord, peripheral motor, and sensory nerves, as well as sympathetic fibers. Loss of consciousness, seizures, amnesia, disorientation, deafness, visual disturbances, sensory deficits, hemiplegia, and quadriparesis occur acutely but may be transient. Vascular damage may produce subdural, epidural, or intraventricular hemorrhage.

Additional problems develop within hours to days after injury. The syndrome of inappropriate antidiuretic hormone secretion may precipitate herniation in rare cases. Electroencephalograms reveal diffuse slowing, epileptiform discharges, or burst suppression patterns, but they may not have prognostic significance. Spinal cord dysfunction yields more motor than sensory deficit. Peripheral neuropathies with patchy distribution may reflect direct thermal injury, vascular compromise, or current flow itself. A variety of autonomic disturbances may resolve spontaneously or persist as reflex sympathetic dystrophy.

Ocular damage is common, particularly after lightning strikes. Direct thermal or electrical injury, intensive light, and confusion contribute to the presentation. Findings include corneal lesions, hyphema, uveitis, iridocyclitis, and vitreous hemorrhage. Choroidal rupture, retinal detachment, and chorioretinitis occur less often. Autonomic disturbances in a lightning victim may cause fixed dilated pupils, which should not serve as a criterion for brain death without extensive investigation of other neurologic and ocular functions. Cataracts and optic atrophy are possible late developments.

Electrical injury may induce direct or indirect complications in other organ systems. Tetanic contractions may cause joint dislocations and fractures, especially of the upper extremity long bones and vertebrae. Fractures of the skull and other long bones may occur when high-tension shock throws the victim from the site of contact. Early cardiopulmonary insufficiency, as well as direct renal effects, may cripple renal function. Damaged muscle releases myoglobin and creatinine phosphokinase (CPK). As in crush injuries, myoglobin may induce renal tubular damage and kidney failure. Pleural damage may cause large effusions, whereas primary lung injury or aspiration of gastric contents may lead to pneumonitis. Gastric dilation, ileus, diffuse gastrointestinal hemorrhage, and visceral perforation may occur immediately or later.

In addition to burns at the site of primary contact, burns are common where current has jumped across flexed joints. Such burns are most common on the volar surface of the forearm and across the elbow and axilla. Arcing current may also ignite clothing and produce typical thermal burns. Entry and exit wounds and arc burns are notoriously poor predictors of internal damage. Tissue that appears viable initially may become edematous and then ischemic or frankly gangrenous over several days. Diminished peripheral pulses may provide immediate evidence of vascular damage, but strong pulses do not guarantee vascular integrity. Blood flow falls to a minimum at about 36 hours, but current or thermal damage may lead to vasospasm, delayed thrombosis, ischemic necrosis, or aneurysm formation and hemorrhage weeks after the injury. Viable major arteries near occluded nutrient arteries may account for apparently adequate circulation and uneven destruction of surrounding tissues.

Young children are vulnerable to orofacial burns, especially of the lips (Fig. 89.8). These full-thickness burns of the upper and lower lips and oral commissure usually involve mucosa, submucosa, muscle, nerves, and blood vessels. The lesion usually has a pale, painless, well-demarcated, depressed center with surrounding pale gray tissue and erythematous border. After a few hours, the wound margin extends and marked edema occurs. Drooling is common. The eschar separates in 2 to 3 weeks and bleeding may occur at this time; granulation tissue gradually fills the wound. Scarring may produce lip eversion, microstomia, and loss of function. Damage to facial or even carotid arteries may result in delayed hemorrhage. Devitalization of deciduous and secondary teeth may occur.

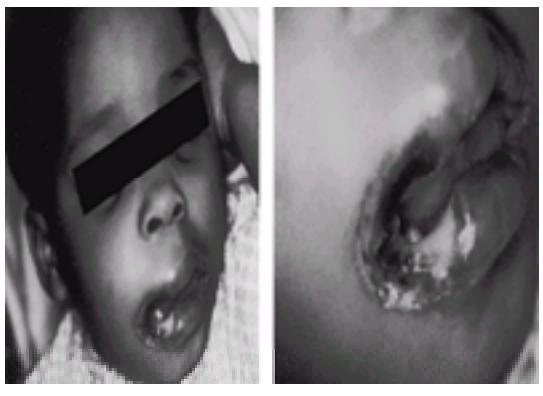


FIGURE 89.8. Patient with electrical burns to the corner of the mouth after biting on an electrical cord. (Courtesy of Evaline Alesandria, MD)

Inadequately debrided burned or gangrenous tissue provides a medium for serious infection. Staphylococcal, pseudomonal, and clostridrial species are common pathogens in the extremities. Streptococci and oral anaerobic organisms may infect mouth wounds.

Management

The first step in emergency management (Table 89.10) is to separate the victim from the current source. The rescuer must be well insulated to avoid becoming an additional casualty. If the current cannot be shut off, wires can be cut with a wood-handled ax or appropriately insulated wire cutters. Contrary to popular myth, a lightning stroke victim does not remain “electrified” and presents no risk to another person.

Table 89.10. Management of Electrical Injuries

Any victim in cardiopulmonary arrest should be resuscitated promptly following the guidelines discussed in Chapter 1. Prolonged efforts to restore adequate cardiopulmonary and cerebral function, especially in the lightning victim, may be appropriate in the context of bizarre neurologic phenomena that inhibit ventilatory efforts, consciousness, or pupillary function. The patient who fails to respond to resuscitative efforts over hours to days and meets standard brain death criteria can be pronounced dead with reasonable certainty.

Any patient who sustains electrical injury deserves a comprehensive physical examination. Bleeding or edema from orofacial burns may compromise the upper airway. The head, particularly eyes, and neck should be examined carefully for evidence of trauma. The skin should be examined carefully for burns and bruises. Limbs should be evaluated for pulses, perfusion, and motor and sensory function, as well as for soft-tissue swelling or evidence of fractures. Burns and deep-tissue injury may progress over hours to days, so repeated examination and monitoring are important.

Neurologic evaluation is especially important in all but the most minor, localized peripheral injuries. Level of consciousness and mental status should be assessed according to the child's developmental level. Cranial nerve, cerebellar, motor, and sensory evaluation are essential.

Children who have sustained minor household electrical injuries and are asymptomatic usually do not require laboratory evaluation, cardiac evaluation, or hospitalization. In one series, investigators were unable to assess the clinical significance of loss of consciousness, tetany, wet skin, or current flow across the heart, and recommended cardiac monitoring if any of these factors is present. If the history is one of a high-tension injury or lightning strike, laboratory evaluation should include ECG, CBC, CPK (with fractionation), BUN, creatinine, and urinalysis, including urine myoglobin. Physical examination that reveals evidence of bruises, bony tenderness, or distorted long bones should prompt appropriate radiographic studies.

Most children who sustain burns of the oral commissure (usually after biting an electrical cord) do not require extensive evaluation or admission. In cases of severe orofacial burns, use of an artificial airway should be considered before progressive edema leads to catastrophe. Mechanical ventilation may be necessary to overcome CNS depression or primary lung involvement.

Patients with persistent coma and loss of protective airway reflexes should be intubated to avoid aspiration. Good oxygenation and ventilation adequate to maintain a normal pH and Pa CO_2 of 35 to 40 mm Hg must be ensured. Seizure activity should be treated as indicated (see Chapter 70 and Chapter 83).

Care of the CNS is of utmost importance. The neck and back should be immobilized if the patient was thrown from the site of injury. If the mechanism of injury was severe, the cervical collar should be maintained in place despite normal cervical spine radiographs. If a child fails to regain consciousness within a short time or shows signs of neurologic deterioration, a computed tomography (CT) scan will help exclude intracranial hemorrhage.

Any patient who has sustained cardiopulmonary arrest, loss of consciousness, or deep-tissue injury should be admitted to hospital for evaluation and treatment. Heart rate, respiratory rate, and blood pressure should be monitored regularly. Doppler evaluation may be helpful in cases of vasospasm, which may complicate assessment of blood pressure and subsequent fluid management. True hypotension may require presser support.

Cardiopulmonary support is nonspecific. Most patients resume good circulatory stability unless severe hypoxia and ischemia have weakened the myocardium. Arrhythmias and acidosis should be treated along usual lines (see Chapter 82

and Chapter 86).

Patients struck by lightning require only maintenance fluids. Patients with ordinary thermal burns should be treated according to standard recommendations (see Chapter 114), although body surface area calculations may seriously underestimate fluid requirements. Extensive vascular and deep-tissue destruction may lead to extensive fluid sequestration. Isotonic fluid should be given in amounts to maintain normal pulse and blood pressure. In all cases, fluids should be given judiciously to avoid CNS complications.

Cerebral edema may develop over hours to days after injury, especially after a lightning strike. If the child's neurologic status fails to improve or deteriorates, intracranial pressure monitoring and treatment, including hyperventilation, osmotic or loop diuretics, and sedation and neuromuscular blockade, may be necessary. Serum and urine electrolytes and osmolality should be followed closely to recognize promptly the syndrome of inappropriate antidiuretic hormone secretion.

Myoglobin in the urine is consistent with muscle breakdown and sets the stage for renal failure. Hydration and brisk diuresis with furosemide and/or mannitol may prevent renal damage but must be undertaken with caution if there is coexistent CNS injury. Extensive muscle damage after lightning injury is uncommon, however, and major CNS injury is common. Treatment should proceed with these relative risks in mind until definitive information is available.

Most burns associated with lightning injury are superficial. Although they may become more apparent after several hours, most remain first- or second-degree burns. Minor burns on the extremities can be treated with antibiotic ointment and should be allowed to slough and heal. Oral and plastic surgeons should evaluate children who sustain oral burns. In most cases, similar conservative management is recommended, but a removable stent may be necessary to minimize scarring.

High-voltage injuries commonly require more aggressive treatment, and early surgical evaluation is essential. Fasciotomy may be necessary to restore adequate circulation to an injured extremity. The approach to debridement of wounds is controversial, but most surgeons rely on repeated examinations to detect nonviable tissue. Approximately 30% of survivors of high-tension injuries ultimately require amputation of some part of an extremity.

The risk of infection in patients with deep-tissue injury is high. Any patient not clearly immunized against tetanus should be given tetanus toxoid. Prophylactic antibiotics have been recommended for oral injuries, but in general, antimicrobial therapy should be reserved for proven or strongly suspected infection.

Suggested Readings

DROWNING AND NEAR-DROWNING

Conn AW, Miyasaka K, Katayama M, et al. A canine study of cold water drowning in fresh versus salt water. Crit Care Med 1995;23(12): 2029–2037.

Ender PT, Dolan MJ. Pneumonia associated with near-drowning. Clin Infect Dis 1997;25:896–907.

Graf WD, Cummings P, Quan L, et al. Predicting outcome in pediatric submersion victims. Ann Emerg Med 1995;26(3):312–319.

Habib DM, Tecklenburg FW, Webb SA, et al. Prediction of childhood drowning and near-drowning morbidity and mortality. Pediatr Emerg Care 1996;12(4):255–258.

Modell JH. Drowning. N Engl J Med 1993;328:253–256.

Spack L, Gedeit R, Splaingard M, et al. Failure of aggressive therapy to alter outcome in pediatric near-drowning. Pediatr Emerg Care 1997;13(2): 98–102.

SMOKE INHALATION

Lin YS, Kou YR. Reflex apneic response evoked by laryngeal exposure to wood smoke in rats: neural and chemical mechanisms. J Appl Physiol 1997;83(3):723–730.

Mlcak R, Cortiella J, Desai M, et al. Lung compliance, airway resistance, and work of breathing in children after inhalation injury. J Burn Care Rehabil 1997;18:531–534.

Thom SR. Smoke inhalation. Emerg Med Clin North Am 1989:7:371–387.

CARBON MONOXIDE POISONING

Hardy KR, Thom SR. Pathophysiology and treatment of carbon monoxide poisoning. Clin Toxicol 1994;32(6):613–629.

Thom SR, Taber RL, Mendiguren II, et al. Delayed neuropsychologic sequelae after carbon monoxide poisoning: prevention by treatment with hyperbaric oxygen. Ann Emerg Med 1995;25(4):474–480.

Tibbles PM, Perrotta PL. Treatment of carbon monoxide poisoning: a critical review of human outcome studies comparing normobaric oxygen with hyperbaric oxygen. Ann Emerg Med 1994;24(2):269–276.

Walker AR. Emergency department management of house fire burns and carbon monoxide poisoning in children. Curr Opin Pediatr 1996; 8:239–242.

ENVIRONMENTAL AND EXERTIONAL HEAT ILLNESS

Cheng TL, Partridge JC. Effect of bundling and high environmental temperature on neonatal body temperature. Pediatrics 1993;92(2):238–240.

Semenza JC, Rubin CH, Falter KH, et al. Heat-related deaths during the July 1995 heat wave in Chicago. N Engl J Med 1996;335:84–90.

Vicario SJ, Okabajue R, Haltom T. Rapid cooling in classic heatstroke: effect on mortality rates. Am J Emerg Med 1986;4:394–398.

White JD, Kamath R, Nucci R, et al. Evaporation versus iced peritoneal lavage treatment of heatstroke: comparative efficacy in a canine model. Am J Emerg Med 1993;11:1–3.

ACCIDENTAL HYPOTHERMIA

Danzl DF, Pozos RS. Accidental hypothermia. N Engl J Med 1994;331(26): 1756–1760.

Delaney KA, Howland MA, Vassallo S, et al. Assessment of acid-base disturbances in hypothermia and their physiologic consequences. Ann Emerg Med 1989;18:72–82.

Goheen MSL, Ducharme MB, Kenny GP. Efficacy of forced-air and inhalation rewarming by using a human model for severe hypothermia. J Appl Physiol 1997;83(5):1635–1640.

Kelman GR, Nunn JF. Nomograms for correction of blood pO2, pCO2, pH, and base excess for time and temperature. J Appl Physiol 1966;21(5):1484–1490.

Otto RJ, Metzler MH. Rewarming from experimental hypothermia: comparison of heated aerosol inhalation, peritoneal lavage, and pleural lavage. Crit Care Med 1988;16:869–875.

ELECTRICAL INJURIES

Bailey B, Gaudreault P, Thivierge RL, et al. Cardiac monitoring of children with household electrical injuries. Ann Emerg Med 1995;25(5):612–617.

Garcia CT, Smith GA, Cohen DM, et al. Electrical injuries in a pediatric emergency department. Ann Emerg Med 1995;26(5):604–608.

Jumbelic MI. Forensic perspectives of electrical and lightning injuries. Semin Neurol 1995;15(4):342–350.

Kleinschmidt-DeMasters BK. Neuropathology of lightning-strike injuries. Semin Neurol 1995;15(4):323–328.

Matthews MS, Fahey AL. Plastic surgical considerations in lightning injury. Am Plastic Surg 1997;39:561–565.

Rabban JT, Blair JA, Rosen CL, et al. Mechanisms of pediatric electrical injuries. Arch Pediatr Adolesc Med 1997;151:696–700.

CHAPTER 90

Radiation Accidents

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BACKGROUND

The international radiation accident registry maintained at Oak Ridge, Tennessee, lists 394 accidents that have occurred worldwide from 1944 until January 1998. Three thousand people were exposed to significant amounts of radiation, and 113 persons died as a direct result of these radiation accidents. Most of the survivors had no permanent injury as a result of the accident. The major concerns for the survivors are a small increase in the risk of cancer and the psychological stress caused by their concerns and those of their community about the long-term effects of radiation exposure. Of the 394 accidents, 239 occurred in the United States, resulting in 30 fatalities. Worldwide, fewer than 10 fatalities have been reported in children. Although the data collected by accident registries are incomplete, it is clear that radiation accidents that result in medically significant injuries are uncommon.

Despite the rarity of medically significant radiation accidents, the emergency physician needs to be aware of the basic principles and management of radiation accidents for four major reasons. First, the Joint Commission on Accreditation of Healthcare Organizations (JCAHO) requires plans for managing environmental accidents, including those involving hazardous materials. Second, although serious radiation accidents are rare, incidents in which radiation is perceived to have an important role are not uncommon. For instance, the discovery of a cardboard box with a radioactive label attached to it in a school play yard is likely to cause considerable concern. Generally, the public perceives radiation to be very hazardous and does not distinguish between amounts of radiation that we are exposed to every day from natural sources and amounts of radiation that have a measurable biologic effect. One definite long-term effect of radiation accidents is psychological stress caused largely by misconceptions and fear of the unknown. If the emergency physician is knowledgeable about the effects of radiation, he or she can correctly counsel the patient and immediately help decrease the psychological trauma. Third, fear of radiation and lack of knowledge about its effects have led to the medical mismanagement of several individuals who were thought to be involved in a radiation accident. Concern about the possible radiation aspects of the accident prevented the medical team from providing the appropriate medical care. Finally, the emergency physician needs to be knowledgeable about the effects of radiation because the rare radiation accident is often not initially recognized. A broad understanding of radiation accidents that have occurred in the past should help physicians recognize situations in which radiation might be considered as a cause. Frequent training and drills are essential to ensure that the emergency department (ED) staff has the procedural skills and supplies to deal with possible radiation accident victims.

PATHOPHYSIOLOGY

Types of Radiation

The source of some of the confusion about the medical consequences of an unintentional exposure to radiation originates in the general lack of understanding of the word *radiation*. *Radiation* is a general term used to describe energy that is emitted from a source (Fig. 90.1). Some forms of radiation are able to deposit a large amount of energy in a small volume of tissue. These energetic forms of radiation are called ionizing radiation because they deposit enough energy to strip electrons from atoms. Other types of radiation are less energetic and are called nonionizing radiation. The

distinction between ionizing and nonionizing radiation is important because their biologic effects are very different.

FIGURE 90.1. Types of radiation.

Ionizing radiation can be further subdivided into types of radiation that have no associated mass (nonparticulate) and those that have mass (particulate). X rays and gamma rays are nonparticulate types of radiation. This type of radiation can penetrate deeply into the body and affect radiation-sensitive tissues (e.g., bone marrow, the lining of the gastrointestinal tract). X rays are emitted by excited electrons, whereas gamma rays are emitted by excited or unstable nuclei. Once an x ray or gamma ray has been emitted, they are indistinguishable.

Particulate radiation can be further divided into particles that are charged and particles that are uncharged. Neutrons, a type of particulate radiation that has no electrical charge, can penetrate the body to depths similar to x rays and gamma rays. Because neutrons deposit their energy in a more concentrated area, neutrons cause more biologic damage than x rays or gamma rays.

Alpha rays and beta rays are charged particles. Alpha rays have a 2+ electrical charge and a large mass (two protons and two neutrons). Beta rays have a single negative charge and a small mass (one electron). Charged particles do not penetrate the body very well. Because of their larger mass and charge, alpha rays cannot penetrate even the dead layers of skin. For example, plutonium-239, an alpha emitter, is a biologic hazard only when it is inhaled, ingested, or otherwise absorbed into the body. Beta rays are more penetrating and can severely damage the skin. Beta rays cannot damage the deep radiation-sensitive organs in the body unless the radioactive source is incorporated into the body. At the Chernobyl nuclear plant accident in Russia, some of the firemen had severe skin damage as a result of intense beta ray exposure. This injury contributed to their deaths.

The words *radiation* and *radioactive* are often confused. An atom that is unstable spontaneously gives off radiation and is radioactive. In contrast, an x-ray machine cannot spontaneously give off radiation. An external power source is needed. Therefore, an x-ray machine is not radioactive. A patient who has been exposed to radiation does not become radioactive. Patients spontaneously give off radiation only if they have radioactive atoms on them (external contamination) or in them (internal contamination).

Amounts of Radiation

Although radiation cannot be perceived by the human senses, Geiger counters can easily measure amounts of radiation that are far below the levels that can be shown to have a measurable biologic effect. Geiger counters are inexpensive and are readily available in the nuclear medicine department at most hospitals. Because a Geiger counter can detect and quantify the radiation hazard, managing a radiation hazard is easier than managing biologic hazards such as human immunodeficiency virus (HIV), meningitis, or methicillin-resistant *Staphylococcus aureus.*

Radiation exposure is commonly measured in three different units: roentgen, rad, and rem. A *roentgen* is a measure of the number of ion pairs that are produced in a volume of air by the radiation from an x-ray machine or from radioactive atoms. The *rad* (roentgen absorbed dose) is a measure of how much energy is deposited per gram of tissue. The absorbed dose depends on the type of radiation and the size, shape, and composition of the object absorbing the radiation. Finally, a *rem* (roentgen equivalent in man) takes into account the biologic effects of various kinds of radiation. Some types of radiation (e.g., neutrons, alpha particles) cause greater biologic harm than x rays or gamma rays. For x rays and gamma rays, a rad and a rem are equivalent. The nomenclature for radiation dose is further complicated by the fact that the international community uses terms that have not yet gained widespread use in the United States. The Standard International Units for radiation dose are listed in Table 90.1.

	Common United States Units		Standard International Units
Exposure	Roentgen, R		None
Dose	Radiation absorbed dose, rad		gray, Gy
	100 rads	=	1 Gy
Dose	Roentgen equivalent in man, rem		sievert, Sv
	100 rems	=	1 Sv
Quantity	Curie, Ci		becquerel, Bq
	(3.7×10^{10} disintegrations/sec)		(1.0 disintegrations/sec)
	1 mCi	=	37 MBq

Table 90.1. Units for Radiation, with Abbreviations

To put these units into perspective, it is helpful to recall that we are exposed to about 300 mrems (1000 mrems = 1 rem) of radiation each year from natural sources. During a 70-year lifetime, the total radiation exposure from natural sources will be more than 20 rems. There is no measurable biologic effect of radiation with acute doses below 10 to 20 rems. Typical radiation exposures that we encounter as part of our daily lives as well as in medicine are listed in Table 90.2.

Sources	Dose
Round-trip intercontinental air flight	2-3 mrems
Chest radiograph	5-10 mrems
Living in brick house	20 mrems/yr
Natural radiation	300 mrems/yr
Angiography	1000 mrems

Table 90.2. Common Radiation Doses

An additional unit, the curie, is used to describe the quantity, or amount, of a radioactive material. The curie is the number of radioactive atoms that are decaying per unit time. One curie of radium equals approximately 1 g of radium. Diagnostic studies in nuclear medicine typically use millicurie amounts of radionuclides. The hazard posed by a radionuclide cannot be inferred easily from knowledge of the amount of the radionuclide. In addition to the amount of the radionuclide (in curies), the hazard posed by a radionuclide depends on its decay scheme, the energies of its emissions, its half-life, and on how long it stays in various organs in the body. If the radionuclide decays only by emitting alpha particles, it is not a hazard if it is kept outside the body. As mentioned previously, alpha rays cannot penetrate even the dead layers of the skin. On the other hand, radionuclides (e.g., iodine-131, hydrogen-3 as water) that are readily absorbed by the body and/or are concentrated by an organ can be a significant hazard even when present in small amounts.

Radiation Safety

The worst radiation accident involving a commercial nuclear power plant in the United States resulted in a radiation dose to off-site medical personnel of 14 mrems. Following the Chernobyl accident, the highest radiation dose to off-site medical personnel was a few rems. These doses should reassure ED personnel that it is extremely unlikely that a radiation accident would ever threaten their own safety.

Patients who have been exposed only to radiation, such as a patient who has just had a chest radiograph taken, are not radioactive and require no special radiation precautions.

Although the radiation doses to ED personnel involved in the care of an accident victim contaminated by radioactive material are likely to be very small, simple protective measures can be used to make the doses even smaller. There are three methods of protection from the radiation exposure: minimizing time of exposure, maximizing distance to the extent practical, and using shielding as appropriate (Fig. 90.2). The amount of exposure received is directly proportional to the time spent near the source of radiation. Distance is the most practical and effective method of reducing radiation exposure because the dose decreases by the square of the distance. Doubling the distance from a source of radiation will reduce the dose by a factor of 4, and tripling the distance will reduce the dose by a factor of 9 (Fig. 90.3). This is known as the inverse square law. Shielding, such as that in the familiar lead apron, although useful in radiology departments where radiation is a hazard because of low-energy scattered radiation, does not play a significant role in radiation accident management. The reason for this is that most sources of radiation that are likely to be encountered have higher energies, and the amount of apron shielding required for useful protection becomes impractical.

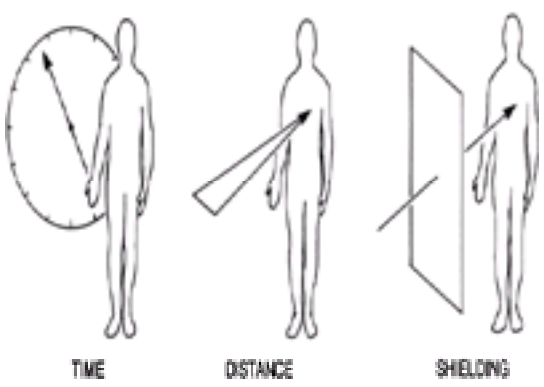

FIGURE 90.2. Three methods of reducing radiation exposure.

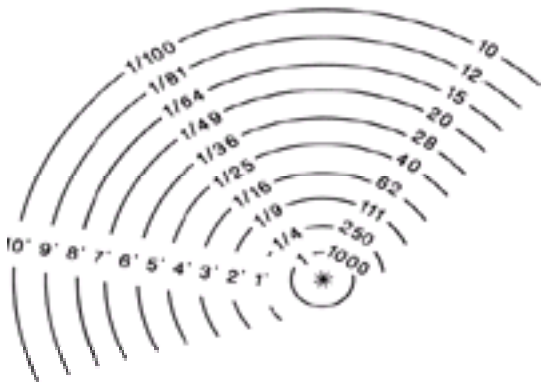

FIGURE 90.3. The effect of distance on radiation exposure from a point source of radiation.

CLINICAL MANIFESTATIONS AND EVALUATION

Recognizing an Accident

Radiation accidents can be recognized by understanding three questions: Who is likely to be affected by a radiation accident? What are the likely sources of radiation? and What are the likely injuries?

Types of Victims

The people who are most likely to be involved in a radiation accident are individuals whose work involves radiation. Although these accidents are usually recognized, several work-related accidents were not initially appreciated. One valuable way to determine whether a radiation accident has occurred is to contact the worker's supervisor and/or the radiation safety officer for the employer. In most instances, they will immediately recognize whether the possibility of a serious radiation exposure exists.

A second group of individuals who may be involved in a radiation accident are members of the general public who inadvertently come into contact with a radiation hazard. An example of this kind of accident occurred in Goiana, Brazil, in September 1987. In this accident, a cesium-137 radiation therapy source was stolen from an abandoned medical clinic. The poachers brought the source to their home, exposing their family and friends to large doses of radiation. Their exposure was worsened when the victims broke open the cesium-137 source, subsequently contaminating their food, their living quarters, and the area surrounding their homes. As a result of this accident, four people, including a 6-year-old girl, died. Hundreds of other people were exposed to nonlethal amounts of radiation. The accident was not recognized until 2 weeks after the source had been opened. When the individuals first became ill, their symptoms were diagnosed as gastroenteritis.

A third group of individuals who may be involved in a radiation accident are people who are unknowingly and intentionally exposed to radiation by another person. An infamous case that happened in the United States involved a 13-year-old boy who was intentionally exposed to radiation by his father. These exposures occurred on multiple occasions during weekend visitations over 6 months. Months later, the son recalled occasionally finding “shiny silver pellets” in the ear pieces of headphones he was told to wear, in a pillow he was told to use, and in a sock he found on his bed. Other injuries suggested that he was exposed at other times while under sedation.

Soon after his first exposure, the boy developed skin lesions, described as bruises, that gradually developed into reddish brown blisters. These lesions were attributed to an infection. Subsequently, he developed lesions on the medial aspects of both thighs, the right ankle, right hand, and left forehead. He also began losing hair from the left side of his head. The lesions became increasingly incapacitating and the boy was admitted to a hospital for 3 weeks. An infectious cause for the lesions could not be established. A psychiatrist suggested a neurodermatitis caused by the conflict between the boy and his father. During a 20-month period, the boy was seen by 16 physicians. Finally, a plastic surgeon recognized the lesions as radiation necrosis. The boy required multiple operations to repair his injuries.

The father, a petroleum engineer, had access to two curie cesium-137 sources. The dose rate at contact for such a source is approximately 500 rads/minute. However, at 1 cm, the dose rate drops to 30 rads/minute and is about 4 rads/minute at 3 cm from the surface. The father was found guilty of child abuse.

Types of Radiation Sources

To cause a significant radiation injury, an intense radiation source is needed. The four major types of possible intense radiation sources are listed in Table 90.3. Because of their different physical properties, different types of sources are likely to cause different types of radiation injuries.

Type of Source	Examples	Likely Injuries
Sealed	Industrial radiography	Contamination unlikely
	Brachytherapy	Local radiation injury with small source
	Some radiation therapy machines	Whole-body exposure with large source
	Industrial sterilizers	
Unsealed	Medical radionuclides (e.g., ^{131}I, ^{99m}Tc)	External and internal contamination likely
	Accidental release by a nuclear power plant	
	Radium dial painters	
Radiation devices	Cyclotron	Local radiation injury likely
	Linear accelerator	
Uncontrolled fission	Nuclear reactor	Large whole-body doses likely
	Uranium enrichment	
	Weapons production	On-site and off-site contamination possible for nuclear reactions

Table 90.3. Intense Radiation Sources

Sealed sources contain a radioactive source in a leak-proof container. Because the containers are leak-proof, these accidents usually cause radiation exposure only. Contamination occurs only if the container is broken open. Examples of sealed sources include industrial radiography sources, some radiation therapy machines, and industrial sterilizers.

By far, industrial radiography sources have caused the most radiation accidents. Thousands of these highly radioactive sources exist in the United States. They are used in industry to radiograph metal parts such as pipe welds. When not in use, the radioactive source is shielded in a thick lead container. To take a radiograph, one must crank the source out of the shield using a cabling system. Until recently, the source was attached to the cable by a simple mechanism that sometimes failed. If the source detached from the cable, it could be lost. The source is about the size of a pen (Fig. 90.4); it is metallic and innocuous appearing. In the past, the source was not labeled; a passerby could find the source and not recognize it to be dangerous. Because this type of source is small, it usually causes a local radiation injury (see the following) involving the hands (from picking up the source) or the buttocks (from putting the source in a pocket).

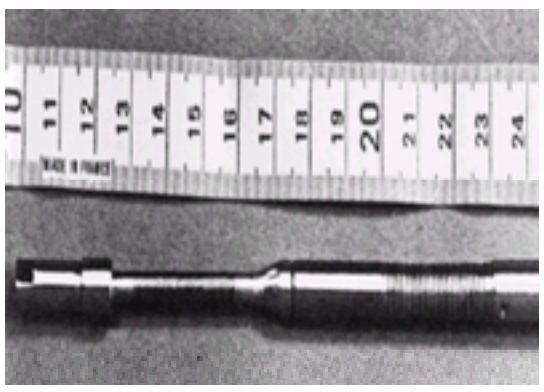

FIGURE 90.4. Source of radiation that affected two Algerian children in 1978.

Sealed sources used in radiation therapy can be small (brachytherapy) or large (cobalt therapy machine). Industrial sterilizers use very large, intense radiation sources to sterilize products (e.g., medical supplies) that would be damaged by other methods of sterilization. Large, sealed sources are more likely to result in whole-body radiation exposure.

Unsealed sources, a second type of radiation source, consist of radioactive material in a form that is dispersed easily (e.g., liquid, powder). The likely injury caused by unsealed sources is external and internal contamination. An unsealed source commonly found in hospitals is a solution of radioactive iodine (iodine-131) that is used to treat some thyroid diseases. If these radioactive atoms are kept on the outside of the body (external contamination), they usually cause only minor harm. The radioactive dirt can be washed from the skin much the way that other kinds of dirt are washed from the skin. If properly managed, external contamination is generally a nuisance rather than a serious health threat to the patient or medical staff. If the radioactive dirt gets into the patient's body (internal contamination) by ingestion, inhalation, or absorption through the skin or a wound, minimizing the radiation dose is more difficult. Other examples of unsealed sources include radium ingested by radium dial painters and radionuclides released during the accident at the Chernobyl nuclear power plant.

Radiation devices, the third potential radiation source listed in Table 90.3, are likely to cause a local radiation injury. Because these devices emit radiation when switched on but are not radioactive, it is unlikely that patients exposed to radiation from radiation devices will be radioactive when they present for treatment.

A fourth possible radiation source is an uncontrolled nuclear reaction (criticality accident). Usually, there is an intense radiation exposure for a very brief period. Criticality accidents have resulted in the largest whole-body radiation exposures that have occurred as the result of radiation accidents. Most of the radiation dose results from neutrons. These accidents can occur at only uranium enrichment facilities and nuclear reactors where there is a critical mass of nuclear material. Fortunately, criticality accidents are rare. Only two criticality accidents have occurred since the late 1960s; one of these was the 1986 accident at Chernobyl. The Chernobyl accident caused the deaths of 31 people who were at the nuclear power plant at the time of the accident. In addition, millions of people were exposed to low levels of radiation when hundreds of square miles of land were contaminated with radioactive debris from the Chernobyl accident.

Types of Radiation Injury

Perceived

There are three major types of radiation injury (Table 90.4). The first and by far the most common injury is the perceived radiation injury. Because of misconceptions about the possible health effects of radiation, members of the general public, fearful of having been exposed, may attribute almost any illness to radiation exposure. Unfortunately, these perceptions are often reinforced by physicians who are not knowledgeable about radiation biology. The psychological stress caused by misdiagnosis can be significant.

Perceived	Contamination
Exposure	External
Whole-body	Internal
Local	Metal fragment
	Hot particle

Table 90.4. Types of Radiation Injuries

Exposure

The second major type of radiation injury is exposure to radiation. Because these patients do not have radioactive dirt on them or in them, they are not radioactive and can be treated without any additional precautions. Two types of radiation exposure are possible. Large doses of penetrating radiation over a short period to a large portion of the body (i.e., whole-body radiation) cause the acute radiation syndrome. Exposure to alpha or beta particles of any source would never cause the syndrome because this type of radiation is nonpenetrating. Large doses of radiation over a short period to a small portion of the body cause a local radiation injury. When only a small portion of the body is exposed, much larger doses of radiation can be tolerated. Analogous medical situations would be whole-body radiation as conditioning for bone marrow transplantation and localized radiation therapy for breast cancer.

Whole-Body Exposure

The signs and symptoms of the acute radiation syndrome (Table 90.5) begin to appear with whole-body radiation doses of 100 rems. Organs with rapidly dividing cells (the bone marrow and the lining of the gastrointestinal tract) are the most susceptible to radiation damage. The amount of damage that occurs depends on the dose as well as on the dose rate. For example, a dose of 100 rads received in 1 minute would probably cause symptoms; however, a dose of 100 rads received at a dose rate of 1 rad/day for 100 days would not cause symptoms. Doses of about 400 rems are lethal in approximately 50% of untreated people. With maximum medical treatment, the dose of radiation that will kill 50% of people may be as high as 650 rems.

Whole-Body Absorbed Dose (rem)	Comments
5	Asymptomatic
10	Asymptomatic (minimal detectable dose using cytogenetics)
50	Asymptomatic (minor depression of white cells and platelets possible)
100	Nausea and vomiting in approximately 15% of patients within 2 days of exposure
200	Nausea and vomiting in most patients Moderate hemopoietic syndrome
400	Nausea, vomiting, and diarrhea within 48 hr; severe hematologic depression; 50% mortality without medical treatment
600	100% mortality within 30 days without medical treatment; 50% mortality with medical treatment
700	Gastrointestinal syndrome; survival unlikely; death within 2–3 wk
5000	Neurovascular syndrome; death in 24–72 hr

Table 90.5. Dose-Effect Relationship after Acute Whole-Body Radiation Exposure

The acute radiation syndrome consists of three distinct phases (Table 90.6). The prodromal phase begins minutes to hours after the radiation exposure and lasts for 2 to 3 days. During the prodromal phase, the patient may have nausea, vomiting, diarrhea, fatigue, and/or headache. The prodromal phase is followed by the latent phase, during which the patient is relatively asymptomatic. The latent phase generally lasts until 3 weeks after the exposure. The third and final phase is the manifest illness phase. During the manifest illness phase, the patient is at greatest risk for infection and bleeding as a result of the bone marrow suppression and gastrointestinal epithelial damage caused by the radiation exposure. As the radiation dose increases, the duration of the prodromal phase increases and the length of the latent

phase decreases.

Prodromal (0–2[a])	Latent (2–20[a])	Manifest Illness (21–60[a])
Fatigue	Asymptomatic	Bone marrow depression
Nausea and vomiting		Sepsis
Diarrhea		Bleeding
Headache		Diarrhea
Dizziness		
Decreased lymphocyte count		

[a]Days after exposure.

Table 90.6. Acute Radiation Syndrome—Signs and Symptoms

With doses of 200 to 400 rems, the primary effect of the whole-body radiation is to depress the bone marrow. Although the lymphocyte count (Fig. 90.5) decreases rapidly within the first 24 hours, there is no need for acute medical treatment. The patient will be at greatest risk 34 weeks after the radiation exposure, when the white cell and platelet counts reach a nadir (Fig. 90.6). At this time, the patient is vulnerable to death from infection and bleeding. If the patient can be supported during this period of vulnerability and if the bone marrow is not irreversibly damaged, a recovery phase ensues (Fig. 90.7).

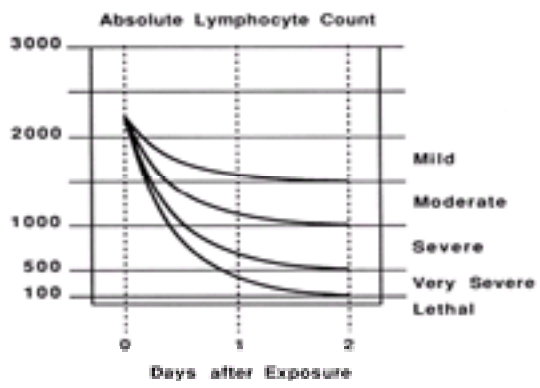

FIGURE 90.5. Effect of whole-body radiation on lymphocytes in the first 2 days after exposure.

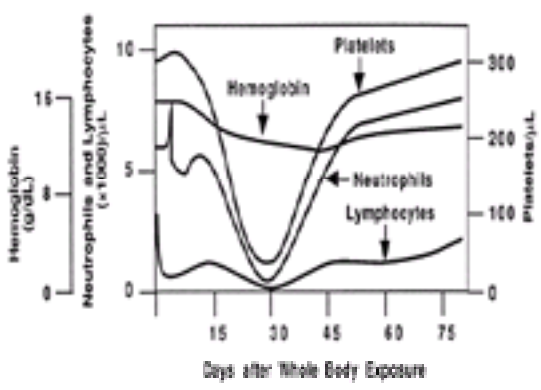

FIGURE 90.6. Effect of whole-body radiation on blood counts in the days after exposure.

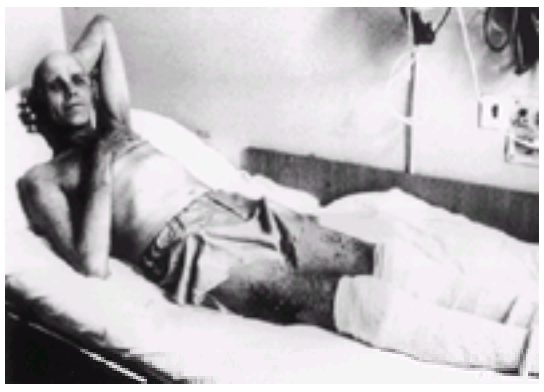

FIGURE 90.7. Chernobyl victim suffering from acute radiation syndrome a few weeks after the accident. Note the hair loss, indicating a radiation dose of several hundred rems. Also note injury to the skin of the lower extremities as a result of high (a few thousand rems) doses of beta (nonpenetrating) radiation. (Courtesy of A.M. Davis.)

The gastrointestinal syndrome occurs from absorbed doses of more than approximately 700 rads. During the prodromal phase of this syndrome, there is prompt onset of severe nausea, vomiting, and diarrhea. There is a latent period of approximately 1 week and then recurrence of gastrointestinal symptoms, sepsis, electrolyte imbalance, and likely death. The patient is very susceptible to infection because of the depressed bone marrow and because pathogens can readily enter the body across the damaged gastrointestinal lining.

At dose levels of more than 5000 rads, the cardiovascular/central nervous system (CNS) syndrome predominates. There is almost immediate nausea, vomiting, prostration, hypotension, ataxia, and convulsions. The permeability of blood vessels increases. The patient experiences CNS symptoms as a result of brain edema and also experiences hypotension caused by the difficulty of maintaining a normal intravascular space. Death usually occurs within 1 to 4 days.

Estimating the whole-body radiation dose may be difficult. The signs and symptoms during the prodromal period are nonspecific except for a rapidly decreasing lymphocyte count. Nausea and vomiting is a sensitive but nonspecific finding. Patients who do not have nausea and vomiting are unlikely to have been exposed to a radiation dose that is large enough to cause the acute radiation syndrome. On the other hand, individuals may have nausea and vomiting for reasons other than exposure to radiation. The whole-body radiation dose from radiation accidents is rarely uniform. The nonuniform nature of the radiation dose makes it more difficult to predict the biologic effects of the exposure. Chromosome analysis (cytogenetic dosimetry) may be helpful in estimating the radiation dose, but the results of this study take about 1 week.

A radiation accident involving whole-body exposure to four young (14 to 20 year old) women occurred in Algeria in 1978. In this accident, a 25 Ci iridium-192 industrial radiography source fell from a truck and was found by two young (3 and 7 year old) boys. They played with the source for several hours before taking it home (Fig. 90.8 and Fig. 90.9). The source was taken away by their grandmother who hid it in the kitchen. The source remained in the room for 5 to 6 weeks, irradiating several people, including the four young women. This accident was not discovered until the four women had severe bleeding from the mucous membranes in the mouth, anorexia, nausea, purpura, and bone marrow depression. The lymphocyte count in all four patients was less than 10% of the normal. The whole-body dose over the 5 to 6 weeks was estimated to have been between 600 and 1000 rads. All four patients injured by this accidental exposure survived.

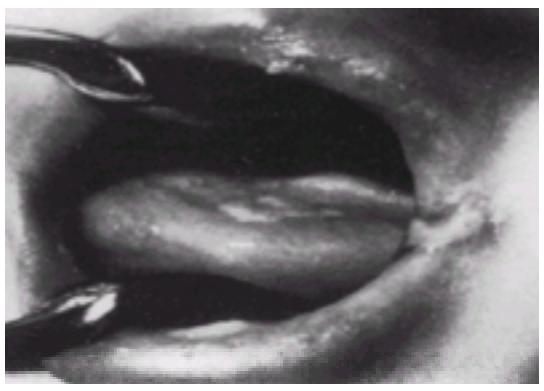

FIGURE 90.8. Mouth lesions caused by radiation of approximately 2500 rads. The lesions healed eventually.

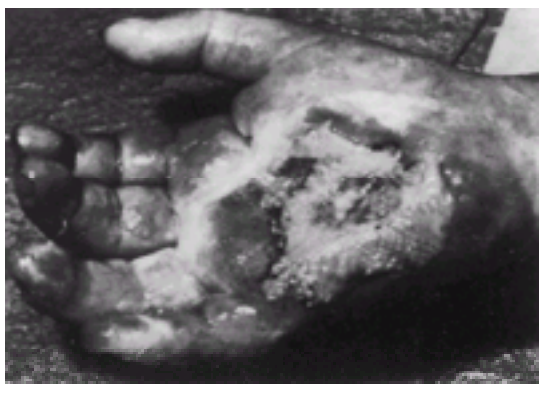

FIGURE 90.9. Radiation burns on the hand of the older Algerian child, from exposure to 1,500 to 10,000 rads. Reconstructive surgery was required.

Local Radiation Exposure

The second type of radiation exposure that can occur involves a large radiation dose to a small part of the body. If only a small part of the body is exposed, much larger doses can be tolerated. Local radiation injuries do not cause bone marrow depression unless they are accompanied by a significant whole-body radiation dose. These injuries are rarely life-threatening, but they are difficult to manage because they often cause a slowly progressive injury that takes months to years to fully evolve. The injury develops slowly because the radiation causes progressive fibrosis of the blood vessels, which, in turn, causes tissue necrosis. The ultimate extent of the injury may not be appreciated initially. Healing after amputation or reconstructive surgery is poor because of deficient blood supply.

The hand is the most common site for localized irradiation injuries because workers carelessly or unknowingly pick up an intense radioactive source or put their hands into an intense radiation field. The next most common sites are the thighs and buttocks because individuals are likely to put things that they find into their pockets. Most of the industrial radiography sources deliver an extremely high dose on direct contact with the skin. For example, the 25 Ci iridium-192 described earlier has a surface dose rate of about 20,000 rads/minute and will cause an absorbed dose of about 12,500 rads/minute at 1 cm depth in tissue. In contrast, analytical x-ray machines, which emit x rays of much lower energy than the photons of iridium-192, are not likely to cause deep blood vessel injury.

Local radiation injuries can be readily differentiated from thermal burns. The effects of a thermal burn are present immediately, and the patient invariably knows when the injury occurred. If a patient presenting with a burnlike injury does not know the cause of the injury, a local radiation injury should be suspected. Table 90.7 lists the dose-related findings expected after a local radiation exposure.

Absorbed Dose (rems)	Findings
300	Threshold for erythema (100 keV diagnostic x-ray)
600	Threshold for erythema (10 MeV therapeutic x-ray)
1500	Moist desquamation
2000	Skin ulceration with slow healing
>3000	Gangrenous changes

Table 90.7. Absorbed Dose to Produce Skin Changes

If erythema is seen within the first 48 hours, ulceration will probably occur. The erythema may come in waves, that is, be present, disappear, and return later. With transepidermal injury, blister formation may occur at 1 to 2 weeks with doses in the range of 10,000 rads and at 3 weeks at dose levels of 3,000 to 5,000 rads. Treatment is required to prevent infection and to relieve pain. Skin grafting, especially musculocutaneous flaps, may be appropriate if the radiation exposure was very localized and superficial. Progressive gangrene, caused by the obliterative changes in the small vessels, will occur if the radiation exposure is large and involves deep structures. Under these circumstances, amputation may be necessary.

The two Algerian boys who found the 25 Ci iridium-192 industrial radiography source described earlier had local radiation injuries. The younger boy was 3 years old and presented with lesions of the mouth and hands (Fig. 90.8). The boy apparently had sucked on the source, receiving an approximate dose of 2,500 rads to the lip surface. The older boy had a necrotic deep ulceration in the hypothenar region of the right hand, apparently from using the source as a drumstick (Fig. 90.9). The estimated dose to the center of the lesion was estimated at 10,000 rads, and at the periphery of the necrosis, 1,500 rads. Ultimately, the oral lesions in the young boy healed. The palmar lesion of the older boy required reconstructive surgery.

Contamination

Contamination represents the third major type of radiation injury. Contamination occurs when radioactive dirt or liquid remains on a patient (external contamination) or, when inhaled or ingested, in the patient (internal contamination). Contamination is the only type of radiation injury that requires the medical staff to take any radiation-related precautions. It should be reemphasized that there is little danger to the medical staff when caring for a contaminated person once he or she has been transported to the hospital. On the other hand, medical personnel who respond to the accident site may be exposed to large, potentially life-threatening doses of radiation. For these rescue workers, 75 to 100 rems is the voluntary limit for lifesaving activities.

External Contamination

External contamination rarely is a significant medical problem. To prevent additional radiation exposure to the patient and unnecessary radiation exposure to the medical staff and the public, external contamination should be removed and dispersal of radioactive materials prevented. Based on the assumption that all radiation exposure is potentially harmful, the goal of the treatment of any contaminated patient is to keep radiation exposures “as low as reasonably achievable.” This is called the ALARA principle and requires advance planning, specific supplies, and appropriate protective clothing. Preventing the dispersal of radioactive materials is accomplished by treating the patient in a single location and controlling access to that location.

Internal Contamination

Internal contamination potentially is a more serious problem. It is much more difficult to eliminate radioactive materials from within the body than it is to remove radioactive dirt on the outside of the body. When external contamination is present, care must be taken to prevent internal contamination. Death resulting from radiation from internal contamination is rare. A few deaths have been caused by medical misadministrations. In the Goiania accident, a 6-year-old girl died from severe internal contamination with cesium-137. Internal contamination, especially with iodine-131, was a concern after the reactor accident at Chernobyl in 1986. A familiar model of internal contamination, for example, is the bone scan

performed in your nuclear medicine department.

Metal Fragment

Another source of contamination that should be separately addressed is a metallic fragment. Metallic fragments can be intensely radioactive. If a radioactive metal fragment is present, it should not be touched with fingers. Tongs or forceps will increase the distance between the radioactive metal fragment and the fingers and thus greatly reduce any radiation dose.

“Hot” Particles

“Hot” particles are microscopic particles that can be intensely radioactive. Typically, they contain cobalt60 or fission products. These particles can be difficult to localize and remove. They may give a large radiation dose to a small volume of tissues. If the particle is trapped under a nail or is in the fold of the skin, routine washings may not dislodge it. The particle can sometimes be localized by using a thick piece of lead. If the lead is placed between the particle and the radiation detector, the exposure rate should decrease substantially. Once the particle is localized, it can usually be removed by using simple mechanical means. Rarely, a punch biopsy of the skin may be necessary.

MANAGEMENT

General Measures

The principles governing the treatment of radiation injuries are similar to the principles governing the treatment of any medical condition, especially those arising from hazardous materials. Treatment objectives must be prioritized (Fig. 90.10). Because no survivable radiation injury requires immediate lifesaving treatment, the medical staff should focus its attention on treating non–radiation-related life-threatening conditions. In the past, some medical personnel were so distracted by the radiation aspects of an accident that routine medical care was delayed.

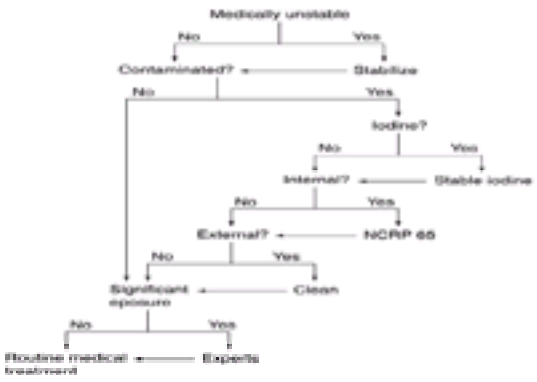

FIGURE 90.10. Treatment of radiation injuries.

Once the patient is stabilized, the radiation-related injuries can be addressed. Because there is no immediate treatment for radiation exposure, the problem of radioactive contamination should be addressed first. In most circumstances, a Geiger counter can be used to determine whether contamination is present. In addition, the probability of contamination can be assessed by obtaining an accurate description of the accident and the likely radiation source.

It must be appreciated that a great deal of the trauma from a radiation accident is related to the psychological stress and concern about cancer risk from the radiation exposure. The emergency physician should make certain that any concerns that the patient has about the long-term effects of radiation will be addressed subsequent to the acute care episode.

Internal Contamination

Treatment of internal contamination is most effective if initiated promptly. The requirement for prompt treatment is a dilemma for the physician. First, it is difficult to determine whether internal contamination is present until the external contamination has been removed. Moistened cotton-tipped applicator can be used to perform nasal swabs. If these have radioactivity on them, inhalation of radioactive materials is possible. The nature of the accident may provide clues to the possibility of internal contamination (e.g., a fire with smoke leading to inhalation of radioactive particles). Second, the most effective treatment requires knowledge of the radionuclide involved and its chemical form. This information is usually not immediately available. Fortunately, there are simple general treatment measures that can be effectively instituted before the magnitude of the internal contamination is fully known.

If given soon after exposure, stable iodine is effective at preventing the uptake of radioactive iodine by the thyroid gland. Prompt administration of stable iodine should be considered if there is the possibility of external contamination with radioactive iodine (Fig. 90.11). Because radioactive iodine is volatile, it is likely to be inhaled. If a contaminated child were brought to the ED after an accident with a radiopharmaceutical truck carrying radioactive iodine, administration of stable iodine would be appropriate. If further investigation revealed no radioactive iodine, little harm would have been done by administering the stable iodine. A single dose of oral iodine is unlikely to cause any adverse reactions even in patients who have serious reactions to iodinated contrast agents or seafood.

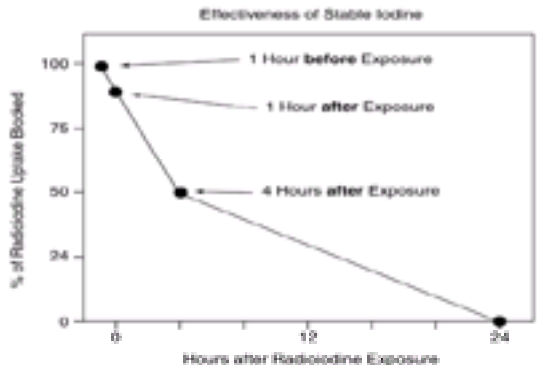

FIGURE 90.11. Stable iodine is most effective if given as soon as possible after the ingestion of radioactive iodine. The dose for children less than 1 year old is 65 mg of potassium iodide per day for 10 days. Children older than 1 year of age should receive 130 mg of potassium iodide a day for 10 days.

After a nuclear reactor accident that results in the release of a large amount of radioactive iodine, three steps can be taken to minimize the adverse effects on the public: 1) stable iodine can be given, 2) the public can be evacuated to prevent further exposure, and 3) the food supply can be monitored carefully to prevent the further ingestion of radioactive iodine. If a reactor accident occurs that involves contamination of members of the public, great panic will ensue. If this happens, emergency medical facilities should try to preserve their valuable resources for patients who need medical treatment. Emergency facilities have no special expertise in the treatment of contamination. If patients are otherwise healthy, they should be referred to other, usually state-designated facilities for the treatment of contamination.

Several simple steps can be taken to treat internal contamination nonspecifically. The goals of treatment are to prevent the absorption of the radionuclide and to enhance its excretion. Safe techniques that prevent the absorption of radionuclides include the administration of activated charcoal and alginate-containing antacids. Enhanced excretion can be achieved by hydration and administration of a purgative. Specific treatment for internal contamination depends on the radionuclide, its chemical and physical form, and the route of internal contamination. Recommendations for many specific treatments can be found in NCRP Report 65. This report should be available to every hospital ED. Initiation of treatments that entail some risk (e.g., pulmonary lavage, intravenous chelating agents) should be undertaken only after consultation with experts. The benefits of the treatment should be significantly greater than the risks of the treatment.

External Contamination

External contamination is treated in the same way as contamination by other hazardous chemical or biologic agents. To make certain that the hazard is treated appropriately, it is easiest to imagine that the patient has been covered with an easily detectable noxious agent (e.g., sewage, bacteria, viruses). Under this circumstance, the caretakers would wear gloves, a gown, shoe covers, and a mask. The purpose of wearing these garments is primarily to keep caretakers clean and to make cleanup easier. The garments do not decrease the exposure to penetrating radiation. The mask is recommended to prevent individuals from inadvertently touching their contaminated fingers to their nose or mouth. If available, film badges or other devices should be worn by the hospital staff who are in close contact with the patient.

If the external contamination is widespread, it may be helpful to cover the floor. If only a small area of contamination is present, spread of the contamination can be prevented by simply wrapping the contaminated area until it can be cleaned. Because it is much easier to detect radioactive contamination than chemical or biologic hazards, cleanup after a radiation accident will be much more effective and documentable. Precautions that are taken to prevent the inadvertent spread of contamination will make cleanup much easier.

External contamination rarely is a significant medical problem; however, logistical problems that must be addressed require preplanning. To minimize the chances of contaminating an unnecessarily large area of the ED, the patient should be admitted through a separate entrance. If this is not possible, the patient can be placed on a clean stretcher outside the ED and wrapped in a cloth (not plastic) sheet and then transported to the desired area of the hospital. Access to the treatment area should be controlled.

Removal of the patient's clothing will usually eliminate 70 to 90% of the external contamination (Table 90.8). Contaminated articles should be placed in labeled plastic bags. Residual contamination is likely to be on the hands, face, and hair. These should be washed with washcloths, using lukewarm water and soap. Particular attention should be paid to folds in the skin, the ears, and the area under the fingernails. Cleaning the skin with damp washcloths is much better than cleaning with running water. The radioactive dirt on the damp washcloth can be contained by placing the cloth in a plastic bag. Radioactive dirt that is in wash water is much more difficult to control. Shaving should not be performed because this may make small cuts and increase absorption through the skin. Excessive rubbing of the skin also may increase transdermal uptake.

Remove clothes.	Cover clean wounds to prevent contamination.
Wash with a damp cloth and tepid water.	Prevent external contamination from becoming internal.
Pay special attention to skin folds and fingernails.	Do not abrade the skin.

Table 90.8. Decontamination

Open, uncontaminated wounds should be covered to prevent them from becoming contaminated. Contaminated wounds should be cleaned like any other dirty wound. All samples from the patient should be saved and labeled.

A Geiger counter should be used to monitor and document the progress of the decontamination efforts. If contamination persists, the source may be fixed to the skin or it may be internal. Radiation experts should be consulted before more aggressive decontamination attempts are made. Some residual contamination may be acceptable.

Exposure

There is no emergency treatment for whole-body or local radiation exposure. Medically significant whole-body radiation exposure is unlikely if the patient does not have nausea and vomiting. Serial complete blood counts are also helpful in excluding the diagnosis of a recent large whole-body exposure to radiation (Table 90.9). In the absence of other major trauma, the absolute lymphocyte count will rapidly fall in patients who have been exposed to a large radiation dose. If a patient has been exposed to a large dose of radiation, very little needs to be done in the ED. The threat to the patient's life will occur a few weeks after the exposure.

In the Emergency Department	Later
Complete blood count, and	Cytogenetics
every 6 hours for 24 hours	Sperm count
Nasal swabs	Eye examination (baseline for cataracts)
Collect all excreta	Human leukocyte antigen typing

Table 90.9. Appropriate Laboratory Tests for Patients Involved in a Significant Radiation Accident

The diagnosis of a local radiation injury requires vigilance. The physician should consider the possibility of a local radiation injury when there is an unexplained painless “burn.” A complete blood count to exclude an accompanying whole-body exposure is indicated. The prognosis of a local radiation injury depends on the dose. The dose can be estimated only by having a qualified physicist reconstruct the accident that led to the exposure.

Suggested Readings

A Guide to the Hospital Management of Injuries Arising from Exposure to or Involving Ionizing Radiation. Chicago: American Medical Association, January, 1984.

Collins VP, Gaulden ME. A case of child abuse by radiation exposure. In: Hubner KT, Fry SA, eds. The Medical Basis for Radiation Accident Preparedness. New York: Elsevier-North Holland, 1980:197–203.

Hubner KF, Fry SA, eds. The Medical Basis for Radiation Accident Preparedness. Proceedings of a conference. Oak Ridge, TN, 1979. New York: Elsevier-North Holland, 1980.

Ionizing radiation exposure of the population of the United States. NCPR Report 93. Bethesda: National Council on Radiation Protection and Measurements, 1987.

Jacobson BP, Plato PA. Toeroek D. Contamination of the home environment by patients treated with I131: initial results. Am J Public Health 1978; 68:225–230.

Jammet H, Gongora R, Jockey P, et al. The 1978 Algerian accident: Acute local exposure of two children. In: Hubner KF, Fry SA, eds. The Medical Basis for Radiation Accident Preparedness. New York: Elsevier-North Holland, 1980:229–245.

Jammet H, Gongora R, Pouillard P, et al. The 1978 Algerian accident: Four cases of protracted whole body irradiation. In: Hubner KF, and Fry SA, eds. The Medical Basis for Radiation Accident Preparedness. New York: Elsevier-North Holland, 1980:113–129.

Leonard RB, Ricks RC. Emergency department radiation accident protocol. Ann Emerg Med 1980; 9:462–470.

Management of persons accidentally contaminated with radionuclides. NCRP Report 65. Bethesda: National Council on Radiation Protection and Measurements, 1980.

The radiological accident in Goiana. Vienna: The International Atomic Energy Agency, 1988.

The radiological accident in San Salvador. Vienna: The International Atomic Energy Agency, 1990.

Ricks RC. The scope of the problem. In: Ricks RC, Berger ME, O'Hara FM, eds. The Medical Basis for Radiation Accident Preparedness. III. The Psychological Perspective. New York: Elsevier, 1991:310.

Ross JF, Holly FE, Zarem HA, et al. The 1979 Los Angeles accident: exposure to Iridium 192 industrial radiographic source. In: Hubner KF, Fry SA, eds. The Medical Basis for Radiation Accident Preparedness. New York: Elsevier-North Holland, 1980:205–221.

Royal HD. Three Mile Island and Chernobyl reactor accidents. In: Mettler FA, Kelsey CA, Ricks RC, eds. Medical Management of Radiation Accidents. Boca Raton: CRC Press, 1990:269–292.

Royal HD. The role of physicians in minimizing the psychological effects of radiation accidents. In: Ricks RC, Berger ME, O'Hara FM, eds. The Medical Basis for Radiation Accident Preparedness III. The Psychological Perspective. New York: Elsevier, 1991:277–285.

Shleien B. Preparedness and Response in Radiation Accidents. FDA Publication 83-8211. Washington, DC: US Department of Health and Human Services, 1984.

Young RW. Acute radiation syndrome. In: Conklin JJ, Walker RI, eds. Military Radiobiology. Orlando: Academic Press, 1987:165–190.

CHAPTER 91

Bites and Stings

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This chapter is oriented to the clinical diagnosis and management of injuries that result from bites and stings, especially as they relate to children. Although the largest proportion of the morbidity and mortality from these injuries occurs in the pediatric age group, little attention has been focused in the pediatric literature on the specifics of treatment.

An overall assessment should include vital signs, location and size of fang or sting marks, pain, swelling, color of surrounding skin, and any systemic symptoms. General care should include relief of pain and itching, tetanus prophylaxis, antibiotics if needed, and emotional support. Animals must be identified as venomous or not. Venomous animals are those that inject their toxin into other animals to produce a harmful effect. Poisonous creatures are those whose tissues are toxic, either in whole or in part. This chapter deals with venomous bites and stings and with wounds inflicted by nonvenomous animals. In evaluating any potential venomous bite or sting, the physician must distinguish between the asymptomatic and the symptomatic bite or sting. Clinical observation may be the only means of distinguishing between the two.

Only those venomous animals found within the continental United States and Canada are discussed here. Of marine life, only that within the tidal zone or commonly washed ashore is considered. This chapter covers marine invertebrates and vertebrates, terrestrial invertebrates, venomous reptiles, and common mammalian bites.

MARINE INVERTEBRATES

Phylum Coelenterata (Cnidaria)

The phylum is divided into three large classes: the *Hydrozoa* (hydras, Portuguese man-of-war), *Scyphozoa* (true jellyfish), and *Anthozoa* (soft corals, stone corals, anemones).

The phylum includes some of the most beautiful and deadly marine creatures. All members of the phylum have specialized organelles called nematocysts, which are used for entangling, penetrating, anchoring, and poisoning prey (Fig. 91.1). When the tentacles touch an object, the nematocysts fire, releasing toxin-coated, barbed threads. Firing of the nematocysts is not fully understood; the process may be protein- or cation-mediated. The nematocysts of most species cannot penetrate human skin. However, those that do may cause severe pain, serious illness, or even death. The severity of envenomation is related to the degree of toxicity of venom, number of nematocysts discharged, and general condition of the victim. Stings from sessile forms are generally not as severe as stings from free-floating forms. The various toxins have been named and described but not completely biochemically characterized. Venom varies from species to species. Paralysis and central nervous system (CNS) effects appear to be related primarily to toxic proteins and peptides. Burning pain and urticaria are secondary to the presence of serotonin, histamine, or histamine-releasing agents in the venom.

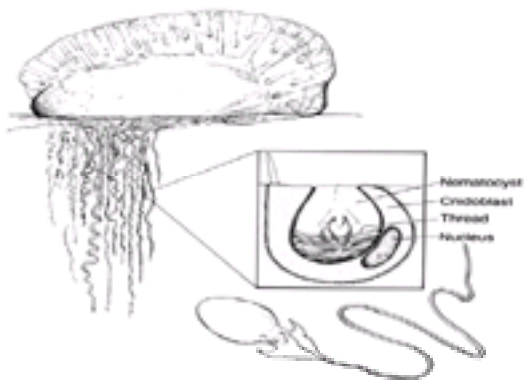

FIGURE 91.1. Marine invertebrate causing human sting.

Class Hydrozoa

Feathered hydroid *(Pennaria tiarelia)* is found from Maine to Florida and along the Texas coast just below the low-tide line. It is attached to solid objects, including pilings and floats. The mild sting that occurs with handling may be treated with local care.

Portuguese man-of-war *(Physalia physalis)*—commonly considered a jellyfish—is in reality a hydrozoan colony. The float can be up to 30 cm in length. The tentacles hang from the float, may reach a length of more than 75 feet, and contain about 750,000 nematocysts each. This pelagic animal is often driven ashore by storms all along the Atlantic coast. Releasing one of the most powerful marine toxins, the nematocysts may discharge even when the animal is dead and on the beach. Because of the length and transparency of the tentacles in the water, swimmers are often stung without seeing the animal. The toxin contains polypeptides and degradative enzymes. Local effects include pain and irritation. Systemic reactions include headache, myalgias, fever, abdominal rigidity, arthralgias, nausea and vomiting, pallor, respiratory distress, hemolysis, renal failure, and coma. Death may occur if the area stung is extensive in relation to the size of the victim. Treatment is discussed in the next section, Scyphozoa.

Class Scyphozoa

The common purple jellyfish *(Pelagia noctiluca)* is only mildly toxic. Local skin irritation is the major clinical manifestation.

Sea nettle *(Chrysaora guinguecinda)* is a common jellyfish found along the Atlantic coast. Clinical manifestations are the same as those for purple jellyfish.

Lion's mane *(Cyanea capillata)* is a highly toxic creature that received considerable publicity as the instrument of death in the Sherlock Holmes classic *Adventure of the Lion's Mane.* The animal reaches a width of 244 cm, with tentacles as long as 61 cm. The shaggy clusters of the golden yellow tentacles that hang from the medusa resemble the mane of a lion. The animal is found along both coasts. Contact with the tentacles produces severe burning. Prolonged exposure causes muscle cramps and respiratory failure.

Treatment of hydrozoan and scyphozoan stings is based on the same general principles. It is directed at three objectives: relieving pain, alleviating effects of venom, and controlling shock. The most important step is to remove the tentacles; as long as the tentacle adheres to the skin, the nematocysts continue to discharge. Inactivate the unexploded nematocysts by topical application for 30 minutes with vinegar (3% acetic acid), a slurry of baking soda, or meat tenderizer (papain). The area should be washed with sea water or normal saline. Any adherent tentacles should be carefully removed with instruments or a gloved hand, and the wound area should be immobilized. There is no antivenin available for *Physalia* or the scyphozoans except for the sea wasp, *Chironex fleckeri,* of Australia. General supportive measures for systemic reactions include oral antihistamines, oral corticosteroids, and codeine or meperidine for pain. Cardiac and respiratory support may be required. Muscle spasms have been treated with 10% solution of calcium gluconate 0.1 mL/kg given intravenously. Local dermatitis should be treated with a topical corticosteroid cream.

Class Anthozoa

The anemones found within United States tidal zones are only mildly toxic at worst. Coral cuts and stings can be a problem for swimmers off the Florida coast. The stinging ability of stony corals is not well defined but is considered to be of minor significance. Coral cuts, however, are important. The severity of coral cuts stems from a combination of factors, including laceration of tissue, nematocyst venom, foreign debris in the wound, and secondary bacterial infection. The clinical picture is one of stinging sensation followed by wheal formation and itching. If the wound is untreated, an ulcer with an erythematous base may form within a few days. Cellulitis, lymphangitis, fever, and malaise commonly occur.

Treatment consists of cleaning the wound and irrigation with copious amounts of saline. Removal of foreign particles must be accomplished, and debridement may be necessary. The sea provides an excellent inoculum for wound infections. Marine bacteria are generally heterotrophic, motile, and facultatively anaerobic, Gram-negative rods. Organisms include *Vibrio* species, *Erysipelothrix rhusiopathiae,* and *Mycobacterium marinum.* Wounds should be left open. Broad-spectrum antibiotic therapy, particularly tetracycline, at a dosage of 40 mg/kg per day in four divided doses, has been advocated but cannot be used in children younger than 8 years old. For children younger than 8 years, cephalexin (50 mg/kg per day in four divided doses) or trimethoprim–sulfamethoxazole should be used.

Phylum Echinodermata

Phylum Echinodermata includes starfish, sea urchins, and sea cucumbers. Of the three classes, only the *Echinoedae*—sea urchins—have clinical relevance for American children. The long-spined urchins (e.g., *Diadema*) are dangerous to handle. They do not appear to possess venom as do some of the tropical urchins, but the spines, composed of calcium carbonate, easily pierce the skin and lodge deep in the flesh. The spines may break off readily into the wound and can penetrate wet suits and sneakers. Most injuries occur during wading in shallow water. Clinically, penetration is accompanied by intense pain followed by redness, swelling, and aching. Complications include tattooing of the skin, secondary infection, and granuloma formation.

In treatment, all spines should be removed as completely as possible. If spines break off in the wound, debridement should be performed with local anesthetic under aseptic conditions, but any spines not reachable will be absorbed in time. Soaking the wound in warm water may be helpful. Systemic antistaphylococcal antibiotics should be used if infection develops.

MARINE VERTEBRATES

Stingrays

Background

Stingrays are the single most important group of venomous fishes, accounting for an estimated 750 attacks per year in North America. Stingrays are bottom feeders that have a habit of burying themselves in sand or mud of bays, shoal lagoons, and river mouths. The animals are found along the Atlantic, Pacific, and Gulf coasts and range from several inches in diameter to more than 14 feet in length. Six different species are represented in North American waters. Envenomations usually occur when an unsuspecting swimmer steps on the back of the animal and causes it to hurl its barbed tail upward into the victim as a reflex defense response. Most injuries are confined to the lower extremities, although wounds to the chest and abdomen have been reported.

The venom apparatus consists of a serrated, retropointed, dentinal caudal spine located on the dorsum of the tail. Spines vary in length, depending on the size of the ray, but may reach a length of 122 cm in some species. The spine is encased in an integumentary sheath that contains specialized secretory cells that hold the venom. When the stingray's barb strikes the victim, it easily penetrates the skin, rupturing the integumentary sheath over the spine and causing the venom to pass along the ventrolateral grooves of the barb into the wound. Laceration of the victim's tissues when the spine is withdrawn facilitates the absorption and distribution of the toxin. The venom is a heat-labile toxin that has been shown to contain at least 15 fractions, including serotonin, 5-nucleotidase, and phosphodiesterase. The toxin depresses medullary respiratory centers, interferes with the cardiac conduction system, and produces severe local pain.

Clinical Manifestations

Because the barb is retropointed, the wound it produces is a combination of puncture and laceration. Wounds may vary in length from 3.5 to 15 cm. The sting is followed immediately by pain, which spreads from the site of injury over the next 30 minutes and usually reaches its greatest intensity within 90 minutes. Pain and edema are most often localized to the area of injury; however, syncope, weakness, nausea, and anxiety are common complaints attributed to both the effects of the venom and the vagal response to the pain. Among other generalized symptoms are vomiting, diarrhea, sweating, and muscle fasciculations of the affected extremity. Generalized cramps, paresthesias, hypotension, arrhythmias, and death may occur. The wound often has a jagged edge that bleeds profusely, and the wound edges may be discolored. Discoloration may extend several centimeters from the wound within hours after injury and may subsequently necrose if untreated. Often, parts of the stingray's integumentary sheath contaminate the wound.

Management

Treatment is aimed at 1) preventing complications evoked by the venom, 2) alleviating the pain, and 3) preventing secondary infection. At the scene, the wound should be irrigated with cold salt water. Flushing can help remove much of the venom. Bleeding should be controlled with direct pressure. On the patient's arrival in the emergency department (ED), shock, if present, should be treated with intravenous (IV) fluids. An attempt should be made to remove any remnants of the integumentary sheath if it can be seen in the wound. The extremity should be placed in hot water (40° to 45°C [104° to 113°F]) for 30 to 90 minutes. After soaking, the wound should be reexplored. Further debridement can then be accomplished and the wound can be closed. Pain relief may be achieved with meperidine (1 to 2 mg/kg). Tetanus prophylaxis should always be considered, but antibiotics are reserved for wounds that become secondarily infected.

Sharks

Background

Fear of sharks is as old as human history. Sensational media reports of shark attacks—and several popular movies—have asked whether it is safe to go into the water. The answer may be no, but not because of sharks. The chance of being assaulted by a shark along the North American coast is roughly 1 in 5 million. Because of the large number of sporting activities that take place in the ocean environment, however, clinicians who practice in coastal areas may be called on to manage a victim of these primitive creatures.

In U.S. waters, most attacks are by the gray reef, great white, blue, and mako sharks. Factors that increase a risk of attack include swimming near sewer outlets, time of day (late afternoon and early evening), murky warm water, increased

commotion, deep channels, and bright objects. Attacks of surfers along the northern California coast were believed to be caused by sharks mistaking surfboard shapes in the water for elephant seals, part of the shark's usual diet.

Clinical Manifestations

Attack victims usually do not see the shark before it strikes. Occasionally, the attack is preceded by one or more "bumps," during which the victim may sustain extensive abrasions from the rough denticles of the shark's skin. Two types of bite wounds are described: tangential injury and a definitive bite. Tangential injury is caused by the slashing movement of the open mouth as the shark makes a close pass. Severe lacerations, incised wounds, and loss of tissue are seen. Definitive bite wounds vary according to the part of the body seized by the shark. Lacerations, loss of soft tissue, amputations, and comminuted fractures are recorded. Most injuries involve only one or two bites and are confined to the extremities.

Management

Hypovolemic shock is the immediate threat to life in shark attacks. Bleeding should be controlled at the scene with direct compression, and intravascular volume should be replaced with crystalloid until blood products are available. The victim should be kept warm and given oxygen when being transported to an ED. Wounds should not be explored in the field. Tetanus toxoid and tetanus immune globulin should be considered, and prophylactic antibiotics with a third-generation cephalosporin or trimethoprim– sulfamethoxazole is suggested.

Scorpaenidae

Background

The 80 species found in the *Scorpaenidae* family include the zebrafish, scorpionfish proper, and the stonefish. In California, the sculpin is commonly involved. *Scorpaenidae* are generally found in shallow water, around reefs, kelp beds, or coral. All members of the family are nonmigratory, slow swimming, and often buried in sand. The venom apparatus consists of a number of dorsal, anal, and pelvic spines covered by integumentary sheaths containing venom glands that lie within anterolateral grooves. The venoms are unstable, heat-labile compounds. Most often envenomation occurs when the fish are handled during fishing excursions.

Clinical Manifestations

The clinical signs and symptoms are essentially the same, varying among the species in degree only. Severe pain at the site of the wound is the first and primary clinical sign for all species. The wound and surrounding area becomes ischemic and then cyanotic. Paresthesia and paralysis of the extremity may occur. Other clinical signs include nausea, vomiting, hypotension, tachypnea proceeding to apnea, and myocardial ischemia with electrocardiographic (ECG) changes.

Management

Treatment involves irrigating the wound with sterile saline. The injured extremity is then immersed in very hot water (40° to 45°C [104° to 113°F]) for 30 to 60 minutes or until the agonizing pain is completely relieved. Meperidine hydrochloride (1 to 2 mg/kg) may be required for pain. The patient should be monitored carefully for cardiotoxic effects and respiratory depression. Antivenin is available only for the stings of the stonefish of Australia.

Catfish

Background

The catfish is a popular food and sport fish found in many lakes and rivers throughout the United States. The venom apparatus consists of a number of spines located in the dorsal and pectoral fins. The integumentary sheaths covering the spines contain venom glands. The venoms are unstable, heat-labile compounds. Most often envenomation occurs when the fish are handled during fishing excursions. A combination of injuries are seen; wounds secondary to puncture and laceration, foreign body reaction, and the effects of venom.

Clinical Manifestations

The spines inflict a puncture wound or laceration. The spines may become imbedded in the flesh of the victim causing soft-tissue swelling, which may become infected or lead to a foreign body reaction. The venom produces a local inflammatory response—local intense pain, edema, hemorrhage, and tissue necrosis.

Management

Treatment involves irrigating the wound with sterile saline. The injured extremity is then immersed in hot water (40° to 45°C [104° to 113°F]) for 30 to 60 minutes or until pain is relieved. Meperidine hydrochloride (1 to 2 mg/kg) may be required for analgesia. The wound should be explored to locate any retained spines. Adequate debridement is essential. Systemic antibiotics to cover Gram-negative organisms are recommended. Wounds may be closed using a delayed primary closure.

TERRESTRIAL INVERTEBRATES

Phylum Arthropoda

The arthropods make up the largest phylum in the animal kingdom. All *Arthropoda* have an exoskeleton with jointed appendages. The phylum is divided into two subphyla: the Chelicerata, which includes scorpions, spiders, ticks, and mites, and the Mandibulata, which includes insects.

Scorpions

Background

Of 650 known scorpion species (class Arachnida), only a limited number are dangerous to humans. In the southwest United States, *Centruroides sculpturatus* is the potentially lethal inhabitant. Although *C*. *sculpturatus* and *C. exilicauda* (Fig. 91.2) have been considered separate species in the past, recent taxonomic classification treats the two as one species. The animal has two pinching claws anteriorly and a tail or pseudoabdomen that ends in a telson (Fig. 91.2). The telson houses a pair of poison glands and a stinger. Normally, scorpions grasp their prey with pincers and then sting the victims by arching their tails over their heads. The animals are nocturnal, and during the day, they seek shelter under stones and debris. They often crawl into sleeping bags and unoccupied clothing. In one report, 80% of stings occurred in children less than 10 years old.

FIGURE 91.2. *Centruroides exilcauda (sculpturatus).* (Courtesy of F. E. Russell.)

Clinical Manifestations

The scorpion's poison gland produces a neurotoxin. The general neurotoxicity is excitatory, affecting the autonomic and skeletal neuromuscular system. Common symptoms include local pain, restlessness, hyperactivity, roving eye movements, and respiratory distress. Other associated signs may include convulsions, drooling, wheezing, hyperthermia, cyanosis, and respiratory failure. The diagnosis may be difficult because a history of a sting may not be forthcoming. There is no laboratory test for confirmation of envenomation.

Management

Numerous treatment modalities have been used in addition to general supportive care. Cryotherapy of the site of sting has been advocated to reduce swelling and local induration. An antivenin has been developed that may be advantageous in decreasing the severity and duration of symptoms. Antivenin should be considered after general supportive care has been instituted only if the following symptoms persist: tachycardia, hyperthermia, severe hypertension, and agitation. Antivenin that is not approved by the Federal Drug Administration (FDA) is available through the Antivenom Production Laboratory in Arizona State University in Tempe, Arizona. Sedative-anticonvulsants, in particular phenobarbital (5 to 10 mg/kg), have been used to treat persistent hyperactivity, convulsions, and/or agitation. Calcium gluconate (0.1 mL/kg of the 10% solution) has been given intravenously to reduce muscular contractions and associated pain, but benefit has not been proved. Corticosteroids and antihistamines have little if any proven benefit.

Spiders

More than 100,000 species of spiders (class Arachnida) are known to exist. All are carnivorous and have fangs and venom by which they immobilize and kill their prey. The risk of serious bites is small, except in a few species. In most species, the fangs are too short and fragile to penetrate human skin, and the venom is mild. Although most spiders are shy and retiring creatures that will not bite people unless provoked, two species in the United States are capable of producing more severe reactions.

Loxoscelism (Bite of the Brown Recluse Spider)

Three species of *Loxosceles* have caused envenomation, primarily in the southern and midwestern states. These small spiders (1 to 1.5 cm in length) are characterized by a brown violin-shaped mark on the dorsum of the cephalothorax. They are found outdoors but will establish nests indoors, especially in closets. The most common species, *Loxosceles reclusa,* is, as its name implies, shy and will only attack when provoked. The venom is cytotoxic and also contains a factor similar to hyaluronidase.

Clinical Manifestations. Clinically, the bite is usually innocuous. Because the bite is initially unnoticed, there is sometimes a delay in seeking medical attention. The spectrum of reaction ranges from minor local reaction to severe necrotic arachnidism (Fig. 91.3). The local reaction is characterized by mild to moderate pain 2 to 8 hours after the bite.

At the site of the bite, erythema develops with a central blister or pustule. Within 24 hours, subcutaneous discoloration appears and spreads over the next 3 to 4 days, reaching a size of 10 to 15 cm. At this time, the pustule drains, producing an ulcerated “crater.” The local reaction varies with the amount of venom injected. Scar formation is rare if there is no evidence clinically of necrosis within 72 hours of the bite. Systemic reaction is most commonly noted in small children. Symptoms are noted 24 to 48 hours after the bite and include fever, chills, malaise, weakness, nausea, vomiting, joint pain, morbilliform eruption with petechiae, intravascular hemolysis, hematuria, and renal failure.

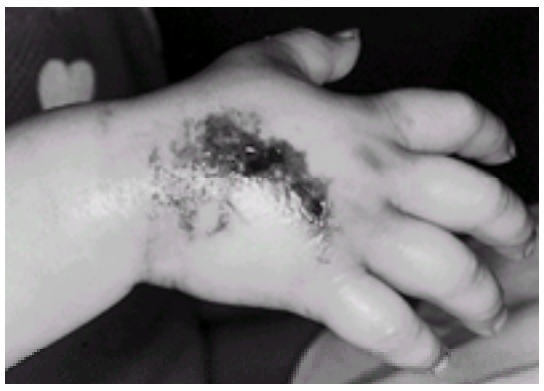

FIGURE 91.3. Spider bite necrotizing.

Management. Because of the delay in initial diagnosis, treatment varies with the clinical stage of the bite. There is no specific serologic, biochemical, or histologic test to diagnose envenomation accurately. Unless all or part of the spider is brought for identification, definitive diagnosis cannot be made. Table 91.1 lists the spiders found in the United States known to cause necrotic lesions. An algorithm for management of suspected bites is shown in Figure 91.4. One recent series of adult patients suggests that serious complications are rare. Most victims will heal with supportive care. Large-dose steroids have been advocated in the past; however, recent studies have found no significant alteration of necrosis from the venom by steroids or heparin. Once large areas of necrosis have become demarcated, surgical excision and skin grafting are required, although the need for grafting is rare. The use of dapsone continues to be controversial. Animal studies do not support the use of dapsone, hyperbaric oxygen, or the two in combination in the treatment of these envenomations. However, current recommendations are to limit the use of dapsone to adults with proven brown recluse bites. Dapsone should not be used in children because of methemoglobinemia. Antivenom is not yet commercially available. For systemic manifestations, vigorous supportive care is needed. Platelet count for evidence of hemolysis is needed as well as monitoring of hemoglobin, urine sediment, blood urea nitrogen (BUN), and creatinine for evidence of hemolysis and renal failure.

Genus Names	Common Name	Geographic Distribution
Argiope	Golden orb weaver	Throughout North America (individual species more restrictive)
Chiracanthium	Running spider	Throughout United States
Loxosole	Brown recluse	Kansas and Missouri to Texas West to California
Lycosa	Wolf spider	Throughout United States
Phidippus	Black jumping spider	Atlantic Coast to Rocky Mountains

Table 91.1. Spider Known to Cause Necrotic Lesions

FIGURE 91.4. Management of suspected brown recluse spider bite.

Latrodectism (Bite of the Black Widow Spider)

The bite of *Latrodectus mactans* is the leading cause of death from spider bites in the United States. The animal is shiny

black with a brilliant red hourglass marking on the abdomen. The marking is found on the mature female and may be present on the male. The male is not a threat because it is only one-quarter the size of the female, meaning its fangs are unable to penetrate human skin. The webs are usually found in corners or out-of-the-way places. The female is not aggressive unless guarding her egg sac or provoked. The venom, a complex protein that includes a neurotoxin, stimulates myoneural junctions, nerves, and nerve endings.

Clinical Manifestations. Reaction is generalized pain and rigidity of muscles 1 to 8 hours after the bite. No local symptoms are associated with the bite itself. The pain is felt in the abdomen, flanks, thighs, and chest and is described as cramping. Nausea and vomiting are often reported in children. Respiratory distress can occur. Chills, urinary retention, and priapism have been reported. There is a 4 to 5% mortality rate, with death resulting from cardiovascular collapse. The mortality rate in young children may be as high as 50%.

Management. Because of the size, color, and distinctive markings of this spider, bites are seldom mistaken if the child is old enough to describe the spider. A child who has severe pain and muscle rigidity after a spider bite should be considered a *Latrodectus* bite victim. A clinical grading scale has been developed by Clark (Table 91.2). Treatment with *Latrodectus* antivenin (Lyovac; Merck, Sharp & Dohme) should be instituted as soon as a bite is confirmed in children who weigh less than 40 kg; the usual dose is 2.5 mL (one vial). Antivenin should be administered following the package insert and after skin testing to determine the risk of hypersensitivity to horse serum. For children who weigh more than 40 kg, it is not as urgent to institute antivenin treatment, but indications for its use include patients who are less than 16 years old, who have respiratory difficulty, or who have significant hypertension (grades II and III). Serum sickness is a possible side effect. Because the dosage is low, however, serum sickness is uncommon, with a rate lower than those reported for other types of antivenom. Calcium gluconate 10% solution is often given for control of leg and abdominal cramps. The dosage is 0.1 mL/kg per dose as IV push over 5 minutes. A recent series found calcium gluconate effective in only 4% of cases. Methocarbamol (Robaxin) does not appear to be as efficacious as calcium gluconate. Muscle relaxants such as diazepam also have been advocated, but they are variably effective and the effects are short lived. Analgesia may be achieved with morphine or meperidine.

Grade	Symptoms
1	Asymptomatic
	Local pain at bite site
	Normal vital signs
2	Muscular pain—localized
	Diaphoresis—localized
	Normal vital signs
3	Muscular pain—generalized
	Abnormal vital signs
	Nausea, vomiting
	Headaches
	Diaphoresis

Table 91.2. Grading Scale for *Latrodectus* Envenomation

Tarantulas and Others

Tarantulas, although fearsome in size and appearance, do not bite unless provoked. The venom is mild, and envenomation is not a problem. The wolf spider (*Lycosa* species) and the jumping spider (*Phidippus* species) also have been implicated in bites. Like the tarantula, they have a mild venom that causes only local reactions. Bites from all three of these spiders should be treated with local wound care.

Ticks

Ticks are responsible for transmitting a variety of infectious agents, including spirochetes, viruses, rickettsiae, bacteria, and protozoa. Examples of tickborne illness include Rocky Mountain spotted fever (RMSF), Lyme disease, tularemia, ehrlichiosis, babesiosis, relapsing fever, and Colorado tick fever (see Chapter 84). In addition, tick paralysis is associated with the bite of the wood tick, *Dermacentor andersonii;* the dog tick, *Dermacentor variabilii;* and the deer tick, *Ixodes scapularii.* The gravid engorged tick releases a neurotoxin that can produce cerebellar dysfunction or an ascending weakness. The mechanism of action of the toxin is not well understood.

Clinical Manifestations. Following tick attachment there is a latent period of 4 to 7 days followed by symptoms of restlessness, irritability, and ascending flaccid paralysis. Respiratory paralysis and death may follow if the tick is not detected. Laboratory data, including cerebrospinal fluid (CSF) are usually normal, but lymphocytic pleocytosis has been reported.

Management. Management is based on general supportive care and a diligent search for the tick. Ticks should be removed using blunt forceps or tweezers. The tick should be grasped as close to the skin surface as possible and pulled upward with a steady even pressure. A twisting or jerking motion may cause the mouth parts to break off. Do not squeeze or crush the body of the tick because this may introduce infective agents. After the tick is removed, the bite site should be cleaned. Once the tick is removed, the paralysis is reversible without apparent sequelae.

Centipedes and Millipedes

Centipedes (class *Myriapode* order *Chilopoda*) are venomous, biting with jaws that act like stinging pincers. Bites can be extremely painful; however, the toxin is relatively innocuous, causing only local reaction. Treatment consists of injection

of local anesthetic at the wound site and local wound care.

American millipedes (order *Diplopoda*) are generally harmless.

Insects

The insects (class Insecta) constitute the largest number of animal species. Hymenoptera is the most important order of the class and includes bees, wasps, hornets, yellow jackets, and ants (Fig. 91.5). Because of differences in venom composition and rate of systemic reactions, ants are covered separately in this chapter. Hymenoptera are responsible for 50% of human deaths from venomous bites and stings. A variety of toxic reactions are seen, but the most common is allergic.

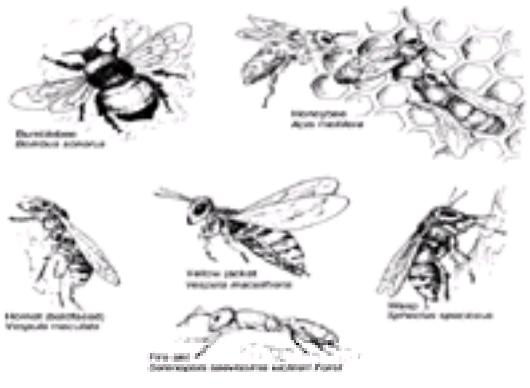

FIGURE 91.5. Hymenoptera capable of causing allergic reactions.

Bee, Hornet, Yellow Jacket, Wasp (Fig. 91.5)

Clinical Manifestations. Clinically, the stings of bees and wasps differ because the barbed stinger of the bee remains in the victim's skin, whereas the wasp may sting multiple times. Reactions may vary. The venoms of the bee, hornet, yellow jacket, and wasp contain protein antigens that can elicit an immunoglobulin E (IgE) antibody response in those who are stung. In addition, venoms contain various biogenic amines, phospholipase, phosphatase, and hyaluronidase. Because of the similarity of the venoms, crossreactivity can occur. The allergic reactions may be grouped by severity. Group I consist of a local response at the site of bite or sting. Group II includes the mild systemic reactions typified by generalized pruritus and urticaria. Group III consists of severe systemic reactions, including wheezing, angioneurotic edema, nausea, and vomiting. Group IV consists of life-threatening systemic reactions, including laryngoedema, hypotension, and shock. Anaphylactic reactions secondary to insect stings occur in 0.5 to 5% of the population.

Management. Because the barbed honeybee stinger with venom sac is avulsed and often remains in the victim's skin, it must be removed if seen. In the past, texts suggested that removal was best accomplished by scraping the stinger and that squeezing or pulling must be avoided. A recent study compared the effects of delays in removing stings with the effects of different methods of removal. The findings showed that the method of removal is irrelevant (scraping versus pulling) but that delays in removal are likely to increase the dose of venom received. Stings do not need to be dried with baking soda.

Treatment of stings is based on the severity of the allergic reaction. Group I reactions can be treated with cold compresses at the site of sting. Group II reactions are treated with diphenhydramine hydrochloride 4 to 5 mg/kg per day (maximum 200 mg) orally in four divided doses for several days. Group III reactions are treated with epinephrine 1:1000 solution 0.01 mL/kg (maximum 0.3 mL) injected subcutaneously followed by diphenhydramine orally. In addition H_2-blockers may provide additional benefit. Ranitidine or cimetidine can be used. These children should be observed in the hospital for 24 hours. Group IV reactions may require intubation if upper airway obstruction is present. Wheezing refractory to epinephrine should be treated with an aminophylline bolus of 6 mg/kg over 20 minutes, followed by a 1.1 mg/kg per hour infusion if needed. Hypotension should be treated with a fluid bolus of saline or lactated Ringer's solution 10 to 20 mL/kg given over 20 to 30 minutes. IV epinephrine (1:10,000) should be considered if hypotension fails to respond to subcutaneous epinephrine and fluid bolus. Hydrocortisone (2 mg/kg) may be given intravenously every 6 hours for 2 to 4 days. All children in this group should be admitted to an intensive care unit (ICU). Children who have had a group III or IV reaction should be followed by an allergist for hyposensitization. Parents of these children should keep an insect sting emergency kit. The EpiPen or EpiPen Jr are spring-loaded autoinjectors triggered by placing pressure on the thigh with the instrument. The pens inject 0.3 or 0.15 mg (EpiPen and EpiPen Jr, respectively) of epinephrine. The pens are used as first aid in the field by the parent or guardian and are not meant to substitute for prompt definitive treatment at a medical facility. Parents should receive information regarding the avoidance of situations and behaviors that would attract stinging insects.

Fire Ants

Clinical Manifestations. An increasing number of bites and envenomations in the South has been accounted for by fire ants (*Solenopsis richteri* and *Solenopsis invicta*). The venom differs from the other Hymenoptera in that it is an alkaloid with a direct toxic effect on mast cell membranes. There is no crossreactivity with other members of the order.

The fire ant bites with well-developed jaws and then uses its head as a pivot to inflict multiple stings. The clinical picture of fire ant sting is one of immediate wheal and flare at the site. The local reaction varies from 1 to 2 mm up to 10 cm,

depending on the amount of venom injected. Within 4 hours, a superficial vesicle appears. After 8 to 10 hours, the fluid in the vesicle changes from clear to cloudy (pustule) and becomes umbilicated. After 24 hours, it is surrounded by a painful erythematous area that persists for 3 to 10 days. Edema, induration, and pruritus at the site occur in up to 50% of patients. Occasionally, systemic reactions occur as with other Hymenoptera.

Management. Treatment of fire ant stings is symptomatic. Local care, such as ice applied to the reactive area, and frequent cleansing of the lesions to prevent secondary infection are all that is usually required. Topical steroids, antibacterial medications, and antihistamines do not appear to be efficacious in prevention of pustule formation. Antihistamines are useful for pruritus. Systemic reactions are rare and should be treated similarly to other Hymenoptera reactions.

TERRESTRIAL VERTEBRATES

Venomous Reptiles

Background

God said to the serpent,

> Be accursed beyond all cattle, all wild beasts. You shall crawl on your belly and eat dust every day of your life. I will make you enemies of each other: you and the woman, your offspring and her offspring.
>
> —Genesis 3:14

Throughout recorded history, serpents and their encounters with humans have evoked strong emotions, folklore, and medicinal practices. Research during the past several decades has lessened the mystique surrounding the venomous substances secreted by 15% of this country's 120 snake species. The recent emphasis on antivenin therapy and expedient supportive medical care has dramatically reduced mortality and morbidity from poisonous snakebites.

In the United States, an estimated 8000 people are bitten annually by poisonous snakes. Predictably, the pediatric population, especially males, aged 5 to 19 years, accounts for a disproportionately large number of these victims. The highest incidence occurs in the Southeast and Southwest between April and October, although venomous snakebite occur at least sporadically in most states. Only 10 to 15 deaths are reported per year, but the morbidity in limb dysfunction and other complications, although unknown, is undoubtedly much higher. With appropriate therapy, most long-term morbidity can be prevented.

The poisonous snakes indigenous to the United States are members of the Crotalidae (pit viper) or *Elapidae* families (Table 91.3). The rattlesnake, water moccasin, and copperhead are pit vipers and are responsible for 99% of venomous snakebites. The coral snake is the only member of the *Elapidae* family in this country and, along with imported exotic snakes, accounts for the remaining 1% of serious snakebites.

Family	Genus	Species	Common Name
Crotalidae			Pit vipers
	Crotalus		Rattlesnakes
		C. adamanteus	Eastern diamondback
		C. atrox	Western diamondback
		C. horridus	Timber rattlesnake
		C. viridis	Western rattlesnake
		C.v. viridis	Prairie rattlesnake
		C.v. helleri	Southern Pacific rattlesnake
		C.v. oreganus	Northern Pacific rattlesnake
		C.v. abyssus	Grand Canyon rattlesnake
		C.v. lutosus	Great Basin rattlesnake
		C. cerastes	Sidewinder
		C. ruber	Red diamond rattlesnake
		C. mitchelli	Speckled rattlesnake
		C. lepidus	Rock rattlesnake
		C. tigris	Tiger rattlesnake
		C. willardi	Ridge-nosed rattlesnake
		C. scutulatus	Mojave rattlesnake
		C. molossus	Black-tailed rattlesnake
		C. pricei	Twin-spotted rattlesnake
	Sistrurus		
		S. catenatus	Massasuga rattlesnake
		S. miliarius	Pygmy rattlesnake
	Agkistrodon		
		A. piscivorus	Water moccasin
		A. contortrix	Copperhead
Elapidae		Micruroides euryxanthus	Sonoran (Arizona) coral snake
		Micrurus fulvius	Eastern coral snake

Table 91.3. Poisonous Snakes Indigenous to the United States

The pit vipers have several characteristic features that distinguish them from the nonvenomous snakes (Fig. 91.6): 1) a pit that contains heat-sensitive organs that assist these poor-visioned reptiles to localize their prey is located on each side of the head between the eye and nostril; 2) the pupils are elliptical and vertically oriented in contrast to the usually round pupil of a harmless snake; 3) two curved fangs or hollow maxillary teeth that are 5 to 20 mm long and, in larger snakes, may be spaced as wide as 3 cm are folded posteriorly against the palate and advance forward when the pit viper strikes; 4) the head is relatively more triangular than that of most nonvenomous snakes; and 5) the scutes, or scales, on the ventral portion caudad to the anal plate continue in a single row, whereas nonpoisonous snakes have a cleft, or double row.

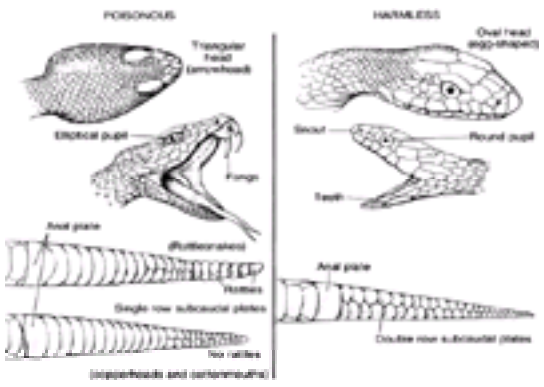

FIGURE 91.6. Comparison of poisonous and nonpoisonous snakes.

The rattlesnake *(Crotalus)* is distributed widely throughout most of the United States and is the culprit in approximately 60% of all pit viper attacks. Several species are notably more menacing and toxic to humans. The large and cold diamondbacks (*Crotalus adamanteus* and *Crotalua atrox*) often stand their ground when approached by humans and inflict most lethal snakebites in North America. Other rattlesnakes that commonly cause the more severe bites include the timber *(Crotalus horridus),* prairie, and pacific *(Crotalus viridus)* rattlesnakes. Several other *Crotalus* species are implicated in less severe human envenomation.

Rattlesnakes vary considerably in size and color, even among species. The eastern diamondback, which inhabits the coastal Southeast, may be as large as 2 m long and 7 kg in weight; it usually has a brightly outlined symmetrical diamond pattern. The timber rattler found in the Northeast and Southeast westward to Texas may be only 1 m long and have nearly black scales (especially in colder climates). Emergency physicians must become familiar with the particular species in their areas.

The pygmy rattler and massasanga are considered rattlesnakes because, in common with *Crotalus* species, they possess a “rattler” on their tail. These two relatively small snakes are members of the genus *Sistrorus,* however, and their bites are not as toxic as those of true rattlesnakes.

The copperhead *(Agkistrodon contortrix)* is a very common poisonous snake that lives in the Southeast and much of the Northeast, extending westward as far as Texas and Nebraska. This snake accounts for approximately 30% of venomous snake bites but, luckily, is seldom a serious threat to life or limb. *A*. *contortrix* is usually 0.6 to 1 m in length and has a light pink to red–brown body with darker brown crossbands shaped like hourglasses. The head has a coppery tinge.

The water moccasin, also known as the cottonmouth *(Agkistrodon piscivorus),* is a semiaquatic pit viper indigenous to the Southeast, including the Gulf states and the Mississippi valley as far north as southern Illinois. These are larger and more belligerent snakes, often traveling with their heads in an aggressive 45-degree angle from the horizontal. Their body is olive brown to black, with darker markings on the sides that often fade over the dorsum. The ventral surface is lighter in color. The oral mucosa is distinctively white, hence the name cottonmouth. Like the copperhead, bites from this species are, in general, less serious than *Crotalus* species.

The relatively passive coral snake is responsible for only 10 to 15 snakebite cases per year in this country. As a member of the *Elapidae* family, it does not share the pit viper's distinctive physical characteristics (i.e., it has round pupils, a blunt head, and ventral caudal scutes, and lack pits). Unlike the nonpoisonous snakes, the coral snake has two small maxillary fangs. The snout of the coral snake is always black and is followed by a yellow ring and subsequent black band. Red and black bands then alternate down the approximately 2-ft length of the coral snake, with narrow yellow rings bordering the red band (Fig. 91.7). The nonvenomous king snake is often confused with the coral; it has red bands directly bordered by black bands. The yellow rings in this snake are within the black bands. The adage about coral snakes holds true:

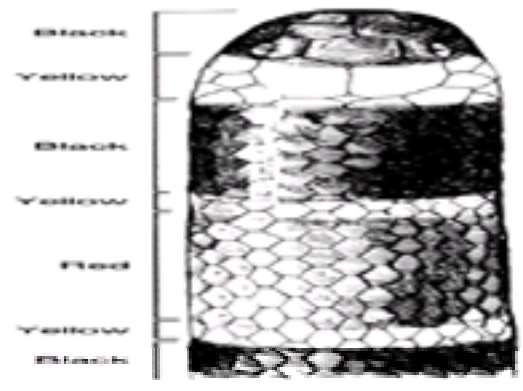

FIGURE 91.7. Coral snake.

Red on yellow, kill a fellow.
Red on black, venom lack.

There are two species of coral snake, the eastern *(Micrurus fulvius)* and the Arizona *(Micruroides euryxanthus). M*. *fulvius* is responsible for most human envenomations and is found in most states east of the Mississippi, with the exception of

the Northeast. *M*. *euryxanthus* is indigenous only to Arizona and New Mexico.

Pathophysiology

Snakebite envenomation is a complex poisoning because of the assorted deleterious effects of venoms and the multiple human and snake variables that influence venom toxicity. Venoms are mixtures of potent enzymes, primarily proteinases, and low-molecular peptides that possess extensive pathophysiologic properties. A crotalid venom often has a combination of necrotizing, hemotoxic, neurotoxic, nephrotoxic, and/or cardiotoxic substances. The neurotoxins make up a large fraction of the venom of the Mojave rattlesnake. These toxins are related to phospholipase A and bind the nicotinic acetylcholine receptors and thus prevent the depolarizing action of acetylcholine. Proteolytic enzymes aided by hyaluronidase cause much of the local tissue destruction. Many venoms induce increased endothelial permeability and venous pooling, creating intravascular depletion. A transient hemoconcentration may be present during this plasma "leak." Hemotoxic effects induce hemolysis, fibrinogen proteolysis, and thrombocytopenia, which, along with activation of plasminogen, can lead to a bleeding diathesis in severe envenomation. Respiratory failure may occur secondary to pulmonary edema or a shock state.

The ultimate toxicity of the snakebite also depends on human and snake variables. Human factors include the victim's size and general health and wound characteristics that affect venom absorption. A small child is more susceptible to a given volume of venom than a larger person. (Unfortunately, young children are commonly bitten more than once.) Fang penetration of a vessel or subfascial compartment ensures a more rapid absorption and serious systemic effects. Likewise, a bite on the head, neck, or trunk (3% of snakebites) hastens systemic absorption. Approximately one-third of snakebites involve the upper extremity and cause a higher long-term functional morbidity than do lower extremity wounds.

Snake variables include the snake's size, the amount of venom injected, and the potency of the particular species' venom. Venom secretion is under voluntary muscular control; any condition that facilitates it (e.g., long, healthy fangs, or full stores of venom) adds to the toxicity of the bite. An angered and hungry rattlesnake unloads more venom than a recently satiated and surprised rattlesnake.

Clinical Manifestations

Pit Viper

Local pain after a *Crotalus* envenomation is typically intense, and a sensation of burning occurs within a couple of minutes. The pain is greater with ensuing edema and presumably increases with larger inocula of venom. Victims of a significant rattlesnake bite often complain within minutes of a perioral numbness, extending to the scalp and periphery. This paresthesia may be accompanied by a metallic taste in the mouth. These patients also may have nausea, vomiting, weakness, chills, sweating, syncope, and other more ominous symptoms of systemic venom absorption. A copperhead or pygmy rattlesnake envenomation produces less local symptoms, and systemic consequences are often minimal or nonexistent unless a small child, multiple bites, or larger than average snake is involved. The water moccasin's effects are more variable.

There is a relative lack of serious pain or swelling with the Mojave rattler bite, although, as in other *Crotalus* bites, the patient may complain of paresthesia in the affected extremity. Within several hours, these patients may develop neuromuscular symptoms such as diplopia, difficulty in swallowing, lethargy, nausea, and progressive weakness from the large portion of neurotoxin in this species.

The wound should be inspected for fang punctures, and if two are present, the distance between them should be noted. An interfang distance of less than 8 mm suggests a small snake; 8 to 12 mm, a medium snake; and more than 12 mm, a larger snake. Fang wounds by small snakes such as the pygmy rattler may be extremely subtle; in larger crotalid snakebites, the fang marks may be hidden within hemorrhagic blebs and edema. Occasionally, only one puncture or two scratches will be present, but both wounds may be potentially venomous. However, not all crotalid bites are envenomated; 10 to 20% of known rattlesnake strikes do *not* inject venom. Other causes of puncture wounds also must be kept in mind—notably rodent bites and thorn wounds. Nonpoisonous snakes sometimes leave an imprint of their two rows of teeth, but the wounds should lack fang puncture marks.

Pit viper envenomations are characterized by intense pain, erythema, and edema at the wound site within 5 to 10 minutes. There may be bloody serosanguinous fluid dripping from the fang punctures. Progressive swelling proportional to the inoculum of venom develops over the next 8 hours and may continue to some degree for an additional 24 hours. Rarely, the venom is deposited predominantly in a muscle compartment, resulting in a deceptively minimal amount of edema. The Mojave rattlesnake bite provides another example of a seemingly innocuous local wound in the setting of a potentially serious envenomation. In a severe diamondback rattlesnake bite, an entire extremity may be swollen within 1 hour.

Local ecchymoses and vesicles usually appear within the first few hours, and hemorrhagic blebs are often present by 24 hours. Lymphadenitis and lymph node enlargement also may become apparent.

Without appropriate therapy, these local manifestations progress to necrosis and may extend throughout the bitten extremity, effectively maiming the victim. Also, as in any animal wound, secondary infection is a risk; the snake's oral flora includes Gram-negative bacteria. Table 91.4 summarizes local characteristics of pit viper bites.

Pain	Ecchymosis
Edema	Vesicles
Erythema	Hemorrhagic blebs

Table 91.4. Local Signs of Pit Viper Envenomation

The dramatic signs of crotalid envenomation are derived primarily from the victim's hypovolemic state, hemorrhagic tendencies, and neuromuscular dysfunction. Table 91.5 outlines the more notable physical signs.

General
Anxiety, diaphoresis, pallor, unresponsiveness
Cardiovascular
Tachycardia, decreased capillary perfusion, hypotension, shock
Pulmonary
Pulmonary edema, respiratory failure
Renal
Oliguria, hemoglobinuria, hematuria
Neuromuscular
Fasciculations, weakness, paralysis, convulsions
Hematologic
Bleeding diathesis

Table 91.5. Systemic Signs of Crotalid (Pit Viper) Envenomation

Coral Snake

Coral snakes leave unimpressive local signs but can neurologically cripple their prey. The bite may have one or two punctures, at most 7 to 8 mm apart, as well as other small teeth marks. There is usually only mild pain and little, if any, swelling. Local wound and, eventually, extremity paresthesia and weakness may be reported. Over several hours, generalized malaise and nausea, fasciculations, and weakness develop insidiously. The patient may complain of diplopia and have difficulty talking or swallowing. Physical examination reveals bulbar dysfunction and generalized weakness. Respiratory failure may ensue.

Management

Pit Viper

As in all medical emergencies, the airway, breathing, and circulation (ABCs) of the patient must be addressed before attending to the snakebite (Fig. 91.8). The first priority of prehospital care of the snakebite victim is rapid transport to a medical facility. Time is of the essence, and all activities in the field must be tempered by this fact.

FIGURE 91.8. Management of pit viper bite. [a]Perform within 5 to 10 minutes of bite; continue suction for 30 to 60 minutes. [b]Complete blood count, platelet count, prothrombin time, partial thromboplastin time, urinalysis, type and hold; in moderate or severe cases, add fibrinogen, arterial blood gases, electrolytes, blood urea nitrogen, and creatinine. [c]Seldom need antivenin; exceptions with large snakes and small children. [d]1:100 dilution if allergy history; saline control; resuscitation medications at hand; antivenin seldom indicated if greater than 12 to 24 hours since bite.

It is important to approach the patient with reassurance and to place him or her at rest. The affected extremity should be stripped of any jewelry or clothing and immobilized in a position of function below the level of the heart. The patient should be kept warm and not allowed anything by mouth.

Tourniquets, inadvertently tightened for prolonged full vascular occlusion, have created more problems than they have solved and therefore cannot be recommended for prehospital care. In experienced hands, however, a *constriction band* that obstructs lymph and venous flow can be valuable when *incision and suction* are indicated or when a long transport is anticipated (longer than 30 to 60 minutes). The band should be at least 2 cm wide and placed 5 to 10 cm proximal to the wound (proximal to the nearest joint if the wound is nearby). The constriction should be loose enough to admit a finger and preserve good distal arterial pulses. Vigilant observation for adequate perfusion is necessary because of progressive edema; the constriction band should be shifted to remain proximal to the swelling. To be effective, the band must be applied initially within 1 hour of the pit viper bite. It may be removed when antivenin therapy is started.

Incision and suction of the pit viper wound are indicated only if they are started within 5 to 10 minutes of the snake's strike. A constriction band should be placed, then liner incisions, approximately 1 × 0.5 cm deep, should be made through the fang marks along the long axis of the extremity, thus avoiding tendons or neurovascular structures. Suction is then applied with a snakebite kit suction cup for the next 30 to 60 minutes. A large syringe with the end cut off also can serve as a suction device. Incision and suction are relatively useless unless initiated within 5 to 10 minutes after the bite occurred. (Note that constriction bands and incision and suction are not recommended in coral snake envenomation.)

In the rare situation in which skilled personnel and supplies are at the scene and a long transport is expected, it would be reasonable to allow one or two attempts at IV access. Many authorities also suggest capturing or killing the snake for later verification, but again, prudence dictates that time not be wasted in this adventure and that an inexperienced person not risk the bite of an agitated snake. If the snake arrives in the ED, treat it with respect—more than one person has been bitten by a “dead” snake, and decapitated snakes can bite reflexly for up to 1 hour.

A complete blood count (CBC), coagulation studies, platelet count, urinalysis, and blood crossmatching should be obtained on all patients with suspected venomous snakebite. (Blood may be difficult to crossmatch after massive hemolysis.) In moderate or severe poisoning, serum electrolytes, BUN, creatinine, fibrinogen, and arterial blood gases also are indicated. Hemolysis, anemia, thrombocytopenia, hypofibrinogenemia, prolonged bleeding times, and metabolic acidosis all may be seen in severe poisoning. Repeat the laboratory studies every 6 hours to ensure no significant changes occur.

Therapy will be based on the clinician's overall grading of venom toxicity. Local and systemic manifestations, as well as laboratory findings, weigh heavily in this judgment. The clinical pattern may change dramatically as the venom's effects unfold; thus, frequent reassessment is crucial. The physician should measure and record the circumference of the injured extremity at the leading point of edema and 10 cm (4 in) proximal to this level every 30 minutes for 6 hours then at least every 4 hours for a total of 24 hours. Table 91.6 is derived from a grading system suggested by the Scientific Review Subcommittee of The American Association of Poison Control Centers.

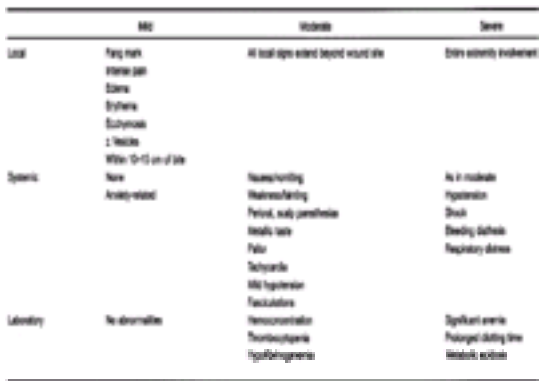

Table 91.6. Grading of Crotalid (Pit Viper) Snakebites

If the history and physical examination on arrival in the ED are consistent with a venomous snakebite, immediate laboratory evaluation and IV access are indicated. Aggressive supportive medical care must be available if signs of major system dysfunction are present. Any prehospital care (e.g., extremity immobilization) should be rechecked. If an occluding tourniquet is inappropriately present, the physician should place a more proximal constriction band and then cautiously remove the tourniquet, being prepared to respond therapeutically to a systemic release of venom.

One antivenin (antivenin crotalidae polyvalent: Wyeth Laboratories) is effective for the rattlesnake, water moccasin, and copperhead. For maximal venom binding, the antivenin should be given within 4 hours of the snake strike. Benefits of antivenin administration after 12 hours are questionable, and use is not indicated after 24 hours (an exception may be continued coagulopathy). The initial recommended dosage varies with the severity of the envenomation. The amount of antivenin is not calculated on a weight basis; children require more than adults as a rule. Dosages in the higher range are used when snake or human variables associated with higher morbidity/mortality are present. For example, a child with two eastern diamondback bites should receive a large dose on the basis of potential severity. On the other hand, a copperhead bite is usually mild and often may be observed for progression without any antivenin given.

Antivenin is highly antigenic horse serum; therefore, skin testing is mandatory (read package insert). Resuscitation equipment, including airways and oxygen, IV epinephrine (1:10,000), antihistamines, and steroids must be kept in close

proximity. The standard skin test involves an intradermal injection of 0.02 mL of 1:10 dilution of reconstituted antivenin. If the history suggests a likely reaction, use a more diluted (1:100 or greater) preparation. A saline control in the opposite extremity is useful for judging a positive-reaction wheal, which is usually apparent within 15 minutes.

If the skin test is negative, the reconstituted antivenin (one vial with 10 mL of saline) is diluted with normal saline in a 1:4 dilution. The antivenin should be infused by IV slowly (1 to 2 mL/hr). During the first 10 to 20 minutes, signs or symptoms of an allergic reaction should be observed for. If no reaction occurs, the rate of infusion should be increased so that the total volume is completed over 2 hours; extremity edema and vital signs should be measured every 15 minutes for evidence of progressive venom toxicity. The initial dose of antivenin should be repeated every 2 hours until the progression of the swelling has stopped. The number of antivenin vials initially anticipated is a rough estimate; more or less antivenin may be required as the clinical reassessment dictates (as many as 75 vials have been used in a child).

Positive skin tests or allergic signs during antivenin infusion warrant consultation with a medical herpetologist. The absolute threat to life must be reassessed, and if present, plans must be made to continue the antivenin. If mild allergic manifestations develop, the infusion should be stopped and diphenhydramine (1 to 2 mg/kg IV) given. Once the allergic symptoms have resolved, a minimum of 5 minutes should pass, then the infusion should be restarted at a slower rate. If symptoms recur, the antivenin should be stopped again; further therapy at this point is controversial. Some authorities recommend an epinephrine drip that is titrated to minimize any allergic phenomena when the antivenin is restarted. If this option is suggested by your consultant, an epinephrine infusion (starting at 0.1 µg/kg per minute) would be a reasonable dosage. Steroids (prednisone 1 to 2 mg/kg per day) are also recommended.

An alternative desensitization method for allergic reactions is described in the product insert but requires at least 3 hours to achieve and thus is not practical in severe envenomations. If life-threatening anaphylaxis occurs, epinephrine 1:10,000 (0.01 mL/kg), diphenhydramine (1 to 2 mg), and steroids (methylprednisolone 2 mg/kg every 6 hours) are given by IV immediately, and other supportive measures are instituted as needed.

Wound care includes irrigation, cleansing, a loose dressing, and consideration of tetanus prophylaxis. The affected extremity should be maintained just below the level of the heart and in a position of function. Cotton padding between swollen digits is useful. Broad-spectrum prophylactic antibiotics are recommended by most authorities. Analgesics for pain may be offered if the cardiorespiratory status is not in question. Surgical excision of the wound, routine fasciotomy, and application of ice are contraindicated. Excision of the wound does not remove significant venom after 30 minutes, and cryotherapy has been associated with increased extremity necrosis and amputations. Fasciotomy should be reserved for the very rare case of a true compartmental syndrome. Necrosis is usually the result of the proteolytic enzymes or inappropriate therapy and is not caused by compartmental pressure. Superficial debridement will be required at 3 to 6 days; a wound care regimen suggested at this stage includes local oxygen, aluminum acetate (1:20 solution) soaks, and triple dye. Physical therapy is beneficial during the healing phase.

The major thrust of supportive care is correction of the intravascular depletion that results from increased venous capacitance, interstitial third spacing, and hemorrhagic losses. Moderate or severe envenomation mandates two IV lines for separate but simultaneous antivenin therapy and volume replacement. Shock usually develops between 6 and 24 hours after the snakebite but may present within the first hour in severe envenomation. Signs of hypovolemia (e.g., decreased capillary perfusion, oliguria, tachycardia, anemia, hypotension) deserve aggressive therapy. Central vascular monitoring and accurate urine output measurements are desirable for optimal therapy. Normal saline or lactated Ringer's solution (20 mL/kg over 1 hour), followed by fresh whole blood or other blood components, often corrects the hypovolemia (see Chapter 3). Vasopressors are usually needed only transiently in the more severe cases. A bleeding diathesis is best managed with fresh whole blood, or blood component therapy, primarily packed cells (10 mL/kg), and fresh-frozen plasma (10 mL/kg). With life-threatening bleeding, platelets (0.2 units/kg) and a more concentrated fibrinogen source (cryoprecipitate—dose 1 bag/5 kg body weight) also should be considered. Abnormal clotting parameters, including fibrinogen and platelet and blood counts, should be reevaluated every 4 to 6 hours. Respiratory support also is commonly required when shock has developed. Renal failure is another potential problem in this setting.

As many as 75% of antivenin recipients may develop a serum sickness syndrome approximately 4 days to 3 weeks after treatment. Serum sickness is almost ensured with doses greater than seven vials of antivenin. Rashes, arthralgias, edema, malaise, lymphadenopathy, fever, and/or gastrointestinal symptoms evolve over several days. High-dose prednisone (2 mg/kg per day, maximum 80 mg) until symptoms abate (and then a tapering schedule) has been used with success in most cases. Diphenhydramine (5 mg/kg per day) is often given as an adjunct.

Coral Snake

When coral snake wounds are present or the history or specimen is consistent with an eastern coral snakebite, antivenin for *Micrurus fulvius* (Wyeth) is administered before development of further symptoms. This is also an equine serum and requires preliminary skin testing (see package insert). The initial recommended dosage is three to five vials by IV; an additional three to five vials may be given as needed for signs of venom toxicity. There is no antivenin available for the Arizona coral snake *(Micruroides euryxanthus).* Supportive care should provide a satisfactory outcome in these cases. Constriction bands, suction and drainage, and other local measures do not retard coral snake venom absorption and, hence, are not indicated.

Exotic Snakes

The clinician confronted with an exotic snakebite or a clinician inexperienced in snakebites should consult a local medical herpetologist, poison control center, or the Oklahoma Poison Control Center (405-271-5454), which indexes the availability of unusual antivenins. Report all illegally possessed reptiles to the police or to the appropriate Fish and Game Agency.

Mammalian Bites

Background

Children suffer the majority of casualties in this country's growing epidemic of mammalian bites. The overall morbidity of mammalian bites is staggering in terms of infectious complications, cosmesis, disability, psychological trauma, and medical expenses. At least 1 million people are bitten each year, and about 1% of ED visits are prompted by bite wounds.

Dog bites account for the overwhelming majority (80 to 90%) of these injuries and thus have been the subject of numerous investigations. The most common dog attack involves a 5- to 14-year-old boy close to home and a large-breed or mixed-breed canine. The dog's owner can be identified 85 to 90% of the time; in fact, in 15 to 30% of cases, the dog belongs to the victim's family. Usually, the dog has never previously bitten anyone and often has been provoked, although unintentionally. However, animal jealousy has been implicated in unprovoked biting of infants. A seldom realized mortality of approximately 10 cases, primarily young children and infants, occurs each year in the United States.

The remainder of the mammalian bites are perpetrated by cats (5 to 10%), rodents (2 to 3%), and other wild or domesticated animals. Another mammalian species, *Homo sapiens,* inflicts approximately 2 to 3% of the bite wounds that present to medical attention. Human bites share with cat wounds a notoriously high infectious complication rate.

Pathophysiology

Anatomic wound characteristics and the microbiologic inoculum of the offending species determines the pathologic consequences of the bite. For example, many dog bites are localized crush injuries with a substantial amount of infection-prone devitalized tissue—a result of the enormous pressures dogs generate in their bites. Forces of 200 to 400 pounds per square inch, sufficient to perforate sheet metal, have been documented in dogs. Overall, however, only 5 to 10% of dog bites becomes infected, probably because the resulting lacerations and abrasions are so accessible to good wound hygiene. The typical feline bite, however, is a deep puncture wound and is difficult to irrigate or cleanse, thus subjecting it to a high infection rate (up to 50% in some series). The penetration of tendons, vessels, facial compartments, and bones also has a high infectious risk with increased morbidity. The hand offers all these anatomic components in a relatively small cross-sectional area and therefore is potentially prone to serious infections regardless of the biting species. Injuries to deeper structures, including major vasculature, the brain, peritoneum, abdominal organs, and airways also have been sporadically reported.

Scores of aerobic and anaerobic bacteria indigenous to mammalian oral flora are inoculated into the wound during biting. Cultures of fresh wounds before clinical infection reflect the variety of these contaminants but are not predictive of the causative organism in later infections. The most commonly isolated bacteria in infected cat and dog bite wounds are *Staphylococcus aureus* and *Pasteurella multocida,* a Gram-negative rod. In one series, *P. multocida* was found in 50 and 80% of infected dog and cat wounds, respectively. Other series have isolated *P. multocida* in only 10 to 20% of infected dog wounds and incriminated other more common bacteria: streptococci, coagulase-negative staphylococci, *S. aureus,* and enteric bacteria. Some unusual isolates in clinical infection have included unnamed bacteria—Center for Disease Control and Prevention types II-J and EF-4. At least 25% of all infected mammalian bites yield mixed cultures of aerobes and often anaerobes, if carefully sought. Anaerobic bacteria usually are recovered (not alone but only in mixed cultures with aerobes).

Human bite infections are mixed bacterial infections, with *Streptococcus viridans* or *S. aureus* being the predominant organism. Anaerobic bacteria, especially *Bacteroides* and *Peptostreptococcus* species, are commonly cultured. The more serious morbidity in infected human bites of the hand has been correlated with *S. aureus* isolation and, more recently, with *Eikenella corrodens*, a facultative anaerobe.

Finally, the multiple systemic diseases that may be transmitted by mammalian bites need to be considered.

Clinical Manifestations

Mammalian bite wounds cause a spectrum of tissue injuries from trivial to life-threatening. Scratches, abrasions, contusions, punctures, lacerations, and their complications are seen commonly in the ED. The complications usually involve secondary infections or damage to structures that underlie the bite.

Although dog bites are insignificant lesions in at least half of the cases that come to medical attention, 5 to 10% warrant suturing and 2%, hospital admission. Approximately 33% of dog bites involve the upper extremity, presumably while the victim is fending off the attack. Another 33% of wounds are located on the lower extremities and 20% on the head and neck area. The remainder of bites involves multiple areas. Other than bites on the hand, the rate of secondary infection in dog bites given good local care approximates that of nonbite wounds.

Predictably, young children suffer more serious canine injuries. The dog strikes the head and neck in 60 to 70% of victims less than 5 years old and in 50% those less than 10 years old. These wounds most often involve the lips, nose, and cheek areas, and on rare occasions, they penetrate the skull, with resulting depressed skull fractures and intracranial lesions. It has been estimated that each year 44,000 children suffer facial dog bite wounds, one-third of them requiring complex repair. The uncommon life-threatening injuries occur almost exclusively in young children and include major vascular injury, visceral penetration, and chest trauma. The deaths are usually secondary to acute hemorrhagic shock.

Cat bites are located in the infection-prone upper extremities in two-thirds of cases and usually are puncture wounds rather than lacerations or contusions. Infections that complicate these wounds result from the same organisms isolated in dog bites, but a higher incidence of *P. multocida* is found. *P. multocida* infections characteristically present within 12 to

24 hours of the injury and rapidly display erythema, significant swelling, and intense pain. These infections often respond slowly to adequate drainage and antibiotic therapy. Local infections from other organisms usually present 24 to 72 hours after the bite in a less fulminant manner. Viridans streptococcal infections are occasional exceptions to this generalization and may resemble a *P. multocida* clinical course.

Cat scratches are most commonly located on the victim's upper extremities or periorbital region and are more likely to develop secondary bacterial infection than scratches from the other common domesticated species. Corneal abrasions also are occasionally associated with the periorbital wounds. Cat-scratch disease, an uncommon complication of these injuries, is characterized by a papule at the scratch site and a subsequent regional lymphadenitis. The primary lesion is typically a crusted, erythematous papule, 2 to 6 mm in diameter, that develops 3 to 10 days after the scratch. A tender regional lymphadenopathy occurs 2 weeks after the primary lesion. Malaise and fever are associated symptoms in approximately 25% of patients. Unusual manifestations of this disease include encephalopathy, exanthem, atypical pneumonia, and parotid swelling. The disease is self-limiting, with resolution of the symptoms within 2 to 3 months. *Bartonella henselae* is considered to be the causative organism in most cases. An indirect fluorescent antibody test to *Bartonella* is useful in the diagnosis and is available through the Centers for Disease Control and Prevention.

Human bites in older children and adolescents are most commonly incurred when a clenched fist strikes the teeth of an adversary. The wound typically overlies the metacarpal-phalangeal joint, and on relaxation of the fist, the bacterial inoculum penetrates more deeply into the relatively avascular fascial layers. Hand infections, regardless of infection site, usually present with mild swelling over the dorsum of the hand 1 to 2 days after injury. Infected hand bites may be superficial and localized to the wound, but if there is pain with active or passive finger motion, a more serious deep compartmental infection or tendonitis should be suspected. Osteomyelitis occasionally occurs in hand infections. In younger children, human bites are more often on the face or trunk than on the hands. Often, a playmate inflicts the wound, but child abuse must always be considered. Systemic diseases that may be spread by human bites include hepatitis B and syphilis.

Rodent bites usually occur in disadvantaged socioeconomic groups or among laboratory workers and have a relatively low incidence of secondary infection (10%). Ratbite fever is a rare disease that may present after a 1- to 3-week incubation period with chills, fever, malaise, headache, and a maculopapular or petechial rash. There are two forms: Haverhill fever *(Streptobacillus moniliformis)* and Sodoku *(Spirullum minus),* both of which are responsive to IV penicillin.

Another uncommon bacterium for which lagomorphs, particularly rabbits, are hosts is *Francisella tularensis.* Tularemia is usually spread to humans by rabbit bites, although contact with or ingestion of contaminated animals or insect vectors is sufficient for transmission. Ulceroglandular tularemia is the most common form of the disease, and streptomycin is the agent of first choice in its treatment.

Serious infections from multiple bacteria, including osteomyelitis, sepsis, endocarditis, and meningitis, have been reported as complications of mammalian bite wounds as well as the more esoteric diseases already mentioned. The risk of rabies or tetanus always must be considered in animal bites.

Management

Meticulous and prompt local care of the bite wound is the most important factor in satisfactory healing and prevention of infection. In more extensive wounds, local anesthesia is achieved before wound hygiene. Then, the skin surrounding the wound should be cleaned with a soft sponge and 1% providone iodine solution can remove obvious contaminants. The wound itself should be forcefully irrigated with a minimum of 200 mL normal saline. A 19-gauge needle or catheter attached to a 30-mL syringe will supply sufficient pressure for wound decontamination and will decrease the infection rate by twentyfold. Stronger irrigant antiseptics—povidone–iodine scrub preparation, 20% hexachlorophene, alcohols, or hydrogen peroxide—may damage wound surfaces and delay healing. Soaking in various preparations has not proved helpful in reducing infections.

Most open lacerations from mammalian bites can be sutured if local care is effected within several hours of the injury and good surgical technique is used. Facial wounds often mandate primary closure for cosmetic reasons and, overall, are low infection risks because of the good vascular supply. In fact, Callaham has demonstrated a lower infection rate in sutured dog bite wounds than in those left open. The exceptions to suturing are minor hand wounds and other high-risk bites. In large hand wounds, hemorrhage should be carefully controlled. We suggest closing the subcutaneous dead space in these wounds with a minimal amount of absorbable suture material. Cutaneous sutures can be placed after 3 to 5 days if there is no evidence of infection.

Extremities with extensive wounds should be immobilized in a position of function and kept elevated as much as possible. This is especially true of hand wounds, which should have bulky mitten dressings and be supported by an arm sling. All significant wounds should be rechecked in follow-up in 24 to 48 hours.

The following wounds may be considered at high risk for infection: puncture wounds, minor hand or foot wounds, wounds given initial care after 12 hours, cat or human bites, and wounds in immunosuppressed patients. As a rule, these wounds should not be sutured, and prophylactic antibiotics are indicated. No single antibiotic is ideal for all of the most common organisms involved in infected mammalian bite wounds. Amoxicillin–clavulanic acid (Augmentin) comes close. It is effective in *P. multocida, Streptococcus, Staphylococcus,* and anaerobe control, as well as in providing staphylococcus coverage. Combination therapy with phenoxymethyl penicillin (penicillin V) and cephalexin or dicloxacillin has been suggested by some authorities. For high-risk wounds, we recommend amoxicillin–clavulanic acid (30 to 50 mg/kg per day) alone for initial therapy. Erythromycin (40 mg/kg per day) is an alternative for the penicillin-allergic patient. The initial dosage of antibiotic should be given in the ED and continued for the next 3 to 5 days. It must be emphasized that local care ultimately prevents infection more effectively than any prophylactic antibiotics. Studies indicate that prophylactic oral antibiotics for low-risk dog bite wounds are not indicated because the differences in the rate of infection

are not significant and the cost benefit ratio not worth the risks of allergic reactions.

Any bite wound with signs of infection deserves aggressive drainage and debridement and antibiotic therapy after aerobic and anaerobic cultures are obtained. Moderate to severe hand infections or other wounds that involve deep structures usually require debridement and exploration under general anesthesia. Culture swabs should sample the depth of the wound; or, in cases of cellulitis, the specimen can be collected by needle aspiration of the leading edge of erythema. While awaiting cultures, a Gram stain is often helpful in differentiating the probability of staphylococci or streptococci from *P. multocida.*

Parenteral antibiotics and admission to the hospital are indicated if the child has systemic symptoms or has wounds with potential functional or cosmetic morbidity. The choice of parenteral antibiotics should be governed by the same factors considered in selection of prophylactic antibiotics and then modified by culture results.

Tetanus immunization status should be checked in every injury that violates the epidermis, regardless of the cause. Recommendations for tetanus immunoglobulin and immunization are noted elsewhere.

Concern for rabies is the factor that prompts many patients to seek medical care. Although the incidence of rabies (1 to 5 cases per year) is extremely low, the physician must always assess the possibility of rabies exposure and promptly initiate prophylaxis when indicated. The history should include the apparent health of the animal and any provocation for attack. Wild carnivores and bats generally should be regarded as rabid; rodents (rats, squirrels) and lagomorphs (rabbits) can usually be considered no risk. Exposure to bats even without bite or scratch should warrant serious consideration of prophylaxis. Rabies prophylaxis is not indicated in bites by a healthy dog or cat with a known owner, assuming the animal's health does not deteriorate over the following 10 to 14 days. Bites by strays and other domesticated mammals should be considered individually and with consultation of the local health department. Scratches, abrasions, and animal saliva contact with the victim's mucous membranes all are capable of rabies spread.

If postexposure antirabies immunization is indicated both passive antibody (RIG) and vaccine (HDCV) should be given. Immunization with RIG (rabies immune globulin, human) is administered only once, in a dose of 20 IU/kg. Half the dose is given intramuscularly, and the remainder is infiltrated locally around the wound. The HDCV immunization (human diploid cell rabies vaccine) should be administered on days 0, 3, 7, 14, and 28 for a total of five doses, each 1.0 mL given intramuscularly.

Suggested Readings

GENERAL

Auerbach PS. Marine envenomation. N Engl J Med 1991;325:486–493.

Auerbach PS, ed. Wilderness Medicine. 3rd ed. St Louis: Mosby, 1995.

Ennik F. Death from bites and stings of venomous animals. West J Med 1980;133:463–468.

Halstead BW. Poisonous and Venomous Marine Animals of the World. Princeton: Darwin Press, 1978.

Halstead BW, Vinci JM. Venomous fish stings. Clin Dermatol 1991;5:29–35.

McGoldrick J, Marx JA. Marine envenomations. Part 1: vertebrates. J Emerg Med 1991;9:497–502.

McGoldrick J, Marx JA. Marine envenomations. Part 2: invertebrates. J Emerg Med 1992;10:71–77.

COELENTERATA

Auerbach PS. Hazardous marine animals. Emerg Med Clin North Am 1984;2:531–544.

Burnett JW, Rubinstein H, Callon GJ. First aid for jellyfish envenomation. South Med J 1983;76:910–912.

Guess HA, Sauiteer PL, Morris C. Hemolysis and acute renal failure following a Portuguese man-of-war sting. Pediatrics 1982;70:979–981.

Koffman MB. Portuguese man-o'-war envenomation. Pediatr Emerg Care 1992;8:27–28.

Stein MR, Marraccini JV, Rothchild NE, Burnett JW. Fetal Portuguese man-o'-war envenomation. Ann Emerg Med 1989;18:312–315.

Turner B, Sullivan P. Disarming the blue bottle: treatment of Physalia envenomation. Med J Aust 1980;2:394–395.

ECHINODERMATA

Baden HP, Burnett JW. Injuries from sea urchins. South Med J 1977; 70:459–460.

Beeching N, Morgan HV. Sea-urchin granuloma of the toe. Practitioner 1982;226:1567–1571.

STINGRAYS

Bitseff EL, Garoni WJ, Hardison CD, Thompson JM. The management of stingray injuries of the extremities. South Med J 1970;63:417–418.

Halstead BW. Poisonous and Venomous Marine Animals of the World. Princeton: Darwin Press, 1978.

Kizer KW. Marine envenomations. J Toxicol Clin Toxicol 1983–1984; 21:527–555.

Russell FE. Stingray injuries: a review and discussion of their treatment. Am J Med Sci 1953;226:611–622.

SHARKS

Auerbach PS. Hazardous marine animals. Emerg Med Clin North Am 1984;2:531–544.

Baldridge HD, Williams J. Shark attack: feeding or fighting? Mil Med 1969;134:130–133.

White JAM. Shark attack in Natal. Injury 1976;6:191–194.

SCORPAENIDAE

Kizer KW, McKinney HE, Auerbach PS. *Scorpaenidae* envenomation: a five-year poison center experience. JAMA 1985;253:807–810.

CATFISH

Baack BR, Kucan JO, Zook EG, Russel RC. Hand infections secondary to catfish spines: case reports and literature review. J Trauma 1991;31:1432–1436.

Zeman MG. Catfish stings: report of three cases. Ann Emerg Med 1989; 18:211–213.

ARTHROPODA

Berg RA, Tarantino MD. Envenomation by the scorpion *Centruroides exilicauda (C. sculpturatus):* severe and unusual manifestations. Pediatrics 1991;87:930–933.

Berger RS. Management of brown recluse spider bite. JAMA 1984;251:889.

Berger RS. The unremarkable brown recluse spider bite. JAMA 1973;225:1109–1111.

Bond GR. Antivenin administration for *Centruroides* scorpion sting: risks and benefits. Ann Emerg Med 1992;21:788–791.

Clark RF, Wethern-Kestner S, Vance MV, Gerkin R. Clinical presentation and treatment of black widow spider envenomation: a review of 163 cases. Ann Emerg Med 1992;21:782–787.

DeShazo RD, Butcher BT, Banks WA. Reactions to the stings of the imported fire ant. N Engl J Med 1990;323:462–466.

Eitzen EM, Seward PN. Arthropod envenomations in children. Pediatr Emerg Care 1988;4:266–272.

Gendron BP. *Loxosceles reclusa* envenomation. Am J Emerg Med 1990; 8:51–54.

Ginsburg CM. Fire ant envenomation in children. Pediatrics 1984;73: 689–692.

Gorman RJ, Snead OC. Tick paralysis in three children. Clin Pediatr 1978;17:249–251.

Haller JS, Fabara JH. Tick paralysis. Am J Dis Child 1972;124:915–917.

Hobbs GP, Anderson AR, Green TJ, Yealy DM. Comparison of hyperbaric oxygen and dapsone on therapy for *Loxosceles* envenomation. Acad Emerg Med 1996;3:758–761.

Hunt GR. Bites and stings of uncommon arthropods. Parts I and II. Postgrad Med 1981;70:91–102, 107–114.

Iserson KV. Methemoglobin from dapsone therapy for suspected brown spider bite. J Emerg Med 1985;3:285–288.

Jerrard DA. ED management of insects stings. Am J Emerg Med 1996; 14:429–433.

Key GF. A comparison of calcium gluconate and methocarbamol (Robaxin) in the treatment of latrodectism (black widow spider envenomation). Am J Trop Med Hyg 1981;30:273–276.

King LE, Rees RS. Dapsone treatment of a brown recluse bite. JAMA 1983;250:648.

Kunkel DB. Arthropod envenomations. Emerg Med Clin North Am 1984; 2:579–586.

Likes K, Banner W, Chavez M. *Centruroides exilicauda* envenomation in Arizona. West J Med 1984;141:634–637.

Milne M, Milne L. Audubon Society Field Guide to North American Insects and Spiders. New York: Knopf, 1980.

Needham G. Evaluation of five popular methods for tick removal. Pediatrics 1985;75:997–1002.

Phillips S, Kohn M, Baker D, et al. Therapy of brown spider envenomation: a controlled trial of hyperbaric oxygen, dapsone and cyproheptadine. Ann Emerg Med 1995;25:363–368.

Rachesky IJ, Banner W Jr, Dansky J, Tong T. Treatments for *Centruroides exilicauda* envenomation. Am J Dis Child 1984;138:1136–1139.

Rauber AP. Black widow spider bites. J Toxicol Clin Toxicol 1983;84:473–485.

Rees R, Campbell D, Rieger E, King LE. The diagnosis and treatment of brown recluse spider bites. Ann Emerg Med 1991;16:945–949.

Rees R, Shack RB, Withers E, Madden J, et al. Management of the brown recluse spider bite. Plast Reconstr Surg 1981;68:768–773.

Reeves JA, Allison EJ JR, Goodman PE. Black widow spider bite in a child. Am J Emerg Med 1996;14:469–471.

Rimsza ME, Zimmerman DR, Bergesson PS. Scorpion envenomation. Pediatrics 1980;66:298–302.

Valentine MD, Lichtenstein LM. Anaphylaxis and stinging insect hypersensitivity. JAMA 1991;258:2881–2885.

Visscher PK, Veller RS, Camazine S. Removing bee stings. Lancet 1996; 348:301–302.

Wingert, WA. Envenomation: snakes and spiders. Audio-Digest Emerg Med 1986;3:10.

Woestman R, Perkin R, Van Stralen D. The black widow: is she deadly to children? Pediatr Emerg Care 1996;12:360–364.

Wong RC, Hughes SE, Voorhess JJ. Spider bites. Arch Dermatol 1991; 123:98–104.

Wright SW, Wrenn KD, Murray L, Segar D. Clinical presentation and outcome of brown recluse spider bite. Ann Emerg Med 1997;30:28–32.

VENOMOUS REPTILES

Burch JM, Agarwal R, Mattox KL, et al. The treatment of crotalid envenomation without antivenin. J Trauma 1988;28:35–43.

Burgess JL, Dart RC, Egen NB, Mayersohn M. Effects of constriction bands on rattlesnake venom absorption: a pharmacokinetic study. Ann Emerg Med 1992;21:1086–1093.

Cruz NS, Alvarez RG. Rattlesnake bite complications in 19 children. Pediatr Emerg Care 1994;10:30–33.

Davidson TM, Schafer SF. Rattlesnake bites: guidelines for aggressive treatment. Postgrad Med 1994;96:107–114.

Lawrence WT, Giannopoulos A, Hansen A. Pit viper bites: rational management in locales in which copperheads and cottonmouths predominate. Ann Plastic Surg 1996;36:276–285.

Picchioni AL, Russell FE, Hardy DL, Kunkel DB. Management of poisonous snakebite. Vet Hum Toxicol 1984;26:139.

Russell FE. Snake Venom Poisoning. Great Neck, NY: Scholium International, 1983.

Russell FE, Picchioni AL. Snake venom poisoning. Clin Tox Con 1983;5:73.

Russell FE, Carlson RW, Wainschel J, Osborne AH. Snake venom poisoning in the United States: experiences with 550 cases. JAMA 1975; 233:341.

Stewart ME, Greenland S, Hoffman JR. First-aid treatment of poisonous snakebite: are currently recommended procedures justified? Ann Emerg Med 1981;10:331–335.

Watt CH. Poisonous snakebite treatment in the United States. JAMA 1978;240:654–656.

Weber RA, White RR. Crotalidae envenomation in children. Ann Plast Surg 1993;31:141–145.

Whitley RE. Conservative treatment of copperhead snakebites without antivenin. J Trauma 1996;41:219–221.

Wingert WA, Chan L. Rattlesnake bite in Southern California and rationale for recommended treatment. West J Med 1988;148:37–44.

Wingert WA, Wainschel J. Diagnosis and management of envenomation by poisonous snakes. South Med J 1975;68:1015.

MAMMALIAN BITES

Aghababian RV, Conte JE Jr. Mammalian bite wounds. Ann Emerg Med 1980;9:79.

Baker MD, Moore SE. Human bites in children. Am J Dis Child 1991;141:1285–1291.

Boenning DA, Fleisher GR, Campos J. Dog bites in children: epidemiology, microbiology and penicillin prophylactic therapy. Am J Emerg Med 1983;1:17.

Brogan TV, Bratton SL, Dowd MD, Hegenbarth MA. Severe dog bites in children. Pediatrics 1995;96:947–950.

Callaham M. Treatment of common dog bites: infection risk factors. J Am Coll Emerg Phys 1978;7:83.

Callaham M. Human and animal bites. Top Emerg Med 1982;4:1.

Callaham M. Prophylactic antibiotics in dog bite wounds: nipping at the heels of progress. Ann Emerg Med 1994;23:577–579.

Centers for Disease Control and Prevention. Rabies prevention. MMWR 1980;29:265.

Centers for Disease Control and Prevention. Dog bite related fatalities— U.S. 1995–1996. MMWR 1997;46:463–467.

Cummings P. Antibiotics to prevent infections in patients with dog bite wounds: a meta-analysis of randomized trials. Ann Emerg Med 1994;23:535–540.

Dire DJ. Cat bite wounds: risk factors for infection. Ann Emerg Med 1991;20:973.

Dire DJ. Emergency management of dog and cat bite wounds. Emerg Med Clin North Am 1992;10:719–736.

Dire DJ, Hogan DE, Walker JS. Prophylactic oral antibiotics for low risk dog bite wounds. Pediatr Emerg Care 1992;8:194–199.

Falling Centers for Disease Control. Dog bite related fatalities in the US. 1995–1996. MMWR 1997;46:463–467.

Karlson TA. The incidence of facial injuries from dog bites. JAMA 1984; 251:3265.

Kizer KW. Epidemiologic and clinical aspects of animal bite injuries. J Am Coll Emerg Phys 1979;8:134.

Lindsey D, Christopher M, Hollenbach J, et al. Natural course of the human bite wound: incidence of infection and complications in 434 bites and 803 laceration in the same group of patients. J Trauma 1991;27:45.

Malinowski RW, Strate RG, Perry JF Jr, Fischer RP. The management of human bite injuries of the hand. J Trauma 1979;19:655.

Mann JM. Systematic decision-making in rabies prophylaxis. Pediatr Infect Dis 1982;2:162.

Marcy SM. Infections due to dog and cat bites. Pediatr Infect Dis 1982; 1:351.

Peeples E, Boswick JA Jr, Scott FA. Wounds of the hand contaminated by human or animal saliva. J Trauma 1980;20:383.

Pinckney LE, Kennedy LA. Traumatic deaths from dog attacks in the United States. Pediatrics 1982;69:193.

Rosen RA. The use of antibiotics in the initial management of recent dog-bite wounds. Am J Emerg Med 1985;3:19.

Schweich P, Fleisher G. Human bites in children. Pediatr Emerg Care 1985;1:51.

Wear DT, Margileth AM, et al. Cat scratch disease: a bacterial infection. Science 1983;221:1403.

Wiseman NE, Chochinov H, Fraser V. Major dog attacks in children. J Pediatr Surg 1983;18:533.

Zook EG, Miller M, Van Beek AL, Wavak P. Successful treatment protocol for canine fang injuries. J Trauma 1980;20:243.

CHAPTER 92

Allergic Emergencies

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ASTHMA

Background

Asthma is the most common chronic disease of childhood, affecting approximately 5 to 10% of children. Many of these children, particularly those who are economically disadvantaged, are treated in the emergency department (ED) only during acute exacerbations and receive no other ongoing, consistent care. This places the additional burden on the emergency physician for facilitating appropriate follow-up care as well as for managing acute exacerbations.

There has been a dramatic increase in hospitalizations for asthma since the 1960s, despite only modest increases in prevalence. More alarming is the increase in asthma-related deaths in children over the past decade despite several "advances" in therapy. The reasons for this apparent increase in the severity of childhood asthma is a subject of active investigation and has not been fully elucidated. However, socioeconomic factors appear to play a major role. Risk factors for life-threatening asthma have been identified and are listed in Table 92.1. Unfortunately, despite these alarming statistics, patients, parents, and physicians often grossly underestimate the life-threatening potential of asthma.

Previous life-threatening exacerbation	Lack of access to emergency medical care
>2 hospital admissions within past year	History of noncompliance and/or medication abuse
>3 visits to the emergency department within past year	Psychological and/or psychosocial problems
Inadequate general medical management	

[a]Note: Although these factors are associated with life-threatening disease in studies of large numbers of patients, individual children with some of these findings may be appropriately discharged after treatment.

Table 92.1. Risk Factors for Life-Threatening Asthma[a]

The approach to the assessment and management of acute asthma has undergone many changes in recent years and remains controversial. This chapter presents an approach based on current knowledge and is intended to provide a set of guidelines that can be adapted, as needed, to meet the needs of the individual patient or ED setting. The approach is in general agreement with the Practical Guide for the Diagnosis and Management of Asthma developed by the Expert Panel 2 Report of the National Asthma Education Program and the National Heart, Lung, and Blood Institutes, published in 1991 and updated in 1997. Areas of controversy and new therapies under investigation also are discussed.

Pathophysiology

Over the past decade, research has further refined the definition of asthma to accentuate the chronic inflammatory nature of the illness. Therapy must ideally reverse the airway narrowing as well as block or modify the impact of the cellular role of mast cells, eosinophils, macrophages, neutrophils, T lymphocytes, and epithelial cells.

The two most important components that impact on bronchospasm in asthma occur through immunoglobulin E (IgE)-mediated mast cell degranulation and the recruitment of other cellular components that contribute to the "late-phase" reaction and the more inflammatory aspect of the disease. Clinically, this separates the immediate redirection in pulmonary function due to antigen stimulation in the first hours from a later redirection beginning at 4 to 12 hours associated with cellular recruitment. Key to this late-phase response is the attraction and impact of inflammatory cells with T-helper lymphocytes contributing greatly. Release of leukotrienes, granular proteins, and other mediators cascade this process.

In patients with hyperreactive airways, it is clear that viral infection, including respiratory syncytial virus (RSV) and rhinovirus, increase the responsiveness of the airways and bronchial inflammation probably by cytokine-triggered upregulation.

The physiologic consequences of asthma are progressive air trapping with dead space ventilation, increased airway resistance, and mismatching of alveolar ventilation-perfusion. As the acute episode progresses, there is further decrease in forced vital capacity (FVC), forced expiratory volume at one second (FEV_1), and peak expiratory flow rate (PEFR). Each breath is initiated at higher lung volumes that lie on the steep, stiff portion of the compliance curve. At this point, high-pressure changes are required to achieve acceptable tidal volumes. Arterial oxygen saturation decreases with ventilation-perfusion abnormalities. Hypoxemia may cause pulmonary artery hypertension and hyperventilation. Hyperventilation occurs early in the course of acute asthma, out of proportion to any respiratory or metabolic demands. It may be caused by stimulation of pulmonary receptors innervated by the vagus nerve through inflammatory irritation or by stretch receptor activity. If the acute asthmatic process continues unchecked, minute ventilation cannot be maintained and $PaCO_2$ ultimately rises as the child becomes fatigued.

Metabolic changes also occur with acute asthma. The increased work of breathing increases oxygen and energy consumption, leading to a metabolic acidosis. Compensation for this acidosis may not be possible because of already maximal respiratory effort. As respiratory muscles fatigue, respiratory acidosis develops. Combined respiratory and metabolic acidosis sets the stage for respiratory failure.

Although the pathophysiology of acute asthma in children and adults is similar, young children are particularly susceptible to status asthmaticus. In the child less than 5 years of age, peripheral airway resistance is substantially higher than in an adult. A small degree of narrowing of peripheral airways results in disproportionate increases in resistance to air flow. Respiratory reserve in children is limited and increases with age as the size of the conducting airways grow larger. Part of this is related to the respiratory surface area being much smaller in the child less than 5 years old than in the adult. Another age-related mechanical factor, decreased elastic recoil, contributes to earlier small airway closure during normal tidal breathing in young children. This leads to airway collapse and atelectasis during asthma exacerbations or respiratory infection. The horizontal insertion of the diaphragm in infancy makes diaphragmatic recruitment during airway obstruction less efficient. In infants and young children, the contribution of bronchial smooth-muscle constriction to airway obstruction may be less important than edema, mucous plugging, hypersecretion, and atelectasis.

Triggers

Triggers for acute asthma attacks may be nonspecific and include acute viral infection, allergy, weather change, cigarette smoke or other inhaled irritants, exercise, and cold air. Low-level infections in the upper airways, such as sinusitis and otitis media (OM), have been implicated in exacerbations of previously stable asthma in children. Allergic reactions not only trigger acute episodes of asthma but also can induce a state of bronchial lability. Allergy may promote acute attacks through immediate mediator release and by chronic airway obstruction through the late-phase inflammatory response. Drug sensitivity, particularly to aspirin products, may induce hyperreactive airways.

Clinical Findings

In the child with known asthma and an obvious exacerbation, clinical evaluation and initial treatment should begin almost simultaneously. However, for the purposes of clarity, these phases of management are considered in sequence.

The evaluation of the child with wheezing begins with a rapid cardiopulmonary assessment to determine the severity of the episode. This includes the child's general appearance, paying particular attention to the degree of respiratory distress, color, and mental status. Simultaneously, oxygen is administered, and if indicated, immediate resuscitative and therapeutic efforts are initiated, as described in the next section. After a baseline evaluation, the child should be reassessed frequently and, ideally, after each therapeutic intervention.

In addition to the history and physical examination, other selected studies may be useful in the assessment of acute asthma. These studies include PEFR, pulse oximetry, arterial blood gases (ABGs), chest radiograph, and other laboratory evaluations. Each aspect of the assessment is discussed in more detail later in this chapter. A general guide to several parameters useful for estimating the severity of the episode can be found in Table 92.2.

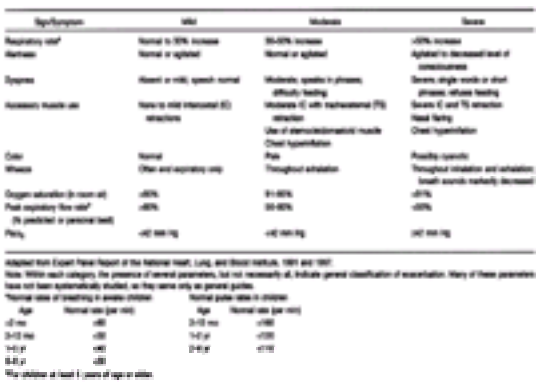

Table 92.2. Estimation of Severity of Acute Asthma Exacerbation

History

The history should include the duration of the current episode and rapidity of onset, the parent's or patient's subjective assessment of severity, other associated symptoms, and the suspected trigger. The child's current medications should be determined, including details of when the medications were started, the dosage, the route, the timing of the last dose, and a history of missed doses from noncompliance or emesis. A history of previous medications, particularly oral or inhaled steroids in the past 6 months, should be elicited. An attempt should be made to estimate the child's severity of disease and risk for a life-threatening episode. Details about past episodes, such as frequency of ED visits, hospital admissions, intensive care unit (ICU) admissions, and episodes of respiratory failure that required mechanical ventilation, should be addressed. For children with their first episode of wheezing, the possibility of a foreign body aspiration or another cause of wheezing should be explored (Table 92.3). A personal or family history of atopic disease is suggestive of asthma. Current hydration status should be evaluated with questions regarding recent fluid intake, emesis, and urine output. The family's ability to cope with the child's disease at home should be assessed in preparation for making a disposition decision.

Congenital	*Allergic*
Cystic fibrosis	Asthma
Lobar emphysema	Anaphylaxis
Tracheobronchomalacia	Allergic pulmonary aspergillosis
Tracheal stenosis	*Acquired*
Bronchial stenosis	Foreign body aspiration
Diaphragmatic hernia	Bronchopulmonary dysplasia
Tracheoesophageal fistula	Bronchiectasis
α_1-Antitrypsin deficiency	Mediastinal bronchial compression (including tumor or lymph node)
Vascular ring	Recurrent aspiration
Infections	*Cardiac*
Bronchiolitis	Congestive heart failure
Pneumonia (viral, bacterial, myco-bacterial, fungal)	Pulmonary edema

Table 92.3. Differential Diagnosis for Wheezing

Physical Examination

As noted previously, the physical examination begins immediately with an overall assessment of the child's degree of respiratory distress. Severe retractions, accessory muscle use, nasal flaring, cyanosis, decreased muscle tone, and altered mental status all are indicative of impending or existing respiratory failure and require immediate intervention. The respiratory rate (RR) and heart rate (HR) should be noted and compared to age-appropriate normals (Table 92.2).

The remainder of the respiratory examination includes auscultation for decreased breath sounds, wheezing, rhonchi, and crackles. Crackles can occur and are most often caused by focal areas of atelectasis. Any asymmetry in the pulmonary findings should be noted.

Peak Expiratory Flow Rate

PEFR can serve as a simple, quantitative, reproducible, and inexpensive measure of airway obstruction in the child with mild to moderate distress. Its utility is limited, however, by the inability of children less than 5 to 7 years old and inexperienced patients at any age, to perform the maneuver reliably. In addition, PEFR is effort dependent and measures predominantly large airway disease. Normal PEFR varies based on sex and height as well as on characteristics unique to each peak flow meter model. With increasing use of PEFR to monitor chronic asthma at home, knowledge of the child's personal best PEFR may be available in the ED. A flow rate of less than 80% of predicted or personal best is considered abnormal, and less than 50% indicates moderate to severe obstruction. The degree of improvement after bronchodilator therapy is more useful than the initial value before therapy. Patients with PEFRs below 60% of predicted after ED therapy are more likely to relapse after outpatient therapy.

Oxygen Saturation and Arterial Blood Gases

Although ABGs have an unquestioned role in the evaluation of a severe exacerbation, they have potential disadvantages, particularly in children. They can be technically difficult to obtain, can be painful, and have less reliable results in the crying child. Pulse oximetry, however, provides a noninvasive, continuous, and generally valid measure of arterial hemoglobin oxygen saturation (Sa_{O_2}). Pulse oximetry provides additional data regarding the need and adequacy of supplemental oxygen, response to therapy, and appropriate disposition. If available, it should be measured initially in all children with acute asthma. It is one of the few objective measures in young children who are unable to perform PEFRs reliably. Supplemental oxygen should be provided to maintain an Sa_{O_2} level greater than 90% (95% in young infants), and if hypoxia persists after ED treatment, hospital admission should be considered. For children with lesser degrees of hypoxia, studies have shown that Sa_{O_2} alone does not predict the need for hospitalization accurately for the individual patient. As a rule, however, children with lower oxygen saturations are more likely to relapse. In any case, confirmation of adequate oxygenation is reassuring and obviates the need for ABGs in most mild to moderate exacerbations.

The ABG provides an objective measure of ventilation as well as oxygenation. It is essential in the evaluation of any child in whom impending or existing respiratory failure is suspected clinically, although it should never delay the initiation of therapy. The ABG must be interpreted in conjunction with the clinical picture. The trend of the Pa_{O_2} and Pa_{CO_2} is more

important than the initial value. Mild hypoxia and hypocarbia are expected early in the course of acute asthma. As obstruction progresses and the child becomes fatigued, the hypoxia becomes more severe and the Pa CO_2 rises to “normal” or above “normal” range, resulting in a mixed metabolic and respiratory acidosis. It should be emphasized that a “normal” PaCO_2 of 40 mm Hg in a child with tachypnea or significant respiratory distress may be a sign of impending respiratory failure and requires aggressive management and close monitoring.

Other Studies

The role of chest radiographs in the emergency management of acute asthma is not well defined. Typical findings on routine films such as hyperinflation, atelectasis, and peribronchial thickening do not correlate with severity and rarely alter management. Patients for whom a chest radiograph may be of a higher yield include those with failure to respond to therapy, persistence of focal findings after bronchodilator therapy, reduced oxygen saturation after therapy, or clinical suspicion of a complication or a cause for wheezing other than asthma. These factors also apply to children who present with wheezing for the first time. Radiographic evaluation of the sinuses may be helpful for the child with chronic, persistent wheezing in whom sinusitis is suspected.

The uncommon child taking theophylline may benefit from a theophylline level to help determine further management. The level should be obtained early in the course of therapy. Serum potassium measurement should be considered in children at risk for hypokalemia secondary to receiving frequent b-agonist therapy. A complete blood count (CBC) generally is not useful and, if drawn after adrenergic therapy, often reveals a leukocytosis with neutrophil predominance that should not be misinterpreted as secondary to bacterial infection.

Differential Diagnosis

Wheezing (see Chapter 80) is a continuous, high-pitched, musical, auscultatory finding generally most prominent on expiration; it is caused by obstruction of intrathoracic airways. It must be distinguished from stridor, a harsh, high-pitched, audible sound most prominent on inspiration. Stridor (see Chapter 72) is associated with diseases that cause upper airway obstruction such as croup and epiglottitis, as discussed in Chapter 84. Croup may present with evidence of both upper and lower airway obstruction.

Some children present with chronic cough (see Chapter 15) as the only symptom of hyperreactive airways or “cough variant asthma.” Typically, the cough is worse at night, and the child is asymptomatic during the day. Many of these children have a family history of atopic disease or will have reduced peak flows. There are also children whose acute exacerbations are characterized by severe coughing without obvious wheezing. In the ED, the most practical approach may be a trial of bronchodilators that can be therapeutic as well as diagnostic.

Although wheezing is most commonly associated with asthma, other problems also should be considered (Table 92.3). Several other entities may cause wheezing and usually can be differentiated from asthma through a careful history, physical examination, and attention to the patient's response to therapy. The two most common problems other than asthma that bring children to the ED with wheezing are bronchiolitis and foreign body aspiration.

Bronchiolitis (see Chapter 84) is an acute infection of the small airways most commonly caused by the RSV but also by parainfluenza, adenovirus, and influenza. Outbreaks usually occur during the winter and peak from January through March. It generally affects children less than 1 year of age most severely and manifests with a history of an upper airway infection followed by fever, feeding difficulty, progressive wheezing, respiratory distress, and occasionally, apnea. Some infants, particularly those with chronic lung disease, respond to bronchodilators.

Foreign body aspiration is seen most commonly in children 6 months to 5 years old (see Chapter 29). Although the presentation is usually more subtle, classically, these children have a history of sudden onset of choking, coughing, or wheezing when eating or playing with a small object. The examination may reveal asymmetric breath sounds or wheezing associated with varying levels of respiratory distress. These children may have some, although generally incomplete, response to bronchodilators. The chest radiograph may be normal or show differential hyperinflation on inspiratory/expiratory or bilateral decubitus views, focal atelectasis, or occasionally, a radiopaque foreign body.

Complications

Throughout the ED stay, the potential for complications of acute asthma should be kept in mind. These may be a consequence of the disease itself, the therapy, or both.

The most common pulmonary complication is atelectasis secondary to mucous plugging. Air leaks that lead to a pneumomediastinum and/or a pneumothorax are potentially life-threatening. Children who require mechanical ventilation are at particular risk for air leak complications. A pneumothorax should be suspected in any child with a sudden deterioration associated with chest pain, asymmetry of breath sounds, or a shift of the trachea.

Cardiac arrhythmias are associated with adrenergic agents and with theophylline alike. When these drugs are used together, particularly in association with hypoxemia and acidosis, the risk of arrhythmias is increased.

Frequent b-agonist therapy can cause hypokalemia. The syndrome of inappropriate antidiuretic hormone secretion (SIADH), which may result in dangerous hyponatremia, also is a potential, although rare, complication of acute asthma.

Management

Initial Approach

The primary goals in the acute management phase of an asthma exacerbation are to correct hypoxemia and to rapidly reverse airflow obstruction. Supplemental oxygen, repetitive b $_2$-agonists, and the early addition of systemic corticosteroids achieve this. Patients should be monitored closely and evaluated serially to determine their response to therapy, to identify those who require more aggressive therapy, and to make a final disposition. For children who are discharged, the emergency physician should prescribe an intensified regimen for a minimum of 3 to 5 days and should recommend appropriate follow-up so that chronic management issues can be addressed.

This section of the chapter presents a stepwise approach to the ED management of acute asthma in children. This approach is generally consistent with the Expert Panel 2 (NIH) 1997 recommendations and is summarized in algorithm form in Figure 92.1, with specific dosage recommendations in Table 92.4.

FIGURE 92.1. Approach to acute asthma in children. *PEF,* peak expiratory flow; *FEV_1,* forced expiratory volume in 1 second. (Modified with permission from Expert Panel 2 Report, NIH, 1997.)

Table 92.4. Emergency Department Acute Asthma Therapy

All children with acute asthma can be assumed to be hypoxemic unless oxygen saturation is measured immediately and indicates otherwise. In addition, b-adrenergic therapy may exacerbate hypoxemia transiently by increasing blood flow to poorly ventilated areas of the lung, thereby increasing ventilation-perfusion mismatch. Hence, unless Sa O_2 is greater than 90%, humidified oxygen should be administered immediately. Oxygen is most effectively delivered by mask or nasal cannula, although some small children tolerate oxygen tubing held by their face best. It should be delivered at a flow rate sufficient to eliminate cyanosis and, ideally, to maintain Sa O_2 levels greater than 90% (95% in infants). In contrast to adults, oxygen-induced suppression of respiratory drive is virtually never encountered in asthmatic children.

Repetitive, inhaled b $_2$-agonists are the mainstay of initial bronchodilator therapy in children. They work by relaxing bronchial smooth muscle directly and may also modulate mediator release from mast cells and basophils. In the ED, these always should be delivered in oxygen. Inhalation therapy has been shown to be as effective as subcutaneous injection for mild to moderate obstruction and is associated with fewer systemic side effects. The highly b $_2$-selective agent albuterol offers the additional benefits of reduced cardiotoxicity, longer duration of action, and potent bronchodilation. Frequent (every 15 to 30 minutes) doses of nebulized albuterol appear to be effective in reversing airway obstruction. Although the ideal dose of albuterol (0.5%) has not been determined, the Expert Panel 2 (NIH) recommends 0.15 mg/kg per dose. From a practical standpoint, patients older than 1 year who weigh less than 30 kg should be given 2.5 mg (0.5 mL) and those who weigh more than 30 kg should be given 5 mg (1 mL). A minimum of 1.25 mg (0.25 mL) is used for infants less than 1 year old. Recent evidence suggests that an albuterol metered-dose inhaler (MDI) and chamber may be just as effective as nebulized treatments. Recommendations for this approach are outlined on Table 92.4.

Subcutaneous injection of epinephrine or terbutaline remains an acceptable alternative in settings in which nebulized therapy is unavailable or delayed, or for the toddler resisting an inhalation treatment. It also may be indicated as initial therapy for the child with severe obstruction, hypoventilation, or apnea in whom the delivery of nebulized medication to the airways is believed to be inadequate. Under these circumstances, the injection can be given simultaneously with the initial aerosol and, if indicated, by mask ventilation during preparation for intubation.

The recognition that inflammation is central to the pathogenesis of acute asthma has meant that corticosteroids have assumed an increasingly important role in the acute and chronic management of asthma. Early treatment of asthma

exacerbations with steroids has been shown to prevent progression of airway obstruction, to decrease the need for emergency treatment and hospitalization, and to reduce morbidity. Steroids are believed to potentiate the effect of b-adrenergic agents within hours, in part through alteration of cell membrane receptors and downregulation of inflammatory mediator generation. They also appear to decrease small airway inflammation and edema within 24 hours. Steroid therapy may be particularly beneficial for infants in whom the primary pathophysiology is small airway edema and whose response to bronchodilators may be limited. Because there is a time lag between the administration of steroids and the onset of clinical effect, the first dose is administered as soon as possible. As a rule, almost all children who have had a significant exacerbation ultimately receive steroids. An exception is made for children with mild symptoms who require either no nebulizer treatments or, at worst, one treatment that immediately produces adequate resolution. Exposure to chickenpox in the unprotected host is one of the rare instances in which steroids may need to be avoided acutely. Depending on the level of distress and the child's ability to tolerate oral medications, the dose of corticosteroid is given either intravenously (methylprednisolone 2 mg/kg or equivalent; maximum 125 mg) or by mouth (prednisone or prednisolone 2 mg/kg; maximum 80 mg). Both forms are equally efficacious. Recent studies suggest that nebulized steroids may have a role in the management of acute asthma exacerbations. In addition, children taking inhaled steroids with minor exacerbations may sometimes be adequately managed by doubling their inhaled steroid dose. In general, this should be done in consultation with the physician who manages the patient's asthma.

Ipratropium bromide (Atrovent) is a quaternary ammonium derivative of atropine that limits systemic absorption and decreases its side effects. Ipratropium appears to act synergistically with albuterol, adding additional bronchodilation particularly for patients with moderate to severe air flow obstruction. Ipratropium may be mixed with albuterol and delivered simultaneously. Recommendations vary, but the NIH guidelines suggest 0.25 mg for children (0.5 mg in adolescents and adults) every 20 to 30 minutes mixed with the first three albuterol treatments, then every 2 to 4 hours as needed.

Theophylline has no role in the management of acute asthma and the Expert Panel 2 (NIH) no longer recommends theophylline for admitted patients.

Some children with acute asthma will be dehydrated. These children should receive an initial fluid bolus calculated to return them to a normovolemic state, followed by a maintenance infusion. There is no evidence that overhydration is advantageous. In addition, SIADH is a potential complication.

Antibiotics are indicated only for specific bacterial infections such as sinusitis or otitis media.

After 2 to 4 hours of frequent bronchodilator treatments and the initiation of corticosteroid therapy, the limit of ED management has been reached and a disposition decision should be made. If appropriate resources are available, continued management of the patient in an “observation unit” or “clinical decision unit” for up to 24 hours may avoid the need for hospital admission.

Approach to the Child with Respiratory Failure

There is no universally accepted approach to the child with severe asthma who presents in respiratory failure or deteriorates in the ED despite the therapy already outlined. The options include continuous nebulized albuterol, intravenous (IV) b_2-agonist therapy, and/or mechanical ventilation. The choice of therapy should be guided by the child's mental status, response to therapy, ability to sustain the increased work of breathing, degree of hypoxia, trend in Pa CO_2, and the physician's familiarity and comfort with the various treatment options. In all cases, these patients require continuous monitoring and constant involvement of ED personnel. Arrangements should be made for admission to an ICU setting with skills to handle the child.

The use of continuous nebulized therapy is well established in the ICU setting and currently is under study in EDs. Continuous terbutaline and albuterol have been demonstrated to reverse respiratory failure and to eliminate the need for mechanical ventilation. Albuterol is administered as 0.5 mg/kg per hour (maximum, 15 mg/hour).

IV b_2-agonist therapy is an option for children who fail continuous nebulized therapy. In the United States, terbutaline is administered as a 10 µg/kg loading dose over 10 minutes followed by an initial infusion of 0.4 µg/kg per minute. The infusion is titrated up to effect in increments of 0.2 µg/kg per minute while the child is monitored for unacceptable tachycardia. The usual effective range is 3 to 6 µg/kg per minute. Although not yet available in the United States, albuterol is the favored IV agent in Canada and Europe.

If the child continues to deteriorate, intubation and mechanical ventilation should be considered. There is no single ideal approach to the intubation of children with acute asthma, although some general recommendations can be made. Intubation of these children can be hazardous and should be performed in the presence of the most skilled personnel available. The airway should be managed assuming a full stomach, as discussed in Chapter 5. Many authorities consider ketamine (1 to 2 mg/kg by IV) to be the induction agent of choice because of its bronchodilating effects. Agents that may increase bronchospasm through histamine release such as meperidine, morphine, D-tubocurare, and atracurium are best avoided. After intubation, all b_2-agonist, anticholinergic, and antiinflammatory drug therapy should continue.

Volume-controlled ventilation is preferred using larger-than-average tidal volumes (10 to 20 mL/kg), normal respiratory rates for age, and high flow rates to ensure long expiratory times. To do this, inspiratory pressures often exceed 50 to 60 cm H_2O. In an effort to minimize barotrauma, incomplete correction of the respiratory acidosis or “controlled hypoventilation” (PaCO_2 greater than 50 mm Hg) should be the goal with much higher PaCO_2 necessary in selected cases. In addition, sedatives and neuromuscular relaxants generally are required. Surveillance for barotrauma is critical.

Investigational Approaches to Acute Asthma Exacerbations

Heliox. Heliox is a mixture of helium and oxygen. The gas mixture has a lower density than oxygen and therefore theoretically reduces turbulent flow and airway resistance. This effect reduces work of breathing, which may in turn limit or delay respiratory muscle fatigue and allow more time for standard therapeutic agents to take effect. The lower gas density may also improve ventilation to alveoli with longer time constants resulting in better matching of ventilation and perfusion and more effective delivery of aerosolized medications. Heliox must contain a high concentration of helium (60 to 80%) that requires a tight-fitting mask, limiting its acceptance in young children. Data are limited and conflicting, particularly in children, but there is some suggestion that Heliox may be an effective adjunct for severe exacerbations.

Intravenous Magnesium Sulfate. Although IV magnesium sulfate has been studied extensively in adults and appears to improve pulmonary function, there is no consensus regarding its role in the management of asthma exacerbations. A recent study of children with moderate to severe exacerbations showed that the group treated with magnesium (25 mg/kg by IV; maximum, 2 g) had greater improvement in FEV_1, were less likely to require admission, and had no significant adverse effects compared with the group treated with placebo. This study suggests that IV magnesium may have a role as an adjunct in treatment of children with moderate to severe acute asthma exacerbations.

Disposition. Criteria for the disposition of acutely ill asthmatic children after ED management are difficult to specifically define. Scoring systems that use various assessment tools have not withstood prospective evaluation. As a guideline, however, admission should be considered for children who meet any of the following criteria:

1. Persistent respiratory distress
2. Sao_2 of 91% or less in room air
3. PEFR less than 50% of predicted levels
4. Inability to tolerate oral medications (i.e., vomiting)
5. Previous emergency treatment in last 24 hours
6. Underlying high-risk factors: congenital heart disease, bronchopulmonary dysplasia, cystic fibrosis, and neuromuscular disease
7. Evidence of air leak

Other factors that should be considered in conjunction with these criteria include access to emergency care and medical advice, family reliability, sophistication of available home therapy, and severity of past exacerbations.

Children who also meet the criteria listed in Table 92.5 should be admitted to an ICU setting.

Severe Respiratory Distress	Significant Complications
Estimate in severe range after therapy[a]	Pneumothorax
Pao_2 <60 mm Hg or Sao_2 <90% in 40% O_2	Arrhythmia
$Paco_2$ <42 mm Hg	Theophylline toxicity

[a]See Table 92.2.

Table 92.5. Criteria for Admission to the Intensive Care Unit

Discharge Management. Children who have an adequate clinical response to ED therapy may be discharged to home. Ideally, they should be observed for 30 to 60 minutes after their last treatment to ensure that they do not relapse immediately. Many of these children will experience persistent mild to moderate symptoms and considerable acute disability. It is important to understand that the basic pathophysiology that consists of small airway obstruction, inflammation, and altered lung mechanics is not immediately reversed in the ED and additional therapy will be required. An effort also should be made to remove the trigger, if possible.

In general, if a child already on a regimen of medication has had an exacerbation that required emergency management, this regimen must be intensified, at least for the next 3 to 5 days. Short-course, high-dose oral steroids (i.e., prednisone, 2 mg/kg per day up to 80 mg/day for 3 to 5 days) should be prescribed and administered for essentially all children who present to the ED with a significant exacerbation. In addition, all patients currently taking or who have recently been taking oral or inhaled corticosteroids must be given steroids as part of the acute management of their exacerbation. Children who experience frequent acute exacerbations, nocturnal symptoms, or multiple absences from school also may benefit from the addition of a systematic or inhaled corticosteroid to their regular regimen.

For children who experience their first episode of wheezing or who are not receiving long-term therapy, an inhaled albuterol is generally well tolerated in the subacute phase following the acute episodes. Children less than 5 years old should use an MDI with a spacer and mask or a nebulizer. The use of a spacer device in all children will improve delivery of medications in MDIs. If the child is receiving other long-term therapy, such as cromolyn sodium or inhaled steroids, it is important to continue it during acute exacerbations. Table 92.6 lists outpatient treatment options.

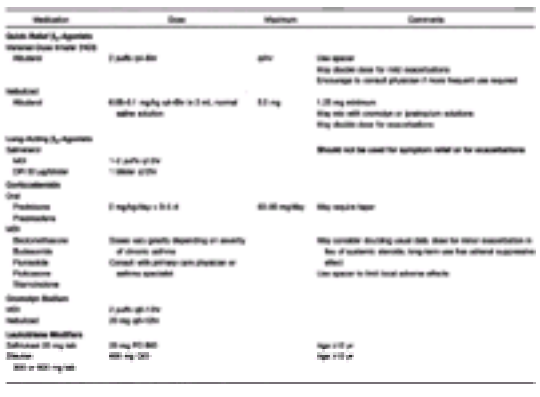

Table 92.6. Outpatient Asthma Therapy

It must be emphasized again that the ED plays a role beyond the management of the acute episode. If possible, ED personnel should take advantage of the educational opportunity and instruct the patient and family on the proper use of the inhaler and peak flow meter, environmental issues, and appropriate use of medications. Patients should leave with specific written instructions and be referred for follow-up appointment in the next 2 to 5 days to ensure resolution of the acute episode and to review long-term management.

ANAPHYLAXIS

Background

Anaphylaxis is a potentially life-threatening manifestation of immediate hypersensitivity. The severity of these reactions varies from mild urticaria to shock and death. Anaphylaxis most commonly involves the pulmonary, circulatory, cutaneous, gastrointestinal (GI), and central neurologic systems.

The classic anaphylactic response is an IgE-mediated reaction that occurs after reexposure to an antigen to which the patient has previously been sensitized. The term *anaphylactoid reaction* sometimes is used to refer to a clinically similar syndrome that is not IgE mediated and does not necessarily require previous exposure to the inciting agent. It has become common practice to use *anaphylaxis* to describe the clinical syndrome regardless of the responsible mechanism.

Any route of exposure, including parenteral, oral, and inhalation, has been associated with anaphylaxis. The most common inciting agents are Hymenoptera stings, drugs, foods, immunotherapy, radiocontrast media, and blood products (Table 92.7). IgE-mediated anaphylaxis to the latex present in gloves, Foley catheters, and endotracheal tubes was first recognized in the late 1970s. Patients who undergo multiple procedures, such as those with myelomeningocele and genitourinary dysplasias, as well as health care workers appear to be at greatest risk.

Insect Venom	*Foods*
Hymenoptera	Peanuts
Fire ants	Milk
Drugs	Seafood—shellfish
Antibiotics—penicillin,	Grains
cephalosporins,	*Blood Products*
sulfonamides	*Immunotherapy*
Local anesthetics—	Allergen extracts
lidocaine	*Other*
Aspirin	Latex
Radiocontrast media	Idiopathic

Table 92.7. Common Causes of Anaphylaxis

Pathophysiology

Currently, there are three well-established mechanisms that lead to anaphylaxis after exposure to a foreign substance. The first is the classic IgE-mediated reaction. The IgE antibodies form when a person is exposed to the foreign antigen (either in its native state or as a hapten attached to a carrier protein) for the first time. IgE binds to high-affinity receptors on mast cells and basophils. On reexposure, the antigen induces bridging of IgE molecules, leading to degranulation of these cells and to the release of various preformed and rapidly generated mediators. Immune complexes or other agents capable of activating the complement cascade induce the second mechanism. This results in the formation of anaphylatoxins such as C3a and C5a, which directly trigger release of mediators from mast cells and basophils. The third mechanism involves the ability of certain agents to stimulate the release of mediators directly by an unknown mechanism that does not involve IgE or complement. Agents capable of direct stimulation include hyperosmolar solutions such as mannitol and radiocontrast media.

The sudden release of numerous mediators from mast cells and, perhaps, from basophils is presumed to be responsible for the pathophysiologic features of anaphylaxis (bronchospasm, increased vascular permeability, and altered systemic and pulmonary vascular smooth-muscle tone). The most notable of these mediators is histamine, but others that have

been implicated include prostaglandin D_2, leukotriene C_4, platelet-activating factor, tryptase, chymase, heparin, and chondroitin sulfate.

Other causes of apparent anaphylaxis for which no clear mechanism has been identified exist. These include reactions after the ingestion of aspirin and other nonsteroidal anti-inflammatory drugs (NSAIDs) and exercise-induced anaphylaxis in which vigorous exercise is the trigger.

Clinical Findings

The time between exposure to the inciting agent and onset of symptoms can vary from minutes to hours, although most reactions occur within 1 hour. This interval depends on the sensitivity of the patient and the route, quantity, and rate of administration of the antigen. Although uncommon, some patients experience a biphasic reaction in which symptoms may recur up to 12 hours after the initial reaction.

The signs and symptoms of anaphylaxis vary in both the spectrum and severity of involvement. Reactions may be limited to the skin, as in a mild urticarial reaction, or catastrophically involve multiple systems, leading to shock and death.

In general, systemic reactions include cutaneous involvement such as pruritus, flushing, erythema, urticaria, and in more severe cases, angioedema. A more detailed discussion of urticaria is found at the end of this section (see also Chapter 66). Mucous membrane involvement may be limited to pruritus and congestion of the eyes, nose, and mouth. Swelling of the lips or tongue can potentially impair swallowing and ventilation.

An immediate life-threatening feature of anaphylaxis is upper airway obstruction that results from edema of the larynx, epiglottis, and other surrounding structures. This may be experienced as subtle discomfort of the throat or as obvious stridor and respiratory distress. Anaphylaxis also can cause lower airway disease secondary to bronchospasm. This leads to findings similar to acute asthma, such as a sense of chest tightness, cough, dyspnea, wheezing, and retractions.

Another potential life-threatening feature of anaphylaxis is cardiovascular collapse and hypotensive shock. Although the mechanisms are not fully understood, these cardiopulmonary manifestations are thought to result from profound vasodilation, increased vascular permeability, capillary leak, and intravascular volume depletion, as well as a possible direct toxic effect of circulating mediators. Arrhythmias and electrocardiographic evidence of myocardial ischemia also may be seen.

Central nervous system (CNS) involvement can include dizziness, syncope, seizures, and an altered level of consciousness. These may occur as a result of hypoperfusion or, possibly, as a direct toxic effect of mediator release.

GI symptoms are relatively common and include nausea, vomiting, diarrhea, and crampy abdominal pain.

Urticaria is a common manifestation of immediate hypersensitivity reactions as well as a number of other disease processes (Table 92.8). In the patient with acute urticaria from an IgE-mediated process, the urticaria may be localized to the area of exposure, such as the area around a sting. In addition to the localized urticaria, there may be a systemic reaction. Urticaria may be associated with angioedema—swelling of the lower dermis and subcutaneous tissues. The angioedema associated with urticaria is pruritic. Angioedema without pruritus usually is secondary to processes other than immediate hypersensitivity.

Dermographism	Familial urticaria
Physical urticaria	Hereditary angioedema
Cold	Familial cold urticaria
Cholinergic	Urticaria secondary to common agents
Pressure	Urticaria secondary to serum sickness
Solar	

Table 92.8. Urticaria: Classification

Urticaria can be separated into those that are acute and those that are chronic. Most immediate hypersensitivity reactions are associated with an acute reaction, but chronic, recurrent urticaria can be mediated by recurrent exposure to an unknown antigen. In determining the cause of an urticarial reaction, the classification shown in Table 92.8 is important because management may vary.

The physical urticarial reactions may be life-threatening, and they should be included in the differential diagnosis of anaphylaxis. Cold urticaria is an acute reaction to cold temperatures with hives at the site of cold exposure. Generalized cold exposure, such as immersion in a cold pool, can precipitate an anaphylactic reaction with hypotension and shock. The cold urticarias often are acquired and may follow viral infections. There is a familial cold urticaria that is rare and associated with a delayed onset, leukocytosis, and pain that distinguishes it from acquired cold urticaria. Cholinergic urticaria is characterized by punctate hives surrounded by an erythematous flare. Exercise, anxiety, shivers, and environmental temperature change can precipitate the reaction. It has been associated with exercise-induced

anaphylaxis and often causes systemic manifestations (e.g., abdominal pain, headaches). Solar urticaria is a reaction to light, often sunlight, with the development of pruritus, erythema, and edema. Solar urticaria can be a manifestation of porphyria. Pressure urticaria is associated with hives that develop at the site of significant prolonged pressure in areas of the body. It often is associated with tight clothing.

Management

Initial Assessment

Immediate resuscitative efforts must be initiated for the child who manifests the full-blown syndrome or any of the independent life-threatening manifestations, such as upper airway obstruction or shock. All patients who complain of an "allergic reaction" should be evaluated expeditiously to determine the extent of involvement, signs of progression, and the need for urgent intervention.

History

The history should be directed toward determining the nature and severity of the reaction, the rapidity with which symptoms evolved, and evidence of ongoing progression. Change in voice, difficulty in swallowing, dyspnea, and a sense of impending doom all are characteristic of potentially serious reactions.

Attempts also should be made to determine the offending agent. This may be obvious as in a reaction to a bee sting. The history should focus on the 1- to 2-hour period before the onset of symptoms. The association of anaphylactic reactions to food often is confusing. Although patients often focus on a particular food as the cause, a more detailed history may implicate something else in the meal. For example, it is common to associate reactions with chocolate, whereas the nuts in many chocolate preparations generally are the offending agents.

Physical Examination

A rapid cardiopulmonary assessment should precede any detailed physical examination. It should be quickly determined whether any evidence of upper airway obstruction, bronchospasm, or shock exists. Once these issues have been addressed, a more detailed physical examination should be performed to evaluate the patient for less serious cutaneous and GI manifestations of the reaction.

Management of a life-threatening anaphylactic reaction requires simultaneous evaluation and management of the airway, breathing, and circulation (ABCs), and the immediate administration of epinephrine as illustrated in Figure 92.2 and discussed in sequence in the following.

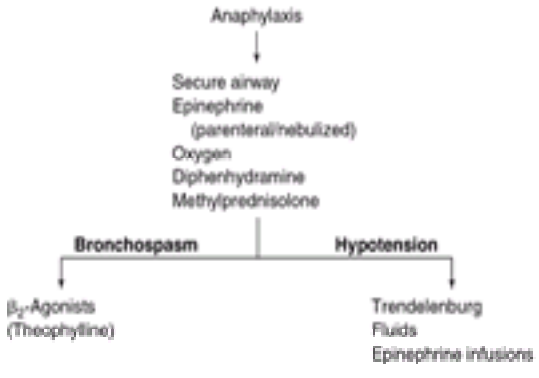

FIGURE 92.2. Management of anaphylaxis.

Maintenance of the Airway and Oxygenation

The physician should administer 100% oxygen with bag-valve-mask ventilation, as indicated, to assist ventilation. If there is complete airway obstruction, immediate endotracheal intubation should be attempted. If intubation is unsuccessful, cricothyrotomy is indicated. Rapid administration of epinephrine may lessen the difficulty of airway management.

Epinephrine, the first-line drug for anaphylaxis, should be administered *simultaneously.* It has several beneficial actions. As an a-adrenergic agonist, epinephrine promotes vasoconstriction, which increases blood pressure and decreases capillary leakage. As a b-adrenergic agonist, it relaxes bronchial smooth muscle, increases cardiac rate and contractility, and inhibits further mediator release. Epinephrine can be administered subcutaneously in the patient with reasonable perfusion (epinephrine 1:1000, 0.01 mL/kg to a maximum of 0.5 mL). If the patient is hypotensive or hypoperfused or if the initial subcutaneous dose is ineffective, the epinephrine should be administered by IV or through an intraosseous needle as a 1:10,000 solution, 10 µg/kg (0.1 mL/kg) over 1 to 2 minutes. In severe cases, this may need to be followed by a continuous epinephrine infusion of 0.1 µg/kg per minute, which can be titrated to effect up to a maximum of 1.0 µg/kg per minute (6 mg epinephrine should be added to 100 mL D5W normal saline solution; 1 µg/kg per minute = 1 mL/kg per hour).

Bronchospasm should be treated aggressively with supplemental oxygen, b_2-agonists such as albuterol or epinephrine, and corticosteroids, as outlined in the previous section on asthma. Some authorities believe that theophylline has a role

in the treatment of bronchospasm secondary to anaphylaxis.

Although not studied specifically in this setting, some authors recommend nebulized epinephrine (2.5 mL undiluted 1:1000) or racemic epinephrine 2.5% (0.5 mL in 2.5 mL normal saline) for relief of both the upper and lower airway obstruction associated with anaphylaxis.

Maintenance of the Circulation

Hypotensive patients should be placed in the Trendelenburg position, and a rapid bolus of 20 mL/kg of a crystalloid solution should be administered immediately and repeated as necessary. Because plasma volume may fall precipitously by 20 to 40%, large amounts of fluid may be necessary. If hypotension persists after epinephrine, normal saline bolus, and positioning, a continuous infusion of epinephrine should be started as previously described.

Other Therapy

The H_1-receptor antihistamines such as diphenhydramine (1 to 2 mg/kg intramuscularly or intravenously; maximum, 50 mg) are indicated in histamine-mediated allergic reactions. They work synergistically with the epinephrine therapy.

Corticosteroids do not take effect during the initial resuscitative phase of anaphylaxis. In significant reactions, however, their early administration may block or reduce the late-phase reactions over the next several hours or days. They can be administered as methylprednisolone 1 to 2 mg/kg by IV (maximum, 125 mg) or prednisone 1 to 2 mg/kg by mouth (maximum, 80 mg).

Many authorities recommend H_2-blocking antihistamines such as cimetidine (5 mg/kg; maximum, 300 mg) or ranitidine (1 to 2 mg/kg; maximum, 50 mg) in addition to H_1-blocking antihistamines, particularly for more severe reactions. These agents may work synergistically with the H_1-blocking antihistamines.

On occasion, in severe reactions from injection, a venous tourniquet above the injection site of the offending agent and local infiltration of epinephrine 1:1000 (0.005 mL/kg) and/or the application of ice may decrease further absorption.

Management of Limited Reactions

Most children with allergic reactions present with involvement limited to a diffuse, pruritic rash; localized swelling; or benign involvement of the mucous membranes. Appropriate management of these children varies according to the specific presentation. Options include subcutaneous epinephrine (e.g., for evolving urticaria), antihistamines, and corticosteroids.

Diphenhydramine (5 mg/kg per day divided every 4 to 6 hours, with a maximum of 300 mg/24 hours) and hydroxyzine (2 mg/kg per day divided every 4 to 6 hours, with a maximum of 200 mg/day) are the antihistamines most commonly prescribed for urticaria. In the case of cold urticaria, cyproheptadine (0.25 to 0.5 mg/kg per day divided every 12 hours, with a maximum of 32 mg/day) is the drug of choice, whereas hydroxyzine is preferred for cholinergic urticaria or most other chronic urticarias.

Disposition

Patients with severe reactions that involve upper airway obstruction or shock generally should be monitored for a minimum of 8 to 24 hours. Children with a history of asthma appear to be at increased risk for delayed and severe reactions and may also require prolonged monitoring.

Patients with less severe manifestations can be discharged home on a course of antihistamines and, in selected cases, corticosteroids. As a rule, therapy initiated in the ED should be continued for a minimum of 48 hours. Follow-up with the child's primary care physician also is advised.

Strategies to avoid exposure to the offending agent should be discussed. All children with a history of significant anaphylaxis with an antigen that cannot be avoided totally (e.g., Hymenoptera venom or food) should be instructed to carry a preloaded syringe of epinephrine to be used in emergencies (EpiPen).

SERUM SICKNESS

Background

Serum sickness is an immune complex–mediated disease. It was first described in 1905 in patients who had received heterologous antisera, which, at that time, was used routinely to treat various infectious diseases. Today, the most common cause of serum sickness is exposure to medication.

The clinical syndromes secondary to an immediate hypersensitivity reaction may appear similar to serum sickness. In the former, IgE is primarily responsible, whereas IgG or IgM immune complexes classically mediate the latter. In many cases, however, elements of both processes are involved. More recently, a clinical syndrome similar to serum sickness but apparently not mediated by immune complexes has been reported. These reactions are referred to as serum sickness-like reactions and appear to be responsible for the relatively common reactions seen in association with cefaclor.

Currently, the most common medications implicated in serum sickness or serum sickness-like reactions include the

penicillins, sulfonamides, cephalosporins, streptomycin, the hydantoins, and thiouracil. Because these drugs are all low-molecular-weight substances, they cannot act as antigens directly but must bind to proteins, usually through their metabolites. Therefore, it is often difficult to substantiate sensitization.

The use of agents made from heterologous serum has decreased dramatically. These preparations currently are found in antithymocyte globulins used to prevent organ transplant rejection and in antitoxins for the management of clostridial infections, diphtheria, tetanus, and specific arachnid and snake envenomations.

Less commonly, exposures to various chemical, infectious agents, or autologous antigens result in a serum sickness-like illness.

Pathophysiology

In the classical serum sickness model, an animal is injected with foreign serum protein. The animal develops the symptoms characteristic of serum sickness 7 to 10 days later. During the initial period after the injection of the foreign protein, there is a period of antigen excess (Fig. 92.3). This is a period during which no antibody has been formed. Antibodies develop approximately 6 to 10 days after the initial antigen is injected, and antibody–antigen complexes then form. These immune complexes deposit in the tissues and also may activate complement. After this period of immune complex formation, there is a period of antibody excess during which antigen clears from the system. Symptoms develop when soluble immune complexes are being cleared by the body.

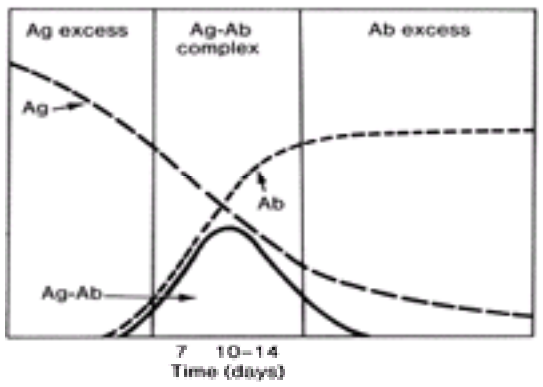

FIGURE 92.3. Time course of immune complex formation.

These immune complexes can activate the classical complement pathway. The classical pathway of activation begins with the formation of the C1q antibody complex, which activates a C1 esterase that cleaves the fourth and second complement components (C4 and C2). The C4 and C2 that have been activated can then cleave C3. It is cleavage of C3 and generation of all of its active components that allow for the activation of the late-acting complement components (C5–C9). Because the immune complex activation involves the classical components, C4, C2, and C3, their serum concentration decreases. The complement activation generates the anaphylatoxins C3a and C5a, which then increase vascular permeability, release histamine, and can produce bronchospasm. They activate many other cells in the inflammatory process and lead to inflammation around deposits of complexes in various tissues.

Clearance of immune complexes depends on their size and the effectiveness of the reticuloendothelial system. The most vulnerable organs to injury include the kidneys and the vascular system.

It also had been shown in experimental animals that immune complex deposition can be enhanced by concomitant activation of an immediate hypersensitivity reaction. The IgE-mediated mast cell degranulation increases vascular permeability, which, in turn, enhances immune complex deposition.

Clinical Findings

The severity of serum sickness and the specific clinical manifestations vary widely. The reaction is characterized by fever, malaise, and a rash that is most commonly urticarial but also may appear as maculopapular or vasculitic. Other manifestations include arthralgias or arthritis, lymphadenopathy, angioedema, and nephritis. Other less common problems include abdominal pain, carditis, anemia, and neuritis. Serum sickness is usually self-limited and typically resolves within 1 to 2 weeks with or without therapy.

Characteristically, the onset of symptoms occurs 7 to 14 days after the primary exposure. If there has been prior sensitization, however, reexposure can result in onset of a few days.

History

The history should be directed toward identifying the offending agent and determining the extent and severity of the symptoms.

The physician should review possible exposures up to 14 days before symptoms to ascertain a possible cause for the process. Because a secondary exposure can produce a more rapid onset of symptoms (1 to 4 days), inquiries about this interval and about previous exposures may also be revealing.

The history should elicit information about the extent of systemic illness and the involvement of specific organ systems. It is important to determine the time course and nature of the rash, the degree of joint pain, swelling or warmth, and evidence of renal involvement such as hematuria, edema, and reduced urine output. In light of the potential for involvement of other organ systems, a complete review of systems is indicated.

Physical Examination

Perform a thorough physical examination to determine the severity and extent of involvement of the reaction. A general inspection will help determine how ill and uncomfortable the child is. Examination of the skin may reveal a maculopapular eruption, urticaria, or the palpable purpura of a cutaneous vasculitis. Painful angioedema is commonly present. Generalized lymphadenopathy often occurs. In more severe reactions, the joints show erythema, warmth, and effusion. Wheezes may be appreciated on auscultation of the lungs, and a pericardial friction rub may be audible if pericarditis is present. The liver and spleen often enlarge. Rarely, neurologic deficits occur secondary to a vasculitis of the central nervous system.

Laboratory

The selection of laboratory studies should be determined by the severity of the reaction, evidence of specific organ system involvement, and the degree of diagnostic uncertainty. For most patients, a urinalysis to look for evidence of renal involvement is all that is required. A list of other studies that may be indicated for individual patients with immune complex–mediated disease is outlined in Table 92.9.

Blood Tests	Hepatitis B screen
Erythrocyte sedimentation rate	Heterophil antibody
Complete blood count with differential	Immune complex assay
CH_{50}, C3, C4	*Other Laboratory Tests*
Blood urea nitrogen, creatinine	Urinalysis
Antinuclear antibody	Electrocardiogram
Rheumatoid factor	Stool Hematest
Hepatic enzymes	Computed tomography scan

[a]Laboratory evaluation should be tailored for each individual patient as noted in text.

Table 92.9. Possible Laboratory Evaluation of Serum Sickness[a]

The erythrocyte sedimentation rate (ESR) may be elevated. A CBC and differential may reveal leukopenia or leukocytosis. The C3, C4, and CH_{50} may decrease because of complement activation. Any of the tests for circulating immune complexes (e.g., cryoglobulins) may be elevated. Stool Hematest should be performed, and an echocardiogram should be considered for patients with abdominal pain or other symptoms involving the GI tract. If carditis is suspected, a screening electrocardiogram should be performed. Severe headache or focal neurologic deficits are indications for a computed tomography (CT) scan.

Management

The treatment of serum sickness is based on the extent and severity of the patient's disease. Keeping in mind that the reaction is usually self-limited, the goal is to provide symptomatic relief and monitor for complications. If possible, the offending antigen should be eliminated. It has been demonstrated in animal models that elimination of the antigen allows more rapid resolution of immunopathologic damage.

Pharmacologic management usually involves one or more of the following: antihistamines, NSAIDs, and corticosteroids. Pruritus, rash, and angioedema can be managed with an antihistamine such as hydroxyzine (2 mg/kg per 24 hours divided every 6 to 8 hours, with a maximum of 200 mg/24 hours). Although experience in the treatment of serum sickness is limited, use of the second-generation nonsedating antihistamines may also be considered (Table 92.10). Urticarial lesions and angioedema that evolves rapidly may respond acutely to subcutaneous epinephrine (1:1000, 0.01 mL/kg subcutaneously, with a maximum 0.30 mL) and/or longer-acting Sus-Phrine (0.005 mL/kg subcutaneously, with a maximum of 0.15 mL). Mild joint involvement and/or fever often improve with use of an NSAID such as ibuprofen (30 to 50 mg/kg per 24 hours divided every 6 to 8 hours up to a maximum 2.4 g/24 hours). In more severe disease or after failure to respond to these measures, a burst of corticosteroids may be indicated. This involves the use of 1 to 2 mg/kg per day of prednisone in divided doses for 7 to 10 days, followed by a taper for 3 to 4 weeks (maximum, 80 mg/day). In life-threatening serum sickness with significant circulating immune complexes, plasmapheresis may play a role, but this procedure has not been used extensively for treatment of this disease.

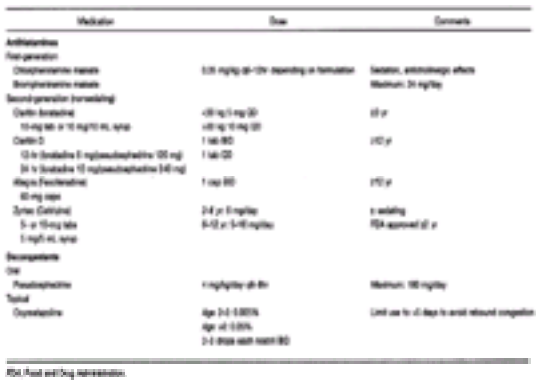

Table 92.10. Emergency Department Management of Allergic Rhinitis

Most children with serum sickness can be managed as outpatients with close follow-up by their primary care physicians. Children with more severe involvement may benefit from hospitalization.

ALLERGIC RHINITIS

Background

Allergic rhinitis is the most common manifestation of atopic disease. Peak incidence occurs in the pediatric age group, affecting up to 30% of children and adolescents. Although not a life-threatening problem, allergic rhinitis can have a significant, often underestimated, negative impact on the quality of life of affected children. The morbidity of allergic rhinitis results from the direct manifestations of the disease as well as from associated complications such as serous OM, sinusitis, acute asthma, sleep disturbances, dysosmia, and the consequences of chronic mouth breathing.

Pathophysiology

Allergic rhinitis is caused by an IgE-mediated hypersensitivity response of the nasal mucosa to foreign allergens. Following sensitization to a foreign antigen, reexposure triggers an immediate hypersensitivity reaction. This early response is characterized by activation of mast cells and release of preformed mediators of inflammation such as histamine and chemotactic factors and newly formed mediators such as prostaglandins and leukotrienes. These mediators cause vasodilation, mucosal edema, mucus secretion, stimulation of itch receptors, and reduction in the threshold for sneezing.

Many children experience a late-phase allergic response that consists of spontaneous release of inflammatory mediators 3 to 12 hours after exposure. There also is a “priming effect” that leads to increased responsiveness to small antigen loads and hyperresponsiveness to environmental irritants.

Seasonal allergic rhinitis is most commonly caused by exposure to tree pollens (early spring), grass pollens (late spring and early summer), and ragweed or other weed pollens (late summer and fall). Allergens responsible for *perennial* allergic rhinitis include animal dander, house dust mites, and mold spores.

Clinical Findings

The classic symptoms of allergic rhinitis include nasal congestion, paroxysmal sneezing, pruritus of the nose and eyes, and watery, profuse rhinorrhea. Other complaints may include noisy breathing, snoring, repeated throat clearing or cough, itching of the palate and throat, “popping” of the ears, and ocular complaints such as redness, itching, and tearing.

The physical examination is variable but may reveal the “gaping” look of a mouth breather, dark discoloration of the infraorbital ridge caused by venous congestion (allergic shiners), and a transverse external nasal wrinkle secondary to chronic rubbing of the nose (allergic salute). Intranasal findings are variable. The mucosa often is edematous and may appear pale or violaceous. The nasal secretions may be clear, mucoid, or opaque.

Management

There are several approaches to the management of allergic rhinitis. These include identification and avoidance of environmental allergens and irritants, an antihistamine generally prescribed together with an oral or topical decongestant, topical corticosteroids, and immunotherapy.

Recognizing that long-term therapy must be highly individualized, the emergency physician will generally limit interventions to those that safely provide rapid, symptomatic relief and then refer the child to the primary care physician for long-term therapy. In addition, topical corticosteroids, first-line therapy for chronic allergic rhinitis, may require as long as 2 weeks to achieve maximal relief. Rapid relief can generally be achieved by prescribing an antihistamine in combination with a decongestant (Table 92.10).

Antihistamines acutely reduce the rhinorrhea, pruritus, and sneezing associated with allergic rhinitis. Because they do not reduce congestion caused by mucosal edema, they are most effective in combination with a decongestant. The newer second-generation H_1-receptor antagonists are preferred over older antihistamines because they are generally

nonsedating, longer acting, and devoid of anticholinergic effects.

Oral decongestants, such as pseudoephedrine, relieve nasal congestion through their a-adrenergic activity. Potential adverse effects include headache, restlessness, insomnia, and decreased appetite. Topical decongestants such as oxymetazoline hydrochloride present another alternative but should be prescribed for only brief periods (less than 5 days) to avoid tachyphylaxis.

Environmental approaches and an antihistamine–decongestant combination may be all that is required for the child with only mild, self-limited episodes of allergic rhinitis. As already noted, children with more severe or chronic allergic rhinitis should be referred to their primary care physicians who can determine the best alternative for long-term management.

The first-line approach for patients who require long-term therapy is topical corticosteroids (Table 92.11) with or without a second-generation oral antihistamine. For completeness, other categories of topical treatment are also listed in Table 92.11. Children with significant ocular symptoms may also benefit from topical ophthalmic treatment (Table 92.12).

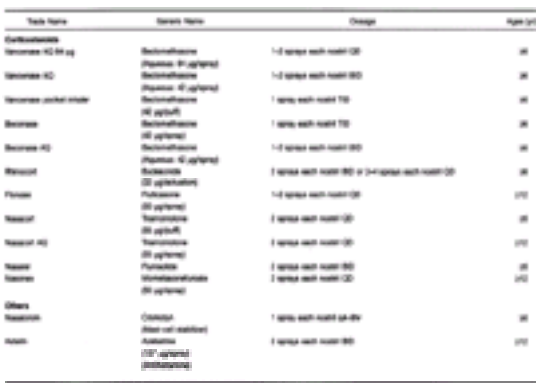

Table 92.11. Topical Treatment for Allergic Rhinitis

Trade Name	Generic Name	Category	Dosage	Ages (yr)
Naphcon-A	Naphazoline and Pheniramine	Antihistamine–decongestant	1–2 gtt QID	≥6
Livostin	Levocabastine	Antihistamine	1–2 gtt QID	≥12
Alomide	Lodoxamide Tromethamine	Mast-cell stabilizer	1–2 gtt QID	≥2
Patanol	Olopatadine	Mast-cell stabilizer and antihistamine	1–2 gtt BID	≥3
Acular	Ketotifen	NSAID	1–2 gtt QID	≥12

NSAID, nonsteroidal anti-inflammatory drug.

Table 92.12. Ophthalmic Drops

Suggested Readings

ASTHMA

Amirav I, Newhouse MT. Metered-dose inhaler accessory devices in acute asthma. Arch Pediatr Adol Med 1997;151:876–882.

Appel D, Karpel JP, Sherman M. Epinephrine improves expiratory flow rates in patients with asthma who do not respond to inhaled metaproterenol sulfate. J Allergy Clin Immunol 1989;84:90–98.

Asthma mortality and hospitalization among children and young adults—United States, 1980–1993. MMWR 1996:45;350–353.

Barnett PLJ, Caputo GZ, Baskin M, Kuppermann N. Intravenous versus oral corticosteroids in the management of acute asthma in children. Ann Emerg Med 1997;29:212–217.

Benatar SR. Fatal asthma. N Engl J Med 1986;314:423–429.

Carter ER, Webb, CR, Moffitt DR. Evaluation of Heliox in children hospitalized with acute severe asthma. A randomized crossover trial. Chest 1996;109:1256–1261.

Ciarallo LM, Saver AH, Shannon MW. Intravenous magnesium therapy for moderate to severe pediatric asthma: results of a randomized placebo-controlled study. J Pediatr 1996;129:809–814.

DeNicola LK, Monem GF, Gayle MO, Kissoon N. Treatment of critical status asthmaticus in children. Pediatr Clin North Am 1994;41:1293–1324.

Folkerts G, Busse WW, Nijkamp FP, et al. Virus-induced airway hyperresponsiveness and asthma. Am J Respir Crit Care Med 1998;157: 1708–1720.

Fuglsang G, Pederson S, Borgstrom L. Dose-response relationships of intravenously administered terbutaline in children with asthma. J Pediatr 1989;114:315–320.

Geelhoed GC, Landau LI, LeSouef PN. Oximetry and peak expiratory flow in assessment of acute childhood asthma. J Pediatr

1990;117:907–909.

Gershel JC, Goldman HS, Stein REK, et al. The usefulness of chest radiographs in first asthma attacks. N Engl J Med 1983;309:336–339.

Harris JB, Weinberger MM, Nassif E, et al. Early intervention with short courses of prednisone to prevent progression of asthma in ambulatory patients incompletely responsive to bronchodilators. J Pediatr 1987;110:627–633.

Kass JE, Castriolta RJ. Heliox therapy in acute severe asthma. Chest 1995;107:757–760.

Kerem E, Levison H, Schuhs, et al. Efficiency of albuterol administered by nebulizer versus spacer devise in children with acute asthma. J Pediatr 1993;123:313–317.

Littenberg B. Aminophylline treatment in severe, acute asthma: a meta-analysis. JAMA 1988;259:1678–1684.

Littenberg B, Gluck EH. A controlled trial of methylprednisolone in the emergency department treatment of acute asthma. N Engl J Med 1986;314:150–152.

Moler FW, Hurwitz ME, Custer JR. Improvement in clinical asthma score on Pa CO_2 in children with severe asthma treated with continuously nebulized terbutaline. J Allergy Clin Immunol 1988;81:1101–1109.

NHLBI—Highlights of the expert panel report 2: guidelines for the diagnosis and management of asthma. NIH publication No 97-4051A; May 1997.

Nadel JA, Busse WW. Asthma. Am J Respir Crit Care Med 1998;157: 5130–5138.

Parrillo SJ. Cough variant asthma. Pediatr Emerg Care 1986;2:97–101.

Scarfone RJ, Fuchs SM, Nager AZ, Shane SA. Controlled trial of oral prednisone in the emergency department treatment of children with acute asthma. Pediatrics 1993;92:513–518.

Scartone RJ, Loiselle JM, Wiley JR, et al. Nebulized dexamethasone versus oral prednisone in the emergency treatment of asthmatic children. Ann Emerg Med 1995;26:480–486.

Schuh S, Johnson DW, Callahan S, et al. Efficiency of frequent nebulized ipratropium bromide added to frequent high dose albuterol therapy in severe childhood asthma. J Pediatr 1995;126:639–645.

Schuh S, Parkin P, Rajan A, et al. High-versus low-dose, frequently administered, nebulized albuterol in children with severe, acute asthma. Pediatrics 1989;83:513–518.

Schuh S, Reider MJ, Canny G, et al. Nebulized albuterol in acute childhood asthma: comparison of two doses. Pediatrics 1990;86:509–513.

Skoner DP, Fischer TJ, Gormley C, et al. Pediatric predictive index for hospitalization in acute asthma. Ann Emerg Med 1987;16:25–31.

Strunk RC. Identification of the fatality-prone subject with asthma. J Allergy Clin Immunol 1989;83:477–485.

Younger RE, Gerber PS, Herrod HG, et al. Intravenous methylprednisolone efficacy in status asthmaticus at childhood. Pediatrics 1987;80:225–230.

ANAPHYLAXIS

Atkinson TP, Kaliner MA. Anaphylaxis. Med Clin North Am 1992;76: 841–855.

Barach EM, Nowak RM, Tennyson GL, Tomlanovich MC. Epinephrine for treatment of anaphylactic shock. JAMA 1984;251:2118–2122.

Bochner BS, Lichtenstein LM. Anaphylaxis. N Engl J Med 1991;324: 1785–1790.

Landwher LP, Boguniewicz M. Current prospectus on latex allergy. J Pediatr 1996;128:305–312.

Marquardt DL, Wasserman SI. Anaphylaxis. In: Middleton E, Reed CE, Ellis EF, et al, eds. Allergy Principles and Practice. 4th ed. St Louis: Mosby, 1995.

Patel L, Radivan S, David TJ. Management of anaphylactic reactions to food. Arch Dis Child 1994;71:370–375.

Patterson R, Valentine J. Anaphylaxis and related allergic emergencies including reactions due to insect stings. JAMA 1982;248:2632–2636.

Stark BJ, Sullivan TJ. Biphasic and protracted anaphylaxis. J Allergy Clin Immunol 1986;78:76–83.

Wood RA. Anaphylaxis: causes and management. Contemp Pediatr 1996; 13:89–96.

SERUM SICKNESS

Cochrane CG, Koffler D. Immune complex disease in experimental animals and man. Adv Immunol 1973;16:186–264.

Evans R, Kim K, Mahr TA. Current concepts in allergy: drug reactions. Curr Prob Pediatr 1991;21:185–191.

Heckbert SR, Stryker WS, Coltin KL, et al. Serum sickness in children after antibiotic exposure: estimates of occurrence and morbidity in a health maintenance organization population. Am J Epidemiol 1990;132: 336–342.

Kearns GL, Wheeler JG, Childress SH, et al. Serum sickness-like reactions to cefaclor: role of hepatic metabolism and individual susceptibility. J Pediatr 1994;125:805–811.

Lawley TJ, Bielory L, Gascon P, et al. A prospective clinical and immunologic analysis of serum sickness in man. N Engl J Med 1984;311:1407–1413.

Lawley TJ, Frank M. Immune complexes and allergic disease. In: Middleton E, Reed CE, Ellis EF, et al, eds. Allergy principles and practice. 4th ed. St Louis: Mosby, 1993.

Platt R, Dreis MW, Kennedy DL, Kuritsky JN. Serum sickness-like reactions to amoxicillin, cefaclor, cephalexin, and trimethoprim-sulfamethoxazole. J Infect Dis 1988:158:474–477.

Striker BH, Tijssen JG. Serum sickness: like reactions to cefaclor. J Clin Epidemiol 1992;45:1177–1184.

Wiggens RC, Cochrane CG. Immune-complex mediated biologic affects. N Engl J Med 1981;304:518–520.

ALLERGIC RHINITIS

Druce HM. Allergic and nonallergic rhinitis. In: Middleton E, Reed CE, Ellis EF, et al, eds. Allergy Principles and Practice. 4th ed. St Louis: Mosby, 1993.

Lemanske RF. A review of the current guidelines for allergic rhinitis and asthma. J Allergy Clin Immunol 1998;101:5392–5396.

Meltzer EO. Treatment options for the child with allergic rhinitis. Clin Pediatr 1998;37:1–10.

Naclario RM. Allergic rhinitis. N Engl J Med 1991;325:860–869.

Rachelefsky GS. Pharmacologic management of allergic rhinitis. J Allergy Clin Immunol 1998;101:5367–5369.

CHAPTER 93

Gastrointestinal Emergencies

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GASTROINTESTINAL BLEEDING

Gastrointestinal (GI) bleeding is a common and occasionally life-threatening condition in infants and children. An orderly approach to this problem is essential and is outlined in Chapter 30. One of the most important initial steps in establishing the cause of GI bleeding in children is determining whether the source of bleeding is the upper or lower intestinal tract. The following discussion of general principles of management, as well as specific diagnoses is, therefore, organized accordingly. Only those conditions most appropriately classified as medical diagnoses are described in this chapter. Additional causes of GI bleeding that are more appropriately classified as surgical diagnoses, including intestinal malrotation with volvulus, intestinal duplications, intussusception, and Meckel's diverticulum, are discussed in detail in Chapter 118.

General Principles of Management

In contrast to the adult experience, most children presenting to the emergency department (ED) with either upper or lower GI bleeding have not experienced significant blood loss. Most children can be managed successfully with judicious laboratory investigation, conservative supportive care, and follow-up with the patient's primary care provider or an appropriate subspecialist. A detailed discussion of pertinent laboratory evaluation and initial management for each of the most common causes of GI bleeding in children is provided in the following sections.

Severe GI bleeding should be considered a potentially life-threatening emergency that may require the cooperation of a team, including the emergency physician, surgeon, and gastroenterologist. Similar to the management of all potentially life-threatening conditions in children, the initial approach to the child with significant GI hemorrhage begins with an assessment of the child's airway, breathing, and circulation. A child with overt hemodynamic instability or suspected significant volume loss should be positioned with legs elevated and given nasal oxygen. Patients who have hematemesis should have the head elevated 30 to 45 degrees to lessen the chance of pulmonary aspiration of blood. In massive upper intestinal bleeding, protecting the airway with an endotracheal tube may be lifesaving. The next priority is the insertion of two large-bore intravenous catheters (14- to 20-gauge in the child and at least 22-gauge in small infants). A percutaneous central venous line or cutdown should be placed if the person administering the intravenous line has difficulty obtaining adequate peripheral venous access; the intraosseous route provides a temporary alternative.

Immediate blood studies in any patient with severe GI bleeding should include 1) type and crossmatch, 2) complete blood count (CBC), 3) platelet count, 4) prothrombin time (PT), and 5) partial thromboplastin time (PTT). Additional laboratory studies may be indicated based on the differential diagnosis of the most likely cause of the patient's bleeding, and are discussed later in this section. These studies should be done at the time of insertion of the intravenous lines. Arterial blood gases are also important parameters to follow in severe blood loss associated with shock. The hematocrit is an unreliable initial index of acute blood loss because it may be normal or only slightly decreased. Its subsequent fall will depend on 1) the rate and type of fluid replacement and 2) the body's own hemostatic mechanisms, resulting in both renal conservation of fluid and electrolytes and gradual shifts of fluid from extravascular to intravascular compartments.

Intravenous therapy has two major objectives: 1) restoration of intravascular volume (reflected in blood pressure or pulse)

and 2) restoration of oxygen-carrying capacity (reflected in hemoglobin and hematocrit values). The former objective can be accomplished both by nonsanguineous crystalloid or colloid solution or blood products, whereas the latter objective can be accomplished solely by the infusion of blood. The practical limitations of time required to properly type and crossmatch blood make nonsanguineous solution the mainstay of early resuscitation. In the rare circumstance of massive, ongoing hemorrhage in which low oxygen-carrying capacity is believed to be an important factor at onset of resuscitation, Rh and type-specific blood can usually be provided in 10 to 15 minutes. If type-specific blood is not immediately available, O-negative blood may be used. In most cases, proper type and crossmatch can be performed while intravascular volume is restored by nonsanguineous solutions. The exact type of solution to be used is controversial. Studies in both animals and humans have shown a reduction in intravascular as well as extravascular volume in acute blood loss; therefore, the preferred method is manual infusion of crystalloid solutions, such as normal saline or Ringer's lactate, in 20 mL/kg boluses until intravascular volume is minimally restored as indicated by a rise in blood pressure and disappearance of clinical signs of peripheral vasoconstriction. Colloid solutions such as albumin, plasma, or Hetastarch should be used only when blood loss is massive and continuous because in this situation respiratory insufficiency or shock lung may develop with a fall in plasma oncotic pressure. Dextran is to be avoided because it may affect platelet function. Patients who are in shock at the time of admission should have the urinary bladder catheterized in the ED to accurately measure urine output and to allow for early detection of acute tubular necrosis.

Overexpansion of intravascular volume is potentially dangerous, particularly in bleeding varices but also in bleeding gastric or duodenal ulcers. Therefore, after correction of shock and restoration of urine flow, further intravenous volume replacement should be titrated to match continuing blood loss. The decision to begin transfusion depends on the level of hematocrit taken at the time of restoration of blood volume and on evidence of ongoing bleeding. For a patient who has stopped bleeding, blood transfusion is given to allow some reserve in case of rebleeding. Under most circumstances, slow transfusion to return hematocrit to approximately 30% is recommended to achieve this objective. In this case, packed cells (10 mL/kg) are used to reduce the volume load to the patient. In addition, packed blood cells contain considerably less ammonia than whole blood, an important factor for patients who have severe liver disease. For a patient who has continuous bleeding, ongoing blood transfusion is the only means of maintaining adequate oxygen-carrying capacity. In this case, the rate of bleeding determines the rate of transfusion. A sustained rate of transfusion is recommended and is best achieved with an electrical infusion pump, not by gravity. Potential complications of massive transfusions include hypercitrinemia, hyperlacticacidemia, hypocalcemia, decreased levels of clotting factors, and thrombocytopenia. The risks inherent in massive transfusions are definitely lowered by using packed red blood cells, fresh-frozen plasma, proper filters, and blood warmers. Any patient with or without a previous history of liver disease who presents with GI bleeding associated with an abnormal PT should receive vitamin K (5 to 10 mg intramuscularly or intravenously) as soon as possible.

UPPER GASTROINTESTINAL BLEEDING

Background

Upper GI bleeding is generally regarded as originating proximal to the ligament of Treitz. Hematemesis or bloody gastric aspirates from a nasogastric tube may originate from the mouth, nasopharynx, esophagus, stomach, biliary tree, or duodenum. The most common causes of upper GI bleeding in children are mucosal lesions, including esophagitis, gastritis, Mallory-Weiss tear, peptic ulcer disease, and duodenitis. Less common but important causes include bleeding esophageal varices and vascular lesions. The profile of common diagnoses has changed recently because increasing use of endoscopy enables specific, often microscopic, diagnoses to be substituted for previously documented “bleeding of unknown origin.” Endoscopy, when performed by a well-trained physician, is the most sensitive and specific diagnostic procedure for determining the cause and site of upper GI bleeding. Specific diagnosis should be pursued in patients who have 1) active bleeding documented by nasogastric lavage; 2) evidence of severe hemorrhage (hemodynamic instability or equilibrated hemoglobin level less than 10 g/dL); 3) conditions that affect healing or clotting, such as catabolic state or serious chronic disease; 4) a history of previous unexplained gross or occult bleeding or unexplained iron deficiency anemia; or 5) a history of chronic dyspepsia (vomiting, abdominal pain, nausea, oral regurgitation, heartburn, dysphagia).

The pathophysiology, clinical manifestations, and specific management issues related to each of the most common causes of upper GI bleeding in children are discussed. As noted in Chapter 30, an upper tract source for GI bleeding is often indicated by a history of hematemesis or by obtaining fresh (red) or old (coffee ground) blood via gastric lavage after placement of a nasogastric tube.

General Principles of Nasogastric Lavage

Not every child with a history of possible upper GI bleeding requires nasogastric lavage. Patients who have a history of acute self-limited hematemesis of streaks of blood or a small amount of coffee-ground material in the context of forceful emesis, recurrent gastroesophageal reflux, symptoms of infectious gastroenteritis, or epistaxis can often be managed presumptively without gastric lavage. However, nasogastric lavage should be performed in all patients suspected of having significant GI bleeding that is indicated by either history or physical examination (e.g., pallor, unexplained tachycardia, poor perfusion). The purpose of gastric lavage is to confirm the level of bleeding and to estimate the rate of bleeding. There is no evidence that gastric lavage has any therapeutic role in controlling hemorrhage. It is important to realize that a clear nasogastric aspirate does not exclude major bleeding from the upper GI tract.

Most patients can be effectively lavaged with a nasogastric sump tube (12 Fr in small children; 14 to 16 Fr in older children). Verification of the location of the tube in the stomach by injection of air and auscultation over the stomach is essential. The recommended volume for each saline infusion depends on age: 50 mL for infants, 100 to 200 mL for older children. With the patient's head elevated 30 degrees, the solution, at room temperature, is rapidly infused into the stomach, allowed to stand for 2 to 3 minutes, and then aspirated by gentle suction. Return volumes should approximate input volumes, and discrepancies should be recorded. If aspiration meets with significant resistance, the physician should either reposition the tube, reposition the patient, or increase the amount of solution introduced. Saline lavage of

the stomach should be performed by two people. One person fills and empties the stomach while the other person empties and fills the syringes.

Blood-flecked gastric aspirate or coffee-ground material indicates a low rate of bleeding. In contrast, bright red blood, especially if it does not clear with repeated lavage for 5 to 10 minutes, suggests a significant or ongoing hemorrhage. No benefit is derived from continuous lavage longer than 10 minutes if return is not clearing. The tube can be left to gravity or low suction and irrigated every 15 to 30 minutes to assess the activity of the bleeding. The presumed lesion causing the bleeding determines the subsequent management of the patient.

Nonspecific Mucosal Lesions

Background

GI bleeding may be a complication of all acute and chronic nonspecific upper GI mucosal lesions (esophagitis, gastritis, Mallory-Weiss tears, and duodenitis). Regardless of the cause, upper GI bleeding usually stops spontaneously, often by the time the patient arrives in the ED. Esophagitis as a result of gastroesophageal reflux (GER) is being diagnosed more often with improved pediatric fiberoptic endoscopes. Increasing use of endoscopic biopsy has documented esophagitis in 60% of patients with clinically significant GER. Exposure to aspirin and nonsteroidal anti-inflammatory drugs (NSAIDs) (e.g., ibuprofen, naproxen) has also been associated with gastritis and mucosal ulceration. A recent large, randomized clinical trial designed to assess the risk of serious GI bleeding (requiring hospitalization) in febrile children after the use of ibuprofen demonstrated no greater risk than with acetaminophen. The study was not able to assess less serious (and clinically inapparent) occurrences of GI bleeding. Mallory-Weiss tears are mucosal lacerations of the gastric cardia or gastroesophageal junction induced by retching or vomiting. These lesions are relatively rare in children, accounting for approximately 5% of cases of upper GI bleeding.

Pathophysiology

The upper GI mucosa bleeds when an ulcerating process erodes into a blood vessel, usually an artery in the base of the ulcer. In most cases, normal mechanisms of thrombosis and healing stop the bleeding and prevent recurrent bleeding. Erosion of larger arteries, however, in which blood flow exceeds the capacity of normal hemostasis, results in continuous hemorrhage. A more common scenario is that thrombosis temporarily stops the bleeding, but aneurysmal dilation of the artery in the recanalization process or continuing arteritis from the chemical irritation of acid digestion facilitate recurrent hemorrhage. Acid and pepsin also produce profound adverse effects on platelet aggregation and plasma coagulation. The pathogenesis of Mallory-Weiss tears involves the production of transient large gradients between the intragastric and intrathoracic pressures at the gastroesophageal junction as a result of forceful retching. The gradient results in dilation of the gastroesophageal junction, and thus, tears.

Clinical Manifestations

Diagnosis is usually suspected by an antecedent history of vomiting and/or abdominal pain and absence of physical findings suggestive of chronic liver disease or portal hypertension. Reflux esophagitis is suspected in infants, usually less than 1 year of age, who have a history of recurrent nonprojectile emesis, “wet burps,” after feeding, or a documented diagnosis of GER and who present with emesis that is blood streaked or contains a small amount of coffee-ground material. Infants may be fussy but consolable and may have been previously diagnosed with colic. Reflux esophagitis should also be suspected in infants with guaiac-positive stools or iron deficiency anemia. A history of repeated aspirin or NSAID use for control of fever and/or pain should also prompt suspicion for gastric mucosal lesions as the cause of upper GI bleeding. Mallory-Weiss tears should be suspected in older children with a history of protracted forceful vomiting and streaks of hematemesis appearing after several episodes of nonbloody emesis.

Management

For patients who have significant bleeding and for whom nasogastric lavage was initiated, if gastric contents clear following initial saline lavage and immediate endoscopy is not planned, gastric irrigation is performed every 15 minutes for 1 hour, then every hour for 2 to 3 hours. Antacid (e.g., Maalox®, 15 mL, infants; 30 mL, preschool-age children; 60 mL, school-age children) is infused into the tube after every lavage. The dosage of antacid is titrated during the hourly monitoring so that gastric pH 1 hour after antacid infusion is greater than 3.5. If the patient is hemodynamically stable and gastric return remains clear for the aforementioned period, the tube is electively removed. Persistent nausea or vomiting or the presence of ileus points to the need for continued drainage.

Patients with nonspecific mucosal lesions theoretically should benefit from neutralization of intragastric acidity by antacids and reduction of gastric acid and pepsin secretion by H_2-receptor antagonists. For patients with significant symptoms or blood loss, H_2-antagonists may be given initially by the intravenous route, switching to the oral route when the nasogastric tube is removed. Either ranitidine (1.0 to 1.5 mg/kg per dose intravenously every 6 hours or 2 mg/kg per dose orally two times a day) or cimetidine (6.0 to 7.5 mg/kg per dose intravenously every 6 hours or 10 mg/kg per dose orally four times a day) is appropriate. For patients who have acute self-limited bleeding and who are not considered candidates for endoscopy, oral H_2-antagonists are continued for 2 to 4 weeks, at which time they are empirically discontinued if the patient is asymptomatic.

In general, all patients with a history suggestive of significant upper GI bleeding should be admitted to the hospital for observation. A clear nasogastric aspirate should never be used as an indication to discharge a patient from the ED if the history suggests significant bleeding. The main reason for admission is the unknown incidence of rebleeding from these lesions in children. Unremitting or recurrent mucosal bleeding requires either therapeutic endoscopy, therapeutic angiography, or surgery. In the hands of a qualified endoscopist, therapeutic endoscopy using either a heater probe or

multipolar electrocoagulation is the treatment of choice.

Esophageal Varices

Background

Portal hypertension may result from either extrahepatic presinusoidal obstruction (50 to 65% of cases in children) or from hepatic parenchymal disorders. Extrahepatic obstruction (e.g., portal or splenic vein obstruction) is associated with omphalitis, dehydration, sepsis, and umbilical vein catheterization. Hepatic parenchymal disease may result from biliary cirrhosis associated with biliary atresia, cystic fibrosis, hepatitis, a $_1$-antitrypsin deficiency, or congenital hepatic fibrosis. Patients with both types of portal hypertension are susceptible to GI hemorrhage from bleeding esophageal varices and from congestive or hemorrhagic gastritis. After development of portal hypertension, the onset of esophageal varices can be variable, from a few months to many years.

Pathophysiology

Portal hypertension results from relative obstruction of portal venous blood flow, leading to the development of portal systemic collateral veins, or varices. Portal-systemic collaterals will develop in any area where veins draining the portal venous system are in close approximation to veins draining into the caval system (i.e., submucosa of the esophagus, submucosa of the rectum, and anterior abdominal wall). Esophageal and gastric fundal varices, connecting branches of the coronary veins with branches of the azygous vein, are the most likely to be the site of spontaneous hemorrhage (Fig. 93.1).

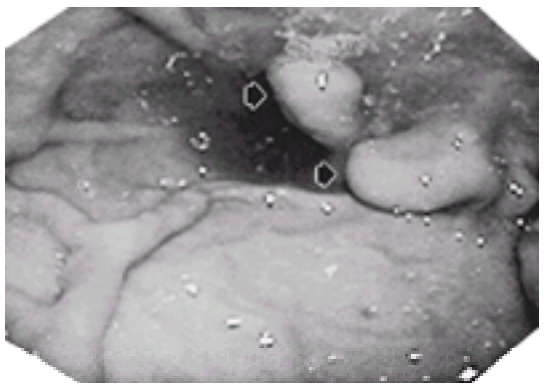

FIGURE 93.1. Gastric varices. The arrows represent 2 large blood-filled varices in the gastric cardia.

Clinical Manifestations

Patients with portal hypertension may have occult bleeding, but more commonly, the bleeding is brisk, and patients will have melena and/or hematemesis. The possibility of bleeding esophageal varices should be considered in any patient with a history of jaundice (beyond the newborn period), hepatitis, blood transfusion, chronic right-sided heart failure, pulmonary hypertension, omphalitis, umbilical vein catheterization, or one of the hepatic parenchymal diseases previously noted. Accordingly, the physical examination may reveal stigmata of the underlying disease leading to portal hypertension, including jaundice, ascites, rectal hemorrhoids, and hepatosplenomegaly.

Management

The initial management of suspected variceal hemorrhage is identical to that of massive upper GI bleeding from any source. Overexpansion of the intravascular volume should be avoided because it contributes to rebleeding. Coagulation abnormalities should be managed aggressively with intravenous vitamin K, fresh-frozen plasma, and platelets. Bleeding varices may be the initial sign of sepsis in patients who have cirrhosis; therefore, any patient who has fever should be started on broad-spectrum antibiotics such as ampicillin 200 mg/kg per day and gentamicin 5 to 7.5 mg/kg per day pending results of blood cultures.

Suspicion of variceal bleeding is not a contraindication to pass a nasogastric tube. If bleeding ceases during the initial gastric lavage, the tube should be managed as previously described. Antacids and H $_2$-antagonists are given in the doses used for mucosal lesions. Pharmacologic therapy of acute variceal hemorrhage uses the splanchnic arterial constrictor vasopressin and the prokinetic agent metoclopramide. Vasopressin administration has been well documented to decrease blood flow and pressure through the portal circulation. Infusion may be initiated before diagnostic endoscopy if variceal bleeding is suspected. The physician should begin infusing 0.1 unit/minute and increase the dosage by 0.05 unit/minute hourly up to a maximum of 0.2 unit/minute in children less than 5 years old, 0.3 unit/minute in children 5 to 12 years old, and 0.4 unit/minute in adolescents more than 12 years old. Side effects can be significant; thus, the child must be monitored carefully. Major complications include myocardial ischemia, life-threatening arrhythmias, and limb vasoconstriction or ischemia. Minor complications include water retention with sodium depletion, benign arrhythmias, and acrocyanosis. The vasopressin is usually given in 5% dextrose in water; the exact dilution is based on overall volumes of fluids being infused. Infusing vasopressin through a large-bore, preferably central venous, line is the safest method. The reported success rate of vasopressin infusion in adults is 50 to 70%. Because of the high rate of rebleeding, once begun, the drug should be continued at the dosage that controls bleeding for a minimum of 12 to 24 hours after all bleeding has stopped. This management plan stems from studies showing sustained vasoconstrictive effects of vasopressin on splanchnic vessels in dogs for more than 24 hours. However, this point is controversial because tachyphylaxis reportedly

also develops with prolonged use of vasopressin.

If bleeding continues after the use of vasopressin, emergency flexible endoscopy should be performed as soon as the patient's vital signs have been stabilized. Actively bleeding esophageal varices or an overlying clot on a varix confirms variceal bleeding. The endoscopist may choose to perform emergency sclerotherapy at that point. Alternatively, sclerotherapy may be delayed until hemorrhaging has been controlled by pharmacologic agents or balloon tamponade, especially if the endoscopist has difficulty obtaining a clear field of vision. Sclerotherapy should not be considered a therapeutic option to control bleeding gastric varices.

Gastroesophageal balloon tamponade is a highrisk procedure. It should be considered only for endoscopically proven gastric or esophageal varices. Indications of these varices include massive, life-threatening hemorrhage or continued bleeding despite 2 to 6 hours of intravenous vasopressin. Either a Sengstaken-Blakemore (S-B) (Fig. 93.2) or a Linton tube may be used. The S-B tube has both gastric and esophageal balloon tubes, whereas the Linton tube has a single lavage gastric balloon. Gastroesophageal tamponade is reported to arrest bleeding initially in 50 to 80% of cases. However, the reported incidence of major complications from use of the S-B tube ranges from 9 to 35%. Death directly attributed to the use of the tube has been reported in 5 to 20% of patients on whom the tube was used. Other major complications include rupture or erosion of the esophageal or gastric fundal mucosa, occlusion of the airway by the balloon, and aspiration of secretions resulting from inadequate drainage of the occluded esophagus.

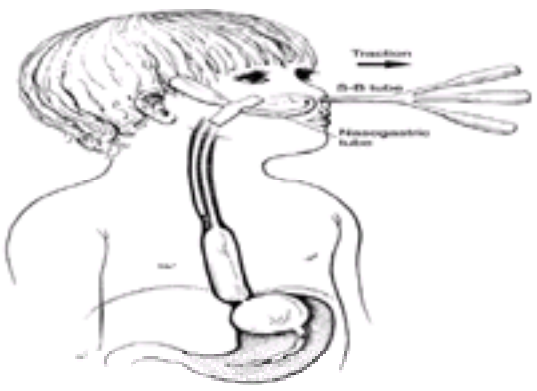

FIGURE 93.2. Gastroesophageal tamponade using a Sengstaken-Blakemore tube.

When a hemostatic tube is used, it should be inserted only by a physician who is skilled in its use. The S-B tube is preferred (Fig. 90.2). A pediatric tube is used for children less than 11 to 13 years old; the adult tube is used in adolescents. Passage through the nose is the procedure of choice, but in small children, passage through the mouth may be necessary. Intranasal oxymetazoline hydrochloride (0.05%) is used for local vasoconstriction. Each balloon is pretested for air leaks before insertion. The tube is heavily lubricated and passed through the nose to its full length; 50 to 100 mL of air is instilled into the gastric balloon and the balloon pulled up until the resistance of the gastroesophageal junction is encountered. An emergency radiograph is then performed to ensure correct balloon position immediately below the diaphragm. Additional air is then instilled into the gastric balloon, up to 150 mL in the pediatric balloon and 250 mL in the adult balloon; the tube is pulled taut and is taped at the nose. In many cases, the gastric balloon alone will stop bleeding by preventing flow of blood from the stomach into the esophagus or by directly occluding a gastric varix. It is absolutely essential to pass a second nasoesophageal tube to aspirate saliva and blood from ongoing esophageal hemorrhage. If esophageal bleeding continues, the esophageal balloon is inflated with air. The volume in the esophageal balloon is determined by the pressure within the balloon, which is measured by connecting its inflow tube to a sphygmomanometer. The pressure is maintained at the minimum required to control bleeding, with the maximum being no more than 40 mm Hg.

If bleeding is controlled, the balloons are kept inflated for 24 hours. The esophageal balloon is deflated first, then the gastric balloon approximately 1 hour later. The gastric tube is irrigated every 30 minutes for 1 to 2 hours more, and then the tube is removed.

A patient with an indwelling S-B tube must be transferred to an intensive care unit. A pair of scissors should be kept at the bedside; if any respiratory embarrassment occurs, the tube should be cut immediately, thereby deflating the balloons.

Endoscopic sclerotherapy should be performed on an elective basis within 6 to 24 hours after active bleeding has been controlled by pharmacologic therapy or balloon tamponade. Recent reports have noted success with endoscopic variceal ligation (EVL) using an elastic band ligature device in children. Randomized studies of endoscopic variceal sclerotherapy (EVS) versus EVL in adults have demonstrated that EVL has a lower complication rate, less recurrent bleeding, and better variceal eradication than EVS. In a recent case series of EVL in children, 55% of children achieved variceal eradication with EVL after an average of four treatment sessions. Urgent shunt surgery is indicated only if two attempts at sclerotherapy fail to control active or recurrent bleeding. An alternative to shunt surgery in patients with cirrhotic portal hypertension is esophageal transection with or without devascularization.

Miscellaneous Causes of Upper Gastrointestinal Bleeding

In the first few days of life, or in breast-fed infants, *swallowed maternal blood* may be the cause of hematemesis or melena in an infant who otherwise appears healthy. Performing a guaiac test on expressed breast milk may suggest the diagnosis. An Apt-Downey test should be performed on a sample of emesis or nasogastric aspirate to definitively diagnose the condition. Blood from the aspirate is placed on filter paper and mixed with 1% NaOH. Adult hemoglobin will be reduced to form a rusty brown or yellow color. Fetal hemoglobin is resistant to denaturation and will retain a bright

pink or red color.

Dieulafoy lesion is an unusual cause of GI bleeding in which massive hemorrhage occurs from a pinpoint nonulcerated arterial lesion, usually high in the fundus of the stomach. The bleeding results from an unusually large submucosal artery that travels a tortuous course through the submucosa and may erode through a mucosal defect. Its characteristic presentation is one of recurrent, massive hematemesis, usually without any prodromal symptoms. This diagnosis is primarily made in adults, but patients as young as 20 months have been diagnosed with a Dieulafoy lesion, and most series contain a number of teenagers. Management is similar to that for any patient with a significant GI hemorrhage. Diagnosis can be made by endoscopy, during which the Dieulafoy lesion can usually be located.

Finally, swallowed foreign bodies can cause significant trauma and GI bleeding. Most swallowed foreign bodies, even those with sharp edges, will pass spontaneously and require no specific therapy. However, on occasion, a sharp foreign body may be the cause of GI bleeding (Fig. 93.3). Removal by endoscopy is indicated if significant bleeding occurs.

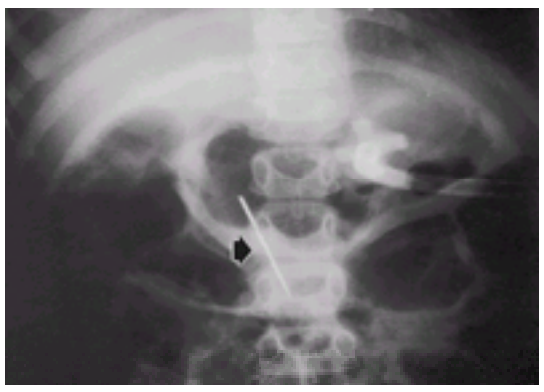

FIGURE 93.3. Gastric foreign body. This 6-year-old developmentally delayed patient ingested a large straight pin that caused bleeding of the gastric antrum. The arrow represents the foreign body. An opaque gastrostomy tube is also radiographically present.

LOWER GASTROINTESTINAL BLEEDING

Background

Rectal bleeding is a relatively uncommon but worrisome complaint in the ambulatory or ED setting. A recent case series of children presenting with rectal bleeding to the ED at Boston Children's Hospital indicated that rectal bleeding was involved in the chief complaint of 0.3% of all ED visits during a 1-year period. The average age of patients was approximately 5 years, with nearly half of the patients less than 1 year old. No patient in the series was judged hemodynamically unstable in the ED, nor did any patient require a blood transfusion. The most common presentation was for hematochezia (98% of patients), with 10% of patients presenting with melena (some patients presented with both complaints). Diarrhea (37% of patients), abdominal pain (43%), and constipation (22%) were the most common associated symptoms, with only 2% of patients presenting with fever. Presumptive diagnoses were made in two-thirds of patients, most of which (81%) did not change with follow-up. Potentially life-threatening disorders (intussusception and Meckel's diverticulum) were found in 4% of cases.

The cause of lower GI bleeding varies with age. Among infants less than 6 months of age, the most common diagnoses are milk-protein sensitivity (allergic colitis), anorectal fissures, and infectious gastroenteritis. Children 1 to 5 years of age are most likely to have infectious gastroenteritis, intussusception, Meckel's diverticulum, colonic polyps, and anorectal fissures. Older children typically have infectious gastroenteritis, inflammatory bowel disease (IBD), and hemorrhoids/rectal varices. The pathophysiology, clinical manifestations, and specific management indicated for the most common conditions causing lower GI bleeding in children are discussed in the following sections.

Anorectal Fissures and Hemorrhoids

Anal fissures are the most common proctologic disorder during infancy and childhood. Most occur in infants less than 1 year of age. Anal fissure may result from diarrhea, which causes perineal irritation, but it is more commonly associated with constipation. The fissure usually starts when passage of a hard stool tears the sensitive squamous lining of the anal canal. Subsequent bowel movements are associated with pain and/or bleeding. Bright red blood is seen coating the stool. The infant begins to withhold stool, leading to increasing constipation and a vicious cycle of hard stools, bleeding, and pain. Anal fissure can be seen by spreading the perineal skin to evert the anal canal. Simply spreading the buttocks to view the anal opening is not sufficient. Treatment consists of local skin care combined with stool softeners. Malt extract (Maltsupex 1 to 3 tablespoons/day) or mineral oil (1 to 3 tablespoons/day) can be given to soften the stool. Local care involves sitz baths four times a day, a perianal cleansing lotion (Balneol) after bowel movements, and an emollient protective ointment (Balmex) after each bowel movement.

Small varicosities of the external hemorrhoidal plexus (i.e., hemorrhoids) may occur in the healing process associated with anal fissure. They rarely cause pain or bleeding. Therapy is directed at treatment of the anal fissure. The presence of external hemorrhoids does not imply associated internal hemorrhoids. The latter may develop in response to portal hypertension and may be a cause of painless rectal bleeding.

All patients with perianal excoriation, multiple anal fissures, recurrent anal fissure, or fissure resistant to conservative management should have perianal cultures for b-hemolytic streptococcus. If this organism is recovered, the patient

should receive a 7-day course of oral penicillin.

Polyps

There are two major types of polyps that may be diagnosed in infancy or childhood: hamartomatous and adenomatous. Hamartomatous polyps are generally benign and are the usual type of polyp found in juvenile polyps, juvenile polyposis coli, and Peutz-Jeghers syndrome. Adenomatous polyps are potentially premalignant and are found in a number of syndromes, including familial adenomatous polyposis and Gardner's syndrome.

Juvenile polyps are the most common of the polyp syndromes in children, found in 15% of patients in one series who had colonoscopy for rectal bleeding (Fig. 93.4). More than one polyp may be found in more than 50% of cases of juvenile polyps. Most (75%) of the polyps are rectosigmoid or in the descending colon, 15% are found in the transverse colon, and 10% in the ascending colon. Autoamputation of juvenile polyps, especially in the rectum, occurs spontaneously in most cases. In juvenile polyposis coli, multiple juvenile polyps are found throughout the colon. Peutz-Jeghers syndrome is the association of mucocutaneous pigmented lesions and hamartomatous polyps. It has autosomal-dominant inheritance with a high degree of penetrance. The macular, melanin-containing pigmented lesions characteristically occur on the buccal mucosa, lips, face, arms, palms and soles, and perianal region. The polyps are typically located in the small intestine but can be found throughout the GI tract.

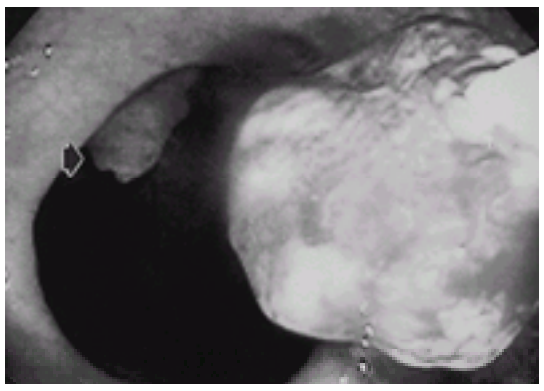

FIGURE 93.4. Juvenile polyp. The arrow demonstrates the cauterized polyp stalk.

Familial adenomatous polyposis is an autosomal-dominant inherited syndrome consisting of multiple adenomatous polyps that are generally confined to the colon but that can be found throughout the GI tract. A 6% incidence of malignant transformation of these lesions is present by age 15 years, prompting recommendations for total proctocolectomy by age 18. Gardner's syndrome is an autosomal-dominant inherited syndrome consisting of hereditary adenomatous polyps of the small and large intestine and soft tissue and bony tumors. The tumors are often epidermoid cysts, fibromas, or osteomas of the skull and mandible and are often the initial manifestation of the disease.

Pathophysiology

Juvenile polyps are proliferations of mature colonic epithelium with aggregates of lymphoid tissue and cystic dilation of normal glandular elements. This histopathology has prompted the use of other terms such as *retention, inflammatory,* or *hyperplastic polyps.* The surface epithelium is often ulcerated, with a loss of mucosal surface. Adenomatous polyps may appear grossly similar to juvenile polyps, although on microscopic examination, adenomatous polyps are distinguished by the amount of cellular atypia seen within colonic epithelial cells.

Clinical Manifestations

The most common presentation for juvenile polyps is painless rectal bleeding, often with blood streaking the outside of the stools. The peak incidence for presentation is between 4 and 5 years of age. Prolapse of the polyp through the rectum may occur. The polyp may also form the “lead point” of an intussusception. All patients with rectal bleeding should have a careful rectal examination because 30 to 40% of polyps are palpable by rectal examination.

Polyps may be part of various inherited syndromes; therefore, a complete physical examination should be performed on any patient with rectal bleeding. A careful search for pigmented lesions or soft tissue and bony tumors may aid in the diagnosis of inherited polyposis syndromes as previously described.

Management

The initial ED management of patients who have suspected polyps is aimed at assessing the amount of blood loss and arranging the appropriate diagnostic study. Blood loss is rarely life-threatening, but significant losses may be noted from chronic intermittent bleeding. All patients should have a CBC performed, and if the history of blood loss is significant, a type and crossmatch may also be indicated. Patients with palpable polyps on rectal examination should have elective flexible colonoscopy. Endoscopic removal of a polyp is safe and effective therapy even in a young child. For patients with painless hematochezia and for whom rectal examination is negative, a Meckel's diverticulum must also be considered. This possibility may prompt a decision to perform a ^{99m}Tc radionucleotide scan before colonoscopy in an effort to identify the possible location of bleeding.

Dietary Protein Sensitivity Syndromes (“Allergic Colitis”)

Dietary proteins are capable of inducing significant bowel injury and may be the cause of several different types of enterocolitis presenting throughout childhood. Each condition, by definition, is induced by a dietary protein and resolves completely after the protein is eliminated from the diet. Immunologic responses may vary from classic allergic mast cell activation to immune complex formation. The development of proctocolitis in response to cow's milk protein exposure was among the first to be described. Subsequently, a similar condition has been described in response to soybean-based formula and among exclusively breast-fed infants, presumably in response to maternal dietary protein intake.

Pathophysiology

The appearance of the rectum and colon on colonoscopic examination characteristically consists of diffuse inflammation, friability, edema, and frequent focal ulcerations. Rectal biopsies demonstrate both acute and chronic inflammatory changes and eosinophilic infiltration is often present.

Clinical Manifestations

The typical presentation of milk-protein sensitivity colitis is that of acute onset of blood-streaked, mucoid diarrheal stools in an otherwise well-appearing infant less than 6 months of age. Mean age of onset among 35 infants in one series was 4.3 + 4.1 weeks. It is unusual to present within the first week of life. Blood loss is typically limited, infants do not appear acutely dehydrated and are afebrile, and weight gain has typically been within normal limits since birth. The differential diagnosis includes anal fissures and infectious enterocolitis. External anal fissures can be ruled out by careful physical examination. Appropriate viral and bacterial cultures of stool may be indicated to rule out infectious causes.

Management

These patients are rarely hemodynamically unstable or seriously ill; therefore, initial ED management is focused on making a presumptive diagnosis based on initial laboratory testing, initiation of appropriate dietary therapy, and arranging adequate follow-up with the patient's primary care physician or a pediatric gastroenterologist. Initial laboratory testing should consist of a CBC with white blood cell (WBC) differential, assessing the hemoglobin as well as assessing for leukocytosis and eosinophilia. Patients with histologically proven milk-protein sensitivity colitis have higher mean peripheral eosinophil counts compared with age-appropriate normal values. However, in the individual patient, a higher-than-normal eosinophil count is actually an insensitive marker (sensitivity = 10%) for histologically proven colitis. In addition, a serum albumin level should be obtained because hypoalbuminemia has been demonstrated to have a sensitivity of approximately 80% for histologic colitis. Examination of stool for blood, fecal leukocytes, and routine bacterial culture should be performed on all infants. Infants who have milk-protein sensitivity colitis will characteristically have leukocytes seen on fecal smear, although eosinophils may not be present in the stool.

Treatment consists of elimination of the offending protein from the infant's diet. The diagnosis is typically confirmed by the rapid resolution of symptoms within 72 hours of the dietary change. Infants receiving cow's milk–based or soy protein formulas should be changed to a formula containing casein hydrolysate as the protein source. (Nutramigen, Pregestimil, and Alimentum are currently available in the United States.) Gross symptoms of allergic colitis respond within a few days to elimination diet therapy, although guaiac-positive stools may persist for several weeks. In exclusively breast-fed infants, elimination of the offending protein from the mother's diet also leads to clinical improvement, and breast-feeding can usually be continued. Persistent evidence of gross bleeding for 5 to 7 days following formula change is indication for flexible proctosigmoidoscopy. Most infants who come to endoscopy are found to have nodular lymphoid hyperplasia. Infants who do respond to dietary elimination should be rechallenged with the original formula in 4 to 6 weeks. Parents should be counseled that symptoms of allergy may change with increasing age such that a positive challenge may evoke vomiting, diarrhea, or GI signs of allergy rather than recurrent rectal bleeding.

Infectious Enterocolitis

Infectious causes of GI bleeding are predominantly a result of bacterial pathogens, including *Campylobacter,* pathogenic *Escherichia coli, Salmonella,* and *Shigella.* Less commonly, infection with *Giardia* or rotavirus is associated with heme-positive stools. A detailed discussion of the pathophysiology, clinical manifestations, and management of bacterial gastroenteritis can be found in Chapter 84.

Pseudomembranous colitis is a form of inflammatory colitis characterized by the pathologic presence of pseudomembranes consisting of mucin, fibrin, necrotic cells, and polymorphonuclear leukocytes. The entity develops as a result of colonic colonization and toxin production by the Gram-positive obligate anaerobe *Clostridium difficile,* in most cases after normal bowel microflora have been altered by antibiotic therapy. All classes of antibiotics have been associated with pseudomembranous colitis. Patients usually present with profuse diarrhea, tenesmus, and crampy abdominal pain, usually beginning during the first week of antibiotic therapy. Frank hematochezia is rare. The diagnosis and management of pseudomembranous colitis is further discussed in Chapter 84.

Miscellaneous Causes of Lower Gastrointestinal Bleeding

Henoch-Schönlein purpura (HSP) (see Chapter 86) is a systemic vasculitis that may cause edema and hemorrhage in the intestinal wall. Peak age of onset is between 3 and 7 years and the male:female ratio is 2:1. The presentation consists of the onset of a purpuric rash, typically confined to the buttocks and lower extremities, followed by arthralgias, angioedema, and diffuse abdominal pain. GI symptoms may precede the usual cutaneous symptoms and include abdominal pain (60 to 70%), occult bleeding (50%), gross bleeding (30%), massive hemorrhage (5 to 10%), and intussusception (3%). In a recent series, thickening of the duodenal wall was noted by ultrasonography in 82% of children who had HSP, with multiple hemorrhagic duodenal erosions noted by endoscopy in two patients. All children with

suspected HSP and GI symptoms should have a stool guaiac test performed, as well as a urinalysis to monitor for the onset of renal involvement (nephritis). Children with HSP limited to involvement of the skin and joints can often be managed as outpatients. However, severe abdominal pain or GI hemorrhage are indications for admission.

Hemolytic uremic syndrome (HUS) (see Chapter 86) is a disorder characterized by the triad of acute microangiopathic hemolytic anemia, thrombocytopenia, and oliguric renal failure. The disease is heralded by a prodrome of intestinal symptoms ranging from diarrhea (in 100% of patients) to hemorrhagic colitis (80%). Fever (20 to 30%), vomiting (75 to 80%), and abdominal pain (60%) are also commonly seen. Acute infectious gastroenteritis or colitis secondary to infection with *E. coli* O157:H7 is now considered the most important initial causative event in both sporadic and epidemic cases of HUS.

All children with HUS require admission to the hospital. Laboratory studies should be obtained, including a CBC, platelet count, PT, PTT, electrolytes, blood urea nitrogen (BUN), and creatinine. Intravenous access needs to be secured immediately for the correction of dehydration and the administration of blood products. As with HSP, the GI manifestations of HUS resolve, usually without sequelae or the need for antibiotic treatment of the initial intestinal infection.

GI *vascular malformations,* including hemangiomas, angiodysplasia, and arteriovenous malformations (AVMs), are rare causes of GI bleeding in children and are often seen as part of congenital syndromes. GI hemangiomas may be part of the Klippel-Trenaunay-Weber syndrome, which consists of a capillary or large vessel hemangioma on an extremity with hypertrophy of that limb. Diffuse visceral hemangiomatosis is rare, often fatal, and is always associated with cutaneous vascular lesions. GI hemangiomatosis should be suspected in any child with unexplained anemia and a syndrome of cutaneous hemangiomata.

Intestinal AVMs are rare in the pediatric age group, may occur both as solitary as well as multiple AVMs, and are typically part of a congenital syndrome (e.g., Osler-Weber-Rendu disease). Many GI vascular malformations, particularly cavernous hemangiomas and AVMs, can be detected using computed tomographic scans with intravenous contrast. Intestinal angiography or tagged red blood cell scans are often used to identify the source of bleeding during an acute hemorrhage. ED management of patients with GI bleeding from vascular malformations is the same as for any patient with potentially significant blood loss. After initial stabilization, referral to an appropriate subspecialist for diagnosis and definitive treatment is warranted.

INFLAMMATORY BOWEL DISEASE

Background

The term *inflammatory bowel disease* (IBD) is used to designate two chronic intestinal disorders of unknown origin: 1) ulcerative colitis, characterized by inflammation and ulceration confined to the colonic mucosa; and 2) Crohn's disease, manifested by transmural inflammation and frequent granulomas that may affect any segment of the GI tract. Approximately 25 to 30% of patients who have Crohn's disease and 20% of those who have ulcerative colitis present before age 20 years, most between the ages of 10 and 16 years, making these diagnoses relatively common in the pediatric age group. Many clinical features are common to both disorders, including diarrhea, GI blood and protein loss, abdominal pain, fever, anemia, weight loss, and growth failure. Extraintestinal manifestations involving the joints (arthritis), skin (erythema nodosum), eyes (uveitis), and liver (chronic hepatitis and sclerosing cholangitis) are seen with both disorders, although they are generally more common with Crohn's disease.

Ulcerative colitis invariably involves the rectum and extends proximally without skip areas (Fig. 93.5). In contrast, Crohn's disease has discontinuous, patchy involvement of the GI tract. The anatomic distribution of Crohn's disease in children is similar to that in adults; 40% of children have small bowel involvement, more than 50% have involvement of the ileum and colon, and about 10% have involvement of the colon alone (Fig. 93.6). Oral, esophageal, and gastric involvement is uncommon in children. The onset of both ulcerative colitis and Crohn's disease is usually insidious, consisting of growth failure, weight loss, diarrhea, and occult rectal bleeding, but may be more dramatic and extensive.

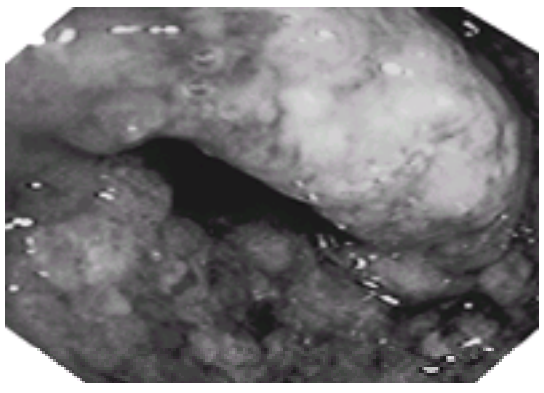

FIGURE 93.5. Severe ulcerative colitis. The mucosa appears granular, nodular, edematous and is actively bleeding.

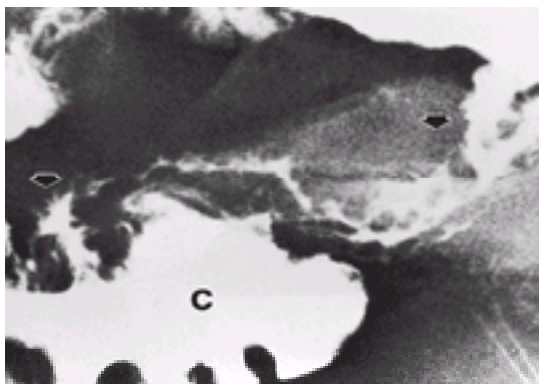

FIGURE 93.6. Crohn's disease of the terminal ileum demonstrated by severe narrowing of the terminal ileum (as shown between the two arrows). The cecum is represented by the “C.”

Pathophysiology

The cause of growth failure in patients with IBD is multifactorial, but inadequate nutrient intake is most likely the final common pathway. Growth failure is twice as likely in children who have Crohn's disease as it is in those who have ulcerative colitis. Malabsorption, especially with small bowel involvement of the disease, may lead to reduced assimilation of fats, vitamins, and minerals. Hematochezia, protein-losing enteropathy, and increased fecal losses of cellular constituents result from chronic inflammation and damage to the intestinal mucosa. The cause of diarrhea is also multifactorial, resulting from extensive mucosal dysfunction, bile acid malabsorption in terminal ileal disease, bacterial overgrowth secondary to strictures and disordered motility, and protein exudation from inflamed surfaces. Extraintestinal manifestations of the disease are often partially the result of a breakdown in the normal barrier and immunoregulatory functions of the GI tract as a result of chronic inflammation. This reaction enables bacterial products and inflammatory mediators (e.g., cytokines) to enter the circulation and subsequently to be deposited in various sites such as the eyes, skin, and joints, leading to localized inflammatory responses.

Clinical Manifestations

Clinical manifestations of IBD can be varied and related to either GI inflammation or the development of either GI tract or extraintestinal complications. Severe abdominal pain is among the most common complaints prompting an ED visit by the patient with IBD. Abdominal pain and diarrhea with or without occult blood are the most common symptoms at presentation. The pain is often colicky and, in Crohn's disease, may localize to the right lower quadrant or periumbilical area, prompting a consideration of acute appendicitis in the differential diagnosis. The abdominal examination may elicit guarding and rebound tenderness. Frank rectal bleeding occurs in fewer than 25% of all cases but is more common in ulcerative colitis. Perianal disease, including fissures, skin tags, fistulae, and abscesses, occurs in 15% of children with Crohn's disease. Perianal disease may precede the appearance of the intestinal manifestations of Crohn's disease by several years.

A low-grade fever and mild leukocytosis commonly occur. Approximately 10% of children with ulcerative colitis and a lesser percentage of those with Crohn's disease present with a fulminant onset of fever, abdominal cramps, and severe diarrhea with blood, mucus, and pus in the stools. A fulminant episode may also occur in the patient who has a known disease. There may be associated anemia and dehydration. IBD occasionally causes massive lower GI bleeding. Rarely, Crohn's disease causes complete intestinal obstruction. The patient always gives a history of antecedent abdominal pain, diarrhea, and weight loss. The presence of abdominal distention, accompanied by diminished or absent bowel sounds, should raise the suspicion of actual or impending perforation, even in the absence of severe pain. Perforation may occur after even minor abdominal trauma and must be ruled out when patients with known IBD complain of abdominal pain after trauma.

The development of massive colonic distension is a rare complication of both ulcerative colitis and Crohn's disease. Toxic megacolon represents a life-threatening emergency that has a reported mortality rate as high as 25%. Approximately 40% of the cases occur with the first attack of IBD; another 40% are seen in patients receiving high-dose steroid therapy for fulminant colitis. Toxic megacolon almost always involves the transverse colon. The pathophysiology is believed to be an extension of the inflammatory process through all layers of the bowel wall with resulting microperforation, localized ileus, and loss of colonic tone. The result is imminent major perforation, peritonitis, and overwhelming sepsis. Antecedent barium enema, opiates, or anticholinergics may all precipitate toxic megacolon. Clinical features include 1) a rapidly worsening clinical course usually associated with fever, malaise, and even lethargy; 2) abdominal distension and tenderness usually developing over a few hours or days; 3) a temperature of 38.5°C (101.3°>F) or higher and a neutrophilic leukocytosis; and 4) an abdominal radiograph showing distension of the transverse colon of more than 5 to 7 cm. The differential diagnosis of acute fulminant colitis includes acute bacterial enteritis, amebic dysentery, ischemic bowel disease, and radiation colitis.

Other potential clinical manifestations of IBD related to extraintestinal complications include thrombosis of cerebral, retinal, or peripheral vessels that may lead to coma, seizures, or focal visual or motor deficits; renal calculi leading to hematuria; and pancreatitis.

Management

The initial ED management of IBD is determined primarily by whether the patient is known to have been previously diagnosed with ulcerative colitis or Crohn's disease and by an assessment of the severity of GI symptoms and systemic toxicity. Several clinical classification systems are used, but in general, mild disease is associated with less than six

stools per day and an absence of systemic signs such as fever and severe anemia. Moderate disease is characterized by more than six stools per day, fever (higher than 38°C [100.4°F]), hypoalbuminemia (serum protein less than 3.2 g/dL), and anemia (hemoglobin concentration less than 10 g/dL). Severe disease is indicated by more than eight stools per day, marked abdominal cramping and tenderness, high fever, significant anemia (hemoglobin concentration less than 8 g/dL), leukocytosis (WBC count greater than 15,000), and occasionally, toxic megacolon.

Initial blood studies most commonly needed to evaluate patients who have known or suspected IBD include a CBC, serum electrolytes, BUN, serum albumin and total protein, transaminases (alanine aminotransferase [ALT] and aspartate aminotransferase [AST]), and depending on the amount of suspected blood loss, a blood type and crossmatch. The erythrocyte sedimentation rate can be a useful marker of inflammation; it is elevated in up to 90% of patients with Crohn's disease and in more than 50% of those with ulcerative colitis. The diagnostic yield of plain supine and upright or decubitus abdominal radiographs is relatively low (10% or less) in terms of positive findings of clinical relevance. Nevertheless, plain films can be useful in establishing the diagnosis of toxic megacolon, bowel obstruction, or perforation and should be strongly considered in the initial management of any patient with known or suspected IBD and who presents to the ED with abdominal pain or tenderness.

Stool examination for occult blood and fecal leukocytes may indicate the presence of active inflammation. For patients who have not been previously diagnosed with IBD, as well as during flare-ups in patients with a known diagnosis, stool should be obtained for culture to rule out infectious colitis, which may often either mimic IBD or complicate a known case. Noninfectious causes of rectal bleeding, including polyps, Meckel's diverticulum, HSP, and HUS, as discussed further in this and other chapters (see Chapter 86 and Chapter 118), may also be considered in some instances, with appropriate diagnostic evaluation tailored accordingly.

Patients with known or previously undiagnosed IBD, who have mild manifestations of disease, and whose initial laboratory and radiographic studies do not reveal significant abnormality can be discharged from the ED after arranging follow-up with an appropriate specialist (pediatric or general gastroenterologist). Further diagnostic studies such as sigmoidoscopy, colonoscopy, or air-contrast barium enema, as well as the institution of medical management with corticosteroids and/or sulfasalazine, can be arranged on an outpatient basis.

The goal of initial management of patients with moderately severe disease is supportive, and intravenous hydration with crystalloid solutions is often necessary to correct acute dehydration. Normal saline may be given as a 20 mL/kg bolus infusion and repeated as necessary to achieve hemodynamic stability. An infusion of a dextrose-containing electrolyte solution may then be initiated based on the initial serum electrolytes. When severe abdominal pain occurs in a patient who is not known to have IBD, surgical consultation is indicated if diagnoses such as acute appendicitis or bowel obstruction are possibilities. Hospitalization of patients with moderately severe disease is often indicated to initiate or modify specific therapy such as systemic corticosteroids or immunosuppressive agents such as azathioprine or 6-mercaptopurine. In addition, improved nutritional intake, either via enteral or parenteral means, is often necessary.

All patients with acute fulminant colitis should be admitted to the hospital. Oral intake should be discontinued and an intravenous infusion begun with normal saline until electrolyte and BUN levels are known. Opiate or anticholinergic drugs should be avoided because they may precipitate toxic megacolon. If toxic megacolon is suspected, arrangements should be made for admission to an intensive care unit. The patient should discontinue all antidiarrheal and anticholinergic medicines. The first priority in the management of children with toxic megacolon is the treatment of intravascular dehydration and shock. Intensive intravenous therapy with normal saline, albumin, or blood must be sufficient to correct hypotension and to ensure adequate urine flow. A nasogastric tube, or preferably a Miller-Abbott tube for small bowel decompression, should be placed. Patients should be started on broad-spectrum antibiotics such as ampicillin (200 mg/kg per day), gentamicin (5 to 7.5 mg/kg per day), and clindamycin (40 mg/kg per day) in combination. Suitable alternative therapies include either ampicillin/sulbactam or cefoxitin in combination with gentamicin.

Management of significant GI bleeding should be performed as described earlier in this chapter. Emergency management of suspected intestinal obstruction includes gastric decompression with nasogastric drainage and intravenous rehydration, initially with normal saline. Patients with fulminant colitis, suspected toxic megacolon, significant GI bleeding, or suspected intestinal obstruction should all receive prompt surgical consultation as part of their initial ED evaluation.

PEPTIC ULCER DISEASE

Background

The term *peptic ulcer disease* describes a group of disorders, consisting of primary and secondary gastric and duodenal ulcers, as well as nodular gastritis, rather than a single disease. With increasing use of endoscopy in children, peptic ulcer disease is a more commonly recognized disorder, although it is still far less common than in adults. Good incidence data in children are generally lacking, although several studies have suggested that large pediatric referral centers diagnose approximately 5 new cases per year or 1 case per 2500 hospital admissions. Primary ulcers have traditionally been defined as those that have no known underlying cause. In contrast, secondary ulcers are those that result from a known ulcerogenic event, such as stress, burns, or the use of NSAIDs.

Primary duodenal ulcers are far more common than primary gastric ulcers in children and typically occur in older children and adolescents (Fig. 93.7). A family history of peptic ulcer disease is typically present in 50% or more of children with duodenal ulcers. Secondary peptic ulceration can be either gastric or duodenal and is most commonly a result of stress from critical illness or injury. Stress ulcers account for 80% of peptic disease in infancy and early childhood. These ulcers often present as medical emergencies as a result of perforation or hemorrhage.

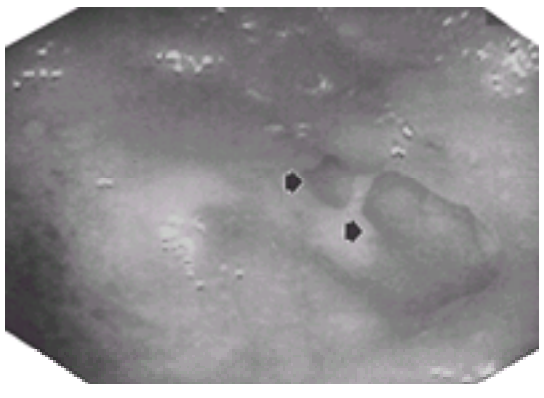

FIGURE 93.7. Duodenal bulb ulcers. The arrows show two individual duodenal nonbleeding ulcers.

Recently, the role of the bacterium *Helicobacter pylori* in the etiology of peptic ulcer disease in children has been vigorously investigated. A recent European study found the prevalence of *H. pylori* infection diagnosed by urea breath test to be 13.7% among healthy preschool-age children. Most children with *H. pylori* infections are asymptomatic. Available evidence to date shows a strong association between *H. pylori* infection and antral gastritis and duodenal ulcer disease in children. However, little to no evidence has been presented for an association with gastric ulcer or recurrent abdominal pain in children. The specific role of *H. pylori* in the pathogenesis and treatment of peptic ulcer disease is discussed in the following section.

Pathophysiology

Traditional teaching regarding the pathogenesis of peptic ulcer disease can be summarized by the adage, “no acid, no ulcer.” Gastric acid is produced by parietal cells in the stomach and is controlled primarily by histamine, acetylcholine, and gastrin. The ability of H_2-receptor antagonists to suppress all modes of gastric acid secretion underscores the central regulatory role of histamine. All agents that stimulate gastric acid secretion also stimulate pepsin secretion. Pepsins are enzymes that hydrolyze proteins, as well as gastric mucus glycoproteins, in an acid pH environment. They are secreted by gastric chief cells as pepsinogens and are converted to active pepsin by gastric acidity.

H. pylori possesses a number of virulence factors that render it particularly pathogenic in the acidic gastric environment. *H. pylori* normally adheres only to gastric mucosa in vivo. It is a flagellated organism with the capacity for active motility, giving it the ability to penetrate the mucous layer overlying the gastric mucosa. It also possesses potent urease activity, converting urea, which is abundant in gastric epithelium, to ammonia and bicarbonate. This capacity has been proposed as both a survival mechanism for the organism (the bicarbonate may moderate the pH of the local environment of the organism), as well as a pathogenic mechanism (the ammonia functions as a gastric irritant). *H. pylori* invariably produces a localized inflammatory reaction that may contribute to epithelial damage either by direct toxic effect or via immunopathologic means (Fig. 93.8).

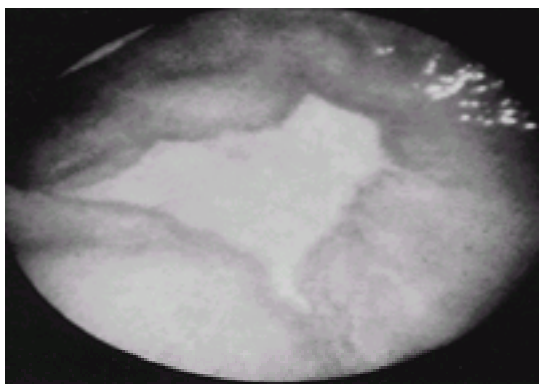

FIGURE 93.8. A large duodenal bulb ulcer secondary to *Helicobacter pylori* infection.

A simplified summary of how duodenal ulcers are formed is that they result when offensive factors, including acid secretion, decreased local blood flow, and *H. pylori* infection, overwhelm protective factors, including the thick layer of mucous gel and the secretion of bicarbonate from surface epithelial cells. The underlying mechanism of gastric ulcer formation is less well understood. Local blood flow, delayed gastric emptying, duodenal reflux, and other factors have all been suggested as important predictors of gastric ulceration. The exact interplay of gastric acid, *H. pylori,* local blood flow, and other factors in the pathogenesis of peptic ulcer disease is the subject of intensive investigation but currently remains unclear.

Clinical Manifestations

Symptoms of peptic ulcer disease vary with the patient's age. Nonspecific signs and symptoms predominate among infants and preschool-age children, with boys and girls affected equally. The older the child, the more specific (and similar to adult patterns of presentation) the signs and symptoms become. Among teenagers with peptic ulcer disease, a male predominance is seen, with boys outnumbering girls nearly 4:1. Infants with peptic ulcer disease (usually secondary to some other condition) may present either with nonspecific feeding difficulties and vomiting, or more fulminantly with upper GI bleeding or perforation. Preschool-age children often complain of poorly localized abdominal pain, vomiting, or

GI hemorrhage and either manifest as hematemesis or melena. Older children and adolescents present almost invariably with abdominal pain, which is described as waxing and waning, sharp or gnawing, and localized to the epigastrium. It may awaken the child at night or in the early hours of the morning. The presence of nocturnal pain may assist in distinguishing recurrent abdominal pain as a result of peptic ulcer disease from functional abdominal pain, which rarely occurs at night. A careful history of the pain as well as a family history of peptic ulcer disease will often suggest the diagnosis of peptic ulcer disease in the older child. History should also be obtained regarding the presence of predisposing factors such as smoking or regular use of NSAIDs.

Physical examination may reveal abdominal tenderness, poorly localized in young children and more commonly localized to the epigastrium or to the right of the midline in older children and adolescents. Stool should be tested for occult blood, and the remainder of the physical examination should be directed toward an assessment of the hemodynamic stability of the patient and, in younger children, to an identification of the underlying condition (e.g., sepsis) that may have predisposed them to the development of peptic ulcer disease. Weight loss may be noted.

Differential Diagnosis

A number of conditions may mimic the presentation of peptic ulcer disease. Abdominal pain is a common symptom during childhood, occurring in 10 to 15% of school-age children. Most children with recurrent abdominal pain have a “functional” cause. These patients typically do not have any weight loss or vomiting and report that their pain is localized to the umbilical area. Further discussion regarding the differential diagnosis of abdominal pain can be found in Chapter 50. It should be noted that the prevalence of *H. pylori* infection in children who have recurrent abdominal pain varies widely in the literature, with most patients studied selected from among those presenting to tertiary hospital gastroenterology clinics. Children with *H. pylori* infection are characteristically asymptomatic. In fact, a recent population-based study found less GI symptoms among children infected with *H. pylori* than in those without infection.

Gastritis, distal esophagitis, *Giardiasis,* and pancreatitis may all cause epigastric pain and tenderness. Biliary tract disease and a ureteropelvic junction obstruction may cause right upper quadrant tenderness. Children who have IBD, HSP, or diabetes mellitus may also present with abdominal pain, tenderness, or GI bleeding.

Diagnosis

Radiologic examination, with either single- or double-contrast (with air) barium upper GI series, is not an effective diagnostic tool to either confirm or rule out the presence of peptic ulcer disease in children. These studies often do not detect superficial ulcers, and conversely, barium trapped in a gastric or duodenal fold may falsely give the impression of an ulcer. However, radiologic examination may be used to rule out other conditions such as malrotation with volvulus or other structural anomalies of the GI tract.

Flexible fiberoptic esophagogastroduodenoscopy with mucosal biopsy is the most accurate method of diagnosing peptic ulcer disease in children. In most tertiary care referral centers, this procedure can be performed safely even on infants. It is typically not performed in the presence of active hemorrhage, although some centers are gathering experience with the use of therapeutic endoscopy to control significant bleeding. When performed, biopsy specimens should routinely be obtained from any area of endoscopic abnormality, as well as from the distal esophagus, antrum, and second part of the duodenum. No clear guidelines exist to indicate which pediatric patients should undergo endoscopy for evaluation of peptic ulcer disease. Suggested guidelines include any child with chronic abdominal pain (longer than 3 months) associated with any of the following signs and symptoms: 1) hematemesis, 2) a history of peptic ulcer disease in a first-degree relative, 3) nocturnal pain, 4) pain occurring within 1 hour of eating or relieved by eating, 5) recurrent vomiting, 6) weight loss, or 7) abdominal tenderness localized to the epigastrium (particularly in older children). In addition, endoscopy should be strongly considered in any patient presenting acutely with significant upper GI bleeding.

All patients for whom an obvious cause of secondary gastric or duodenal ulceration (e.g., stress, sepsis, burns) does not exist should undergo diagnostic evaluation for the presence of *H. pylori* infection. *H. pylori* infection can be confirmed in a variety of ways in patients with primary peptic ulcer disease. Histologic examination of biopsy specimens obtained during endoscopy should routinely be performed because *H. pylori* is readily seen using a variety of staining techniques. In centers with appropriate facilities for culturing the organism, biopsy specimens may also yield growth of the organism, which can assist in the choice of appropriate antibiotic therapy, particularly in recalcitrant infections. A variety of commercially available assays take advantage of the urease activity of the organism for diagnostic purposes. A biopsy specimen is mixed with the assay, which typically contains urea and an indicator dye that changes color when the urea is converted to ammonia by the organism. The urease activity can also be detected through the use of breath tests in which radiolabeled (^{13}C or ^{14}C) urea is ingested by the patient. Degradation of the urea by *H. pylori* results in the release of the radiolabeled carbon, which can be detected in the expired air. Finally, enzyme-linked immunosorbent assays are available for the detection of immunoglobulin G (IgG) antibodies to *H. pylori* in serum. Noninvasive tests such as serology and breath tests, are useful but should not be promoted as the sole method of diagnosing *H. pylori*–associated peptic ulcer disease in children because they cannot distinguish between incidental infection and the presence of peptic ulceration. At this time, therefore, the presence of suspected peptic ulcer disease should be confirmed by endoscopy using the guidelines for performance of endoscopy previously suggested.

Management

The focus of ED management of patients with suspected peptic ulcer disease is on the detection and stabilization of life-threatening complications such as perforation and major GI hemorrhage and on ruling out other potential serious or life-threatening conditions that may require urgent intervention. Depending on the suspected amount of blood loss, all patients with GI bleeding should have a CBC and blood type and screen obtained. If vomiting has been prominent, electrolytes, BUN, creatinine, serum amylase, and lipase should also be obtained. If physical examination findings suggest significant abdominal tenderness with guarding or rebound tenderness, plain radiographs of the abdomen should be obtained to rule out a perforation or bowel obstruction. Intravenous access should be obtained in all patients who

have significant emesis, dehydration, weight loss, or concerning abdominal examination findings. An initial bolus of normal saline (20 mL/kg) should be given and vital signs monitored frequently, with additional boluses given as needed to achieve hemodynamic stability.

A number of approaches are available for treatment of peptic ulcer disease in children. Therapies can be categorized as those that either neutralize acid, block acid secretion, are cytoprotective, or are anti-infective. Antacids are a low-cost, safe, and effective means of treating peptic ulcer disease in children and can be used in patients of any age. Side effects of antacids are related to the cation present in the preparation: magnesium-containing products cause diarrhea, whereas aluminum-containing products cause constipation. Some products are available combining the two to minimize these effects. The usual dosage for children is 0.5 mL/kg, given 1 hour after eating and before bed. Patients with food-related or nocturnal abdominal pain without associated signs of serious illness can be started on empirical therapy with antacids, assuming good follow-up with a primary care physician. Referral to a pediatric gastroenterologist can then be made if the patient fails to respond to 2 weeks of therapy.

H_2-receptor antagonists are the most common agents used to treat peptic ulcer disease. Patients with significant GI bleeding, vomiting, or abdominal tenderness should be admitted to the hospital and begun on intravenous therapy with an H_2-receptor antagonist. Currently, more physicians have pediatric experience with the use of cimetidine and ranitidine than with famotidine. All three agents are competitive H_2-receptor antagonists that reduce gastric acid output, thereby raising gastric pH. Structural differences among the three agents render famotidine the most potent and longest acting, followed by ranitidine and then cimetidine. In addition, ranitidine and famotidine generally have fewer side effects than cimetidine. The recommended oral dosage for cimetidine is 7 mg/kg given every 6 to 8 hours. The dosage of ranitidine commonly used to treat peptic ulcer disease is 1 to 2 mg/kg given every 8 hours in infants and younger children, every 12 hours in older children. Less published experience exists with the use of famotidine in pediatrics. Available data suggest a starting dosage for children older than 1 year of age of 0.5 mg/kg given intravenously every 8 to 12 hours. Patients for whom initial outpatient therapy is appropriate can be started on an H_2-receptor antagonist following an ED visit, but this therapy is best done in consultation with either the patient's primary care physician or pediatric gastroenterologist, who will establish appropriate follow-up for the patient.

Sucralfate is an aluminum salt that “coats” damaged gastric mucosa, effectively insulating it from further damage by acid, pepsin, or bile. It is typically given as a slurry and can be used with H_2-receptor antagonists, provided the drugs are given at least 1 hour apart. The usual adult dosage is 1 g four times a day; this can be titrated down for pediatric patients, although firm dosage guidelines for pediatric use are currently unavailable.

Results of a national consensus conference on *H. pylori* in peptic ulcer disease recommend that ulcer patients with *H. pylori* infection receive antimicrobial treatment as well as antisecretory drugs, both at first presentation as well as for recurrent disease. Eradication of *H. pylori* infection has been shown to reduce the recurrence of primary duodenal ulcers in children. Most children with *H. pylori* infection are asymptomatic, and no convincing evidence that *H. pylori* causes symptoms in the absence of ulceration has been presented; therefore, antimicrobial therapy is currently not recommended for children without ulcers or gastritis who harbor the organism. Several antimicrobial agents, either alone or in combination, have been studied. Although optimal treatment guidelines have not been established for children, several studies have demonstrated that omeprazole in combination with clarithromycin and amoxicillin or metronidazole is an effective regimen for clearance of the organism in those infected. Optimal length of therapy has also not been clearly established. Most authorities currently recommend a 2- to 3-week course of therapy.

REYE SYNDROME

Background

Reye syndrome is a distinct, reversible, clinicopathologic syndrome of unknown origin characterized by severe noninflammatory encephalopathy and fatty degeneration of the viscera. The incidence has dramatically decreased since the early 1980s, when a series of case-control studies established a link between antecedent aspirin exposure and the onset of Reye syndrome. The causality of the relationship between the subsequent decline in aspirin use and the incidence of Reye syndrome in the United States, however, remains controversial.

Pathophysiology

The pathogenesis of Reye syndrome centers around a primary mitochondrial injury in all tissues of the body. Abnormally low mitochondrial enzyme activities parallel histopathologic observations of mitochondrial degeneration in virtually every tissue studied by electron microscopy, including liver, brain, kidney, skeletal muscle, pancreas, and heart. The mitochondrial injury results in decreased activities of enzymes involved in the Krebs' cycle, gluconeogenesis, and urea biosynthesis. Most of the clinical features of Reye syndrome, including lactic acidosis, elevated fatty acids, nitrogen wasting, hyperammonemia, cellular fat accumulation, and cytotoxic cerebral edema, may be explained in the context of primary mitochondrial damage.

Clinical Manifestations

Reye syndrome affects children of all ages. No sex difference is apparent. A biphasic clinical history is remarkably constant (Fig. 93.9). First, the child has a history of a recent, usually febrile, illness that is waning or has resolved. Approximately 90% of the children have an antecedent upper respiratory infection, and 57% of cases are associated with antecedent varicella. The abrupt onset of protracted vomiting usually starts within 1 week following the prodromal illness. The vomiting is unresponsive to restriction of oral intake or to antiemetic therapy.

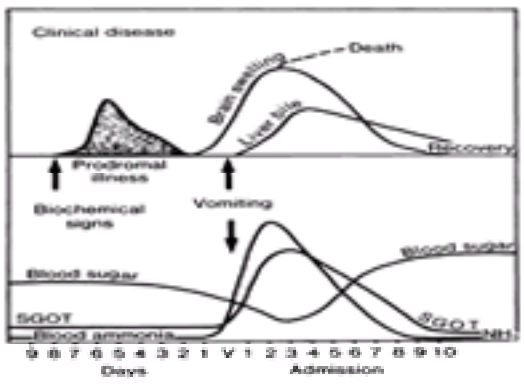

FIGURE 93.9. Clinical history in Reye syndrome.

Coincident with the onset of vomiting (or shortly thereafter), signs of encephalopathy appear. At first, encephalopathy may be manifested by unusual quietness or disinterest. However, a rapid sequential progression to irritability, combativeness, confusion, disorientation, delirium, stupor, and coma may occur. Seizures are a late sign in older children but may occur during early stages of encephalopathy in infancy (usually secondary to hypoglycemia).

In the ED, patients are usually afebrile. Tachycardia and hyperventilation commonly occur. At the initial presentation, only 50% of patients have hepatomegaly. The liver usually increases in size during the first 24 to 48 hours after the diagnosis is made. Absence of jaundice and scleral icterus is characteristic and is the major mitigating clinical sign against hepatic encephalopathy secondary to acute fulminant hepatitis. Despite evidence of encephalopathy, no focal neurological signs or signs of meningeal irritation are apparent.

The diagnosis of Reye syndrome is suggested by the clinical presentation, supported by characteristic biochemical findings, and confirmed by characteristic histologic findings on liver biopsy. The hallmark of the acute encephalopathy of Reye syndrome is the associated evidence of liver abnormality. Transaminases (ALT and AST) and blood ammonia are almost always elevated at the time of the onset of protracted vomiting. The range of transaminase elevation is highly variable and has not been shown to correlate well with severity of the disease. Ammonia levels greater than 300 µg/L have been shown to be an indicator of a poor prognosis. The PT is less than 50% of control in at least one-half the patients, although clinical bleeding is rare and evidence of disseminated intravascular coagulation is absent. The serum bilirubin may be greater than 2 mg/100 mL in 10 to 15% of patients; however, the highest reported value in an accepted case of Reye syndrome is only 3.5 mg/100 mL. The direct reacting fraction of the total bilirubin usually is greater than 15% of the total. Hypoglycemia is rare, except in children who present in stage IV coma and in infants less than 1 year old, in whom the incidence is reported to be as high as 70 to 80%. Azotemia is seen 30 to 40% of the time and ketonuria, 80%. Both reactions are believed to be secondary to starvation and dehydration from vomiting and poor oral intake. Patients most often have a mixed respiratory alkalosis and mild metabolic acidosis. The metabolic acidosis correlates with the level of ammonia elevation and reflects the degree of mitochondrial dysfunction.

Management

Once the diagnosis is suspected, immediate plans should be made to admit the child to a center with a staff and facilities to monitor intracranial pressure. A hospital without such facilities should not observe a patient in the early stages of Reye syndrome because too often the progression of the encephalopathy may proceed rapidly, resulting in increased morbidity and mortality. During the first 72 hours, Reye syndrome should always be managed in an intensive care unit.

Despite the generalized nature of the mitochondrial insult in Reye syndrome, the brain is the principal organ affected by the syndrome. Increased intracranial pressure (ICP) secondary to cerebral edema is the major factor contributing to morbidity and mortality in Reye syndrome. The effectiveness of accurate ICP monitoring via a subarachnoid bolt or intraventricular catheter is now well established. With the ability to monitor ICP, a number of different invasive therapies have been introduced in an attempt to rapidly reduce and control cerebral edema. None of these therapies, including hyperventilation and muscle paralysis using neuromuscular blocking drugs, hyperosmolar agents, high-dose barbiturates, exchange transfusions, or hypothermia, has been clearly proven to protect the brain from progressive ischemic insult.

The proper initial staging of coma is essential (Table 93.1). The survival of the patient is definitely related to the stage of the disease upon admission. The management of Reye syndrome is supportive because no specific curative therapy is currently available. Patients presenting in stage I coma should have initial blood and cerebrospinal fluid studies sent (Table 93.2), and be admitted to an intensive care unit where they should be closely monitored and receive intravenous hydration at a rate of two-thirds maintenance with a 10% dextrose solution to protect against hypoglycemia.

Stage I	Vomiting, lethargy, and sleepiness
Stage II	Disorientation, delirium, combativeness; hyperventilation, hyperreflexia, appropriate responses to noxious stimuli
Stage III	Obtunded, coma, hyperventilation; inappropriate response to noxious stimuli; decorticate posturing; preservation of pupillary, light reflexes and oculovestibular reflexes (doll's eyes)
Stage IV	Deeper coma, decerebrate rigidity, loss of oculovestibular reflexes, dilated fixed pupils, dysconjugate eye movements in response to caloric stimulation
Stage V	Seizures, absent deep tendon reflexes, respiratory reflexes, flaccid paralysis

Table 93.1. Clinical Staging of Reye Syndrome (Lovejoy)

I. Abnormal Studies	F. Ketonuria
A. Evaluation of SGOT, SGPT (at least 2 times normal)	G. Hypoglycemia
B. Elevation of blood ammonia (at least 1.5 times control)	H. Decreased serum bicarbonate
C. Prolongation of prothrombin time	I. Decreased arterial Pco_2
D. Hyperaminoacidemia (particularly glutamine, alanine, lysine)	J. Bilirubin <3.0 mg/100 mL
E. Elevated blood urea nitrogen	II. Normal Studies
	A. Spinal fluid cell count, protein, Gram stain
	B. Platelet count and blood smear
	C. Drug toxic screen
	D. Amylase

Table 93.2. Biochemical Evaluation of Reye Syndrome

In addition to these supportive interventions, stage II patients with blood ammonia less than 300 µg/L should receive lactulose syrup (0.67 g/mL) 0.20.4 g/kg per dose by nasogastric tube every 6 hours to reduce blood ammonia. In addition, mannitol (20% solution) 1 g/kg intravenously over 30 minutes should be given every 6 hours. Mannitol is a hyperosmolar agent that dehydrates the brain by withdrawing fluid from the tissues. Dexamethasone may be used at a loading dose of 0.2 mg/kg intravenously, followed by 0.1 mg/kg every 6 hours. The site of action of steroids on brain edema is still unknown.

For stage II coma with blood ammonia greater than 300 µg/L, and stage III, stage IV, and stage V coma, treatment includes elective intubation and hyperventilation.

ACUTE BILIARY TRACT DISEASE

Background

Acute biliary disease occurs occasionally in children (more often in adolescents) and is associated with a wide spectrum of clinical manifestations. Acute cholecystitis is typically a complication of cholelithiasis, which is primarily associated with hemolytic anemias (pigment stones), such as sickle cell disease and hereditary spherocytosis. Adolescent girls develop cholecystitis more often than boys (cholesterol stones). Gallstones may be asymptomatic and their prevalence increases with age. Acalculous cholecystitis, or acute inflammation of the gallbladder in the absence of gallstones, is actually more common than cholelithiasis in children and has been associated with bacterial enteric infections (typhoid, shigellosis, *E. coli,* scarlet fever, pneumonia, Kawasaki disease, leptospirosis, hepatitis, polyarteritis nodosa, and parasitic infections [ascaris and *Giardia*]). Acute cholangitis resulting from an ascending biliary infection or obstruction is seen primarily in the pediatric patient who has had surgical correction of congenital biliary tract obstruction (biliary atresia, choledochal cyst). Finally, hydrops of the gallbladder, causing jaundice and a right upper quadrant mass effect with pain, is a complication of Kawasaki disease.

Pathophysiology

Biliary colic results from acute transient obstruction of the cystic duct or common bile duct by gallstone(s). Cholecystitis is an aseptic inflammatory process that develops as a reaction to chemical injury triggered by obstruction to the cystic duct by a gallstone. This inflammation is mediated by 1) lysolecithin, which is formed from biliary lecithin by refluxed pancreatic enzyme phospholipase A; 2) refluxed proteolytic pancreatic enzymes; and 3) unconjugated bile salts. The cause of acalculous cholecystitis is unknown. The condition is commonly associated with gallbladder distension, called acute hydrops of the gallbladder. In infectious syndromes, inflammation of the cystic duct and/or enlargement of mesenteric lymph nodes may result in obstruction to bile flow. In vasculitis syndromes, such as mucocutaneous lymph node syndrome (Kawasaki disease) or polyarteritis nodosa, there may be a reactive serositis or vasculitis with increased mucus secretion by the gallbladder that, when coupled with factors that contribute to bile stasis such as fever, prolonged fasting, ileus, or dehydration, may result in gallbladder distension that in turn may kink the cystic duct. Cholangitis results from secondary bacterial infection by enteric organisms in the face of biliary tract obstruction or after surgical manipulation of the biliary tract. Acute cholangitis may be mild and superficial, producing only short-lived symptoms, or it may be severe, causing suppurative cholangitis with septic shock and formation of hepatic abscesses.

Clinical Manifestations

The pain of biliary colic is acute in onset, often follows a meal, and is usually localized to the epigastrium or right upper quadrant. Some children may localize the pain to the periumbilical area. Characteristically, the pain increases to a plateau of intensity over 5 to 20 minutes, typically after meals, and persists for a variable duration, usually less than 4 hours (although less than 1 hour in 50% of patients). In contrast to the colicky pain of intestinal or ureteral origin, biliary colic does not worsen in relatively short cyclic paroxysms or bursts but instead is characterized by its sustained, intense quality. Unlike pancreatitis, the patient tends to move about restlessly and the pain is not improved by changes in position. In addition, referred pain is common, particularly to the dorsal lumbar back near the tip of the right scapula. Nausea and vomiting are commonly associated with biliary colic but are not severe and protracted as seen with pancreatitis. Mild jaundice occurs in 25% of patients, but the serum bilirubin rarely exceeds 4 mg/dL. An attack of acute

cholecystitis begins with biliary colic, which increases progressively in severity or duration. Pain lasting longer than 4 hours suggests cholecystitis. As the inflammation worsens, the pain changes character, becoming more generalized in the upper abdomen and increased by deep respiration and jarring motions. The temperature is usually mildly elevated, ranging from 37.5° to 38.5°C (99.5° to 101.3°F).

In contrast, acute cholangitis should be suspected in the patient who has right upper quadrant abdominal pain, shaking chills, and spiking fever (temperature higher than 39°C [102.2°F]) with jaundice (Charcot's triad). These patients usually have a history of abdominal surgery. The danger of this disorder is that overwhelming sepsis can develop rapidly. Listlessness and shock are characteristic of advanced or severe cholangitis and usually reflect Gram-negative septicemia. Cholangitis can evolve rapidly before development of significant jaundice. Clinically apparent jaundice may be absent even in postsurgical biliary atresia patients. Hydrops of the gallbladder is associated with a palpable right upper quadrant mass and pain. Fever and jaundice generally do not occur.

In addition to scleral icterus, nonspecific physical findings that suggest gallbladder disease include right upper quadrant guarding; Murphy's sign (production of pain by deep inspiration or cough when the physician's fingers are depressing the abdomen below the right costal margin in the midclavicular line and abrupt cessation of inspiration because of pain); and production of pain or tenderness by a light blow applied with the ulnar surface of the hand to the subcostal area. In about one-third of patients with cholecystitis, the gallbladder is palpable as a sausage-shaped mass lateral to the midclavicular line. A rigid abdomen or rebound tenderness suggests local perforation or gangrene of the gallbladder.

Laboratory tests are typically nonspecific. A CBC and blood smear may show evidence of hemolysis. The leukocyte count averages 12,000 to 15,000/mm^3 with a neutrophilic leukocytosis. Leukocyte counts greater than 15,000 ^{3} suggest cholangitis. The serum bilirubin may be elevated but rarely exceeds 4 mg/dL. Higher values are more compatible with either complete common duct obstruction or cholangitis. Serum transaminases, ALT and AST, and alkaline phosphatase may be mildly elevated but are often normal. Marked elevation of transaminases may occur with acute, complete common duct obstruction. Serum amylase may be mildly elevated without other evidence of pancreatitis. Abdominal flat and upright radiographs may show right upper quadrant calcification of gallstones, particularly in patients with hemolytic anemia (pigment stones), or a right upper quadrant mass. Abdominal radiographs are particularly important to rule out perforation. The erythrocyte sedimentation rate is often elevated in children with cholangitis, and organisms may be recovered by blood culture.

Abdominal ultrasound is the most commonly used test to confirm gallbladder disease. This test is noninvasive, easily performed, and provides information on the surrounding organs such as the liver, pancreas, and kidneys. Ultrasound can determine the presence of most gallstones, dilated bile ducts, a thickened gallbladder wall or hydropic gallbladder, sludge, and hepatic abscesses. Other radiographic tests, such as cholecystograph or radionuclide testing, are not typically used in the emergency setting.

Other conditions to be considered in the differential diagnosis of biliary tract disease include perforated peptic ulcer, pneumonia, intercostal neuritis, pancreatitis, hepatitis, and hepatic and abdominal sickle cell crisis. Therefore, evaluation should also include stool guaiac, chest radiograph, amylase:creatinine ratio, and a peripheral blood smear.

Management

All patients with suspected acute biliary tract disease and acute symptoms should be admitted to the hospital. The exception is patients with biliary colic that has resolved spontaneously, in which case an urgent outpatient evaluation by ultrasound can be pursued. Conditions associated with acalculous cholecystitis should be evaluated and treated if identified. General ED management includes discontinuation of oral intake, support with intravenous fluids, and surgical consultation. Cholecystitis and cholangitis associated with gallstones are general indications for surgery. The patient should be made NPO (nothing by mouth) and given intravenous fluids, pain medication, and antibiotics, if cholangitis is considered. Antibiotic coverage should include Gram-negative organisms as well as enterococcus. Ampicillin (200 mg/kg per dose) and gentamicin (5 to 7.5 mg/kg per day) provide good coverage; ampicillin and sulbactam (ampicillin 200 mg/kg per day) can also be used. Narcotics are useful to alleviate the pain and to reduce gallbladder mucosal secretion; however, pain medication should be withheld until a tentative decision regarding early surgery is reached. When given, meperidine 1 to 2 mg/kg per dose is the treatment of choice.

In all patients with suspected cholangitis, blood cultures should be drawn before antibiotics are administered. When possible, antibiotics should be withheld pending a liver biopsy for definitive culture. However, the exception is the clinically septic child in whom antibiotic coverage should be immediately instituted with ampicillin (200 mg/kg per day) or a cephalosporin, such as cefazolin (100 mg/kg per day), and gentamicin (5 to 7.5 mg/kg per day). In these cases, a liver biopsy performed after the institution of antibiotics may still show histologic evidence of cholangitis.

ACUTE PANCREATITIS

Background

Although uncommon, the diagnosis of pancreatitis is often overlooked because no specific pathognomonic symptoms are associated with the condition. Pancreatitis should be considered in any child with acute or chronic epigastric abdominal pain and vomiting, ascites of obscure origin, and following upper abdominal trauma. Table 93.3 lists the causes of pancreatitis. In 30% of cases, the precipitating factor is unknown. Approximately 50% of cases are associated with an infectious agent or blunt trauma. Mumps pancreatitis is seldom severe and rarely occurs in children less than 5 years old; clinical mumps is present in only 50 to 60% of cases. Most blunt injuries to the pancreas are the result of automobile crashes or falls from bicycles; however, because the pancreas is a fixed retroperitoneal structure, mild trauma from small pointed objects, such as sticks, handlebars, or fence posts, may transmit injury directly to the organ.

I	Trauma: blunt, penetrating, surgical
II	Infectious: mumps, coxsackievirus B, hemolytic streptococcus, salmonella, hepatitis A and B
III	Obstructive: cholelithiasis, ascaris, congenital duodenal stenosis, duplications, tumor, choledochal cyst
IV	Drugs: steroids, chlorothiazides, salicylazosulfapyridine, azothiaprine, alcohol, valproic acid, tetracyclines, borates, oral contraceptives
V	Systemic: systemic lupus erythematosus, periarteritis nodosa, malnutrition, peptic ulcer, uremia
VI	Endocrine: hyperparathyroidism
VII	Metabolic: hypercholesterolemia, cystic fibrosis, vitamin A and D deficiency
VIII	Hereditary
IX	Idiopathic

Table 93.3. Causes of Acute Pancreatitis in Children

Pathophysiology

Regardless of the initiating event, the pathophysiology of acute pancreatitis is probably similar. Activation of the numerous pancreatic enzymes, including proteolytic enzymes, lipase, amylase, elastase, and phospholipase A, produces autodigestion of the gland. The process may be focal or diffuse. In mild cases, there is interstitial edema and inflammatory infiltrate without significant cell necrosis. This type of pancreatitis, called acute edematous pancreatitis, is by far the most common form seen in children and is usually self-limiting and associated with complete recovery. When the autodigestive process intensifies with increased inflammation, fat necrosis, and hemorrhagic changes, the process is called necrotic or hemorrhagic pancreatitis. This type of pancreatitis is associated with a 20 to 40% mortality and significant morbidity. It is unclear why the autodigestive process is arrested in some cases and not others, but one factor may be the magnitude of the initial triggering mechanism.

The morbidity and mortality associated with pancreatitis is related to complications from the autodigestive process. Fat necrosis of neighboring tissue and saponification of calcium often result from release of pancreatic lipase. Released proteolytic enzymes may extend the inflammatory process into the retroperitoneum as well as the peritoneal cavity. Proteolytic enzymes may activate kallikrein, a potent vasoactive polypeptide that may mediate systemic vasodilation and increase vascular permeability, producing severe hypotension, shock, and renal and/or pulmonary insufficiency that may prove fatal. Secondary infection may lead to abscess formation, and walling off the autodigestive process may result in pseudocyst formation.

Clinical Manifestations

Epigastric abdominal pain is the most consistent symptom of pancreatitis and may vary from tolerable distress to severe incapacitating pain. Symptoms may be chronic and insidious, but they typically progress rapidly, building to a crescendo over several hours. The pain is usually localized to the epigastrium and may radiate to the back (left or right scapula) or the right or left upper quadrants. The pain is usually described as knifelike and boring in quality and is aggravated when the patient lies supine. Classically, the pain of pancreatitis is constant, as opposed to colicky pain, which waxes and wanes. Nausea and vomiting are the most common associated symptoms. Vomiting may be severe and protracted. Low-grade fever (temperature below 38.5°C [101.3°F]) is present in 50 to 60% of cases. In cases of severe necrotic pancreatitis, patients may complain of dizziness. Mental aberrations are common in necrotic pancreatitis; patients may act overtly psychotic or present in coma.

Early in the course of the disease, there may be a discrepancy between the severity of the patient's subjective pain and the objective physical findings. During the examination, patients are usually quiet and prefer sitting or lying on their side with knees flexed. The abdomen may be distended but is usually not rigid. There may be mild to moderate voluntary guarding in the epigastrium. A palpable epigastric mass suggests pseudocyst. Ascites is rare. Bowel sounds may be decreased or absent. Associated physical findings may include signs of parotitis, mild hepatosplenomegaly, epigastric mass, pleural effusions, and mild icterus. Although rare, rebound tenderness or a rigid abdomen are poor prognostic signs if present. A bluish discoloration around the umbilicus (Cullen's sign) or flanks (Grey Turner's sign) is also a poor prognostic sign and evidence of hemorrhagic pancreatitis. Signs of overt hemodynamic instability are rarely evident at initial presentation. It is particularly important to evaluate patients for clinical signs of hypocalcemia (Trousseau and Chvostek signs).

The clinical diagnosis is often tentative because the same constellation of symptoms (abdominal pain, vomiting, and low-grade fever) and signs (abdominal tenderness and guarding) may be mimicked by several other conditions, including penetrating peptic ulcer, gastritis, esophagitis, biliary colic, acute cholecystitis, intestinal obstruction, and appendicitis.

Currently, two easily attainable laboratory tests are used to make a diagnosis of pancreatitis: serum amylase and serum lipase. The combination of the aforementioned clinical symptoms and an elevation of the level of one or both of these enzymes strongly points to pancreatitis. In acute pancreatitis, the serum amylase increases hours after the onset of the autodigestive process and returns to normal within 3 to 5 days; elevated serum triglycerides may interfere with the assay and result in false-normal values. The degree of serum amylase elevation rarely corresponds to the severity of pancreatic inflammation. Although controversial in the past, lipase assays are now accurate and commonly used to diagnose pancreatitis. Serum lipase may remain elevated for up to 14 days after the onset of acute pancreatitis. Generally, because amylase is rapidly cleared by the kidneys, serum amylase may return to normal after several days even though pain persists. In these cases, following the serum lipase may be more beneficial. Normalization of serum amylase typically indicates resolution of disease, but occasionally hemorrhagic or necrotizing pancreatitis may develop in patients with normal amylase.

Serum amylase and lipase are not pathognomonic for pancreatitis. Many situations, including penetrating or perforated ulcer, intestinal obstruction or infarction, Crohn's disease, pneumonia, hepatitis, liver trauma, acute biliary tract disease, salpingitis, salivary adenitis, renal failure, diabetic ketoacidosis, and benign macroamylasemia, can cause an amylase elevation. Causes for an elevated serum lipase include perforated peptic ulcer and bone fracture with pulmonary fat embolism.

Radiographically, the abdominal ultrasound provides a noninvasive, direct view of the pancreas and is probably the most useful test in diagnosing pancreatitis. Ultrasound can assess pancreatic size, contour, and the presence of calcifications and pseudocyst formation. Ultrasound should be considered in all cases of suspected pancreatitis. Abdominal computed tomography and endoscopic retrograde cholangiopancreatography (ERCP) are being used more often to assess the severity of pancreatitis and pseudocyst formation and to determine possible causes of pancreatitis. However, ERCP should not be performed in the acute phase or in patients with acute pseudocyst formation or pancreatic abscess formation but should be reserved for patients with chronic, recurrent pancreatitis. Rarely, ERCP may be indicated in acute pancreatitis if an obstructing gallstone is present in the common bile duct.

Management

All patients with evidence of pancreatitis or suspected pancreatitis should be admitted to the hospital. Treatment, however, should begin in the ED. The goals of medical treatment include suppression of pancreatic secretion and relief of pain. Morbidity and mortality in pancreatitis are directly related to complications that may already be present at the time of initial presentation. Therefore, aggressive early maintenance of intravascular volume and treatment of hypocalcemia, respiratory distress, and suspected infection are mandatory.

Intravenous fluids should be immediately started, and the patient's oral intake should be discontinued. The patient should be assessed for hypotension. When the patient is judged stable, intravenous fluids should be given at 1.5 times the maintenance rate. Vital signs and urine output should be monitored frequently. Continuous nasogastric suction should be started; aspiration of gastric contents is based on the premise that prevention of delivery of gastric acid into the duodenum will diminish hormonal stimulation of the pancreas. Nasogastric suction also relieves pain and prevents development of ileus. Use of anticholinergics or cimetidine to reduce gastric secretions is controversial and is not recommended in the initial management of patients. A crucial part of management is the treatment of abdominal pain. Pain should be treated with Nubain (0.1 mg/kg per dose intravenously, maximum of 20 mg) or meperidine (1 to 2 mg/kg intravenously, maximum 100 mg). Morphine or codeine should not be used because they increase spasm at the sphincter of Oddi.

Blood studies that should be performed in the ED include amylase, lipase, CBC, electrolytes, BUN, calcium, glucose, SGOT, SGPT, bilirubin, alkaline phosphatase, triglyceride, PT, and PTT. Arterial blood gases should be obtained in patients with tachypnea. A chest radiograph should be obtained and evaluated for pleural effusion, interstitial pneumonic infiltrates, and basilar atelectasis. A flat and upright abdominal radiograph is needed to rule out perforation, ascites, and pancreatic calcifications. In severe cases or in those cases of questionable diagnosis, an abdominal ultrasound should be obtained.

In most cases, maintenance of intravascular volume and relief of pain will result in rapid resolution of symptoms. Prognostic indicators of necrotizing or hemorrhagic pancreatitis include hypocalcemia (less than 8.0 mg/dL), hyperglycemia (greater than 200 mg/dL), clinical shock, elevated hematocrit or BUN, ascites, and oxygen partial pressure less than 60 mm Hg. Such patients should be admitted to an intensive care unit, given sufficient colloid (albumin 0.25 g/kg) to maintain normal intravascular volume, and have more extensive monitoring with an arterial line and urinary catheter. A PaO_2 lower than 60 mm Hg is an indication for elective intubation. Early peritoneal dialysis should be started if rapid clinical deterioration occurs.

Antibiotics are not indicated in the initial management of pancreatitis. Pancreatic abscess should be considered if the patient's temperature is higher than 38.5°C (101.3°F). In those cases, broad-spectrum antibiotic coverage with ampicillin (200 mg/kg per day) and gentamicin (5 to 7.5 mg/kg per day) is indicated pending the results of blood cultures and diagnostic ultrasound. Emergency surgery is rarely necessary in acute pancreatitis; however, indications for surgery include active intraperitoneal bleeding, suspected abscess, biliary duct obstruction, and suspected traumatic transection. Therapeutic surgery for acute, necrotizing pancreatitis has been reported in adults, but this approach has not been accepted in pediatrics.

FULMINANT LIVER FAILURE

Background

Fulminant liver failure occurs when the vital functions of the liver fail, including the development of a coagulopathy, hypoglycemia, hyperbilirubinemia, hypoproteinemia, and encephalopathy. Liver failure can develop acutely or it may be chronically progressive. The cause of liver failure is diverse and includes infectious processes (e.g., viral hepatitis), metabolic diseases (e.g., Wilson's disease), pharmacologic agents, ischemia, and malignancy. Acute liver failure can be a life-threatening problem that causes a severe coagulopathy, hypoglycemia, and encephalopathy. Aggressive supportive medical management is required in most cases.

Pathophysiology

The pathogenesis of fulminant liver failure requires the progression of several key steps that lead to irreversible hepatocyte injury. The initiating step is the exposure of the susceptible person to the inciting agent, which leads to widespread hepatocyte injury. Hepatocyte necrosis may occur secondary to an infectious agent (viral hepatitis), a toxin (various pharmacologic substances), or a metabolic byproduct. Following hepatocyte death, the potentiation of the

responsible agent is necessary to continue the hepatic destructive process. Normally, the liver is capable of regeneration; however, the regenerative process is inhibited in patients who develop liver failure. These steps may lead to terminal hepatic failure in which the liver becomes incapable of supporting those events required for life.

Although infectious agents and toxins are responsible for most proven cases of liver failure, in most cases, no cause is determined. Common drugs and toxins that cause liver failure include acetaminophen, salicylates, solvents, valproic acid, amiodarone, isoniazid, NSAIDs, tetracycline, and chlorinated hydrocarbons. Rarely, metabolic diseases can lead to liver failure. These diseases include galactosemia, tyrosinemia, Wilson's disease, neonatal hemochromatosis, disorders of fatty acid oxidation, bile acid synthetic disorders, and hereditary fructose intolerance.

Clinical Manifestations

Many patients do not exhibit serious clinical features of acute liver failure. Typically, pediatric patients who develop acute liver failure were previously healthy and had no prior medical problems. Patients may initially complain of fatigue, nausea, vomiting, and diffuse abdominal pain. Occasionally, right upper quadrant pain may be severe. Commonly, a history of a prodromal viral illness can be elicited. The presence of jaundice usually initiates the first visit to the physician. As liver failure progresses, patients become more jaundiced and lethargic and begin to develop tremors. In a short time, they become confused or somnolent and may begin to have problems with easy bruising or bleeding.

The onset of encephalopathy occurs in conjunction with the severity and progression of liver failure. Encephalopathy is graded on a scale from I to IV. Grade I is manifested by a coherent individual who shows mild or episodic drowsiness, poor concentration, and impaired intellect. In grade II, the patient continues to be coherent and conversant but also becomes disoriented and fatigued. Agitation and aggressive behavior in conjunction with extreme drowsiness is manifested in grade III encephalopathy. Unresponsive patients who respond only to painful stimuli and who have evidence of cerebral edema are labeled as having grade IV encephalopathy. The clinical features of increased ICP include systemic hypertension, “decerebrate posturing,” hyperventilation, abnormal pupillary responses, and impairment of brainstem reflexes. Cerebral edema is associated with increased mortality and requires aggressive supportive management. Finally, bleeding esophageal and gastric varices as well as ascites may rapidly develop secondary to increased portal hypertension.

Laboratory Findings

Because it may be difficult to diagnose patients clinically, biochemical evidence may be collected that provides evidence of liver failure. The liver plays an important role in hemostasis because the liver synthesizes a number of coagulation factors. An uncorrectable coagulopathy is usually the first laboratory manifestation of liver failure. Other factors may have a shorter half-life, but the PT is the most commonly used marker of the severity of liver disease. A prolonged PT despite intravenous supplementation of vitamin K should alert the physician to impending liver failure. Other laboratory markers suggestive of liver failure include evidence of increasing cholestasis manifested by a rising serum bilirubin, hypoalbuminemia, and hypoglycemia.

It is also important to monitor serum transaminases. Falling transaminases usually indicate resolving liver disease, whereas a decrease in transaminases in association with increasing jaundice and coagulopathy indicates hepatocyte death rather than hepatocyte repair. Monitoring for hypoglycemia is extremely important because the liver is the primary organ for gluconeogenesis. Serum fibrinogen is usually decreased in patients with liver failure. In cases in which the patient has splenomegaly, thrombocytopenia and leukocytopenia may be present.

Hypoglycemia almost always accompanies acute liver failure and may complicate the signs of encephalopathy. Portal hypertension may cause bleeding from esophageal varices or ascites. Hepatorenal syndrome occurs in approximately 75% of patients who reach grade IV encephalopathy. The cause of hepatorenal syndrome is unclear; however, the result is oliguria in the presence of near normal intravascular pressures. Metabolic acidosis occurs in approximately 30% of patients who have liver failure, and the risk of sepsis is increased secondary to the patient's compromised immune function.

Management

All patients suspected of having liver failure should undergo a complete physical examination, including a thorough neurologic evaluation. Laboratory testing should include serum glucose, transaminases, total and direct bilirubin, albumin, PT, GGTP, CBC with differential, electrolytes, blood culture, and fibrinogen. Patients with hypoglycemia should be given intravenous fluids with 10% dextrose and should undergo frequent blood glucose monitoring (every 1 hour) until their blood sugar stabilizes. Metabolic acidosis should be corrected; however, correction of hyponatremia should be gradual in patients with ascites. Patients who have a coagulopathy should be given intravenous vitamin K (2.5 mg in infants; 5 mg in older children and adolescents). A repeat PT should be performed 6 to 8 hours after administration. An uncorrectable PT is suggestive of severe hepatocyte damage. The management of bleeding esophageal varices has been previously discussed in this chapter. Therapeutic management of ascites should occur only in the face of respiratory distress or renal failure; otherwise, the introduction of a diuretic (Aldactone) to achieve a slow, gradual change in ascites is all that is initially required.

Patients with encephalopathy should be frequently monitored for changes in neurologic function. In cases in which the patient has developed cerebral edema, management consists of an intensive care setting, insertion of a subdural transducer, mechanical ventilation (hyperventilation), and administration of mannitol (0.3 to 0.4 mg/kg) to maintain near normal levels of intracranial pressure.

ACUTE VIRAL HEPATITIS

Background

The existing alphabet of viral hepatitis is now up to E, with new variants awaiting discovery. Hepatitis A (HAV), the cause of “infectious” or epidemic hepatitis, is transmitted by the fecal–oral route. On a worldwide scale, fewer than 5% of cases are clinically recognized. HAV is a rare cause of fulminant hepatitis. No chronic carrier state exists. Maintenance of the virus in the human population is through person-to-person spread. Hepatitis B (HBV) is endemic in the human population. Although predominantly transmitted by the parenteral route or sexual contact, the high incidence of infection in family contacts suggests that the virus may also be spread by saliva or breast milk. The ability of HBV to produce a chronic carrier state in 5 to 10% of infected subjects allows maintenance of an infectious pool without serial transmission. Hepatitis C (HCV) accounts for about 95% of hepatitis infections in recipients of blood transfusion and 50% of cases of sporadic non-A, non-B hepatitis. Most of these patients will progress to chronic hepatitis, and about 20% develop cirrhosis. Hepatitis D (HDV) requires hepatitis B helper functions for propagation in hepatocytes, and may occur simultaneously with hepatitis B infection (coinfection), or as superinfection in chronic hepatitis B carriers. Hepatitis E is an enterically transmitted virus responsible for large epidemics of acute hepatitis in Asia, the Middle East, and parts of Africa.

Clinical Manifestations

Most childhood cases of acute hepatitis produce minimal symptoms, are anicteric, and unless suspected by palpation of tender hepatomegaly, are usually confused with a GI flulike illness. Clinical hepatitis classically consists of a 5- to 7-day prodrome of variable constitutional symptoms (low-grade fever, anorexia, nausea, vomiting, malaise, fatigue, and epigastric or right upper quadrant abdominal pain), followed by acute onset of scleral icterus, jaundice, and passage of dark urine. Pruritus and diarrhea are rare. Physical examination after the onset of jaundice may reveal tender hepatomegaly. Mild splenomegaly is present in 25 to 50% of patients. HBV patients may also present with extrahepatic signs and symptoms, such as arthralgia, arthritis, or papular acrodermatitis (on face, buttocks, and extensor surfaces of arms and legs). When the rash is associated with lymphadenopathy and fever, it is called the Gianotti-Crosti syndrome. Onset of the icteric phase of acute hepatitis most commonly is temporarily associated with improvement in the constitutional symptoms. In up to 15% of cases, severe fatigue, anorexia, nausea, and vomiting persist. The icteric period usually lasts 1 to 4 weeks. Occasionally, the jaundice is prolonged for 4 to 6 weeks with increasing pruritus at 2 to 3 weeks.

Differential Diagnosis

A number of infectious agents may mimic a viral hepatitislike illness. The most common are Epstein-Barr virus (EBV, infectious mononucleosis) and cytomegalovirus (CMV). Both agents rarely produce clinical jaundice, and high fever and diffuse adenopathy are more characteristic. Less common agents include herpes, adenovirus, coxsackievirus, rheovirus, echovirus, rubella, arbovirus, leptospirosis, toxoplasmosis, and tuberculosis.

Diagnostic Evaluation

The following laboratory tests are usually performed in all cases of suspected viral hepatitis: serum transaminases (AST and ALT), alkaline phosphatase, total and direct bilirubin, CBC, PT, electrolytes, BUN, glucose, total protein, albumin, globulin, and in patients who are older than 5 years of age, ceruloplasmin. AST and ALT are the best indicators of ongoing hepatocellular injury. Alkaline phosphatase levels are usually less than two times the upper limit of normal for age. Levels greater than three times normal should raise suspicions of EBV or CMV hepatitis or biliary tract disease. Hepatitis classically produces direct fractions of serum bilirubin in excess of 30% of total, indicating definite liver disease. Hyperbilirubinemia may be present in the absence of scleral icterus or jaundice because these signs usually cannot be appreciated until levels of total bilirubin exceed 3 to 4 mg/dL. Serum bilirubin levels peak 5 to 7 days after the onset of jaundice. The initial biochemical screen may reveal several indicators of severe hepatocellular injury, including 1) total bilirubin greater than 20 mg/dL, 2) serum transaminases that exceed 3,000 units/L, 3) WBC count greater than 25,000/mm^3, 4) elevated PT, and 5) hypoglycemia.

Serum albumin and globulin are usually normal. Decreased albumin or increased globulin should suggest an acute flare of chronic liver disease. Serum ceruloplasmin level should be drawn in all patients older than 5 years of age who have suspected hepatitis to rule out Wilson's disease. A chest radiograph may reveal cardiomegaly if any suspicion of low cardiac output states exists. Figure 93.10 and Figure 93.11 contrast the sequence of clinical, biochemical, and serologic events in typical HAV and HBV infection. The serodiagnosis of acute hepatitis is best approached by first testing for anti-HAV IgM, HB surface antigen, HB e antigen, HB serum DNA, anti-HB core Ab, anti-HCV, anti-CMV, and EBV serology. The finding of serum IgM anti-HAV is diagnostic of acute HAV infection because the antibody is present at the time of clinical symptoms. A positive HB Surface antigen suggests the diagnosis of HBV in a symptomatic patient. A positive HB e antigen or anti-HB core Ab are helpful in the rare patient who rapidly clears HB surface antigen from the serum. It is also important to note that in chronic HB surface antigen carriers who have HDV superinfection, the suppression of HBV replication may lead to a transient absence of HBV markers in the serum; unless HDV markers in the serum are sought, the diagnosis may be missed. Anti-HCV does not appear in the patient's circulation until 1 to 3 months after onset of acute illness, and in rare cases, detectable levels may not be demonstrated for up to 1 year. Thus, unless the acute presentation is actually a flare of chronic HCV, serodiagnosis will await long-term follow-up.

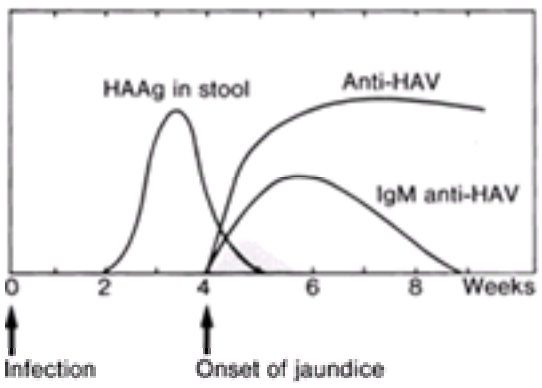

FIGURE 93.10. Serologic changes in hepatitis A. *HAAg,* hepatitis-associated antigen; *HAV,* hepatitis A virus.

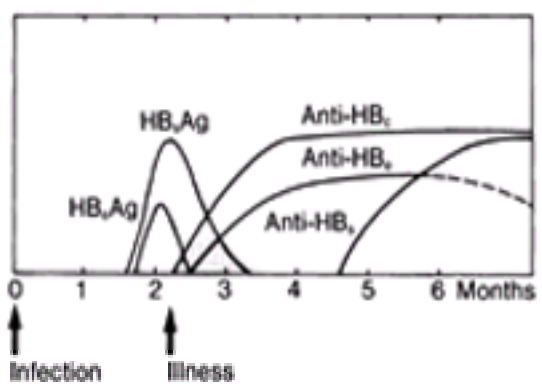

FIGURE 93.11. Serologic changes in hepatitis B. *HB_sAg,* hepatitis B_s antigen; *HB_c,* hepatitis B_c; *HB_e,* hepatitis B_e; *HB_eAg,* hepatitis B_e antigen.

Management

No specific treatment is available for acute viral hepatitis. Most patients can be managed at home. No restrictions in diet or ambulation are necessary. The traditional recommendations of a low-fat, high-carbohydrate diet and bed rest are now recognized to have no effect on the symptoms or duration of the disease. Parents should be told that anorexia and fatigue are common symptoms. Small, frequent feedings may be helpful. Drugs should be strictly avoided. The key for both the patient and other household contacts is personal hygiene. Infants and children should avoid contact with the patient even after they have received immunoprophylaxis. In HAV, shedding of the virus may occur for up to 2 weeks after the onset of jaundice. Patients should be kept at home during this time. After this, they may return to school. Indications for hospitalization of a patient who has acute hepatitis include the following:

1. Dehydration secondary to anorexia and vomiting
2. Bilirubin level greater than 20 mg/dL
3. Abnormal PT
4. WBC count greater than 25,000/mm^3
5. Level of transaminases greater than 3,000 units/L

Patients who have acute hepatitis and who are hospitalized should be isolated. Follow-up studies of all patients with acute hepatitis should be performed to document biochemical resolution. Follow-up serology may also establish a specific cause in cases of apparent non-A, non-B hepatitis (fourfold increase in CMV serology, development of anti-HCV). Reevaluation of patients with HBV is especially important to ensure clearance of HB surface antigen, or to recognize the development of the HB surface antigen carrier state.

Postexposure Prophylaxis

Hepatitis A

The mean incubation period for HAV infection is about 4 weeks (range 15 to 45 days). Conventional immune serum globulin (ISG 0.02 mL/kg intramuscularly) confers passive protection against clinical HAV infection if given during the incubation period up until 6 days before the onset of symptoms. Individuals exposed during the late incubation period or early acute phase of illness should also be promptly immunized. Seventy-five percent of this group will develop detectable levels of anti-HAV IgM, suggesting passive–active immunity. Postexposure immunoprophylaxis is suggested for 1) household and close personal contacts, 2) institutionalized contacts, and 3) contacts within a daycare facility. Grade-school classroom contacts of an isolated case and routine play contacts do not require ISG. However, a second case within a class is indication for immunoprophylaxis of the rest of the class. An alternative method for determining who should receive ISG is to test high-risk contacts for anti-HAV IgG.

Hepatitis B

Prophylactic treatment to prevent infection after exposure to HBV should be considered in the following situations seen in the ED: 1) sexual exposure to the HBV surface antigen–positive patient; 2) inadvertent percutaneous or permucosal

exposure to HBV surface antigen–positive blood; and 3) household exposure of an infant less than 12 months of age to a primary caregiver who has acute HBV. Before treatment in the first two situations, testing for susceptibility is recommended if it does not delay treatment beyond 14 days after exposure. Testing for anti-HBV core Ab is the most efficient prescreening procedure. All susceptible persons should receive a single dose of hepatitis B immunoglobulin (0.06 mL/kg) intramuscularly and hepatitis B vaccine in recommended doses.

Suggested Readings

GASTROINTESTINAL BLEEDING

Bharucha AE, Gostout CJ, Balm RK. Clinical and endoscopic risk factors in the Mallory-Weiss syndrome. Am J Gastroenterol 1997;92(5):805–808.

Countryman D, Norwood S, Andrassy RJ. Mallor-Weiss syndrome in children. South Med J 1982;75(11):1426–1427.

Harris JM, DiPalma JA. Clinical significance of Mallory-Weiss tears. Am J Gastroenterol 1993;88(12):2056–2058.

Katz PO, Salas L. Less frequent causes of upper gastrointestinal bleeding. Gastroenterol Clin North Am 1993;22(4):875–889.

Latt TT, Nicholl R, Domizio P, et al. Rectal bleeding and polyps. Arch Dis Child 1993;69:144–147.

O'Hara SM. Acute gastrointestinal bleeding. Radiol Clin North Am 1997;35(4):879–895.

Oldham KT, Lobe TE. Gastrointestinal hemorrhage in children: a pragmatic update. Pediatr Clin North Am 1985;32(5):1247–1263.

Price MR, Sartorelli KH, Karrer FM, et al. Management of esophageal varices in children by endoscopic variceal ligation. J Pediatr Surg 1996;31(8):1056–1059.

Teach SJ, Fleisher GR. Rectal bleeding in the pediatric emergency department. Ann Emerg Med 1994;23(6):1252–1258.

Thompson EC, Brown MF, Bowen EC, et al. Causes of gastrointestinal hemorrhage in neonates and children. South Med J 1996;89(4):370–374.

Vinton NE. Gastrointestinal bleeding in infancy and childhood. Gastroenterol Clin North Am 1994;23(1):93–122.

COLITIS

Anveden-Hertzberg L, Finkel Y, Sandstedt B, Karpe B. Proctocolitis in exclusively breast-fed infants. Eur J Pediatr 1996;155:464–467.

Hirsch BZ. Breast milk-induced allergic colitis. J Pediatr Gastroenterol Nutr 1995;20:(4):480.

Kato S, Ebina K, Ozawa A, et al. Antibiotic-associated hemorrhagic colitis without *Clostridium difficile* toxin in children. J Pediatr 1995;126(6):1008–1010.

Kelso JM, Sampson HA. Food protein-induced enterocolitis to casein hydrolysate formulas. J Allergy Clin Immunol 1993;92:909–910.

Machida HM, Catto Smith AG, Hall DG, et al. Allergic colitis in infancy: clinical and pathologic aspects. J Pediatr Gastroenterol Nutr 1994;19(1):22–26.

Odze RD, Wershil BK, Leichtner AM, Antonioli DA. Allergic colitis in infants. J Pediatr 1995;126(2):163–170.

Wolf DC, Giannela RA. Antibiotic therapy for bacterial enterocolitis: a comprehensive review. Am J Gastroenterol 1993;88(10):1667–1683.

INFLAMMATORY BOWEL DISEASE

Aideyan UO, Smith WL. Inflammatory bowel disease in children. Radiol Clin North Am 1996;34(4):885–902.

del Rosario JF, Orenstein SR, Bhargava S, Putnam PE. Thrombotic complications in pediatric inflammatory bowel disease. Clin Pediatr 1994;33(3):159–161.

Gore RM, Ghahremani GG. Radiologic investigation of acute inflammatory and infectious bowel disease. Gastroenterol Clin North Am 1995;24(2):353–384.

Grand RJ, Ramakrishna J, Calenda KA. Inflammatory bowel disease in the pediatric patient. Gastroenterol Clin North Am 1995;24(3):613–632.

Levine JB, Lukawski-Trubish D. Extraintestinal considerations in inflammatory bowel disease. Gastroenterol Clin North Am 1995;24(3):633–646.

Manoop SB, Rostami G, Gopalswamy N. Hepatobiliary complications of inflammatory bowel disease. Am Fam Physician 1995;52(5):1440–1444.

Motil KJ, Grand RJ, Davis-Kraft L, et al. Growth failure in children with inflammatory bowel disease: a prospective study. Gastroenterology 1993;105:681–691.

PEPTIC ULCER DISEASE

Bode G, Rothenbacher D, Brenner H, Adler G. *Helicobacter pylori* and abdominal symptoms: a population-based study among preschool children in southern Germany. Pediatrics 1998;101(4):634–637.

Bourke B, Jones N, Sherman P. *Helicobacter pylori* infection and peptic ulcer disease in children. Pediatr Infect Dis J 1996;15(1):1–13.

Bujanover Y, Reif S, Yahav J. *Helicobacter pylori* and peptic disease in the pediatric patient. Pediatr Clin North Am 1996;43(1):213–234.

Chong SKF, Qinyuan L, Asnicar MA, et al. *Helicobacter pylori* infection in recurrent abdominal pain in childhood: comparison of diagnostic tests and therapy. Pediatrics 1995;96(2):211–215.

Gremse DA, Shakoor S. Symptoms of acid-peptic disease in children. South Med J 1993;86(9):997–1000.

Gryboski JD. Peptic ulcer disease in children. Med Clin North Am 1991;75(4):889–902.

James LP, Kearns GL. Pharmacokinetics and pharmacodynamics of Famotidine in paediatric patients. Clin Pharmacokin

1996;August(3):103–110.

Judd RH. *Helicobacter pylori,* gastritis and ulcers in pediatrics. Adv Pediatr 1992;39:283–306.

Kelly DA. Do H_2 receptor antagonists have a therapeutic role in childhood? J Pediatr Gastroenterol Nutr 1994;19(3):270–276.

Macarthur C, Saunders N, Feldman W. *Helicobacter pylori,* gastroduodenal disease, and recurrent abdominal pain in children. JAMA 1995;273(9):729–734.

Mahony MJ, Wyatt JI, Littlewood JM. Management and response to treatment of *Helicobacter pylori* gastritis. Arch Dis Child 1992;67:940–943.

Mezoff AG, Balistreri WF. Peptic ulcer disease in children. Pediatr Rev 1995;16(7):257–265.

Nord KS. Peptic ulcer disease in the pediatric population. Pediatr Clin North Am 1988;35(1):117–140.

Sherman PM. Peptic ulcer disease in children: diagnosis, treatment and the implication of *Helicobacter pylori.* Gastroenterol Clin North Am 1994;23(4):707–725.

REYE SYNDROME

Arrowsmith JB, Kennedy DL, Kuritsky JN, Faich GA. National patterns of aspirin use and Reye syndrome reporting, United States, 1980 to 1985. Pediatrics 1987;79:858.

Heubi JE, Partin JC, Partin JS, Shubert WK. Reye's syndrome: current concepts. Hepatology 1987;7:155.

Treem WR, Witzleben CA, Piccoli DA, et al. Medium-chain and long-chain acyl-CoA dehydrogenase deficiency: clinical, pathologic and ultrastructural differentiation from Reye's syndrome. Hepatology 1986; 6:1270–1278.

ACUTE BILIARY TRACT DISEASE

Boey JH, Way LW. Acute cholangitis. Ann Surg 1980;191:264.

Earnest DL. Manifestations and complications of gallstones. Pract Gastroenterol 1981;5:20.

Holcomb GW Jr, O'Neill JA, Holcomb GW III. Cholecystitis, cholelithiasis, and common duct stenosis in children and adolescents. Ann Surg 1980;191:626.

Sean HF, Golden GT, Horsely JS III. Cholecystitis in childhood and adolescence. Arch Surg 1973;106:651.

Way LW, Sleisenger MH. Acute cholecystitis. In: Sleisenger MS, Fordtran JS, eds. Gastrointestinal Disease: Pathophysiology, Diagnosis, Management. Vol. 2. Philadelphia: WB Saunders, 1978:1302.

ACUTE PANCREATITIS

Arvanitakis C, Cooke AR. Diagnostic tests of exocrine pancreatic function and disease. Gastroenterology 1978;74:932.

Brandborg LL. Acute pancreatitis. In: Sleisenger MS, Fordtran JS, eds. Gastrointestinal Disease: Pathophysiology, Diagnosis, Management. Vol. 2. Philadelphia: WB Saunders, 1978:1409.

Gauthier A, Escoffier JM, Camatte R, Sarles H. Severe acute pancreatitis. Clin Gastroenterol 1981;10:209.

Jordan S, Ament M. Pancreatitis in children and adults. J Pediatr 1977; 91:211.

Levitt MD. Clinical use of amylase clearance and isoamylase measurements. Mayo Clin Proc 1979;54:428.

Mallory A, Kern F. Drug-induced pancreatitis: a critical review. Gastroenterology 1980;78:813.

Perrault J. Hereditary pancreatitis. Gastroenterol Clin North Am 1994; 4:743–751.

Weizman Z, Durie P. Acute pancreatitis in childhood. J Pediatr 1988; 113:24–29.

LIVER FAILURE

Kirsh BM, Lam N, Layden TJ, Wiley TE. Diagnosis and management of fulminant hepatic failure. Compr Ther 1995;21:166–171.

Lidofsky SD. Fulminant hepatic failure. Crit Care Clin 1995;11:415–430.

Tygstrup N, Ranek L. Assessment of prognosis in fulminant hepatic failure. Semin Liv Dis 1986;6:129–137.

Whitington PF. Fulminant hepatic failure in children. In: Suchy FJ, ed. Liver Disease in Children. St Louis: Mosby, 1994:180–213.

VIRAL HEPATITIS

Balistreri WF. Viral hepatitis. Pediatr Clin North Am 1988;35:375.

Lau JYN, Alexander GJM, Alberti A. Viral hepatitis. Gut 1991(Suppl):547.

Sherlock S, Dusheiko G. Hepatitis C virus updated. Gut 1991;32:965.

CHAPTER 94

Pediatric and Adolescent Gynecology

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INTRODUCTION

Evaluation of Premenarchal Girls

Among premenarchal girls with gynecologic complaints, the most commonly encountered specific entities are vaginal infections, urethral prolapse, trauma, and suspected sexual abuse. Many other patients have nonspecific genital irritation. In assessing any child with a gynecologic complaint, the physician must be alert to the possibility that sexual abuse is the underlying problem.

The premenarchal girl should not receive a standard pelvic examination, including the use of a speculum and vaginoabdominal palpation, because such an examination is uncomfortable and unnecessary for diagnosis. An exception to this rule is the girl with vaginal bleeding caused by an injury. If an external source for the bleeding cannot be identified, speculum examination of the vaginal vault under sedation or anesthesia is warranted to allow visualization of the injury. Some major vaginal lacerations produce only mild pain or minimal bleeding. For most premenarchal girls, the history, a general physical examination including inspection of the vulva (Fig. 94.1) and visualization the vagina, and culture of a vaginal discharge, if one is present, will lead promptly to a diagnosis.

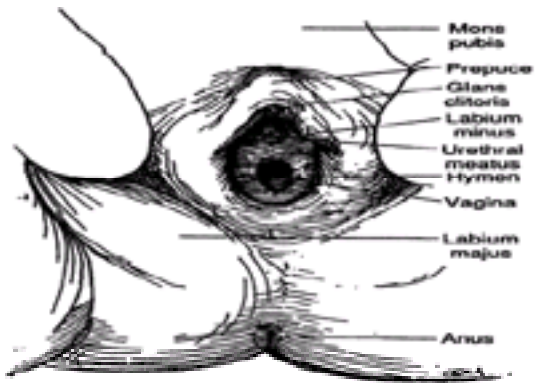

FIGURE 94.1. Anatomy of the normal female external genitalia.

Most young children will cooperate readily for initial inspection of the external genitalia either on the examining table or while held on a parent's lap. The child should be placed in the supine position with flexed hips and knees and with heels touching (Fig. 94.2). In most cases, the examiner can obtain a good view of the child's introitus by grasping the labia majora firmly and exerting gentle laterocaudal traction. For inspection of the vaginal vault, the knee–chest position is helpful. Most children more than 4 years old can cooperate for this maneuver. The child is first asked to “get up on your hands and knees like you are going to crawl.” She is then instructed to rest her head on her folded arms, facing her parent. The examiner or an assistant gently presses the child's buttocks and labia upward and outward. If the child

relaxes her abdominal muscles and back at this point, her vagina will usually fall open, permitting inspection of the vault using an otoscope as a light source (Fig. 94.2). If the child has a vaginal discharge or bleeding, she should then be returned to the supine position so that specimens for culture can be obtained, using either a soft plastic medicine dropper or a cotton-tipped swab moistened with nonbacteriostatic saline solution. A rectal examination should be performed if there is abdominal pain or a lower abdominal mass. Rectoabdominal palpation may be helpful if a hard vaginal foreign body is suspected, but nearly all vaginal foreign bodies are composed of toilet tissue and are therefore not palpable. On rectal examination, a child's cervix is normally felt as a firm, midline button of tissue, but the uterus and ovaries should not be palpable.

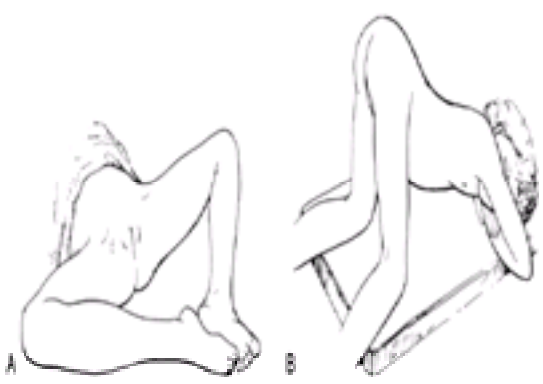

FIGURE 94.2. A. Girl in the frog-leg position for examination of the external genitalia. **B.** Girl in the knee–chest position with exaggerated lordosis and relaxed abdominal muscles. The examiner can inspect the interior of her vagina by gently separating her buttocks and labia, using an otoscope without an attached speculum for illumination.

Evaluation of Adolescent Girls

The differential diagnosis of gynecologic symptoms and signs in an adolescent girl who has experienced sexual intercourse includes a number of major entities (e.g., pregnancy, pelvic inflammatory disease [PID], tubo-ovarian abscess) that do not pertain to the teenager who is not sexually experienced. Therefore, considering the substantial proportion of American teenagers who are sexually experienced (Fig. 94.3), the emergency physician evaluating an adolescent girl must routinely inquire about sexual activity and, if the response is positive, about contraceptive use, prior sexually transmitted infections, pregnancies, and abortions. A detailed menstrual history—age at menarche, characteristics of the menstrual cycle, date of last menstrual period, presence or absence of dysmenorrhea—should be obtained from every adolescent patient.

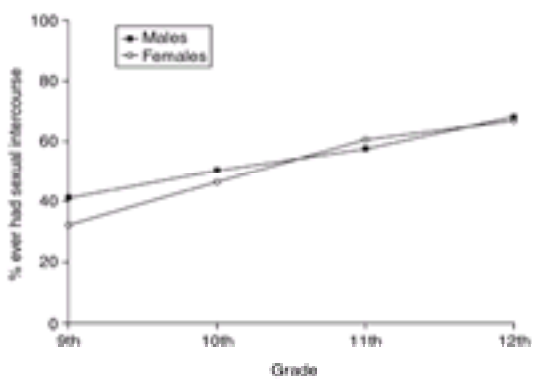

FIGURE 94.3. Percentage of U.S. high school students who have ever had sexual intercourse. (Adapted from Kann L, Warren CW, Harris WA, et al. Youth risk behavior surveillance—United States, 1995. In: CDC Surveillance Summaries, Sept 27, 1996. MMWR 1996;45[SS-4]:66.)

Obtaining a candid history of sexual activity from adolescent girls is not always a simple matter, but the emergency physician can maximize honest reporting by using some basic interviewing principles. First, the teenage girl who asks for a female physician is stating directly what will make her more comfortable. Her request should therefore be honored if it is possible to do so. Second, questions to the teenager about sexual activity (as well as other potentially sensitive subjects like contraception and substance abuse) should not be asked while a parent is present. This is accomplished by asking the parent to leave the room during the daughter's physical examination. Third, before any questions are asked, it is helpful to assure the teenager that if she wishes, her answers will be kept confidential. [1] Finally, the physician who adopts an empathetic, nonauthoritarian interviewing style is most likely to win the teenager's trust.

Most virginal adolescents with menstrual cramps, mittelschmerz, or vaginal discharge do not require full pelvic examinations because the likelihood that occult pelvic pathology will be found is small. Rectoabdominal palpation can be used to evaluate the virginal patient with undiagnosed lower abdominal pain or a mass. However, trauma and vaginal bleeding are indications for pelvic examination, even among patients who have never been sexually active.

Every sexually experienced adolescent girl who comes to the emergency department (ED) for abdominal pain or a gynecologic complaint must receive a pelvic examination because such patients have high rates of pregnancy and sexually transmitted disease (STD). Before the examination, the patient should be given a chance to empty her bladder.

If the physician is male, he should always be accompanied by a female chaperone during the examination. In the emergency setting, a chaperone may also be considered for female physicians. After the patient is situated in the lithotomy position and draped, her vulva is inspected, and a narrow speculum is inserted for visualization of her vagina and cervix. A sterile cotton-tipped swab is used to collect endocervical secretions for culture of *Neisseria gonorrhoeae.* A second endocervical specimen is obtained to test for *Chlamydia trachomatis* antigen. After a sample of vaginal discharge for microscopic examination has been taken with a third swab, the speculum is removed. If the physician suspects gonococcal infection, urethral and rectal swabs may also be taken. The endocervical specimens should be obtained before bimanual palpation is done because lubricating jelly can inhibit growth of *N. gonorrhoeae.* During the bimanual examination, the cervix is assessed for softness, patency of the os, and pain elicited by lateral cervical movement. The size and consistency of the uterus are determined, and the adnexal areas are palpated for masses and tenderness. Last, rectovaginal palpation is performed, checking again for masses and local tenderness.

The emergency physician can add an ounce of prevention to the care of the sexually active teenage girl if he or she not only provides treatment for the patient's current problem but also determines whether she has been receiving routine outpatient gynecologic care. The ED should maintain a list of local programs that provide health care to adolescents so that referral can be made easily. Many communities have specialized services for teenagers sponsored by hospitals, health departments, or private agencies. Similarly, because previously unrecognized pregnancy is a common ED diagnosis, procedures should be established to facilitate prompt referral of teenagers who need counseling, prenatal care, and therapeutic abortion to the appropriate services.

GYNECOLOGIC DISORDERS OF CHILDHOOD

Congenital Vaginal Obstruction

Background

Definition

In normal females, the vagina provides an outlet for genital secretions and menstrual blood. If the vagina is obstructed, accumulating fluid will eventually distend it, causing symptoms to develop either during infancy or after menarche. During infancy, vaginal distension with mucus secreted as a result of stimulation by maternal hormones is called hydrocolpos or mucocolpos. If the volume of secretions is so large that the uterus is also distended, this condition is called hydrometrocolpos. If an obstructing congenital malformation is not recognized before menarche, menstrual blood will gradually fill the vagina, producing hematocolpos or, less commonly, hematometrocolpos.

Etiology

For hydrocolpos or one of its variations to arise, a female must have vaginal obstruction, a uterus, and a patent cervix. The two most common anomalies with these features are transverse vaginal septum (sometimes called vaginal atresia), and imperforate hymen. Although the embryologic origin of these malformations is not fully understood, they are probably produced between the 16th and 20th weeks of gestation if the developing vaginal plate fails to perforate at its junction with either the fused paramesonephric (Müllerian) ducts proximally or the urogenital sinus caudally. Most patients with complete agenesis of the vagina (Rokitansky-Küster-Hauser syndrome) have rudimentary uteri or none at all, so hydrocolpos does not occur.

Epidemiology

Transverse vaginal septum occurs sporadically, with an estimated incidence of 1 in 2,000 to 84,000 girls. However, a few inbred kindreds have been described with transverse vaginal septum, polydactyly, and congenital heart disease (McKusick-Kaufman syndrome). In one survey, imperforate hymen occurred in 0.1% of term female neonates. Epidemiologic studies may be confounded because the two conditions can be confused with each other and because a congenitally imperforate hymen occasionally may open spontaneously during infancy.

Clinical Manifestations

Infancy

Although vaginal obstruction should properly be identified during the initial examination of the newborn female, infants with hydrocolpos often go unrecognized until days or weeks later when they develop the three hallmarks of this condition: 1) a lower abdominal mass, 2) difficulty with urination, and 3) a visible bulging membrane at the introitus. Except in mild cases, the lower abdominal mass consists of the bladder as well as the hydrocolpos itself. The infant strains to micturate or has urinary retention because the urethra is obstructed extrinsically. In more severe cases, infants may also have constipation, hydronephrosis, edema of the lower extremities, and hypoventilation. Inspection of the perineum should immediately indicate the proper diagnosis.

Adolescence

The girl with congenital vaginal obstruction who escapes notice during infancy will not come to attention until late in puberty when she presents with either primary amenorrhea or lower abdominal pain. She will have had satisfactory pubertal development until her menarche apparently fails to occur. Accumulating menstrual blood will then eventually produce vague lower abdominal pain that is not necessarily cyclic. As the hematocolpos grows, it will finally interfere with comfortable micturition, producing symptoms of urgency, frequency, or dysuria. The history of amenorrhea and the finding of a lower abdominal mass may lead the physician to suspect a tumor or even pregnancy, but the characteristic appearance of the introitus covered by a bluish bulging membrane is diagnostic of hematocolpos with imperforate hymen

(Fig. 94.4). Patients with a high transverse vaginal septum will not be so easily diagnosed because the introitus will appear normal. However, palpation of the vagina will promptly show that it is obstructed and that the cervix cannot be felt.

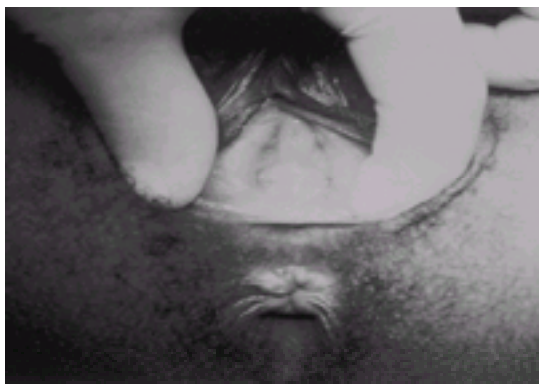

FIGURE 94.4. Hematocolpos in a 15-year-old patient with an imperforate hymen. The membrane bulges at the introitus underneath the labia minora (see also Fig. 94.5).

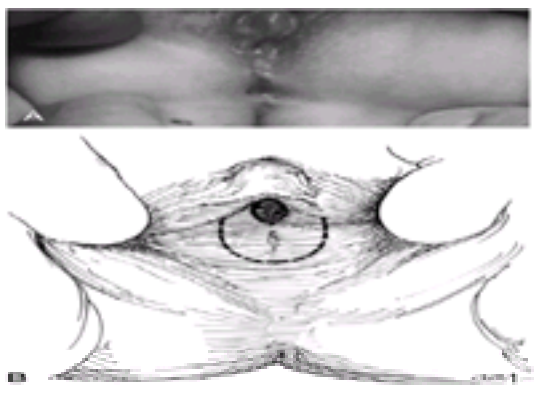

FIGURE 94.5. A. Labial adhesion in an asymptomatic 3-year-old girl. **B.** A flat surface, a dense central line of fusion, and an opening below the clitoris are characteristic features of labial adhesions, which cover the introitus (see also Fig. 94.4).

Complications

The complication of congenital vaginal obstruction most likely to require urgent attention among both infants and adolescents is acute urinary retention. This condition can be managed readily after the primary diagnosis has been recognized. Patients without complete urethral obstruction can instead have variable degrees of hydroureter or hydronephrosis as a result of the chronic extrinsic pressure. Rarely, an infant may have respiratory insufficiency or inferior vena caval obstruction because of the large mass. Imperforate hymen is usually an isolated anomaly, but other types of obstruction, chiefly transverse vaginal septum, are regularly associated with renal malformations, including hypoplastic or single kidneys, and duplicated or ectopic collecting systems. Therefore, the patient's laboratory evaluation should include assessment of both renal function and urinary tract anatomy. Endometriosis can be a late complication of severe hematocolpos.

Differential Diagnosis

The differential diagnosis of hydrocolpos and its variations includes patients with either a lower abdominal or pelvic mass but no vaginal obstruction, and patients with apparent vaginal obstruction. In the former category, the physician simply needs to demonstrate that the patient has a patent genital tract. The usual measures for diagnosis of the mass can then be undertaken. The latter group includes girls with microperforate hymen, labial adhesions, Gartner's (mesonephric) duct cysts, and rarely, complete agenesis of the vagina and testicular feminization. The microperforate hymen has a tiny orifice just below the urethra and requires only careful inspection for its diagnosis. Adhesions of the labia minora are superficial to the plane of the hymen and are characterized by a central vertical line of fusion (Fig. 94.5). A large vaginal Gartner's duct cyst can resemble an imperforate hymen, but it can be seen to protrude through the hymenal ring, which is itself patent. Patients with complete agenesis of the vagina have only a rugated dimple or shallow indentation at the introitus. A short blind vagina also occurs in testicular feminization, a disorder characterized by end-organ insensitivity to androgen. These patients are phenotypic females with an XY karyotype who undergo breast development at puberty but lack pubic hair and female reproductive organs.

Management

Patients with congenital vaginal obstruction need surgical treatment. Hydrocolpos or hematocolpos complicated by respiratory insufficiency, compression of the inferior vena cava, or hydronephrosis must be corrected without delay. The management of simple imperforate hymen can be modified according to the patient's age. Surgery should be scheduled promptly for adolescents but can be performed electively for asymptomatic infants and children.

Labial Adhesions

Background

Labial adhesions are an acquired attachment of the medial surfaces of the labia minora to each other. The terms *labial fusion, synechiae,* and *agglutination* are also often used to describe this condition. Labial adhesions are a common gynecologic condition, occurring in approximately 3 to 7% of girls between the ages of 3 months and 5 years. Most patients are between the ages of 1 and 6 years, but adhesions have been reported in girls as young as 6 weeks of age. The median age at diagnosis is 2 years.

Pathophysiology

A cause for labial adhesions has never been established. However, it is generally agreed that they are a concomitant of the child's normally low levels of endogenous estrogen. Without estrogen, the genital epithelium is relatively thin and susceptible to irritation. Possibly following some episode of inflammation, the two labial surfaces gradually stick together, with the line of fusion usually advancing anteriorly from the posterior frenulum of the labia minora.

Clinical Manifestations

The parent who notices a daughter's labial adhesions at home usually brings her to the ED with a chief complaint that the child's vagina is “closing up.” Alternatively, a physician may notice the adhesions during the child's routine physical examination. The situation is much more difficult when, as occasionally happens, the child is brought to the ED for evaluation because another clinician has mistakenly informed the parents that their daughter has congenital absence of the vagina or ambiguous genitalia. Imperforate hymen is another notable misdiagnosis. In these cases, primary focus must be to provide a careful explanation and reassurance for the distressed parents because the condition is essentially minor.

The diagnosis of labial adhesions can be made promptly and confidently by simple inspection of the child's genitalia. When the labia majora are gently retracted laterally, a flat plane of tissue marked by a central vertical line of adhesion obstructs the view of the introitus within (Fig. 94.5B). This thin vertical raphe is pathognomonic of labial adhesions. It is occasionally difficult to detect if the child's adhesions are old and dense. The length of the adhesions is variable, and they can be perforated. They are usually thickest posteriorly and stop below the clitoris. Even when adhesions appear to have closed the vulva completely, a pinpoint opening generally permits the egress of urine.

Most girls with labial adhesions are asymptomatic. A few have associated dysuria, frequency, or refusal to void that may be a result of either the obvious mechanical obstruction or concurrent urinary tract infection. Whether associated urinary tract infections are a cause or an effect of adhesions is uncertain, but they are a recognized complication of the condition. Girls with urinary tract symptoms should receive urine cultures and appropriate medical follow-up. Because vaginal infection is not associated with adhesions, vaginal cultures are not indicated except in patients who have concurrent vaginal discharge. Asymptomatic girls need no laboratory evaluation.

Management

Treatment is not indicated for asymptomatic girls with labial adhesions because the condition spontaneously remits early in puberty as a result of increasing endogenous estrogen. Some parents, however, prefer that their daughters be treated. Of girls with labial adhesions, 90% can be treated successfully with a small amount of estrogen cream (Premarin or Dienestrol) dabbed onto the adhesions at bedtime for 2 to 4 weeks. After the labia have separated, an inert cream (zinc oxide, Vaseline, Desitin) is applied nightly for an additional 2 weeks to keep the labia apart while healing is completed. Because pharmacies often supply estrogen cream in large tubes, parents should be warned specifically that prolonged use of the hormone can stimulate breast growth in children. Vulvar hyperpigmentation is a common, transient side effect of treatment. Labial adhesions should never be manually separated. The procedure is painful and usually results in relapse when the raw, newly separated labia adhere again. Topical estrogen cream is a painless, inexpensive, and effective therapeutic alternative.

Urethral Prolapse

Background

Urethral prolapse is the protrusion of the distal urethral mucosa outward through its meatus, with a cleavage plane between the longitudinal and circular-oblique smooth muscle layers of the urethra. The prolapsed segment is constricted at the meatus, and venous blood flow is impaired, so the involved tissue becomes swollen, edematous, and dark red or purplish. If the process is not corrected, the tissue can become thrombosed and necrotic.

About half of affected females are prepubertal children, and most of the remainder are postmenopausal women. Most prolapses during childhood occur in girls between the ages of 2 and 10 years. Nearly 95% of affected girls reported in the English-language literature since 1937 have been African-American. This racial disparity among affected children remains unexplained. In contrast, race does not appear to be a risk factor for prolapse among postmenopausal women.

Although the cause of urethral prolapse is not well understood, a sudden or recurrent increase in intra-abdominal pressure (severe coughing, seizure, constipation, lifting heavy objects) has been noted to precede some cases. Other proposed causes of prolapse include local trauma, redundant urethral tissue, neuromuscular dysfunction, and inadequate pelvic support of the bladder neck and urethra, but no convincing evidence has been presented to support any of these hypotheses.

Clinical Manifestations

Vaginal bleeding or spotting is the chief complaint of 90% of children with significant urethral prolapses. The bleeding is painless, occasionally misinterpreted as hematuria or menstruation, and accompanied by urinary frequency or dysuria in about one-fourth of cases. Only a minority of girls or their parents are aware of the presence of a vulvar mass. On the other hand, it is not rare for the physician simply to note a small prolapse during the routine examination of an asymptomatic child.

On examination of the child's vulva, a red or purplish, soft, doughnut-shaped mass is seen (Fig. 94.6). Most prolapses are not tender and measure 1 to 2 cm in diameter. By retracting the labia majora posterolaterally, the examiner can often demonstrate that the mass is separate from and anterior to the vaginal introitus, but this process may be difficult if the prolapse is large. A small central dimple in the mass indicates the urethral lumen. This dimple can be missed if lighting is inadequate, bleeding is active, or mucosal edema is significant. In most cases, the appearance of the prolapse is diagnostic. However, if the diagnosis is in doubt, sterile straight catheterization of the bladder through the mass can be performed to demonstrate the anatomic relationships safely and rapidly. No other test is needed. If urinalysis is performed on a spontaneously voided specimen, red blood cells are likely to be found, but urine cultures are routinely sterile. Urethral polyps, prolapsed ureterocele, sarcoma botryoides, and urethral carcinoma may be included in the differential diagnosis, but these entities are rare in children and lack the characteristically annular appearance of a urethral prolapse.

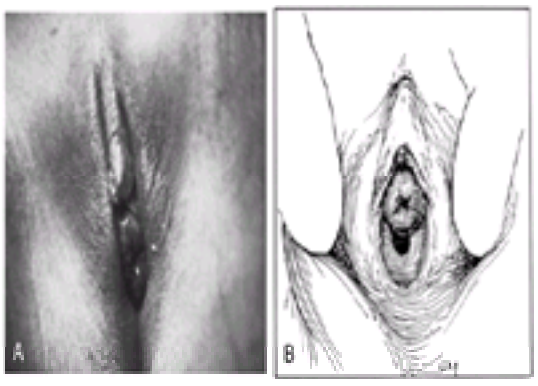

FIGURE 94.6. A. Urethral prolapse in a 6-year-old girl with “vaginal” bleeding. The vaginal orifice cannot be seen. **B.** The smooth doughnut shape and central lumen are characteristic features of a urethral prolapse, which, if large or swollen, often conceals the vagina below it.

Management

For the symptomatic patient with a small segment of prolapsed mucosa that is not necrotic, warm moist compresses or sitz baths, combined with a 2-week course of topical estrogen cream, may be prescribed. Most patients treated in this way have improved within 10 to 14 days and remained normal thereafter, thus avoiding surgery. Patients with dark red or necrotic mucosa should be treated surgically within several days by reduction of the prolapse and/or excision of necrotic tissue. After the diagnosis is confirmed by cystoscopy, the prolapse is excised and the cut edges are sutured together. The procedure is simple and can be carried out in a day surgery unit.

GYNECOLOGIC DISORDERS OF ADOLESCENCE

Dysmenorrhea

Background

Definition

Dysmenorrhea means painful menstruation. It is primary if the pain cannot be attributed to a specific pelvic abnormality such as endometriosis, PID, or a uterine malformation. Menstrual pain resulting from an underlying disorder is termed secondary dysmenorrhea.

Epidemiology

Among all women, and among adolescents in particular, primary dysmenorrhea is far more common than secondary dysmenorrhea. Estimates of the incidence of primary dysmenorrhea have varied, depending on both the age of the women surveyed and the criterion used for a positive response (any pain, pain that interferes with normal activity, or pain that prompts a visit to a physician). In the U.S. National Health Examination Survey (1966 to 1970), 60% of postmenarchal girls between the ages of 12 and 17 years reported having menstrual pain or discomfort. The prevalence of dysmenorrhea increases with increasing chronologic and gynecologic (postmenarchal) age, reflecting the strong association of dysmenorrhea with ovulatory menstrual cycles. At gynecologic age 1, approximately 30% of girls have dysmenorrhea. By gynecologic age 5, the proportion increases to nearly 80%.

Pathophysiology

A pathophysiologic basis for dysmenorrheic pain could not be demonstrated until the 1970s, when numerous investigations indicated that at the end of ovulatory menstrual cycles, prostaglandins $F_{2}a$ and E_2 are synthesized by and released from endometrial tissue. The prostaglandins cause increases in both the uterine resting tone and the amplitude and frequency of myometrial contractions. Uterine contractions that exceed systolic blood pressure produce tissue ischemia and perceptible pain. Intravenous administration of prostaglandins can reproduce the systemic discomforts—vomiting, diarrhea, headache—that often accompany dysmenorrhea. A dose–response relationship has been demonstrated in studies comparing the prostaglandin content of menstrual fluid from women with and without dysmenorrhea and from individual women during painful and pain-free cycles.

The role of ovarian hormones in endometrial prostaglandin production is not completely understood. Progesterone or its withdrawal appears to enhance prostaglandin synthesis during ovulatory cycles. Conversely, progesterone-impregnated intrauterine devices and the inhibition of ovulation by birth control pills are both associated with decreased prostaglandin production and with relief from dysmenorrhea.

Clinical Manifestations

Typical primary dysmenorrhea consists of cramping, dull, midline, or generalized lower abdominal pain at the onset of a menstrual period. The pain may coincide with the start of bleeding or may precede the bleeding by several hours. Many women have associated symptoms including backache, thigh pain, diarrhea, nausea or vomiting, and headache. The discomfort usually abates within 48 hours. Because dysmenorrhea is a hallmark of ovulation, adolescents characteristically do not experience dysmenorrhea until after several months of painless, anovulatory cycles. Menstrual pain that begins either at menarche or more than 4 years after regular cycles have been established is less common. The patient with such early or late dysmenorrhea should be assessed carefully for a possible underlying disorder, but she is still more likely to be having simply a particularly early or late onset of fertile cycles. Because they inhibit ovulation, both combined oral contraceptive pills and contraceptive progestins almost uniformly abolish dysmenorrhea.

Patients with straightforward dysmenorrhea have normal physical examinations and no associated abnormalities on routine laboratory evaluation. PID and congenital abnormalities (rudimentary uterine horn, partially obstructed genital duplications) must be included in the differential diagnosis for sexually active adolescents and those with atypical pain or a pelvic mass. Endometriosis and postoperative adhesions are also potential explanations for chronic, cyclic, undiagnosed pelvic pain in teenagers.

Management

The virginal adolescent with a typical history of dysmenorrhea should undergo a routine physical examination, but a pelvic examination is unnecessary. However, virginal patients with atypical or severe pain should undergo recto-abdominal or one-finger vaginoabdominal palpation. Sexually experienced adolescents with pelvic pain cannot be adequately evaluated without a complete pelvic examination to screen for pelvic infection and unsuspected pregnancy. No specific laboratory or radiologic evaluation is needed for otherwise healthy virginal adolescents. Pelvic ultrasound or laparoscopy may be helpful in the assessment of patients with uncertain diagnoses or with pain unresponsive to adequate treatment (see the following section).

Nonsteroidal anti-inflammatory drugs (NSAIDs) are the treatment of choice for patients with moderate or severe dysmenorrhea, providing pain relief for 60 to 80% of symptomatic women. Aspirin (650 mg four times a day) can be recommended for patients with mild discomfort but is several hundred times less potent an inhibitor of prostaglandin synthesis. Like aspirin, but to a lesser extent, NSAIDs cause gastrointestinal (GI) irritation, inhibit platelet aggregation, and prolong the bleeding time. NSAIDs may also reduce the volume of menstrual blood loss, a little recognized but potentially beneficial side effect. Both NSAIDs and aspirin are contraindicated in patients with aspirin hypersensitivity, GI ulcers, and bleeding disorders. Examples of commonly used NSAID treatment regimens are as follows:

- Mefenamic acid: 500 mg once orally, followed by 250 mg four times a day
- Ibuprofen: 400 to 600 mg orally four times a day
- Naproxen: 500 mg once orally, followed by 250 mg four times a day
- Naproxen sodium: 550 mg once orally, followed by 275 mg four times a day

For sexually active adolescents with dysmenorrhea, birth control pills are an attractive and effective alternative to NSAIDs because they provide both contraception and pain relief. Other agents commonly recommended in the past for the treatment of dysmenorrhea—acetaminophen, caffeine, propoxyphene—lack specific antiprostaglandin action and have only limited effectiveness.

Dysfunctional Uterine Bleeding

Background

Dysfunctional uterine bleeding (DUB) is best characterized as irregular, prolonged, or excessive menstrual bleeding associated with anovulation and unrelated to pregnancy. Ovulatory cycles occur in about 20% of adults with DUB, but this phenomenon is uncommon during the teenage years. Terms often used to categorize patterns of DUB are *metrorrhagia* (irregular or acyclic bleeding) and *menorrhagia* (excessive duration or quantity of bleeding). Menstrual bleeding that persists beyond 9 days, that recurs at intervals of fewer than 21 days, or that produces anemia is abnormal and warrants attention.

DUB is prevalent at the beginning and end of the reproductive years, paralleling the times when anovulatory cycles are most common. In girls during the first year after menarche, about half of menstrual cycles are anovulatory. This proportion decreases gradually, so only 5% of cycles are anovulatory 10 years or more after menarche. Of course, most

adolescents with anovulatory cycles nevertheless experience self-limited, reasonably cyclic bleeding episodes. Why more girls do not develop DUB is not well understood.

Pathophysiology

From the standpoint of ovarian function, the normal ovulatory menstrual cycle is divided into an initial follicular and a subsequent luteal phase. The parallel phases of endometrial development are termed, respectively, *proliferative* and *secretory.* At the start of an ovulatory cycle, pituitary follicle-stimulating hormone (FSH) promotes the growth of ovarian follicles. In turn, the rising concentration of estradiol from these follicles stimulates the proliferation of endometrial stroma and glands, has a negative feedback effect on the secretion of FSH, and finally induces a midcycle surge of luteinizing hormone (LH) that triggers ovulation. The duration of the preovulatory phase of the menstrual cycle is variable but generally lasts about 14 days. After ovulation has occurred, the ruptured ovarian follicle forms a corpus luteum that secretes progesterone as well as estradiol, and levels of both FSH and LH gradually decline. Although progesterone limits the ultimate thickness of the endometrium, it also promotes further growth of the endometrial secretory glands and spiral blood vessels, so they become coiled and tortuous. At the end of the corpus luteum's life span (a highly consistent 14 days unless conception occurs), it degenerates, and circulating levels of both estrogen and progesterone fall, eventually stimulating a resurgence of LH and FSH. As hormonal support wanes, blood flow to the secretory endometrium diminishes, and the spiral arterioles constrict and relax rhythmically under the influence of local prostaglandins. The resulting progressive ischemia leads to endometrial necrosis. Menstrual sloughing begins, and the cycle starts over again.

In contrast, during intervals of anovulation, luteal progesterone is not present to limit the endometrium's thickness or to promote its structural integrity. Parts of the endometrial surface undergo growth and sloughing sporadically, without cyclic coordination. The amount of estrogen secreted by ovarian follicles fluctuates unpredictably, and bleeding can occur either because of a fall in estrogen level (withdrawal bleeding) or despite a sustained level of production (breakthrough bleeding). Relatively constant low levels of estrogen tend to produce intermittent spotty bleeding (metrorrhagia). Larger amounts of estrogen cause greater endometrial proliferation and a cyclic pattern of amenorrhea followed by profuse bleeding (menorrhagia) whenever either the endometrial vessels and glands outstrip their stromal support, or hormone levels spontaneously fall. Compared with the 35 to 75 mL of blood lost during a normal menstrual period, dysfunctional bleeding often results in the loss of 100 to 200 mL each month. It is no surprise, therefore, that iron deficiency with depleted marrow stores or outright anemia is often seen in patients with DUB.

Clinical Manifestations

DUB has a substantial capacity to disrupt the everyday activities of adolescent patients discomfited by an unpredictable, urgent need for bathroom facilities and the risk of visible bloodstains. Large amounts of bleeding often provoke considerable fear in both patients and their parents. These concerns can overshadow the history of the bleeding itself, but the details of the problem's chronology and an estimate of blood loss (in pads per day) will help the physician assess the severity of the bleeding, follow the patient's clinical course, and gauge her prognosis. The symptoms that characteristically accompany only ovulatory menstrual cycles—mittelschmerz, premenstrual breast tenderness, bloating, mood changes, and dysmenorrhea—should be absent. DUB is classically painless, but occasionally, a patient with active bleeding may experience crampy pain if a large quantity of blood is passed rapidly. Weakness or fainting should alert the examiner to the possibility of significant blood loss. Pertinent questions should include whether the patient is pregnant, whether she uses contraception if she has been sexually active, and whether she has an underlying platelet disorder (e.g., thrombocytopenia, von Willebrand's disease).

The physical examination starts with measurement of the patient's vital signs, including a check for orthostatic changes in the pulse and blood pressure. Pertinent signs, including pallor, petechiae or bruises that might indicate a bleeding disorder, and hirsutism or obesity consistent with the polycystic ovary syndrome, should be sought and noted. The pelvic examination is likely to be normal except for the presence of bleeding but should be performed to evaluate the patient for pelvic infections, previously unrecognized pregnancy, and functional ovarian cysts. If necessary, rectoabdominal palpation can be substituted for the standard bimanual examination. Ovarian enlargement is an uncommon finding even among adolescent patients with clear-cut polycystic ovary syndrome. The differential diagnosis of DUB is discussed at greater length in Chapter 76.

Management

A determination of the hemoglobin or hematocrit is essential for the emergency evaluation of patients with DUB because historical estimates of blood loss are imprecise. A platelet count should also be obtained because thrombocytopenia is the most common hematologic disorder that produces menorrhagia. The history and physical examination should be used to guide the choice of additional laboratory tests. Sexually active adolescents should receive a pregnancy test, a cervical culture for gonorrhea, and an antigen-detection test for chlamydial infection because STD-associated endometritis is a common cause of otherwise unexplained uterine bleeding. Patients with menorrhagia beginning at menarche, severe hemorrhage, or a history of bleeding problems should undergo further evaluation for possible disorders of platelet number or function.

In order of decreasing urgency, management of DUB includes the identification and treatment of the following problems: shock or acute hemorrhage, moderate bleeding usually accompanied by anemia, and minor bleeding that produces distress but no imminent danger for the patient (Table 94.1). For patients with brisk hemorrhage or hypotension, prompt hospitalization and volume resuscitation as necessary (see Chapter 3) are the first order of business.

Clinical Situation	Treatment	Comments
Shock, acute hemorrhage	Volume resuscitation Conjugated estrogens 20–25 mg IV every 4 hr with maximum of 6 doses Curettage if estrogen unsuccessful	Add progestin promptly Prescribe iron
Moderate bleeding	Oral estrogen plus progestin, QID regimen Intravenous estrogen if oral treatment unsuccessful	Anemia common Prescribe iron
Minor bleeding	Oral estrogen plus progestin, BID regimen Observation without treatment acceptable	

Table 94.1. Management of Dysfunctional Uterine Bleeding

Control of the bleeding itself is accomplished with hormonal treatment. Regimens vary according to the severity of bleeding and individual preference, but each is designed first to stop the bleeding, second to convert the unstable proliferative uterine endometrium to the secretory state, and finally to allow a self-limited endometrial slough under controlled conditions. (Pregnancy must be excluded in every case before hormonal treatment is begun.) Estrogen is used to support the endometrium acutely and to stop the bleeding. A progestational agent must be administered simultaneously to produce a secretory endometrium; otherwise, the problem will recur predictably whenever the estrogen is stopped. Any of the oral contraceptive pills with 35 or 50 µg of either ethinyl estradiol or mestranol and a progestin provides a convenient means of administering the two hormones together. The dosage for patients with active bleeding and anemia is 1 estrogen–progestin tablet orally four times a day for 5 days. In almost every case, bleeding will decrease substantially within 24 hours and stop within 2 to 3 days. A rarely needed alternative treatment for hospitalized patients consists of conjugated estrogens 20 to 25 mg intravenously every 4 hours until the bleeding stops, with a maximum of six doses. This treatment must be accompanied by a progestational agent (medroxyprogesterone 10 to 20 mg orally per day for 5 to 12 days). If hormonal treatment fails to arrest the bleeding, dilation and curettage should be performed, but the procedure is almost never necessary.

For patients with light but prolonged bleeding and a normal hemoglobin, the oral regimen can be reduced to 1 estrogen–progestin tablet twice a day for 5 days. Nausea is a common side effect of estrogen in each of these regimens and can be treated symptomatically. Vomiting rarely precludes oral therapy. A progestin alone in higher dosages (norethindrone acetate 10 to 20 mg/day for 5 to 12 days) can be used if estrogen is contraindicated or not tolerated, but the resulting hemostasis is less prompt and less predictable.

Treatment with combined estrogen–progestin pills is stopped at the end of 5 days; progestin-only treatment is stopped after 5 to 12 days. After the cessation of treatment, a self-limited, heavy menstrual period will follow within 2 to 3 days. The family must be forewarned so that they anticipate this episode and do not misinterpret it as a recurrence of the DUB. This withdrawal bleeding will stop spontaneously within several days. Subsequent therapy must be tailored to the individual patient. A course of medroxyprogesterone (10 mg orally, daily for 10 to 12 days) can be used every 6 to 8 weeks to produce a secretory endometrium and a controlled withdrawal flow, if spontaneous menstruation has not intervened. For sexually active adolescents and those with chronic or recurrent DUB, long-term combined oral contraceptive pills are an excellent therapeutic choice.

Adolescents with severe, chronic, or recurrent DUB should receive diagnostic investigation to address the question of an underlying endocrinologic disorder (see Chapter 48). Iron supplementation is prudent for all patients with DUB; those without frank anemia are likely to have depleted marrow stores of iron. Finally, outpatient follow-up is an essential component of management because treatment may be needed for months or years and because chronic anovulation is a risk factor for both infertility and the late development of endometrial carcinoma.

Sexual Abuse and Assault

The four major gynecologic aspects of treating a girl who has been sexually abused or assaulted are 1) collection of evidence from the genital area, 2) screening and treatment for STDs, 3) management of injury, and 4) prevention of pregnancy. The overall management of sexually abused children is discussed in Chapter 128.

Police and prosecutors ask physicians to document observations and to collect evidence that may corroborate a patient's history of sexual assault or abuse. This evidence may consist of a child's exact statement concerning the abuse; the finding of prostatic acid phosphatase in vaginal secretions, indicating the presence of seminal fluid; a small bruise on the labia majora; or the discovery of leaf or grass fragments in the underwear of a child who states that she was assaulted in a park. As a rule, any material that might be helpful should be collected if the child has had sexual contact within 72 hours of her evaluation in the ED. A number of commercially available “rape kits” contain swabs, tubes, and evidence tape to simplify this process. However, the clinician should not feel compelled to follow rape kit instructions slavishly because many of the specimens called for (e.g., pubic hair sample, fingernail scrapings) are rarely relevant to the circumstances of victimized children. If material is collected that may be used in court, its movement from physician's possession to locked storage or police officer's custody should be documented with signatures, times, and dates to preserve the chain of evidence that will allow the material's origin to be verified in court. When, as is often the case, a sexually abused child is not examined until several weeks after the most recent episode of sexual contact, the likelihood of the physician's finding physical evidence is low.

STDs are seen in about 5% of abused children, generally mirroring their relative prevalence in the adult population. Although trichomoniasis is among the most common STDs in adults, it is not seen in prepubertal abused girls because trichomonads do not proliferate in the absence of an estrogenic milieu. Syphilis certainly can be transmitted by sexual

assault and abuse, but occurs only rarely. Sexual abuse has been reported as the suspected mode of transmission for some children infected with human immunodeficiency virus (HIV).

In the past, most authorities recommended universal screening of sexually abused children for syphilis and for gonorrhea and *C. trachomatis* from three sites (pharynx, vagina or urethra, and rectum). However, the yield of infections from such surveillance is low, and the cost is substantial. An alternative strategy accepted as appropriate by most experts is to restrict STD screening to high-risk children (children victimized by more than one assailant or by an assailant with a known or suspected STD) and to adolescents. All adolescents are screened because they are at higher risk than younger children of acquiring STDs and, especially, of subsequently developing PID if cervicitis is present. Serologic screening for syphilis and HIV infection should be offered to all children. Before a child is screened for HIV, the parent must be advised that a positive test may indicate vertical transmission and parental infection, rather than acquisition via the assault. Screening for hepatitis B infection can be limited to adolescents who have not received hepatitis B vaccine.

As is true for STD screening, antibiotic prophylaxis after sexual abuse is best limited to children victimized in high-risk situations and to adolescents. The antibiotic regimen should include coverage for both chlamydia and gonorrhea. For adolescents, coverage for trichomonas and bacterial vaginosis (and for hepatitis B if the patient is unvaccinated) should be added. Giving the first vaccine dose (followed in 1 and 6 months by the subsequent doses) is adequate prophylaxis for hepatitis B. As of this writing, the Centers for Disease Control and Prevention (CDC) make no specific recommendation for HIV prophylaxis after sexual assault. However, consideration should be given to offering antiretroviral prophylaxis to children and adolescents when the circumstances of an assault seem to pose a high risk of HIV infection.

For sexually abused children with symptoms or signs of an STD, the question of screening is irrelevant; diagnostic tests should be directed at identifying all suspected infections. The diagnosis and treatment of STDs are reviewed elsewhere in this chapter and in Chapter 84 and Chapter 85. When sexual abuse is involved, for the diagnosis of gonorrhea, chlamydial infection, and herpes simplex virus infection, cultures should be used either in place of or as confirmation of indirect diagnostic methods (e.g., Gram stain, DNA probe, other immunologic tests) because the risk of false-positive presumptive tests is substantial and because test results that may be presented in legal proceedings should be definitive.

Few girls sustain serious physical injuries as a result of sexual abuse or assault. Any sexually abused girl who has vaginal bleeding that cannot be attributed to a clearly visible injury or infection must be examined carefully to determine the source of the bleeding. This will usually require the use of sedation or general anesthesia in premenarchal girls. The management of girls with vaginal bleeding is discussed at greater length in Chapter 76.

The emergency physician must consider the possibility of pregnancy in every postmenarchal girl who has been sexually abused or assaulted. A pregnancy test should be conducted in the ED to ascertain whether the patient is pregnant when she is first evaluated. If an adolescent is not pregnant and is not using hormonal contraception, her risk of becoming pregnant as a result of rape must be assessed. The risk of pregnancy from a single unprotected coitus at midcycle is estimated to be 15%. The risk from coitus occurring more than 6 days before or after ovulation is negligible. Postcoital contraception—100 µg ethinyl estradiol and 1 mg D1-norgestrel (2 Ovral tablets) given once immediately and a second time 12 hours later—reduces the likelihood of pregnancy by about 50% and should be offered to patients seen within 72 hours of a rape. An antiemetic should also be prescribed because the estrogen often produces nausea. The effectiveness of this regimen for preventing pregnancy decreases as the time from coitus to treatment increases. The patient can expect to have her next menstrual period within 21 days after treatment and should be given an appointment for follow-up about 3 weeks after the ED visit.

GENITAL TRACT INFECTIONS

Vaginitis

Background

Definition

Vaginitis, or inflammation of the vagina, can be produced by chemical and mechanical irritants, foreign bodies, and a variety of infectious agents, including viruses, chlamydia, bacteria, fungi, protozoa, and helminths. During childhood, vaginitis is characterized by the presence of vaginal discharge, bleeding, or both. After puberty has begun, girls normally have an asymptomatic vaginal discharge; vaginitis is then indicated by the discomfort it produces or by a change in the character of the discharge. The etiology, clinical manifestations, diagnosis, and treatment of common vaginal infections are presented in this chapter. For a review of the differential diagnosis of vaginal bleeding and discharge, see Chapter 76 and Chapter 77. Table 94.2 and Table 94.3 summarize the treatment of common vaginal infections.

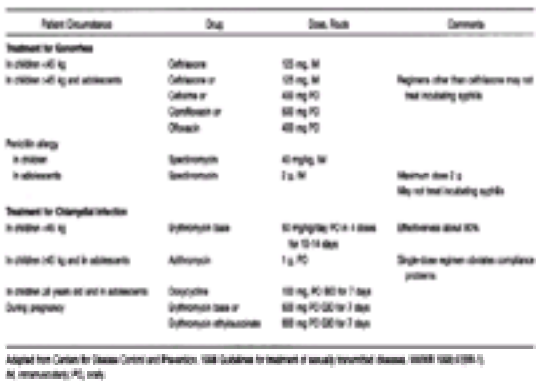

Table 94.3. Summary of Treatment Regimens for Lower Genital Tract Gonorrhea and Chlamydial Infection

Epidemiology

At least half of all symptomatic premenarchal girls with vaginal discharge visible on physical examination will prove to have specific vaginal infections that warrant antimicrobial treatment. Among prepubertal girls in the United States, *N. gonorrhoeae* causes the greatest number of these specific infections. Less common offenders include *Shigella* species, *Streptococcus pyogenes,* and in infants and after puberty has begun, *Trichomonas vaginalis.* Although staphylococci and *Haemophilus influenzae* usually colonize the lower genital tract without producing symptoms, they are associated with vaginal discharge in only a small proportion of patients. *Candida albicans* is the most common vaginal pathogen among both pubertal (but premenarchal) and postmenarchal girls.

The relative prevalence of vaginal infections in a population of postmenarchal adolescents depends primarily on how many of them are sexually active. The proportion of never-married teenagers who have experienced coitus increases steadily with age (Fig. 94.3) and is accompanied by a parallel increase in the prevalence of sexually transmitted infections. Among adult women, bacterial vaginosis is the most common vaginal infection, followed by vulvovaginal candidiasis and trichomoniasis. Bacterial vaginosis is found commonly and nearly exclusively among sexually active adolescents. Diabetes mellitus, pregnancy, immunodeficiency, and use of broad-spectrum antibiotics and corticosteroids predispose patients to developing *Candida* vulvovaginitis, but the infection is most often seen in patients who lack any of these risk factors. Trichomoniasis is transmitted only by sexual contact. Up to one-third of patients with trichomoniasis have concurrent gonorrhea, but there is no increased rate of infection with *C. trachomatis*.

Trichomonal Vaginitis

Clinical Manifestations A small proportion of vaginally delivered female neonates acquire trichomonal vaginitis from their infected mothers. Infants harboring only a few trichomonads may never develop clinical disease, but the remainder will have a thin whitish or yellowish vaginal discharge that appears within 10 days after birth and may persist for several months if untreated. Infected babies may be irritable but are otherwise well.

The classic vaginal discharge of trichomonal vaginitis after puberty is pruritic, frothy, and yellowish. However, many infected women do not complain of excessive discharge, and the discharge may be scant or nondescript. The so-called strawberry cervix with multiple punctate areas of hemorrhage is pathognomonic for trichomoniasis but is visible without colposcopy in only about 2% of infected patients.

For patients of all ages, the diagnosis is made easily and rapidly if characteristically motile, flagellated trichomonads are seen in a saline suspension of discharge examined microscopically within about 15 minutes after the specimen has been obtained (Fig. 94.7). If a longer delay occurs, the organisms will gradually lose their mobility and normal shape, making them much more difficult to identify. The false-negative rate for wet mount examinations can be as high as 40%. More sensitive methods for detecting trichomonal infection are culture and colorimetric probes for trichomonal DNA.

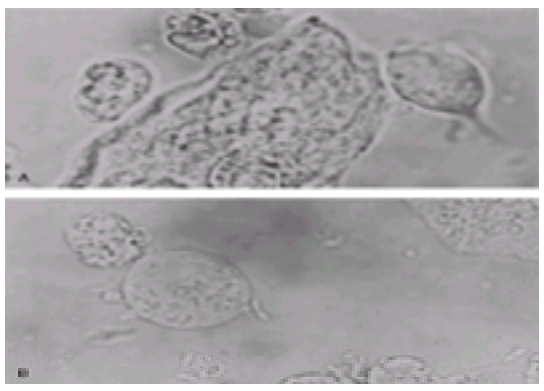

FIGURE 94.7. A. Trichomonad in the vaginal discharge of a 17-year-old patient with gonococcal pelvic inflammatory disease. The flagellated protozoan is elliptical and somewhat larger than the adjacent polymorphonuclear leukocytes (×225 magnification). **B.** After suspension in saline solution for microscopy, trichomonads gradually become swollen and immobile. This balloon-shaped trichomonad is barely recognizable (×225 magnification).

Management Metronidazole is effective for the treatment of vaginal trichomoniasis. The dosage for infants is 15 mg/kg per day orally in three divided doses for 7 days. A single oral dose (2 g) is prescribed for adolescents. Because trichomoniasis is sexually transmitted, the adolescent patient's partner(s) must also be referred for treatment.

Nausea and an unpleasant taste are common side effects of metronidazole. Alcohol should be avoided during treatment to prevent the occurrence of more severe abdominal pain, vomiting, flushing, and headache (disulfiram reaction). Recent data indicate that metronidazole is not a teratogen, but many clinicians prefer to postpone treatment of pregnant patients until the second trimester. Intravaginal clotrimazole (2 intravaginal tablets at bedtime for 7 days) can provide symptomatic relief for pregnant patients but will cure only 10 to 20%.

Shigella Vaginitis

Clinical Manifestations Shigella flexneri, S. sonnei, S. boydii, and *S. dysenteriae* can produce vaginal infections in infants and children but do not appear to cause genital disease after puberty. The vaginitis is characterized by a white to yellow discharge that is bloody in three-quarters of cases. Associated pruritus and dysuria are uncommon. One-third of patients have diarrhea that precedes, accompanies, or follows the vaginal discharge. On inspection, the vulvar mucosa is often inflamed or ulcerated. The diagnosis is established by culture of a specimen of vaginal discharge. Rectal cultures are positive in some cases.

Management Patients with *Shigella* vaginitis should be treated with oral antibiotics chosen on the basis of sensitivity testing. If the antibiotic sensitivity is unknown, trimethoprim–sulfamethoxazole (8 mg/kg per day orally of trimethoprim in two doses for 5 days) should be used.

Streptococcal Vaginitis

Clinical Manifestations S. pyogenes can be identified in cultures of vaginal specimens taken from about 14% of prepubertal girls with scarlet fever. Most of these vaginal infections produce either no symptoms or minor discomfort, but a few patients develop outright vaginitis with a purulent discharge. Streptococcal vaginitis can accompany or follow symptomatic pharyngitis and occurs uncommonly in girls with neither symptomatic pharyngitis nor scarlet fever. Most of these latter patients are pharyngeal carriers of the organism. Streptococcal vaginitis causes genital pain or pruritus and can mimic candidal or gonococcal vaginitis. A swab of the patient's discharge should be cultured to verify the clinical diagnosis, as well as to exclude gonococcal infection.

Management As for any other infection with group A b-hemolytic streptococci, penicillin is the preferred antibiotic. Intramuscular benzathine penicillin G is an alternative if poor compliance with oral treatment is anticipated. Oral erythromycin ethylsuccinate or azithromycin can be prescribed for children who are allergic to penicillin (see Table 94.2 for dosages).

Table 94.2. Treatment of Vaginal Infections

Candida Vulvovaginitis

Clinical Manifestations C. albicans frequently colonizes the vagina after the onset of puberty, when estrogen stimulates local increases in glycogen stores and acidity that both appear to enhance its growth. If the ecologic balance of the vagina is changed either by inhibition of the normal bacterial flora, by impaired host immunity, or by an increase in the availability of nutrients (broad-spectrum antibiotics, immunodeficiency states, corticosteroids, diabetes mellitus, pregnancy), the resulting proliferation of *Candida* will produce symptoms in a fraction of affected patients. However, most patients with candidiasis have no identifiable predisposing risk factor for infection. Because of the importance of estrogen in promoting fungal growth, candidal vulvovaginitis is rare among prepubertal girls.

The most common clinical manifestation of vulvovaginal candidiasis is vulvar pruritus. In severe infections, vulvar edema and erythema can occur. “External” dysuria is produced when urine comes in contact with the inflamed vulva. Vaginal discharge is variable in quantity and appearance. In severe cases, the vaginal vault is red, dry, and has a whitish, watery, or curdlike discharge that may be relatively scanty. Patients with mild disease may have only intermittent itching and an unimpressive discharge.

Microscopic examination of a sample of vaginal discharge suspended in 10% potassium hydroxide solution to clear the field of cellular debris can provide a rapid diagnosis of candidiasis if hyphae are seen (Fig. 94.8). However, in as many as 50% of cases, wet mounts are falsely negative. Gram-stained smears of discharge are somewhat more sensitive because hyphae and yeast cells are Gram positive and more easily visible. Culture can only corroborate or fail to corroborate the clinical impression of candidiasis because the vaginal flora includes *C. albicans* in up to 25% of young women who have no symptoms or signs of infection. Similarly, cultures from patients with classic signs of candidal infection may yield only a light growth of the organism, making heavy growth an inadequate criterion for diagnosis. From these considerations, it is apparent that although the presence of *C. albicans* can be confirmed by laboratory tests, the diagnosis and subsequent treatment of this infection should be guided by the presence or absence of clinical disease.

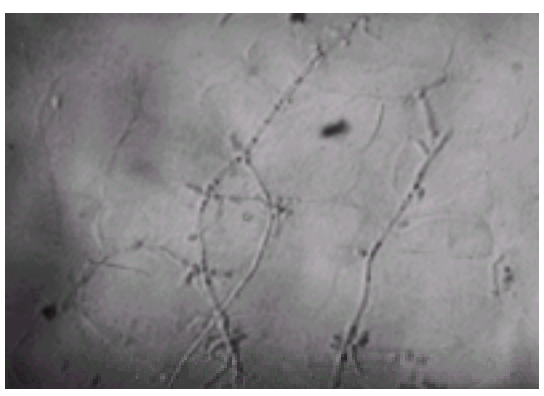

FIGURE 94.8. Branching hyphae of *Candida albicans* in vaginal discharge suspected in 10% potassium hydroxide. Ghosts of vaginal epithelial cells are also visible (×100 magnification).

Management Topical imidazoles will promptly cure 80 to 90% of patients with candidal infections. Most are available without prescription. The creams are packaged with intravaginal applicators, but many premenarchal and virginal girls can be treated adequately and more comfortably by applying cream to the vulva alone. Effective, nonprescription, short-course treatments for patients with mild to moderate candidal vulvovaginitis include butoconazole 2% cream (one full applicator at bedtime for 3 nights); clotrimazole 500-mg tablets (1 tablet intravaginally as a single dose); miconazole 200-mg suppositories (1 suppository at bedtime for 3 nights); and tioconazole 6.5% ointment (1 full applicator as a single dose). For patients with severe discomfort, one of the 5- or 7-day formulations of a topical agent is likely to be more effective. Fluconazole, an oral fungicide, treats candidal vulvovaginitis as effectively as the topical preparations, and many patients prefer oral to topical treatment. However, the potential for promoting fungal resistance and the risks, albeit low, of systemic toxicity and allergy are important disadvantages of oral antifungal agents.

Nonspecific Vaginitis in Children

Clinical Manifestations The term *nonspecific vaginitis,* referring to a disorder of prepubertal girls, encompasses a variety of genitourinary symptoms and signs that are sometimes caused by poor perineal hygiene but that in other cases have no readily identifiable cause. Genital discomfort, discharge, itchiness, and dysuria are relatively common childhood complaints. When a girl with such symptoms has either a normal vulva and vagina or only mild vulvar inflammation on physical examination, a specific vaginal infection is unlikely, and other possible explanations for the complaint—smegma, pinworms, urinary tract infection, a local chemical irritant, or sexual abuse, for example—should be sought with appropriate questions and laboratory tests. (It should be noted that commercially available bubble bath is not often the culprit.) If, on the other hand, a vaginal discharge *is* present on physical examination, the specific vaginal infections discussed in this chapter are diagnostic possibilities, and cultures should therefore be obtained. In reported series of premenarchal girls with vaginitis who have been systematically evaluated, between 25 and 75% are ultimately categorized as having nonspecific vaginitis. The diagnosis should not be made until other entities have been excluded. (A more comprehensive discussion of the differential diagnosis of genital complaints is presented in Chapter 76 and Chapter 77.)

Management General measures to promote cleanliness and comfort should be initiated for the girl with nonspecific vaginitis. Daily soaking in a bath of warm water, either plain or with some baking soda added, gentle perineal cleaning with a soft washcloth, and the use of cotton underwear can be recommended. The girl should be taught to wipe toilet paper anteroposteriorly after having a bowel movement. Using these suggestions, most girls with perineal irritation will be improved within 2 weeks. The remaining patients should be reevaluated to exclude any specific but previously unrecognized disorder. If none is found, these girls may benefit from a brief course of topical estrogen cream (a small amount dabbed onto the vulva nightly for 2 to 4 weeks) to stimulate thickening of the vaginal mucosa so that it is more resistant to local irritation. Parents should be cautioned that estrogen cream is capable of producing breast growth if it is used for a prolonged period of time.

Bacterial Vaginosis

Background Bacterial vaginosis is a syndrome characterized clinically by the presence of three of the following four signs: 1) a homogeneous, white adherent vaginal discharge; 2) vaginal pH above 4.5; 3) a fishy, aminelike odor released when 10% potassium hydroxide solution is added to a sample of the discharge; and 4) the presence of clue cells (Amsel criteria). The syndrome occurs when lactobacilli that normally predominate in the genital tract are displaced by an overgrowth of mixed flora, including *Gardnerella vaginalis, Mobiluncus* species, other anaerobes, and *Mycoplasma hominis.* What accounts for this change in the vaginal microflora is not understood. The high prevalence of the syndrome in sexually active women and in women attending STD clinics suggests that a wide range of epidemiologic and microbiologic factors may contribute to its pathogenesis. Bacterial vaginosis has been implicated as an important cause of endometritis and chorioamnionitis in women and of preterm delivery of low-birth-weight infants.

Clinical Manifestations The symptoms of bacterial vaginosis—malodor and discharge—are not distinctive and can resemble those of trichomonal infection. A complaint of dysuria or pruritus goes against the diagnosis. As many as half of women who have signs of vaginosis are asymptomatic. The vaginal discharge is moderate or copious, grayish-white, and homogeneous. On examination, the vulva, vagina, and cervix are not inflamed, but concomitant infection with trichomonas or gonorrhea can complicate this picture.

Compared with the composite Amsel criteria, use of single tests (e.g., pH, clue cells, or whiff test alone) produces lower positive and negative predictive values for the diagnosis of bacterial vaginosis. When a wet mount of vaginal discharge is examined, epithelial cells are seen to be studded with large numbers of small bacteria and have a granular appearance with shaggy borders (Fig. 94.9). The ratio of epithelial cells to polymorphonuclear leukocytes in the discharge is 1 or higher. Lactobacilli (long rods) are sparse. Gram stain can be used to confirm the presence of clue cells and the scarcity of long Gram-positive rods (lactobacilli). Because 35 to 55% of women without bacterial vaginosis have positive cultures for *G. vaginalis,* culture is not a useful diagnostic test. Trichomonal infection is the major diagnostic alternative for patients suspected of having bacterial vaginosis.

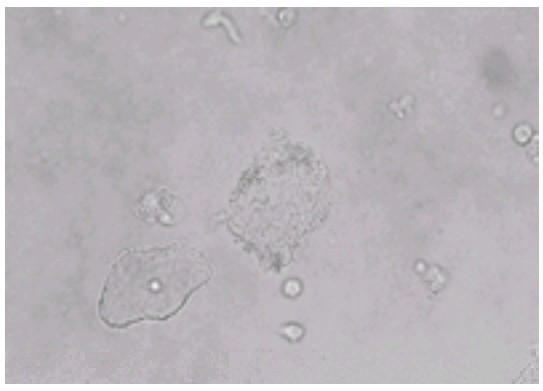

FIGURE 94.9. A clue cell. The vaginal epithelial cell on the right has shaggy borders obscured by coccobacilli (×100 magnification).

Management The standard treatment for bacterial vaginosis is oral metronidazole. Regimens are 500 mg twice daily for 7 days in nonpregnant women and 250 mg three times daily for 7 days in pregnant women. These regimens are moderately effective, yielding a recurrence rate of up to 30% within 3 months. Treatment of patients' sexual partners does not reduce the recurrence rate and is not recommended. Common side effects of metronidazole include GI upset, headache, and a metallic taste. A recent meta-analysis indicated that metronidazole in standard doses is not a human teratogen. However, some clinicians prefer to postpone treatment of pregnant women until the second trimester. Intravaginal clindamycin cream and metronidazole gel are alternative treatments for nonpregnant women but are not recommended during pregnancy. Oral clindamycin (300 mg twice a day for 7 days) is an alternative treatment regimen for pregnant patients with bacterial vaginosis.

Gonococcal Infections

Clinical Manifestations

Prepubertal girls with vaginal gonorrhea uniformly have an obvious whitish to greenish purulent discharge that can be pruritic. Because the child's vaginal flora is normally fairly sparse, a Gram-stained smear of the vaginal discharge can provide a rapid presumptive positive diagnosis if, on microscopic examination, at least eight pairs of typical gonococci can be seen in each of at least two polymorphonuclear leukocytes. However, because the social and legal implications of the diagnosis are major and because this fastidious microorganism can fail to grow in the laboratory, *all management decisions about gonorrhea in a child, including informing the parents of the diagnosis, reporting the case to the state child protective services agency, and instituting antibiotic treatment, should be delayed until after the suspected diagnosis has been confirmed by bacterial culture*.

After puberty, higher estrogen levels stimulate the growth of a thick vaginal mucosa that is relatively resistant to infection by gonococci, and lower genital tract infection is localized to the cervix and/or urethra. A vaginal discharge, excessive menstrual bleeding, or symptoms of cystitis may prompt an infected patient's ED visit, but most infections will produce no symptoms at all. On examination of the cervix, there may be marked central erythema, a purulent discharge, or no abnormality. A culture of a single specimen taken from the endocervical canal with a cotton-tipped swab, plated on a selective medium, and incubated properly will identify about 85% of women with uncomplicated gonorrhea. The remaining 15% includes patients with urethral, rectal, and pharyngeal infections without cervicitis, and those with cervicitis not detected with a single culture.

Management (Table 94.3)

Girls with culture-proven gonococcal vaginitis should be treated with a single intramuscular dose of ceftriaxone. Treatment for *C. trachomatis* should not be given presumptively for children, but rather should be withheld pending the results of screening chlamydial cultures. Pharyngeal and rectal swabs for culture of *N. gonorrhoeae* should be obtained from every child in whom gonococcal vaginitis is confirmed by culture. The finding of gonorrhea in a child mandates detailed psychosocial evaluation and a report to the state child protective services agency for suspected sexual abuse in every case. All household contacts of the child, both children and adults, should be cultured because epidemiologic studies indicate that about 25% of them are likely to have gonorrhea also. Hospitalization of the child may facilitate this investigation in some cases.

Adolescents with uncomplicated genital gonorrhea should receive one of the treatments listed in Table 94.3. The oral treatment regimens have the advantages of easy administration and avoiding both a painful intramuscular injection and the risk of accidental needle stick. Since 1982, the CDC have suggested that every adolescent patient treated for lower genital tract gonorrhea also be treated presumptively for coexisting chlamydial infection because of the substantial likelihood of dual infection.

Every patient with gonorrhea should receive a screening serologic test for syphilis and should be offered screening for HIV infection. None of the treatment regimens for gonorrhea will cure established syphilis, but ceftriaxone is likely to eradicate incubating syphilis. The management of a case of gonorrhea is never complete until the patient's sexual partner(s) has been notified and treated presumptively. Test-of-cure cultures are not indicated because the treatment regimens for gonorrhea are efficacious. However, follow-up remains desirable for adolescents with gonorrhea to review their contraceptive behavior, to counsel them about risk reduction, to identify reinfections, and to obtain Papanicolaou smears.

Chlamydial Infections

Background

Lower genital tract infection with the intracellular bacterium *C. trachomatis* can be acquired at any age by means of sexual contact or during infancy as a result of either direct inoculation from an infected mother during vaginal delivery or spread from the nasopharynx after vertical transmission. Vertically acquired infections may persist in children for a year or more. Chlamydiae infect squamocolumnar epithelial cells and thus can be found in specimens from the urethra, the rectum, the vagina in prepubertal girls, and the cervix after puberty. Infections during childhood are uncommon, usually asymptomatic, and rarely detected. Infections among adolescents and adults are also often asymptomatic but common, occurring in up to 15% of sexually active teenagers. Simultaneous infection with gonorrhea is common in both children and adolescents with lower genital tract chlamydial infection.

Clinical Manifestations

Most prepubertal children with urethral, vaginal, and rectal infections are asymptomatic, although a few complain of dysuria, enuresis, or vaginal discharge. The diagnosis should be suspected in prepubertal girls with vaginal discharge that persists after therapy for a prior gonococcal infection and in girls with urethritis or vaginitis who have had sexual contact. Culture of a urethral or vaginal sample is advisable to confirm the diagnosis in children because the rapid antigen-detection tests can yield false-positive results and because the social and legal implications of a positive test are serious.

In adolescents, *C. trachomatis* can cause dysuria-pyuria syndrome and mucopurulent cervicitis but is often asymptomatic. Untreated lower genital tract infection can result in bartholinitis, perihepatitis, PID, and infertility (see the following section). On speculum examination, the cervix may be erythematous and friable when swabbed. Infected secretions collected from the cervical os may appear yellowish on a cotton swab. The diagnosis in an adolescent should be confirmed by an antigen or DNA detection test of the cervix or urine. These tests have good sensitivity and specificity.

Management

Antibiotic treatment regimens for children and adolescents with lower genital tract chlamydial infections are summarized in Table 94.3. Children should be treated despite the absence of symptoms because complications of the infection may develop after puberty. Adolescents with chlamydial infections should be counseled about the importance of treatment for their sexual partner(s), abstinence until 1 week after all sexual partners have completed treatment, and barrier methods of contraception. Patients should receive follow-up examinations 4 to 6 weeks after treatment to screen for reinfection, coexisting STDs, and cervical dysplasia.

Bartholin Gland Abscess and Cyst

Background

Abscesses and symptomatic cysts of the Bartholin glands are relatively uncommon disorders during adolescence. The Bartholin glands lie at about the 4 and 8 o'clock positions in the vestibule and drain through ducts that open into folds between the hymen and the labia minora. Many Bartholin gland cysts are asymptomatic and need no treatment. The clinical distinction between a symptomatic cyst and an abscess can be arbitrary in some cases.

Clinical Manifestations

The patient with a Bartholin gland abscess presents with a painful, tender, fluctuant mass bulging on the involved side of the vestibule inferior to the labium minus. Pus can sometimes be milked upward from the gland to the duct orifice. Cultures of pus from abscessed glands yield *N. gonorrhoeae* in 10 to 50% of cases. Most of the remaining cases contain mixed growths of facultative, aerobic, and anaerobic organisms, often including *Bacteroides* species. Cultures from a small number of abscesses yield *C. trachomatis,* and in 15 to 30% of cases, the pus is sterile. The patient with a cyst complains of vulvar discomfort; the unilateral mass is typically 1 to 3 cm in diameter and mildly tender. Cyst fluid is usually sterile.

Management

Abscesses and symptomatic cysts are treated similarly, with either placement of a Word catheter, marsupialization, or a “window operation.” Each of these procedures opens the cyst cavity widely and facilitates prolonged drainage. Many experts recommend against simple incision and drainage because of the high recurrence rate associated with this procedure. All patients with abscesses or purulent exudate should be treated for presumed gonorrhea (as outlined in Table 94.3) and their sexual partners referred for concurrent treatment. Complications of Bartholin gland abscess after treatment include slow healing, persistent discomfort, dyspareunia, and recurrence.

Pelvic Inflammatory Disease

Background

Definition

PID is infectious inflammation of the upper genital tract, variably involving the endometrium, fallopian tubes, ovaries, adjacent structures, and the pelvic peritoneum. The following discussion concerns acute PID caused by sexually

transmitted microorganisms ascending from the cervix, and excludes infections associated with childbirth, spontaneous abortion, or pelvic surgery.

Epidemiology

Each year between 1979 and 1988, among American women between the ages of 15 and 44 years, acute PID accounted for more than 165,000 hospitalizations and 1.2 million physician office visits. In 1994, nearly 400,000 women were diagnosed in EDs as having PID. Adolescents account for approximately 16 to 20% of all PID cases in the United States and have the highest age-specific case rates of PID after adjustment is made for the prevalence of sexual activity.

Young age, a large number of sexual partners, and nonbarrier contraceptive methods are risk factors for infection with *N. gonorrhoeae* and *C. trachomatis,* the microorganisms responsible for initiating most cases of acute PID. Recent douching is a risk factor for the development of PID, but the mechanism by which it increases risk is unknown. Overall, reported gonorrhea in the United States has declined steadily since the 1980s. However, teenagers and young adults consistently have the highest age-specific rates of gonorrhea (in 1996, 757 cases per 100,000 women between 15 and 19 years and 523 cases per 100,000 men between 20 and 24 years). In 1994, reported cases of chlamydial infection exceeded reported cases of gonorrhea in the United States for the first time. In 1996, the rate of chlamydial infection for women ages 15 to 19 years was 2,069 per 100,000. In family planning clinic screening programs in 1996, the rate of cervical chlamydial infection in women 15 to 24 years ranged as high as 11%.

Compared with no contraceptive method, barrier methods—male and female condoms, diaphragms, and spermicides—decrease the overall risk of acquiring gonococcal and chlamydial cervicitis. Oral contraceptive pills decrease the likelihood that users with cervical gonorrhea will develop ascending infection. The effect of oral contraceptives on chlamydial disease is less uniform. The increased cervical ectropion produced by oral contraceptives increases the likelihood of chlamydial cervicitis, but the thickened cervical mucus and reduced menstrual flow appear to inhibit ascending infection. A principal reason for strongly discouraging adolescents from using the intrauterine device (IUD) as a contraceptive method is the twofold higher risk of PID associated with IUD use.

The sequelae of PID—chronic and recurrent pelvic pain, dyspareunia, tubo-ovarian abscesses, ectopic pregnancy, and involuntary infertility—affect large numbers of women. It has been estimated that 18% of women with PID will have chronic pelvic pain. The rate of ectopic pregnancy per 1000 reported pregnancies in the United States increased dramatically, from 4.5 in 1970 to 14.3 in 1986, and (based on outpatient as well as inpatient hospital data) to an estimated 19.7 in 1992. It is estimated that half of all ectopic pregnancies result from tubal damage produced by salpingitis. Infertility occurs in about 15% of women who have had a single episode of gonococcal or nongonococcal salpingitis. Repeated bouts of PID substantially increase the likelihood of infertility.

Etiology

Nearly all first episodes of PID in adolescents are the result of gonococcal or chlamydial infection, or both, ascending from the cervix. If lower genital tract infection is not treated, approximately 10 to 40% of women with gonococcal cervicitis and 20 to 40% of women with chlamydial cervicitis will eventually develop PID. *C. trachomatis* is estimated to be responsible for 20 to 50% of all PID cases in the United States.

Subsequent episodes of PID may be produced either by repeated gonococcal or chlamydial lower genital tract infections or by endogenous genital or respiratory flora (most notably *Peptostreptococcus, Prevotella,* and *Bacteroides* species and *Hemophilus influenzae*). Tubal damage caused by concurrent or previous gonococcal or chlamydial infection probably contributes to secondary, polymicrobial PID involving endogenous microorganisms. *M. hominis* produces parametritis and salpingitis experimentally in grivet monkeys and has been isolated from the upper genital tracts of women with PID, but its role in the etiology of PID remains uncertain. Tuberculous PID is fortunately rare in the United States.

Pathophysiology

C. trachomatis was first isolated from the fallopian tubes of patients with acute salpingitis in 1976. The development of salpingitis in grivet monkeys whose cervices were inoculated with *C. trachomatis* confirmed the pathogenesis of this obligate intracellular bacterium. Both chlamydiae and gonococci infect columnar epithelial cells and reach the fallopian tubes by ascending from the cervix along the endometrial surface of the uterus. Menstrual blood facilitates this extension of the infection. In acute chlamydial infection, a polymorphonucleocytic infiltrate occurs, but during repeated or persistent infection, plasma cell endometritis is a prominent feature. An autoimmune response to repeated infection, involving the 60-kD heat shock chlamydial protein, seems likely to account for the severe tubal and peritubal scarring that is particularly associated with chlamydial PID.

Gonococci attach themselves to mucosal secretory cells and invade the submucosal connective tissue, stimulating a marked inflammatory reaction. In the tubal lumen, gonococcal lipooligosaccharides stimulate the production of tumor necrosis factor, endotoxin is released, and IgM–lipopolysaccharide immune complexes activate complement. In either chlamydial or gonococcal infection, pus may spill out from the fimbriated ends of the tubes, producing peritoneal symptoms and signs.

Clinical Manifestations

Although the constellation of symptoms and signs associated with PID—abdominal pain, irregular uterine bleeding, abnormal vaginal discharge, and lower abdominal and pelvic tenderness—is well known, no single symptom or sign or combination of symptoms and signs is both sensitive and specific. Clinical findings that improve the specificity of the diagnosis of PID (i.e., increase the likelihood that the diagnosis is correct) do so only at the expense of sensitivity (i.e., exclude patients who do in fact have PID). Minimum, additional, and definitive criteria for the diagnosis of PID suggested by the CDC are shown in Table 94.4. However, it is noteworthy that approximately one-fourth of women who have pelvic

pain but who do not meet the CDC's minimum criteria have been shown nevertheless to have PID on laparoscopy or endometrial biopsy.

Minimum criteria	Lower abdominal tenderness
	Tenderness produced by motion of the cervix
	Bilateral adnexal tenderness
Additional criteria	Oral temperature >38°C (100.4°F)
	Abnormal vaginal discharge on examination
	Erythrocyte sedimentation rate >15 mm/hr
	Elevated C-reactive protein
	Documented gonococcal or chlamydial cervical infection
Definitive criteria	Fallopian tube visible or fluid-filled on ultrasonography
	Tubo-ovarian abscess on ultrasonography
	Laparoscopic abnormalities consistent with pelvic inflammatory disease
	Endometritis on endometrial biopsy

Adapted from Centers for Disease Control and Prevention. 1998 Guidelines for treatment of sexually transmitted diseases. MMWR 1998;47(RR-1):80; and Case definitions for infectious conditions under public health surveillance. MMWR 1997;46(RR-10):52.

Table 94.4. Minimum, Additional, and Definitive Criteria for the Diagnosis of Pelvic Inflammatory Disease

In patients with suspicious symptoms and signs, a fever higher than 38° C (100.4°F) is about 80% specific for PID but will incorrectly exclude the diagnosis in about 60 to 75% of patients who do have it (25 to 40% sensitivity). A peripheral white blood cell count of more than 10,000/mm^3 is approximately 90% specific but will incorrectly exclude the diagnosis of PID in about 40% of patients with PID (60% sensitivity). A C-reactive protein greater than 5 mg/dL and an erythrocyte sedimentation rate greater than 15 mm/hour are each approximately 50 to 80% specific for PID but will each incorrectly exclude the diagnosis in about 10 to 30% of patients who have it. Perihepatitis (Fitz-Hugh–Curtis syndrome), consisting of right upper quadrant pain and tenderness produced by inflammation of the liver capsule in association with PID, occurs in 5 to 10% of patients with either chlamydial or gonococcal PID. On transvaginal ultrasonography, about one-third of patients with PID will have visible fallopian tubes, and about one-fifth will have a demonstrable tubo-ovarian abscess (Fig. 94.10).

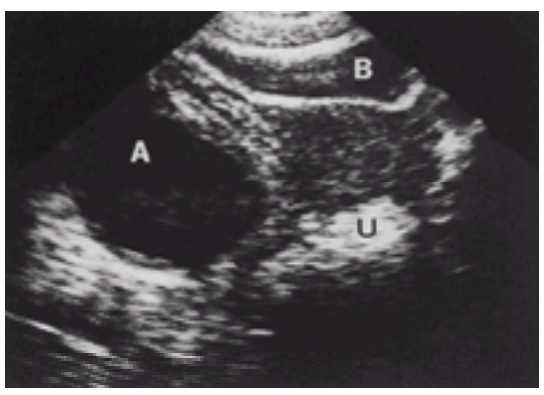

FIGURE 94.10. Transverse real-time sonogram of the pelvis of a 15-year-old patient with gonococcal salpingitis, demonstrating a 6 × 8 cm right tubo-ovarian abscess *(A)* containing a fluid-debris level. *U,* uterus; *B,* bladder.

A complication of PID that warrants prompt diagnosis is ruptured tubo-ovarian abscess. About 15% of tubo-ovarian abscesses rupture spontaneously. The symptoms and signs of a ruptured abscess may be mild if only a small amount of pus has leaked out, but the usual clinical picture includes peritonitis and shock. A pelvic mass is palpable in fewer than one-half the cases. Prompt surgical intervention can be lifesaving.

Laparoscopy confirms the diagnosis of PID in only about 60% of all patients who are suspected, either by gynecologists or by primary care physicians, on clinical grounds of having the disease. Conditions most often mistaken for PID are acute appendicitis, endometriosis, hemorrhagic and nonhemorrhagic ovarian cysts, and ectopic pregnancy. In up to 25% of women judged clinically to have PID, no abnormality can be identified laparoscopically.

The emergency physician must consider the possibility of pregnancy in adolescents with presumed PID for two reasons. First, ascending genital tract infection is rare during pregnancy. As a result, alternative diagnoses to PID, including ectopic pregnancy, should be considered, and hospitalization of the patient is recommended (see the following sections). Second, treatment with fluoroquinolones should be avoided during the first trimester of pregnancy. (Pregnant patients may receive metronidazole for PID because, in a recent meta-analysis, first-trimester exposure was not associated with an increase in the rate of major congenital malformations among live-born infants.)

An important pathophysiologic irony is the observation that tubal occlusion is associated more often with a relatively unimpressive clinical presentation of PID (i.e., long duration of symptoms, no signs of peritonitis, normal peripheral leukocyte count) than with “hot” clinical disease (i.e., short duration of symptoms, fever, peritoneal signs, leukocytosis). Similarly, chlamydial PID is associated with both a longer duration of pain at patient presentation and a higher risk of infertility than is gonococcal PID. Thus, if the diagnosis of PID is allowed to depend substantially on patients' appearance—as either “well” or “sick”—clinicians may be tempted to reject the diagnosis of PID and to withhold antibiotic treatment from those patients at highest risk of subsequent ectopic pregnancy and infertility.

The Kahn approach to the clinical diagnosis of PID is recommended for the emergency physician (Fig 94.11). This

strategy emphasizes diagnostic sensitivity for women with relatively mild illness, encouraging clinicians to err on the side of providing rather than withholding antibiotic treatment, and diagnostic specificity for women with relatively severe illness, focusing on the consideration of major competing diagnoses.

FIGURE 94.11. Strategy for diagnosis of pelvic inflammatory disease. Minimal laboratory evaluation should include tests for gonococcal and chlamydial cervicitis. Expanded laboratory investigation may include, in addition to the minimal evaluation, complete blood count, C-reactive protein or erythrocyte sedimentation rate, and pelvic or transvaginal ultrasonography. (Adapted from Kahn JG, Walker CK, Washington E, et al. Diagnosing pelvic inflammatory disease. JAMA 1991;266:2594–2604.)

Management

The CDC guidelines for the treatment of PID (1998) are summarized in Table 94.5. The antibiotics listed were selected for their effectiveness in combination against *N. gonorrhoeae, C. trachomatis,* and the aerobes and anaerobes responsible for polymicrobial PID. Principal differences between the 1998 treatment guidelines and previous versions of these guidelines are 1) ultrasonographic findings of fallopian tube fluid and thickening are included as definitive criteria for the diagnosis of PID; 2) the previous recommendation that all adolescents with PID be hospitalized is eliminated; 3) 24 hours, rather than 48 hours, of parenteral treatment is recommended after clinical improvement; 4) clindamycin is omitted as an alternative to metronidazole in the oral ofloxacin/metronidazole treatment regimen; and 5) repeat evaluation for *N. gonorrhoeae* and *C. trachomatis* 4 to 6 weeks after the completion of therapy is presented as optional.

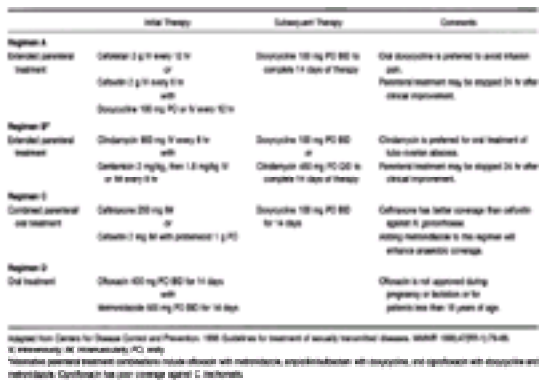

Table 94.5. Treatment Regimens for Pelvic Inflammatory Disease

Hospitalization is recommended for any patient with PID whose diagnosis is uncertain, particularly if ectopic pregnancy or appendicitis seems likely, for patients with severe clinical illness, including those with fever or suspected pelvic abscess, and for patients who are either immunodeficient or pregnant. Parenteral treatment, on either an outpatient or an inpatient basis, is recommended for patients likely to fail a course of oral antibiotics because of either poor compliance or vomiting and for those whose illnesses have not responded to prior oral antibiotics.

The follow-up of outpatients should include a return visit after about 3 days of treatment. The average duration of symptoms among women with gonococcal salpingitis treated with oral antibiotics is 3 to 4 days; the corresponding interval for nongonococcal salpingitis is 4 to 6 days. A poor response to therapy should alert the physician to the possibilities of inadequate compliance, abscess formation, or an alternative diagnosis.

Follow-up for all patients should include reexamination at the end of antibiotic therapy to check for residual pelvic tenderness and adnexal masses. To identify patients with persistent or repeated infection resulting from noncompliance with antibiotics or an untreated sexual partner, follow-up tests for gonococcal and/or chlamydial cervicitis should be scheduled 3 to 4 weeks after the end of treatment. The importance of identifying and treating sexual partners of women with PID cannot be overemphasized. About 25% of such men have asymptomatic urethritis and are unlikely to seek treatment on their own. If contact tracing fails, these men become part of the reservoir of undetected carriers of STDs. All patients with gonococcal and chlamydial infections should be counseled about the HIV and offered serologic screening for both syphilis and HIV infection.

Genital Warts

Background

Genital warts, or *condylomata acuminata,* are multicentric, exophytic tumors on anogenital skin (Fig. 94.12) caused by the DNA-containing human papillomavirus (HPV), most commonly by HPV types 6 and 11. Although HPV infections are not reportable, many venereologists believe they are the most common STDs in the United States. National Disease and Therapeutic Index survey data show that consultations with private physicians for genital warts numbered approximately 60,000 visits in 1966, peaked at nearly 360,000 visits in 1987, and totaled 253,000 visits in 1995. The HPV types that produce most anogenital warts are considered to have low potential for malignant change. HPV infection of the uterine cervix, which is associated with cervical dysplasia and cancer, occurs at an annual incidence of 9 to 20% in college-age women.

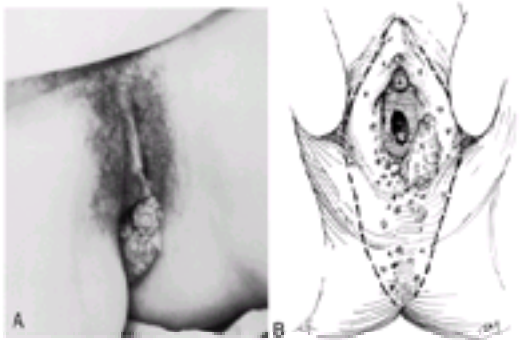

FIGURE 94.12. A. Large vulvar and perianal genital warts in a 15-year-old patient. **B.** In females, genital warts are most commonly located along the posterior margin of the introitus, inside the vagina, and on the labia minora.

Although HPV type 2 (associated with cutaneous, common warts) has been identified in anogenital warts in children, suggesting autoinoculation or heteroinoculation, nearly all genital warts in adolescents and adults, and some in children, are transmitted from person to person by sexual contact. The time from contact with an infected partner to the appearance of genital warts is estimated to be 1 to 3 months. However, the concept of an incubation period does not apply readily to the large number of infections that remain subclinical for long periods.

Pathophysiology

Grossly, genital warts are hyperplastic lesions that occur on squamous epithelium. On microscopic examination, the stratum granulosum contains foci of vacuolated cells. Acanthosis, parakeratosis, and hyperkeratosis are characteristic findings. The presence on Papanicolaou smear of koilocytes—intermediate, often multinucleated squamous cells with perinuclear halos, pyknotic nuclei, and dense peripheral cytoplasm—indicates cervical HPV infection.

Clinical Manifestations

Most patients with genital warts either have no complaint or report noticing “bumps” in the genital area. Uncommonly, large perianal warts can be painful and interfere with defecation. Prepubertal girls with vulvovaginal warts may have a bloody vaginal discharge. Because HPV infection in males is so consistently asymptomatic, most female patients are not aware of their exposure to an infected partner.

Warts can occur anywhere on the perineum, but their growth seems to be encouraged by moisture. The most common locations in females are the posterior fourchette, adjacent areas of the labia minora and majora, and the lower vagina (Fig. 94.12). Single warts 1 cm or more in diameter and clusters of seedlings, each a few millimeters across, are both common. Warts can be velvety and flat or papillomatous. Large warts often contain distinct cauliflowerlike lobulations. On the cervix, acetowhite infected areas are usually seen only with colposcopy. Most visible lesions in men involve the penile frenulum or coronal sulcus. Immunodeficient patients are particularly susceptible to extensive or severe disease with an increased risk of malignant change. Genital warts must be differentiated from *condylomata lata,* a contagious manifestation of secondary syphilis (Fig. 94.13), *molluscum contagiosum,* and, in men, from pearly penile papules.

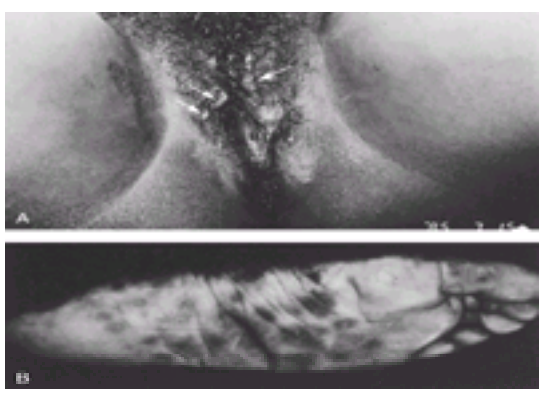

FIGURE 94.13. A. Vulvar condylomata lata of secondary syphilis in a 15-year-old patient. **B.** Macular rash on sole of foot of same patient.

Management

The diagnosis of genital warts is easy to make in patients with obvious lesions but can be more difficult in patients with flat warts or “microwarts” of the vulva. A magnifying lens or colposcope can be used to inspect suspicious areas that have been soaked in 5% acetic acid for 5 minutes. Infected skin, areas of nonspecific inflammation, and skin treated with podophyllin will turn white after soaking. To exclude syphilis, patients should also receive serologic screening.

Although anogenital warts in young children can result from vertical transmission or nonsexual contact with common warts, sexual abuse is another possible source of infection. The management of a child with genital warts should include either consultation with an expert in child abuse and neglect or a report to the state child protective service agency (see Chapter 128). Parents of children with genital warts should be examined for both common and genital warts. Excisional biopsy is preferred to ablative treatment for warts in children because histologic examination of the biopsied tissue can confirm the clinical diagnosis. Viral typing of anogenital warts in children may suggest abuse if a condylomatous HPV type is identified but cannot exclude sexual contact if a cutaneous type is found because fondling is a common manifestation of child sexual abuse.

The goal of treatment is the removal of bothersome tissue. Eradicating visible lesions does not end the viral infection. Whether it reduces contagiousness is uncertain. Spontaneous improvement or resolution of genital warts occurs in a minority of patients. Each of the several available treatment methods has advantages and disadvantages, none is more than about 50 to 60% effective, and recurrences are common. Podophyllin resin and trichloroacetic acid (TCA) are destructive agents that are applied topically. Podophyllin resin, in solutions of up to 25% concentration in tincture of benzoin, and TCA 80% solution are applied by the clinician. Podofilox in a 0.5% gel or solution and 5% imiquimod cream, an immune response modifier that induces cytokines, are available for self-application by patients.

Systemic absorption of podophyllin can produce bone marrow suppression, peripheral neuropathy, coma, and death. It is a teratogen and has been associated with stillbirths. Podophyllin should not be used during pregnancy or on warts that are large, bleeding, or located on mucosal surfaces. Safe maximum doses of 10% podophyllin solution are 4 mL for patients weighing more than 40 kg, and 0.1 mg/kg for children. TCA can be applied to mucosal surfaces and can be administered to pregnant patients. However, TCA has a viscosity lower than water and can spread to unaffected skin rapidly, producing patient discomfort during application more often than does podophyllin. Imiquimod commonly produces local itching, erythema, and burning but is generally tolerated well by patients.

For extensive or recurrent disease or when repeated applications of podophyllin, TCA, or imiquimod are not successful, alternative treatments include surgical removal, cryotherapy, and intralesional interferon. All women with genital warts should be referred to a primary care clinician for gynecologic care, including Papanicolaou screening.

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[1] All 50 states have acknowledged the confidential nature of communications between an adult and a physician. This right of confidentiality is extended to minors seeking treatment for certain sex-related problems. All states allow adolescents to receive outpatient treatment for STDs without parental consent or notification (limitation: if 14 years or older in Hawaii, Idaho, New Hampshire, North Dakota, and Washington). In 1977, the Supreme Court ruled that minors have a right to contraceptive privacy (Carey *v.* Population Services International). This ruling has been extended both by federal agencies that provide contraceptive services and by many states to permit minors to obtain prescription contraceptives without parental consent or notification (exceptions: if 14 years or older in Hawaii, if patient has ever been pregnant in Oklahoma). Many states require minors to obtain parental consent or approval from a judge to receive an abortion. Emergency physicians should familiarize themselves with the relevant current laws of their own states.

Suggested Readings

GENERAL

Centers for Disease Control and Prevention. 1998 guidelines for the treatment of sexually transmitted diseases. MMWR 1998;47(RR-1).

Emans SJH, Goldstein DP. Pediatric and Adolescent Gynecology. 4th ed. Boston: Little, Brown, 1998.

Gans JE, ed. Confidential Health Services for Adolescents. Chicago: American Medical Association, 1993.

Hatcher RA, Trussell J, Stewart F, et al. Contraceptive Technology. 17th ed. New York: Irvington, 1998.

Holmes KK, Mårdh PA, Sparling PF, Wiesner PJ, eds. Sexually Transmitted Diseases. New York: McGraw-Hill, 1990.

Neinstein LS. Adolescent Health Care: A Practical Guide. 3rd ed. Baltimore: Williams & Wilkins, 1996.

CONGENITAL VAGINAL OBSTRUCTION

Dewhurst J. Malformations of the genital tract. In: Practical Pediatric and Adolescent Gynecology. New York: Marcel Dekker, 1980:51–82.

Kahn R, Duncan B, Bowes W. Spontaneous opening of congenital imperforate hymen. J Pediatr 1975;87:768–770.

Spence JEH. Vaginal and uterine anomalies in the pediatric and adolescent patient. J Pediatr Adolesc Gynecol 1998;11:3–11.

Wenof M, Reyniak V, Novendstern J, Castadot MJ. Transverse vaginal septum. Obstet Gynecol 1979;54:60–64.

LABIAL ADHESIONS

Aribarg A. Topical oestrogen therapy for labial adhesions in children. Br J Obstet Gynecol 1975;82:424–425.

Capraro VJ, Greenberg H. Adhesions of the labia minora: a study of 50 patients. Obstet Gynecol 1972;39:65–69.

Soifer H. Adhesions of the labia minora in infants and children. Int Pediatr 1991;6:347–353.

URETHRAL PROLAPSE

Anveden-Hertzberg L, Gauderer MWL, Elder JS. Urethral prolapse: an often misdiagnosed cause of urogenital bleeding in girls. Pediatr Emerg Care 1995;11:212–214.

Lowe FC, Hill GS, Jeffs RD, et al. Urethral prolapse in children: insights into etiology and management. J Urol 1986;135:100–103.

Mercer LJ, Mueller CM, Hajj SN. Medical treatment of urethral prolapse in the premenarcheal female. Adolesc Pediatr Gynecol 1988;1:181–184.

DYSMENORRHEA

Klein JR, Litt IF. Epidemiology of adolescent dysmenorrhea. Pediatrics 1981;68:661–664.

Lennane KJ. Social and medical attitudes toward dysmenorrhea. J Reprod Med 1980;25(Suppl):202–206.

Owen PR. Prostaglandin synthetase inhibitors in the treatment of primary dysmenorrhea. Am J Obstet Gynecol 1984;148:96–103.

DYSFUNCTIONAL UTERINE BLEEDING

Bayer SR, DeCherney AH. Clinical manifestations and treatment of dysfunctional uterine bleeding. JAMA 1993;269:1823–1828.

Claessens EA, Cowell CA. Acute adolescent menorrhagia. Am J Obstet Gynecol 1981;139:277–280.

Falcone T, Desjardins C, Bourque J, et al. Dysfunctional uterine bleeding in adolescents. J Reprod Med 1992;39:761–764.

Smith YR, Quint EH, Hertzberg BS. Menorrhagia in adolescents requiring hospitalization. J Pediatr Adolesc Gynecol 1998;11:13–15.

Van Look PFA, Hunter WM, Fraser IS, et al. Impaired estrogen-induced luteinizing hormone release in young women with anovulatory dysfunctional uterine bleeding. J Clin Endocrinol Metab 1978;46:816–822.

SEXUAL ABUSE AND ASSAULT

Atabaki S, Paradise JE. The medical evaluation of the sexually abused child: lessons from a decade of research. Pediatrics 1999;104(supplement): 178–186.

Dowd MD, Fitzmaurice L, Knapp JF, et al. The interpretation of urogenital findings in children with straddle injuries. J Pediatr Surg 1994;29:7–10.

Emergency Contraception Web Site (Office for Population Research. http://opr.princeton.edu/ec/) and Hotline (1-800-NOT-2-LATE).

Glaser JB, Schachter J, Benes S, et al. Sexually transmitted diseases in postpubertal female rape victims. J Infect Dis 1991;164:726–730.

Glasier A. Emergency postcoital contraception. N Engl J Med 1997;337: 1058–1064.

Neinstein LS, Goldenring J, Carpenter S. Nonsexual transmission of sexually transmitted diseases: An infrequent occurrence. Pediatrics 1984; 74:67–76.

Paradise JE, Finkel MA, Beiser AS, et al. Assessments of girls' genital findings and the likelihood of sexual abuse. Arch Pediatr Adolesc Med 1997;151:883–891.

Siegel RM, Schubert CJ, Myers PA, et al. The prevalence of sexually transmitted diseases in children and adolescents evaluated for sexual abuse in Cincinnati: rationale for limited STD testing in prepubertal girls. Pediatrics 1995;96:1090–1094.

VAGINITIS

Al-Salihi FL, Curran JP, Wang JS. Neonatal *Trichomonas vaginalis:* report of three cases and review of the literature. Pediatrics 1974;53:196–200.

Boisvert PL, Walcher DN. Hemolytic streptococcal vaginitis in children. Pediatrics 1948;2:24–29.

Burtin P, Taddio A, Ariburnu O, et al. Safety of metronidazole in pregnancy: a meta-analysis. Obstet Gynecol 1995;172:525–529.

DeMeo LR, Draper DL, McGregor JA, et al. Evaluation of a deoxyribonucleic acid probe for the detection of *Trichomonas vaginalis* in vaginal secretions. Am J Obstet Gynecol 1996;174:1339–1342.

Gryngarten MG, Turco ML, Escobar ME, et al. *Shigella* vulvovaginitis in prepubertal girls. Adolesc Pediatr Gynecol 1994;7:86–89.

Hay PE, Taylor-Robinson D. Defining bacterial vaginosis: to BV or not to BV, that is the question. Internat J STD AIDS 1996;7:233–235.

Paradise JE, Campos JM, Friedman HM, Frishmuth G. Vulvovaginitis in premenarchial girls: clinical features and diagnostic evaluation. Pediatrics 1982;70:193–198.

Schwebke JR, Hillier SL, Sobel JD, et al. Validity of the vaginal gram stain for the diagnosis of bacterial vaginosis. Obstet Gynecol 1996;88:573–576.

Sobel JD. Controversial aspects in the management of vulvovaginal candidiasis. J Am Acad Dermatol 1994;31:S10–13.

Sobel JD. Vaginitis. N Engl J Med 1997;337:1896–1903.

Wolner-Hanssen P, Krieger JN, Stevens CE, et al. Clinical manifestations of vaginal trichomoniasis. JAMA 1989;261:571–576.

GONOCOCCAL INFECTIONS

American Academy of Pediatrics Committee on Child Abuse and Neglect. Gonorrhea in prepubertal children. Pediatrics 1998;101:134–135.

Farrell MK, Billmire E, Shamroy JA, et al. Prepubertal gonorrhea: a multidisciplinary approach. Pediatrics 1981;67:151–153.

Friedland LR, Kulick RM, Biro FM, et al. Cost-effectiveness decision analysis of intramuscular ceftriaxone versus oral cefixime in adolescents

with gonococcal cervicitis. Ann Emerg Med 1996;27:299–304.

Moran JS, Levine WC. Drugs of choice for the treatment of uncomplicated gonococcal infections. Clin Infect Dis 1995;20(Suppl):S47–65.

CHLAMYDIAL INFECTIONS

Beck-Sague CM, Farshy CE, Jackson TK, et al. Detection of *Chlamydia trachomatis* cervical infection by urine tests among adolescent clinics. J Adol Health 1998;22:197–204.

Bell TA, Stamm WE, Kuo CC, et al. Delayed appearance of *Chlamydia trachomatis* infections acquired at birth. Pediatr Infect Dis J 1987; 6:928–931.

Hammerschlag MR, Rettig PJ, Shields ME. False positive results with the use of chlamydial antigen detection tests in the evaluation of suspected sexual abuse in children. Pediatr Infect Dis J 1988;7:11–14.

Scholes D, Stergachis A, Heidrich FE, et al. Prevention of pelvic inflammatory disease by screening for cervical chlamydial infection. N Engl J Med 1996;334:1362–1366.

BARTHOLIN GLAND ABSCESS AND CYST

Anderson PG, Christenson S, Detlefsen GU, et al. Treatment of Bartholin's abscess: marsupialization versus incision, curettage and suture under antibiotic cover—a randomized study with 6 months' followup. Acta Obstet Gynecol Scand 1992;71:59–62.

Bleker OP, Smalbraak DJ, Schutte MF. Bartholin's abscess: the role of *Chlamydia trachomatis.* Genitourin Med 1990;66:24–25.

Downs MC, Randall HW. The ambulatory surgical management of Bartholin duct cysts. J Emerg Med 1989;7:623–626.

Hill DA, Lense JJ. Office management of Bartholin gland cysts and abscesses. Am Fam Physician 1998;57:1611–1616.

PELVIC INFLAMMATORY DISEASE

Division of STD Prevention: Sexually transmitted disease surveillance, 1996. US Department of Health and Human Services, Public Health Service. Atlanta: Centers for Disease Control and Prevention, September 1997.

Eschenbach DA, Buchanan TM, Pollock HM, et al. Polymicrobial etiology of acute pelvic inflammatory disease. N Engl J Med 1975;293:166–171.

Jacobson L, Weström L. Objectivized diagnosis of acute pelvic inflammatory disease. Am J Obstet Gynecol 1969;105:1088–1098.

Kahn JG, Walker CK, Washington E, et al. Diagnosing pelvic inflammatory disease. JAMA 1991;266:2594–2604.

Laufer MR, Goitein L, Bush M, et al. Prevalence of endometriosis in adolescent girls with chronic pelvic pain not responding to conventional therapy. J Pediatr Adolesc Gynecol 1997;10:199–202.

Peipert JF, Boardman L, Hogan JW, et al. Laboratory evaluation of acute upper genital tract infection. Obstet Gynecol 1996;87:730–736.

Soper DE, Brockwell NJ, Dalton HP, et al. Observations concerning the microbial etiology of acute salpingitis. Am J Obstet Gynecol 1994;170:1008–1017.

Walker CK, Kahn JG, Washington AE, et al. Pelvic inflammatory disease: meta-analysis of antimicrobial regimen efficacy. J Infect Dis 1993;168:969–978.

GENITAL WARTS

Edwards L, Ferenczy A, Eron L, et al. Self-administered topical 5% imiquimod cream for external anogenital warts. Arch Dermatol 1998;134:25–30.

Handley J, Hanks E, Armstrong K, et al. Common association of HPV 2 with anogenital warts in prepubertal children. Pediatr Dermatol 1997;14:339–343.

Ho GYF, Bierman R, Beardsley L, et al. Natural history of cervicovaginal papillomavirus infection in young women. N Engl J Med 1998;338: 423–428.

Stone KM, Becker TM, Hadgu A, et al. Treatment of external genital warts: a randomised clinical trial comparing podophyllin, cryotherapy, and electrodesiccation. Genitourin Med 1990;66:16–19.

CHAPTER 95

Pulmonary Emergencies

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ACUTE RESPIRATORY FAILURE

Background

Respiratory disease accounts for almost 10% of pediatric emergency department (ED) visits. Of all pediatric hospital admissions, approximately 20% result from respiratory illness in children. Respiratory illnesses continue to be a major cause of mortality (Table 95.1). Fortunately, newborn mortality from respiratory illness secondary to prematurity, which was as high as 10,000 deaths per year in the 1980s, has decreased in the last decade. New illnesses such as human immunodeficiency virus (HIV) have accounted for a small but significant increase in mortality as shown in Table 95.1.

Age (yr)	<1	1–4	5–9	10–14	15–19	All
SIDS	5746	5399	410	1	6	7562
RDS/other	3310	47	5	0	1	3363
Congenital	984	26	5	5	0	1016
Asthma	13	21	35	95	119	164
Pulmonary infection*	538	171	73	52	84	918
HIV	8	22	19	7	14	70
Others*	208	78	19	26	26	357
Suffocation*	376	162	76	59	67	742
Respiratory failure	15	14	7	3	10	49

From Centers for Disease Control and Prevention, US Health Statistics 1995—ICD9 code by age. (1995 Mortality Tables on Web Site).
SIDS, sudden infant death syndrome; *RDS*, respiratory distress syndrome; *HIV*, human immunodeficiency virus.
*Pulmonary infection includes viral pneumonia, bronchiolitis, influenza, pneumococcal infection, others, and nonspecified pulmonary infection.
†Other includes assorted alveolar disease, pneumothorax, and unspecified pulmonary causes of death.
‡Suffocation includes choking on foreign body or food and mechanical suffocation.

Table 95.1. Deaths from Pediatric Respiratory Disorders, 1995

Many different diseases may lead to respiratory failure (Table 95.2), including disorders outside the respiratory tract. This section discusses the pathophysiology and clinical manifestations of respiratory failure as well as a general approach to treatment. Management of specific disorders can be found in subsequent sections of this chapter as well as in Chapter 84, Chapter 92, and Chapter 119, and other areas of the text.

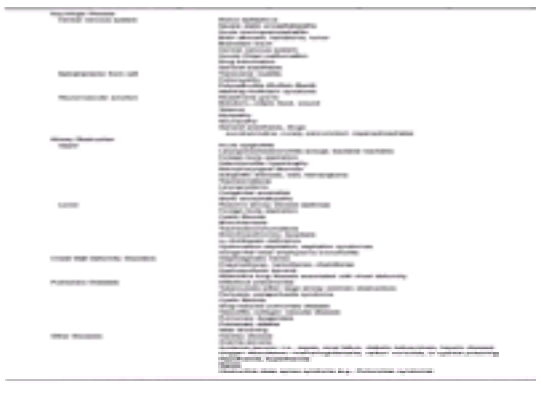

Table 95.2. Causes of Acute Respiratory Failure in Children

Pathophysiology

Acute respiratory failure may occur through a variety of mechanisms. By definition, failure indicates an inability of the respiratory system to provide sufficient oxygen for metabolic needs or to excrete the CO_2 produced by the body. Table 95.2 describes the causes of acute respiratory failure by anatomic location. The most common causes of respiratory failure are related to premature birth, with acquired pneumonitides second.

The neurologic system plays a major role in the control and maintenance of respiration. Children may suffer from either reversible or irreversible causes of central nervous system (CNS) respiratory failure. CNS disease may depress either the respiratory drive or the protective airway reflexes. Alternatively, neurologic disease may directly affect the peripheral nerves or muscles, impairing normal gas exchange through obstruction, fatigue, or ventilation-perfusion mismatch.

Most causes of upper airway obstruction in children are reversible but may lead to respiratory failure if untreated. The treatment approach should be both diagnostic and therapeutic because relief may be simultaneous in many instances (e.g., epiglottitis or aspirated foreign bodies). High-risk patients, such as those with severe static encephalopathy or anatomic head and neck problems but who usually maintain a patent airway when well, may have a partially reversible obstruction during respiratory infection, a seizure, or other acute medical problems.

Lower airway disease is a common cause of acute respiratory failure. Asthma accounts for the largest percentage of this group, but infections such as bronchiolitis or viral pneumonia are also common. Foreign body aspiration can involve the lower airway and continues to be a continued significant problem.

Chest wall deformities and mechanical impairments often play a role in respiratory failure. These entities act by diminishing the vital capacity in a restrictive pattern. The extra energy expended by the inefficient respiratory pattern may lead to both hypoxemia and hypercapnia. Often, children with mechanical problems (e.g., scoliosis) develop significantly increased effort to maintain normal minute ventilation during even mild upper respiratory infection.

Pulmonary parenchymal disease is often the cause of acute respiratory failure. Most children with acute parenchymal disease that causes respiratory failure are less than 1 year of age. In the presence of underlying cardiopulmonary disease, acute pulmonary infection may induce respiratory failure. This correlation is important in children with conditions such as bronchopulmonary dysplasia, congenital heart disease, cystic fibrosis, or other chronic lung processes.

In addition, numerous nonrespiratory diseases may precipitate respiratory failure. The pathophysiologies of the diseases listed in Table 95.2 are varied, but each alters the balance of O_2 consumption and CO_2 production such that they cannot be maintained by the respiratory system, leading to secondary respiratory failure.

Clinical Manifestations

Acute respiratory failure represents the severe end of the spectrum of respiratory distress; it signifies an imbalance of O_2 consumption and CO_2 production. Table 95.3 outlines the clinical findings and laboratory abnormalities. Few clinical manifestations appear early in the progression of respiratory failure. It is important to remember that prevention of the "blood gas"–proven respiratory failure should be the goal of the emergency physician. Therefore, in many cases, therapy should be initiated before the laboratory criteria have been fulfilled.

Clinical findings
Vital signs: tachycardia, tachypnea
General appearance: cyanosis, diaphoresis, confusion, restlessness, fatigue, shortness of breath, apnea, grunting, stridor, retractions, decreased air entry, wheezing
Blood gas abnormalities
$Paco_2$ ≥50 with acidosis (pH <7.25)
$Paco_2$ ≥40 with severe distress
Pao_2 <60 (or Sao_2 <90%) on 0.4 Fio_2
Pulmonary function abnormalities
Vital capacity (<15 mL/kg)
Inspiratory pressure (<25–30 cm H_2O)

Table 95.3. Diagnosis of Acute Respiratory Failure from Pulmonary Causes in Children

Management

Table 95.4 outlines a plan for management of acute respiratory failure. Resuscitation and basic life support are discussed in Chapter 1. The therapeutic approach must relieve or modify alveolar hypoventilation or arteriolar hypoxemia, while the physician simultaneously strives to establish an etiologic diagnosis.

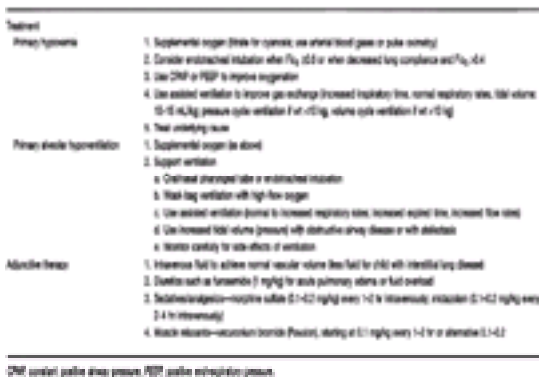

Table 95.4. Management of Acute Respiratory Failure

Treatment of respiratory failure is divided into three categories (Table 95.4). First, the physician should always assume that hypoxemia is present and should give sufficient supplemental oxygen (starting at 1.00 Fi O_2 in severe situations) to improve arteriolar oxygen levels. The goal of this procedure should be to achieve a minimal acceptable Pa O_2 of 60 mm Hg (SaO_2 greater than 90%) in newborns and 70 mm Hg (SaO_2 greater than 93 to 95%) in older children. If hypoxemia persists after adequate supplemental oxygen is administered, assisted positive-pressure ventilation should be initiated (mask bag reservoir, then proceeding to endotracheal intubation) to improve the efficiency of gas exchange. The amount of ventilation provided should be that which produces adequate chest wall expansion and delivery of gas. Usually, this will be achieved with a tidal volume of 10 to 15 mL/kg and a respiratory rate appropriate for the child's age (i.e., 20 to 30 breaths/minute in the young infant and 12 breaths/minute in the adolescent). Inspiratory flow rates need to be set to control the inspiratory:expiratory (I:E) ratio, which is normally around 1:2. Using a manometer in the endotracheal tube reservoir circuit may assist in determining whether unduly low or high airway pressures are generated with each artificial breath. When the patient has a stiff respiratory system or collapse of airways (atelectasis), higher pressures must be generated to sufficiently ventilate the child. If air trapping occurs (i.e., asthma, severe bronchiolitis, or bronchopulmonary dysplasia), the tidal volume and inflating pressure must be increased to adequately ventilate the alveoli. Although past practice strove to achieve normocapnia, more recent practice dictates a pressure-limited ventilation with peak pressures at 35 to 40 mm Hg to minimize the risk of barotrauma. This process maintains oxygenation while allowing varying degrees of permissive hypercapnia and is a safe alternative if the pH is maintained in a reasonable range. When the lung volume is diminished (i.e., neurogenic disease, hyaline membrane disease, interstitial pneumonia in immunocompromised hosts), positive end-expiratory pressure (PEEP) or constant positive airway pressure (CPAP) is indicated. This procedure will increase the end-expiratory lung volume (functional residual capacity) to a position on the compliance curve that allows easier alveolar ventilation.

In general, pressure-cycle or pressure-limited ventilators are used for children who weigh less than 10 kg (most children with acute respiratory failure). The machine settings should replicate the manual settings that produce adequate chest movement and alveolar ventilation. The I:E ratio is set in accordance with the type of disorder. Increased I:E ratios of 1:5 to 1 are used in alveolar-interstitial disorders to improve oxygenation. A normal or decreased I:E ratio is used in lower obstructive airway disease to improve exhalation time to excrete CO_2. From the emergency physician's perspective, the goal is to improve respiratory function, delivering oxygen and minimizing acidosis, without creating significant risk of the complications of ventilation.

Intravenous fluids should be titrated to maintain normal vascular volume as determined by observation of heart rate, blood pressure, peripheral perfusion, and urine output. In severely ill children who require more prolonged therapy in the ED, the measurement of central venous pressure may provide a more exact guide. Children with severe pulmonary interstitial involvement or pulmonary-capillary leak may benefit from a decreased vascular volume. This condition may reduce the need for high FiO_2 concentrations and CPAP and improve the mechanics of the ventilator. At other times, intravenous fluids may need to be transiently increased to improve cardiac filling, which has decreased secondary to reduction in venous return from elevated transpulmonary pressure.

The efficient use of assisted ventilation may require the use of sedation even in the early phases of emergency management. If the child is neurologically depressed, sedation may be contraindicated. In other cases, morphine sulfate 0.1 to 0.2 mg/kg every 1 to 2 hours may be given. A benzodiazepine is also a useful alternative if additional sedation is required. Midazolam 0.1 to 0.2 mg/kg intravenously can be given. Additional doses can be given as needed, generally on a 2- to 4-hour basis. Muscle relaxants are occasionally required to optimally ventilate children with severe respiratory failure. This treatment may be necessary for children with stiff lungs (e.g., severe interstitial pneumonia) or stiff chest wall (e.g., status epilepticus). Muscle relaxants improve chest wall compliance and reduce oxygen consumption. Vecuronium may be given initially in doses of 0.1 mg/kg every 1 to 2 hours (see Chapter 5).

BRONCHOPULMONARY DYSPLASIA

Background

Bronchopulmonary dysplasia (BPD) is a chronic lung disorder that may follow moderate to severe hyaline membrane disease (HMD) (Fig. 95.1) or other acute lung insults around birth. Children with apnea, complex congenital heart disease, or other illnesses requiring prolonged ventilation in the first weeks of life are at risk. The term *BPD* should be limited to children with neonatal lung pathology who are more than 28 days of postnatal age and who have significant clinical, radiologic, or blood gas abnormalities. The overall rate of BPD has dramatically decreased with the advances in newborn care and use of surfactant. In most industrialized nations, BPD is exceedingly rare in infants more than 32 weeks' gestation, but it remains common in infants who are very immature and weigh less than 1000 g at birth. Etiologic

factors have not been proven, but the following have been implicated: lung immaturity, oxygen therapy, positive-pressure ventilation (lung stretch), infection and inflammation, abnormal nutrition, and lung healing.

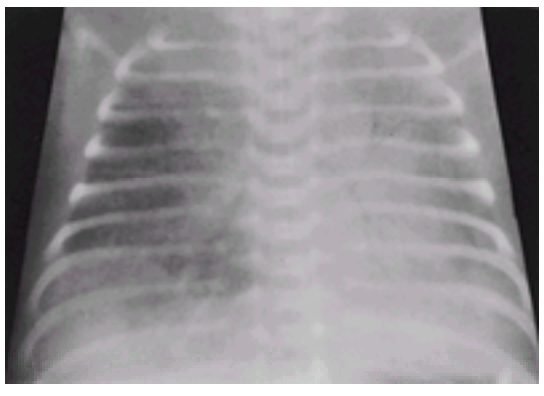

FIGURE 95.1. Hyaline membrane disease. This is one of twins born at 31 weeks' gestation. The chest film shows hypoaeration, diffusely opaque lung fields, air bronchograms, and loss of normal vascular shadows.

Clinical Manifestations

Typically, infants with BPD are discharged from the nursery initially at 3 to 6 months of age for home therapy, although more severely affected patients may require assisted ventilation for much longer intervals. These children have tachypnea and retractions at rest or during the mildest respiratory infections or fever. Their lungs are hyperinflated (increased anteroposterior chest diameter), and they may have crackles, wheezes, or decreased breath sounds in areas of the thorax. Some patients will manifest dyspnea and moderate to severe failure to thrive. Arterial blood gas (ABG) tensions show Pa_{O_2} less than 60 mm Hg (Sa_{O_2} less than 90 to 92%) and/or Pa_{CO_2} more than 45 mm Hg in room air, often despite respiratory rates of greater than 60 to 80 breaths/minute. Chest roentgenograms (Fig. 95.2) demonstrate varying amounts of hyperinflation; several patterns occur, including cystic areas with signs of fibrosis, which are often confused with congenital lobar emphysema; severe cystic fibrosis; and new infiltrates when previous radiographs are not available for comparison.

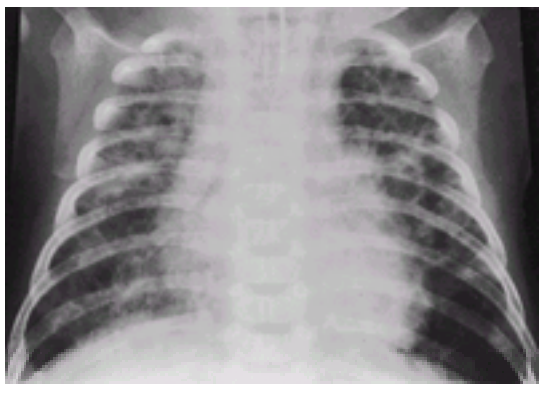

FIGURE 95.2. Bronchopulmonary dysplasia. This 2-month-old child was treated with mechanical ventilation during the first days of life for hyaline membrane disease. The chest film shows generalized overaeration and coarse modularity with multiple cystlike areas throughout both lung fields.

Management

Emergency physicians most often will evaluate children with BPD accompanied by acute respiratory infections. More than 50% of infants with BPD require admission within a year of their diagnosis for respiratory illness. Of particular importance is respiratory syncytial viral (RSV) infection, which occurs most often in winter months. RSV infections can lead to a deterioration in respiratory status or manifest with apnea only without significant acute pulmonary change. Management is primarily limited to supportive care: ensuring oral or intravenous intake, providing relief of hypoxemia, and when necessary, providing assisted ventilation for hypercarbia and acidosis. Although infants with BPD can often be treated as outpatients, care must be taken to determine the severity of the episode. Pulse oximetry can be beneficial in assessing the degree of oxygen saturation and may obviate the immediate need for arterial puncture in mild illness (in general, A saturation 90% or greater is equivalent to a Pa_{O_2} more than 60). ABG is always necessary when signs and symptoms may be the result of hypercapnia or when cyanosis, respiratory distress, or deterioration from baseline cannot be easily reversed. ABGs may be misleading in that decreases in Pa_{O_2} may reflect hypoxia from crying rather than from worsening pulmonary function. A chest radiograph is helpful in most episodes but often merely corroborates clinical changes. Indications for admission include a respiratory rate above 70 to 80 breaths/minute (or significant change from baseline), increasing hypoxia or hypercarbia, poor feeding associated with respiratory symptoms, apnea, or new pulmonary infiltrates. Parental fatigue or stress are also important factors to consider. If the acute exacerbation is mild, outpatient therapy may be indicated. Home therapy may include supplemental oxygen, bronchodilators, and corticosteroids. Most children with BPD have had trials of b-agonists. Nebulizer delivery is favored as it may be easier to use during the infant's sleep time. Some children benefit from diuretic therapy and have a need for increased oxygen supplementation during the acute illness. Antibiotic therapy should be considered when the risk of bacterial infection appears higher. Studies have demonstrated that seasonal use of RSV immunoglobulin may reduce the clinical picture in the most

high-risk infants with BPD.

Interstitial Pneumonia

Background

The group of pulmonary diseases best known collectively as interstitial pneumonia (often with pulmonary fibrosis) is categorized by cause in Table 95.5. The multiple causes, often associated with no known insults, may result in end-stage lung disease requiring lung transplant for cure. Patients may have a disorder that includes multiple cystic areas in the periphery of the lung fields, separated by fibrosis of connective tissue. Usual interstitial pneumonia (UIP—cryptogenic or idiopathic fibrosing alveolitis) is uncommon in children and requires a biopsy for diagnosis. Desquamative interstitial pneumonitis (DIP) may also occur rarely in children. It is not diminishing in frequency as it is in adults and has a familial form. Pathologically, the difference is seen on biopsy; cellular infiltrates predominate in DIP, whereas fibrosis predominates in UIP. Lymphoid interstitial pneumonia (LIP), with a lymphocytic infiltrate, is somewhere between UIP and DIP pathologically and is more recently associated with HIV infection in up to 30% of perinatally transmitted disease (see Chapter 85). The prognostic implications are crucial to the differences among the disorders. DIP is usually sensitive to corticosteroids, with dramatic clinical improvement of pulmonary disease.

Environmental irritants	Sarcoidosis
Inorganic dusts	Inherited disorders
Organic dusts (hypersensitivity)	Neurofibromatosis
Noxious gases	Miscellaneous causes
Drugs	Celiac disease
Radiation	Whipple's disease
Collagen vascular disease	Weber-Christian disease
Rheumatoid arthritis	Histocytosis
Scleroderma	Hermansky-Pudlak syndrome
Systemic lupus erythematosus	

Table 95.5. Causes of Interstitial Lung Disease

Clinical Manifestations

Interstitial pneumonias are rare in children but may manifest in older patients or in immunocompromised children initially with dyspnea on exertion and later at rest. Children often have a concomitant nonproductive cough, and systemic symptoms, including weight loss, anorexia, and fatigue, may occur. Occasionally, hemoptysis or a “spontaneous” pneumothorax may be the first event. Physical findings may include tachypnea and tachycardia, with shallow excursions and bibasilar end-inspiratory rales. In severe cases, digital clubbing and cyanosis may be present. The chest roentgenogram (Fig. 95.3 and Fig. 95.4) may be normal or may show a diffuse reticulonodular infiltrate, especially in the lower lobes. Eventually, the restrictive disease will progress with decreased lung volume and a cystic “honeycomb” appearance. Pulmonary function testing reveals reduced vital capacity with reduced forced expiratory volume in 1 second (FEV_1) but normal FEV_1:FVC (forced vital capacity) ratio. ABG tensions or oxygen saturation predict severity by initial reduction during exercise and with progression during rest. The $PaCO_2$ usually remains normal until late in the course. Polycythemia may be present secondary to hypoxemia in severe cases.

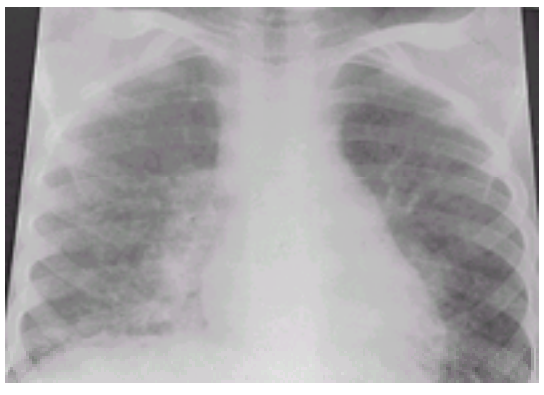

FIGURE 95.3. Sarcoid. A 9-year-old child with hepatosplenomegaly but no pulmonary complaints. The chest film shows interstitial lung disease with hilar adenopathy.

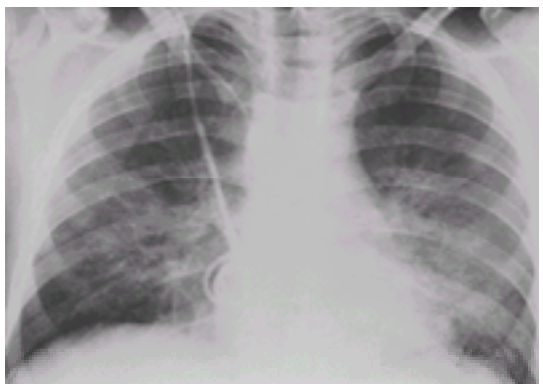

FIGURE 95.4. Idiopathic interstitial pneumonia. An 18-year-old boy with chronic granulomatous disease, after bone marrow transplant, with insidious onset of shortness of breath. The chest film shows bilateral interstitial disease in the lower lung field more on left side.

Management

Because the diagnosis requires biopsy to separate the entities, clinical concerns include 1) how to manage the new patient with suspected UIP, DIP, or LIP and 2) how to manage the child with known interstitial disease and acute illness. The child with characteristic history and physical findings will need admission to the hospital and assurance that the oxygen saturation is satisfactory at rest (by ABG or pulse oximetry). Referral to a pediatric pulmonologist should be made. The workup will usually include pulmonary functions (with diffusion capacity and exercise testing). High-resolution computed tomography (CT) has become an important adjunct because it produces much greater detail on abnormal lung morphology than does a routine chest radiograph. A ventilation-perfusion scan is often obtained, and in some instances, a cardiology consultation may be beneficial. Transthoracic lung biopsy (now often done by thoracoscopy) is the preferred procedure to establish the diagnosis and begin therapy. The older child with only mild impairment may not have to be admitted immediately but does need close follow-up and initial ABG and complete blood count (CBC) (to detect polycythemia), as well as studies, such as antinuclear antibody (ANA) and serum immunoglobulins, to detect vasculitis. Of concern in the differential diagnosis with presumed LIP would be the possibility of an immunocompromised host or HIV infection, which may necessitate a bronchoalveolar lavage to rule out infection, such as *Pneumocystis carinii.* In general, therapy consists of low-flow oxygen and a trial of systemic corticosteroids. Final data on use of other immunosuppressants are still pending, although in many cases, the results appear promising.

The child who presents in relapse of a diagnosed interstitial pneumonia should be assessed in a similar fashion. The physician should test for hypoxemia and treat it. The specific history revealing premature discontinuation of steroids in DIP may account for an acute relapse. Unfortunately, these diseases may progress despite aggressive therapy. Admission for supportive therapy may be helped by use of gallium scan or bronchoalveolar lavage, which measure severity of disease through the finding of active quantitative results of macrophage activation. Patients with active interstitial disease require close follow-up by a pediatric pulmonologist.

Aspiration Pneumonia

Background

Pneumonia is an inflammation of the lung tissue that may follow either a noninfectious or infectious insult, often involving the pathogens entering directly down the respiratory tract. The common infectious causes of pneumonia are discussed in Chapter 84. Although the pathophysiology of these pneumonias may directly involve aspiration of various pathogens, the term *aspiration pneumonia* has become commonly associated with the consequences of inhalation of oropharyngeal or stomach contents. In contrast to episodic pneumonia in an otherwise healthy child, aspiration pneumonia is most common in children with debilitation, altered consciousness, and CNS disorders that impair normal swallowing or protective airway reflexes. Other important causes of aspiration pneumonia are disorders of esophageal motility or problems with gastric emptying, and obstructive lesions such as tracheoesophageal fistula or, rarely, duodenal stenosis. Institutionalized children are often afflicted with this disorder. However, aspiration pneumonia can also occur in healthy children who have full stomachs when undergoing emergency procedures, in otherwise healthy children following seizures, or in children with decreased intestinal motility caused by pain, trauma, or analgesic administration.

Pathophysiology

Wynee and Modell have comprehensively reviewed the pathophysiology of aspiration pneumonias, which may be classified in two groups, based on the pH of the aspirate. A critical pH (2.5), below which severe lung damage is likely to occur, has been described.

Acid Aspiration

Aspirations in humans are considered acidic if the pH of the aspirate is less than 2.5. Experimentally, as the pH decreases below this level, the severity of lung injury increases. The volume and distribution of the aspirate are also important: large, poorly localized aspirates are most damaging. Likewise, aspirates with high bacterial content, such as those in patients with bowel obstruction, are associated with high morbidity. Acid aspiration causes a severe chemical pneumonitis with direct injury to alveolar-capillary membranes. A hemorrhagic, granulocytic, necrotizing reaction generally follows.

Hypoxia from multiple causes can occur within minutes of acid aspiration. These causes include reflex airway closure;

destruction of surfactant, resulting in atelectasis; interstitial and alveolar edema after exudation of fluid and protein across damaged membranes; and alveolar hemorrhage and consolidation.

Nonacid Aspiration

Nonacid aspirates (pH greater than 2.5), such as from the oropharynx or with mixed aspiration, can cause either transient or sustained pulmonary damage. Many of the early effects seen after acid aspiration may also result from nonacid aspiration. Pulmonary edema, endothelial cell alteration, and peribronchial neutrophilic infiltration have all been described. However, alveolar neutrophilic infiltration and necrosis are minimal. The exact nature and extent of lung damage from nonacid aspirates depend greatly on the composition of the aspirate. In contrast to the rapid resolution of nonacid, clear liquid (saline, water) aspiration, a prolonged pathologic response follows aspiration of partially digested meat, vegetable, or dairy products in which small food particles may be present. Unlike the neutrophilic response to acid aspiration, a widespread mononuclear and granulomatous reaction is seen in these cases. When repeated aspirations of irritating food particles occur over an extended period, roentgenograms may show granuloma formation similar to that of miliary tuberculosis.

Infection

The role of infection in the pathogenesis of aspiration pneumonia is unclear. Most physicians agree that infection plays little or no role in the initial pulmonary complications as a result of aspiration. However, following acid aspiration, the injured lung is potentially vulnerable to bacterial infection. Bacterial pulmonary infection is a complication in up to half of these cases. Why most patients do not develop pulmonary infection is not well understood. Although normal gastric contents contain few, if any, bacteria, oropharyngeal secretions may contain up to 10^8 bacteria/cm^3. It is conceivable that bacteria in oropharyngeal secretions either do not always gain access to susceptible areas or are diluted by the aspirate to concentrations that can be cleared by local defense mechanisms.

Two distinct patterns of infection are seen in patients who aspirate. A localized necrotizing bacterial pneumonia, abscess, or empyema may result from a heavily infected inoculum. In nonhospitalized children, anaerobic organisms are generally responsible for these infections; in hospitalized patients, facultative anaerobes and aerobic organisms are more common. The second pattern of infection is that which follows large aspirates, usually of the acid type. Aerobic, rather than anaerobic, organisms predominate here; Gram-negative organisms such as *Pseudomonas aeruginosa* and Gram-positive organisms such as staphylococci are often isolated. Of particular risk are children with bowel obstruction who develop aspiration syndromes.

Clinical Manifestations. Aspiration pneumonia should be suspected in any at-risk child who has signs of respiratory distress. The actual aspiration is often witnessed or vomitus is present in the immediate vicinity, suggesting the possibility of aspiration.

The clinical manifestations of pneumonia are discussed in Chapter 84. Most often, after the aspiration of gastric contents, a brief latent period occurs before the onset of respiratory signs and symptoms. More than 90% of patients are symptomatic within 1 hour, and almost all patients have symptoms within 2 hours. Fever, tachypnea, and cough are usually seen. Hypoxia is common. Apnea and hypotensive shock are less common. Sputum production is usually minimal.

The physical findings in patients with aspiration pneumonia are not dissimilar from those in patients with pulmonary infections resulting from either bacterial or viral causes. Diffuse crackles and wheezing are common; cyanosis appears with progression of the diseases. Chest roentgenograms (Fig. 95.5A and Fig. 95.5B) may show either localized or diffuse infiltrates, which are often bilateral. The chest roentgenogram of a patient who has aspirated stomach contents may evolve suddenly from normal to complete bilateral opacification within 8 to 24 hours.

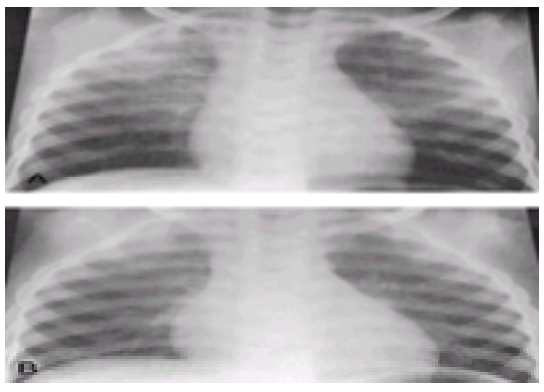

FIGURE 95.5. A. Blood aspiration. A 3-year-old boy with tachypnea 1 day after surgery for enlarged adenoids/tonsils. Chest film shows an infiltrate in the right upper lobe and left lower lobe. **B.** Blood aspiration (see **A**). The chest film 2 days later shows clearing of the infiltrate in the right upper lobe and left lower lobe.

Management. The suspicion of aspiration should be confirmed with a chest radiograph. Children with a significant aspiration pneumonia (lobar infiltrates, moderate to severe respiratory distress) require admission to the hospital. Table 95.6 outlines therapeutic modalities that may be useful. Some children who aspirate may have radiographs that are significantly abnormal in the face of minimal clinical symptoms. A common example is that of mild hydrocarbon aspiration.

Proven measures	Optional modalities
Suction	Corticosteroids
Airway protection	Antibiotics
Oxygen	

Table 95.6. Initial Treatment of Aspiration Pneumonia

In the acute-care setting, children who aspirate stomach contents require primarily supportive care. Specifically, prevention of further aspiration by adequate oropharyngeal suctioning and proper positioning should be the rule. The pulmonary signs and symptoms associated with aspiration of stomach contents may resolve quickly with supportive care or progress to respiratory failure, with the subsequent development of bacterial superinfection over a period of days. Intubation of the trachea is indicated if the airway reflexes are inadequate or if respiratory failure ensues. Supplemental oxygen should be administered, as determined by pulse oximetry or direct measurement of oxygenation with an ABG. The management of subsequent bacterial infection is addressed in the following section.

The use of corticosteroids in the treatment of aspiration pneumonia is controversial. Because experimental evidence indicates at best minimal benefit from steroids in acid aspirations and because these drugs may be contributing factors in the development of secondary bacterial pneumonia, their administration is not indicated in the ED.

Another consideration in the therapy of aspiration pneumonias is the role of prophylactic antibiotic administration. Because fever, purulent sputum, leukocytosis, and pulmonary infiltrates all may result from chemical pneumonitis alone and because no strong data exist that support the use of prophylactic antibiotics in children who acutely aspirate stomach contents, a reasonable initial approach is to defer antibiotic treatment in favor of careful observation.

Assuming that prophylaxis is not given following the aspiration of stomach contents, and clear signs of infection later develop, the choice of antibiotics can be guided by both the clinical setting and the results of properly obtained specimens for culture. Community-acquired pneumonias generally involve anaerobes and are adequately treated with penicillin, whereas nosocomial infections require antibiotics effective against both aerobes (including *Staphylococcus aureus* and Gram-negative bacilli) and anaerobes. A combination such as clindamycin and gentamicin is often used. In neurologically impaired children, with either aspiration or tracheostomy-associated pneumonia, antibiotics effective against penicillin-resistant anaerobic bacteria (i.e., clindamycin or ticarcillin-clavulanate) have been shown to produce clinical and microbiologic responses superior to those associated with agents less effective against these organisms.

Pulmonary Embolism

Background

Pulmonary embolism is largely a disease of adults, afflicting more than 500,000 patients per year in the United States. In autopsy series, the reported incidence of this disorder ranges from 1 to 4% in children and up to 8% in teenagers. In the adolescent population, the incidence of pulmonary embolism is reportedly approximately 1 case per 1,300 hospital admissions. In Cincinnati, over a 20-year period, a retrospective review identified only 20 cases, with an average patient age of 18 years (range, 10 to 20 years).

Perhaps one reason for the uncommon occurrence of pulmonary embolism in children is that generally healthy individuals are rarely afflicted. Major risk factors identified in children include ventriculoatrial shunt(s), trauma, congenital heart disease, infection (e.g., bacterial endocarditis), neoplasia, central venous catheters, prolonged immobilization, surgery, and severe dehydration. Adolescents have a unique set of risk factors for pulmonary embolism, which are a mixture of those for younger children and for adults. They include oral contraceptive use, trauma (particularly spinal injury), elective abortion, surgery, prolonged immobilization, infection, collagen vascular disease, intravenous drug abuse, rheumatic heart disease, severe dehydration, obesity, and renal transplantation.

Pathophysiology

Embolism is not a primary phenomenon, but it is by definition a complication of thrombosis elsewhere in the body. In adult populations, approximately 90 to 95% of pulmonary emboli originate within the deep venous systems of the pelvis and thigh. In contrast, the associated venous thromboses described in autopsy series in children are more commonly located in the chambers of the right side of the heart, mesenteric and cerebral vessels, or the inferior or superior vena cava. In adolescents, deep vein thrombosis is a common concomitant physical finding. In addition to venous thrombi, pulmonary emboli can arise from tumor cells, amniotic fluid, air, or fat (following fractures).

Obstruction of the pulmonary arteries by an embolus affects the pulmonary and bronchial circulation, the airways, and the function of the right and left sides of the heart. The degree of hemodynamic compromise correlates with the extent of arterial obstruction in patients without preexisting cardiopulmonary disease. In patients with underlying heart or lung disease, lesser degrees of obstruction may trigger severe pulmonary hypertension or cardiovascular collapse.

The cause of acute hypoxemia, which is seen in more than 80% of patients with pulmonary embolism, is not well defined.

Alveolar hypoventilation, intrapulmonary shunting, decreased diffusion capacity, and alveolar collapse secondary to a reduction in surfactant may all contribute.

Clinical Manifestations

The classic presentation of massive pulmonary embolism with severe circulatory compromise is easily recognized. However, most patients have nonspecific signs and symptoms and no pathognomonic laboratory abnormalities (Table 95.7). The most common presenting abnormalities in children and adolescents are pleuritic pain (which may radiate to the shoulders), dyspnea, cough, and hemoptysis. Additional findings may include apprehension, nonproductive cough, fever, sweats, and palpitations.

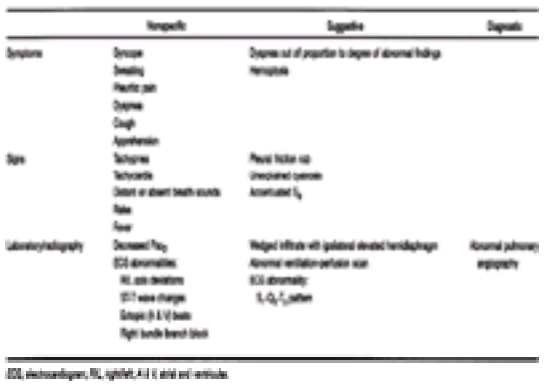

Table 95.7. Clinical Manifestation of Pulmonary Embolism

Aside from tachycardia, abnormalities on physical examination are often lacking. If a sufficiently large associated infarction is identified, there may be decreased resonance over the lung fields and a pleural friction rub. Breath sounds may be distant or absent, and rales may be heard. The presence of hypoxemia not completely explained by the underlying disease process or clinical state should suggest the possibility of pulmonary embolism.

The diagnosis of pulmonary embolism can be established with high probability based on ventilation-perfusion lung scan findings. The characteristic pattern is normal ventilation of poorly perfused areas of lung. Many physicians consider pulmonary angiography to be the preferred diagnostic method in previously healthy individuals. However, the reliability of this method diminishes with time after the acute embolism. The electrocardiogram (ECG) is neither a specific nor a sensitive indicator of pulmonary embolism. The S_1-Q_3-T_3 pattern that has been described with pulmonary embolus may be seen in other conditions, including a pneumothorax. Although the presence of a pulmonary infiltrate with an ipsilateral elevated hemidiaphragm is suggestive of a pulmonary embolism, there are no pathognomonic radiographic signs in the acute-care setting. ABGs generally indicate a decreased partial pressure of oxygen. However, about 15% of patients have a PaO_2 greater than 80 mm Hg and 5%, greater than 90 mm Hg. In one series, all patients had a decreased A-a gradient, if measured.

Management

In all patients strongly suspected of having a pulmonary embolism, a chest radiograph, ECG, and ABG should be obtained. If the clinical suspicion is high, regardless of the results, the patient should be admitted for initiation of definitive treatment. When the patient is vaguely suspected of having pulmonary embolism, all the aforementioned tests are normal, the patient's clinical condition permits, and the likelihood of pulmonary embolism appears low, the patient may be discharged with close follow-up. When abnormalities are uncovered, further diagnostic workup (i.e., ventilation-perfusion scan) and admission to the hospital should be considered.

Initial therapy includes supplemental oxygen, ventilatory support as indicated, and achievement of venous access. Intravenous heparin remains the mainstay of definitive therapy for pulmonary embolism because its onset of action is immediate and it is rapidly metabolized. However, it should be kept in mind that heparin is a common cause of in-hospital drug-related deaths in reasonably healthy adults, and it has been cited as a source of in-hospital complications in adolescents. The initial dosage is 500 units/kg daily given as a continuous intravenous infusion, which is adjusted to maintain the partial thromboplastin time (PTT) at 1.5 to 2 times the baseline value. Warfarin sodium is the orally administered anticoagulant usually used to maintain long-term anticoagulation. This drug may be initiated either at the time of initial treatment with heparin or 1 to 2 days thereafter. The required daily dose varies, depending on concomitant medical illness and other drug ingestion. In adults, the dose is adjusted to maintain the prothrombin time (PT) at twice normal. Warfarin is usually continued for 2 months beyond the time of diagnosis. However, the role of fibrinolytic agents in the treatment of children has not been established.

Pulmonary Edema

Background

Pulmonary edema refers to the abnormal accumulation of fluid within the alveolar spaces and bronchioles. Alterations of normal intravascular and interstitial hydrostatic and colloid osmotic pressures, changes in the permeability characteristics of the fluid-exchanging membranes in the lungs, and impairment of lymphatic drainage have been found to be instrumental in the generation of this condition. Pulmonary edema may occur in a variety of disorders, including adult respiratory distress syndrome (ARDS), left-sided heart failure, congenital heart malformations such as ventricular septal

defects, severe malnutrition, extensive burns, nephrosis, upper airway obstruction, asthma, bronchiolitis, pneumonia and other infections, hypervolemia, and poisoning with barbiturates, narcotics, alcohol, paraquat, epinephrine, hydrocarbons, and gases such as oxides of sulfur and nitrogen, hydrocyanic acid, and smoke.

Pulmonary edema can also develop in some highland dwellers who return home after a brief stay at sea level and in some sea-level dwellers soon after arriving at high altitude. This “high-altitude” pulmonary edema characteristically affects young people who are exposed to altitudes above 2700 m.

Pathophysiology

As previously noted, fluid accumulation in the lung is largely determined by the balance of vascular and interstitial hydrostatic and colloid osmotic pressures, vascular permeability, and lymphatic drainage. Edema may develop following the alteration of one or more of these factors, which vary according to the disease state involved. Many disorders are associated with increased pulmonary vascular bed hydrostatic pressures, resulting from elevation of vasculature pressures distal to the lung. These disorders include congenital conditions such as hypoplastic left heart syndrome, cor triatriatum, mitral stenosis, and left-sided heart failure, which may be seen with severe aortic stenosis, coarctation of the aorta, large arteriovenous fistulas, or myocardial disease. Left-to-right vascular shunting as seen in patent ductus arteriosus, ventricular septal defects, or iatrogenic shunts (e.g., Waterston) may lead to edema via increasing pulmonary vascular blood flow. Hydrostatic pressures are also raised by overaggressive administration of intravenous fluids.

A breakdown in the alveolar-capillary barrier with accumulation of protein-rich fluid in the interstitium and alveoli is the major and the initial manifestation of ARDS. This increased capillary permeability edema is the result of an assault on the pulmonary vasculature by various inflammatory cells. Cellular enzymes, metabolic products, and cytokines seem to be the major factors responsible for tissue destruction.

Decreased plasma colloid osmotic pressure is associated with pulmonary edema. This condition is seen in children with significantly lowered levels of plasma proteins, such as those with nephrosis, protein-losing enteropathies, massive burns, and severe malnutrition.

In patients with upper airway obstruction (Fig. 95.6A, Fig. 95.6B, Fig. 95.6C), exaggeration of the transmural pulmonary vascular hydrostatic pressure gradient is the most likely pathogenic mechanism. The highly negative pleural pressures that accompany upper airway obstruction tend to increase venous return (causing increased pulmonary vascular volumes), impair ejection of left ventricular blood, and cause highly negative pulmonary interstitial pressures, all of which contribute to transudation of fluid across pulmonary capillaries. In some patients, increased lung volumes may mask pulmonary edema on radiograph. The latter becomes manifest when lung volumes decrease following relief of the obstruction.

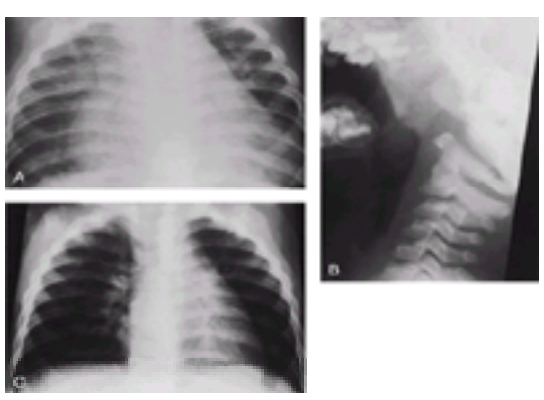

FIGURE 95.6. Cor pulmonale secondary to upper airway obstruction. **A.** This is a 2-year-old boy with tachypnea and dyspnea. The chest film show a large hematoma and mild interstitial edema. **B.** The lateral view of the neck shows obstructing enlarged adenoids and tonsils. **C.** The chest film 2 days after adenoidectomy shows a decreased heart size and improvement in interstitial edema.

A variety of clinical conditions are thought to alter the permeability of the alveolar-capillary membrane, presumably by damage to epithelial or endothelial cells. In some disorders, chemical mediators such as prostaglandins, histamine, and bradykinin may be influential in the genesis of pulmonary edema. Such is the case with asthma, hypersensitivity pneumonitis, Goodpasture's syndrome, and systemic lupus erythematosus. Pulmonary edema caused by denaturation of proteins and cellular damage is seen after the inhalation of several noxious gases, particularly those generated during fires. The inhalation of herbicides (i.e., paraquat) has also been associated with development of pulmonary edema. Other entities that alter alveolar-capillary membrane permeability include circulating toxins, such as snake venom and endotoxins from Gram-negative sepsis, and perhaps severe salicylate poisoning. The mechanism responsible for pulmonary edema that may accompany intracranial pathology is not entirely understood. Acute sympathetic discharge has been documented after head injury or sudden increases in intracerebral pressure. This reaction results in profound generalized vasoconstriction, increasing pulmonary-capillary pressure. The same mechanism might account for the pulmonary edema occasionally reported after seizures in children.

Clinical Manifestations

The onset of pulmonary edema is variable but may be rapid. Tachypnea, cough (often producing frothy, pink-tinged sputum), dyspnea, shortness of breath, and chest pain are commonly seen. Grunting often occurs in an effort to prevent lung collapse. On physical examination, the child may appear pale or cyanotic and have a rapid pulse. Decreased breath

sounds and moist (“bubbly”) rales are the most common auscultatory findings. However, these are generally absent with small increases in lung fluid. Indeed, auscultatory and roentgenographic findings may not manifest until the interstitial and extravascular fluid has doubled or tripled in volume.

Unless it is massive, acute fluid accumulation may not be detectable by chest roentgenogram. Lymphatic and interstitial fluid accumulations may be visible as Kerley A and B lines (septal lines) (Fig. 95.7). These lines represent interlobular sheets of abnormally thickened or widened connective tissue tangential to the radiograph beam. The A lines are located near the hilum of the lung, and the B lines lie in the periphery. Although other processes may produce these lines, when transient they are usually caused by edema. Flattening of the diaphragm on radiograph may also be a finding with pulmonary edema. This is presumably caused by air trapping that results from airway narrowing as a result of bronchiolar fluid collections.

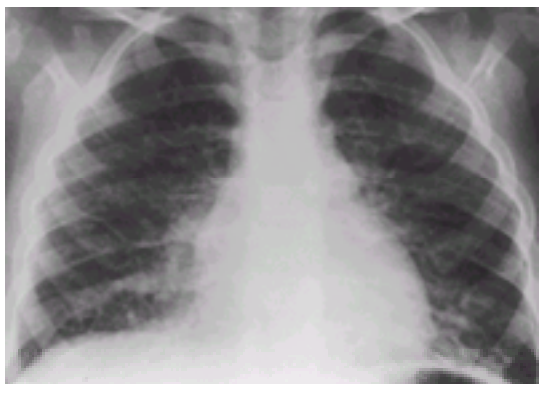

FIGURE 95.7. Interstitial fluid from volume overload. This is a 2-year-old child with paraspinal sarcoma removed 6 months earlier. Before chest radiation he received a large fluid load. The chest film shows interstitial edema with Kerley's lines and bilateral small pleural effusions.

It should be kept in mind that if pulmonary edema is superimposed on another pulmonary process, the clinical and roentgenologic findings may be obscured by those of the primary illness. Similarly, once pulmonary edema is severe enough, it may be difficult to separate edema, atelectasis, and inflammation on the roentgenogram.

Management

The management of patients with pulmonary edema should ultimately be directed toward correction of the primary disorder. Initial efforts (Table 95.8) should be directed toward reversal of hypoxemia by the administration of oxygen and by mechanical ventilation if necessary. The latter has several beneficial effects, including reduced oxygen consumption through reduced work of breathing, improved oxygenation by prevention of alveolar collapse, and reduced fluid filtration in the lung by decreased pulmonary vascular volume and pressure from the positive intrathoracic pressures generated. CPAP therapy delivered via face mask has also been shown to be effective in some patients. In addition to satisfying the patient's oxygen demands, reversal of hypoxemia is often useful in relieving chest pain and is important to the metabolism of vasoactive mediators that affect microvascular permeability.

Oxygen	Venodilation
Diuresis	Morphine 0.1 mg/kg IV
Furosemide 1 mg/kg IV	Digitalis (see Table 82.7, for dosage)

Table 95.8. Treatment of Pulmonary Edema

Other therapeutic measures should be tailored somewhat to fit the patient's individual needs. When heart failure is the cause of pulmonary edema, in addition to oxygen and ventilation, diuretics (to decrease plasma volume), digitalis (to improve contractility), and bronchodilators (to improve contractility and afterload and to produce bronchodilation) are useful. Morphine dilates the venous system and may be helpful in relieving anxiety and dyspnea. In patients with ARDS, clinical studies have shown that the use of methylprednisolone does not improve outcome and may in fact increase the mortality and incidence of secondary infections.

The ability of mammalian lungs to gradually absorb instilled fluid has been known for some time. The process responsible for fluid removal has been identified as active sodium transport by alveolar epithelium. Alveolar fluid clearance can be augmented with b-adrenergic agonists, which enhance sodium transport. Long-acting b-adrenergic agonists have been demonstrated to increase alveolar fluid clearance in animals. When decreased plasma colloid osmotic pressure is an issue, albumin administration may be helpful. To minimize initial rises in vascular pressures,

colloids should be infused slowly, usually in conjunction with diuretics.

Pulmonary Hemorrhage

Background

Bleeding into the lung manifests clinically with hemoptysis, or coughing up blood, and pathologically with pulmonary hemosiderosis, the presence of hemosiderin in lung macrophages. Although hemoptysis is uncommon, it has many possible causes, some of which may manifest in a dramatic and life-threatening manner. Table 95.9 provides a differential diagnosis for pulmonary hemorrhage by category. The cause of hemorrhage should be sought to provide a specific diagnosis. Children with recurrent hemorrhage may have isolated cow's milk allergy or an idiopathic cause. Rarely, isolated hemangiomatosis manifests with hemorrhage. Airway and parenchymal causes include infection, infarction, and congenital anomalies.

Primary	Associated with Other Organ Dysfunction	Secondary	Airways	Parenchymal
Cow's milk allergy	Nephritis	Congestive heart failure	Bronchitis	Trauma
Idiopathic	Wegener's granulomatosis	Clotting disorders	Bronchiectasis/cystic fibrosis	Infection—tuberculosis, other
Hemosiderosis	Vasculitis—Henoch-Schönlein purpura	Malignancy	Airway anomalies	Infection
	Collagen vascular disease	Alveolar injury	Vascular anomalies, including hemangiomatosis	Neoplasm
		Drug	Foreign body	Cavitary lesion
		Radiation		
		Smoke injury		
		Acid aspiration		

Modified from Boat TF. Pulmonary hemorrhage and hemoptysis. In: Chernick V, Boat TF, eds. Kendig's Disorders of the Respiratory Tract in Children. 6th ed. Philadelphia: WB Saunders, 1998:53.

Table 95.9. Causes of Pulmonary Hemorrhage in Children

Clinical Manifestations

The hallmark of pulmonary hemorrhage is recurrent intrapulmonary bleeding with lung injury and secondary depletion of body iron stores. Therefore, the symptoms and signs include hemoptysis, recurrent pneumonias (manifest by fever, tachypnea, tachycardia, and coarse or fine crackles), and pallor. Emesis of blood arising in the pulmonary tree may mislead the clinician to investigate the gastrointestinal tract. Associated symptoms of fatigue and poor weight gain are common.

Laboratory findings after recurrent hemorrhage most characteristically include a microcytic, hypochromic anemia with low serum iron. Leukocytosis and eosinophilia may be present, and the stool usually tests positive for blood. Roentgenograms (Fig. 95.8) may show alveolar infiltrates that may be transient, localized processes or that may be diffuse and chronic. Cardiorespiratory embarrassment ensues in children with severe anemia. Severely affected patients may develop secondary restrictive lung disease with retention of CO_2.

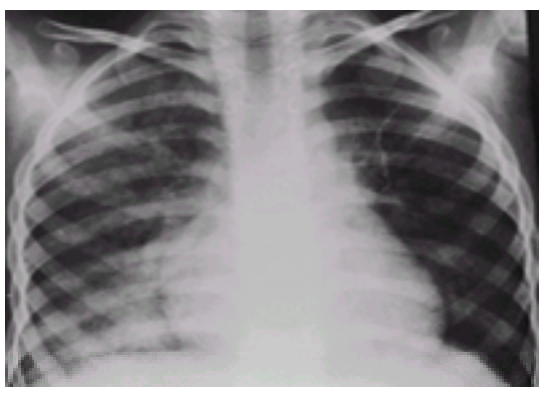

FIGURE 95.8. Idiopathic pulmonary hemosiderosis. A 5-year-old child with repeated bouts of pulmonary hemorrhage. The chest film shows diffuse radiopacities throughout both lungs (more on right side), with a well-defined alveolar opacity in the right lower lobe. Note the surgical sutures in left upper lobe.

A presumptive diagnosis can be made by finding siderophages (iron-laden microphages) in nasogastric washings; these macrophages will stain blue with the Prussian blue reaction (i.e., potassium ferrocyanide and hydrochloric acid). More definitive diagnosis requires the identification of typical macrophages obtained by bronchial lavage or lung biopsy.

Pulmonary function testing may reveal an obstructive pattern secondary to bronchial irritation from blood and, over time, a restrictive pattern from scarring and fibrosis.

Management

Most children with pulmonary hemorrhage have a chronic disease requiring supportive therapy for hypoxia and anemia in

the form of blood transfusions and supplemental O_2. Occasionally, pulmonary hemorrhage is so severe that it causes respiratory insufficiency or hypotension. Positive-pressure ventilation with PEEP is the preferred treatment in this situation; bleeding is usually not rapid or well enough localized to be identified and controlled by bronchoscopy. In allergic, vasculitic, and idiopathic hemorrhage, the use of corticosteroids is indicated as either methylprednisolone (2 mg/kg per day) or hydrocortisone (8 mg/kg per day) intravenously in three to four divided doses. When hemorrhage is caused by infection, especially tuberculosis, antimicrobial therapy should be instituted and steroids avoided. Admission is necessary to support the child until the cause of the process has been determined or acutely until the hemorrhage has been controlled. Bronchoscopy can be useful diagnostically to determine infectious causes and may localize bleeding sites. Occasionally, when the bleeding is brisk as in bronchiectasis with erosion to a bronchial vessel as in cystic fibrosis, embolization of vessels may be needed to stop the bleeding.

Pleuritis

Background

Pleuritis or pleurisy refers to inflammation of the pleural membranes, usually as a result of diseases elsewhere in the body. This inflammation may be associated with minimal or considerable accumulation of liquid in the pleural cavity, and remains a major cause of pulmonary morbidity in children. Specific references to the incidence of pleural effusions in various respiratory infections are made in Chapter 84. The surgical approach to pleural effusions is reviewed in Chapter 119.

The causes of pleural inflammation are varied (Table 95.10). Viral (e.g., coxsackievirus, Epstein-Barr virus, herpes zoster), mycoplasmal, bacterial (e.g., *S. aureus, Streptococcus pneumoniae, Haemophilus influenzae,* group A streptococcus, *Mycobacterium tuberculosis*), and fungal (e.g., histoplasmosis, coccidioidomycosis) infections from pulmonary, subdiaphragmatic, or more distant sites may all eventually involve the pleura. Neoplastic involvement may be primary or metastatic and may result in obstruction of lymphatic drainage, thereby promoting the accumulation of pleural fluid. Pulmonary embolism may cause pleural inflammation with or without effusion as a result of focal parenchymal necrosis. Trauma, both accidental and following diagnostic and therapeutic procedures in the chest, can irritate the pleura and lead to secondary infection. Pleuritis with or without effusion is seen in more than half of patients who have a systemic vasculitis such as systemic lupus erythematosus or sarcoidosis.

Transudative Pleural Effusions	Gastrointestinal diseases
Congestive heart failure	Pancreatitis
Cirrhosis	Esophageal rupture
Nephrotic syndrome	Subphrenic abscess
Acute glomerulonephritis	Hepatic abscess
Myxedema	Whipple's disease
Peritoneal dialysis	Diaphragmatic hernia
Hypoproteinemia	Peritonitis
Meigs' syndrome	Trauma
Sarcoidosis	Hemothorax
Vascular obstruction	Chylothorax
Ex vacuo effusion	Drug hypersensitivity
Exudative Pleural Effusions	Nitrofurantoin
Infectious diseases	Methysergide
Tuberculosis	Miscellaneous diseases
Bacterial infections	Asbestos exposure
Viral infections	Pulmonary and lymph node sarcoidosis
Fungal infections	Uremia
Parasitic infections	Postmyocardial infarction syndrome
Neoplastic diseases	Trapped lung
Mesothelioma	Congenital abnormalities of the lymphatics
Metastatic disease	Postradiation therapy
Collagen vascular diseases	Drug reactions
Systemic lupus erythematosus	
Rheumatoid pleurisy	
Pulmonary infarction/embolization	

From Light RW. Pleural effusions. Med Clin North Am 1977;61:1339.

Table 95.10. Differential Diagnosis of Pleural Effusion

Pathophysiology

The pleura is a double-layered, thin membrane that separates the lung from the chest wall, diaphragm, and mediastinum. The parietal pleura (outer layer) is adherent to the chest wall, and the visceral pleura (inner layer) completely covers the lungs except at the hili. In the healthy child, the two layers of pleura are in apposition, separated by only a thin layer of serous fluid. Fluid seems to enter the pleural space from a parietal pleura and to exit via the lymphatics and vasculature of the visceral pleura.

Several mechanisms have been linked by cause to abnormal pleural fluid accumulation, including changes in hydrostatic or oncotic pressures and diseases of the pleural surface that alter capillary permeability or affect lymphatic reabsorption of protein. On the basis of these mechanisms, pleural effusions can be classified as either transudates or exudates. Transudates occur as a consequence of altered capillary hydrostatic pressure or colloid osmotic pressure, as seen in congestive heart failure or hypoproteinemic states. Exudates result from any disease of the pleural surface that produces increased capillary permeability or lymphatic obstruction, such as pleural infection or tumor.

The determination of the nature of pleural fluid may be helpful from a diagnostic standpoint (Table 95.10). Traditionally, an effusion has been classified as an exudate if the total protein concentration exceeds 3.0 g/dL. However, use of this parameter alone results in inaccurate classification of more than 10% of effusions. Better separation of transudates and exudates can be made by comparing the concentrations of total protein and lactate dehydrogenase (LDH) in serum and pleural fluid. Although several investigators have tested modified versions, the criteria of Light et al. have been the most reliable in distinguishing exudative from transudative effusions. These criteria include a pleural fluid:serum protein ratio greater than 0.5, a pleural fluid:serum LDH ratio greater than 0.6, and a pleural LDH concentration more than two-thirds normal upper limit for serum. If any one of these critical values is exceeded, the effusion is an exudate.

Clinical Manifestations

The hallmarks of pleural disease are pain, shortness of breath, fever, and an abnormal chest roentgenogram. Inspiratory

chest pain from pleural inflammation is the most characteristic symptom.

In “dry” pleurisy, which is caused by a minor pulmonary infection, the patient is febrile with an irritating, nonproductive cough. The symptoms often follow an upper respiratory infection and often last only a few days. Patients are acutely ill appearing, with grunting respirations as a result of pain. Pressure over the involved area elicits tenderness. Upon palpation, a coarse vibration may be felt.

A pleural friction rub is most apt to be heard in pleural inflammation that is associated with little or no effusion. The sound has been variously described as low pitched with either a grating or squeaking quality. It is usually loudest on inspiration, but often it is also audible during expiration. Sometimes, the rub is confused with a low-pitched, nonmusical wheeze (rhonchus) produced by secretions partially blocking the airway. A vigorous cough will eliminate these secretions and sounds but will not affect the pleural friction rub.

Although symptoms of pleural effusion are varied and relate to the primary cause of the effusion, most patients complain of some degree of dyspnea. Pleuritic chest pain is also a common complaint and can occur before the accumulation of fluid. Characteristic physical findings include restriction of movement of the chest wall on the affected side, flatness to percussion, diminished to absent tactile and vocal fremitus, and decreased to absent breath sounds.

The physical findings of atelectasis of an entire lung and large pleural effusion are similar with one exception: both conditions produce dullness to percussion and absent transmission of voice and breath sounds. However, pleural effusion decreases the size of the hemithorax, causing the trachea to deviate away from the diseased side. On the other hand, atelectasis causes the trachea to deviate toward the diseased side.

Pleural effusion is the most common radiographic manifestation of pleural disease. The first roentgenographic sign of a pleural effusion is usually blunting of the costophrenic angles, producing wedgelike menisci that extend upward along the lateral chest wall. Similar collections are seen in the posterior costophrenic angles on lateral views. Larger effusions may be seen to extend up the entire lateral chest wall or retrosternally.

Pleural effusions may alternatively present with what at first appears to be unusual prominence or thickening of the interlobar fissures, or by wedge-shaped accumulations of fluid at either end of these fissures. The latter may appear as focal infiltrates or segmental atelectasis on some views. Although small effusions are easily overlooked on radiograph, with the proper technique, effusions as small as 5 to 25 mL can be demonstrated. In adults, pleural effusions are visible as a meniscus on lateral chest radiograph at a volume of approximately 50 mL. At a volume of 200 mL, the meniscus can be identified on the posteroanterior radiograph, whereas at a volume of 500 mL, the meniscus obscures the hemidiaphragm.

Management

The management of pleural disease is aimed at determining the cause, treating the primary disorder, and relieving associated functional disturbances. When no effusion is present, relief of chest pain is one of the most pressing issues. Analgesics, bed rest, and/or mild sedatives may be indicated. It should be kept in mind that irritability and restlessness may be a result of pain, which, in dry pleurisy, can occur with every phase of respiration.

The accumulation of pleural fluid usually provides relief from pain. Thoracentesis is indicated when fluid accumulation is extensive enough to cause dyspnea and/or for diagnostic purposes (Figure 95.9). Sonographic guidance can simplify and enhance the success of this procedure with small effusions. The complications of thoracentesis include pneumothorax, hemothorax, reexpansion pulmonary edema, and rarely, air embolism. The recommended technique for thoracentesis is given in Section VII, and details of management of fluid in the pleural space are reviewed in Chapter 119.

FIGURE 95.9. Approach to pleural effusion. [a]Consider simultaneous placement of small gauge (8–10 F) chest tube by Seldinger technique for large, free-flowing effusions.

Thoracentesis is the most commonly used method of external drainage of pleural fluid, but other techniques are also available. These techniques include image-guided catheter drainage, thoracostomy tube placement (which may be performed using the Seldinger technique), thoracotomy with debridement and directed chest tube placement, open pleural decortication, and video-assisted thoracoscopic pleural surgery (pleuroscopy). Of these available techniques, image-guided catheter drainage is the only one that might be routinely performed in the ED.

Image-guided percutaneous transthoracic drainage of infected pleural collections can be used in patients who have a diagnostic thoracentesis showing frank empyema. Fluoroscopy, sonography, CT, or any combination of these techniques

can accurately guide catheter placement. Image-guided catheter drainage is most effective in patients with short duration of symptoms, free-flowing or unilocular effusions, absence of thick pleural peel on CT scans, and fluid that can be easily aspirated by needle.

Pleuroscopy, another procedure generally not performed in the ED, may be used either as a diagnostic or a therapeutic procedure. Among the therapeutic purposes of pleuroscopy are 1) lysis of pleural adhesions in the management of pleural effusions, 2) removal of blood after trauma, and 3) performance of pleural toilet in the fibrinopurulent stage of empyema.

Diagnostic tests of pleural fluid should include gross and microscopic examination; Gram stain; and protein, glucose, LDH, and pH determinations. Cytology should also be performed if malignancy is known or suspected. On gross examination, empyema fluid is opaque and viscous, fluid high in cholesterol has a characteristic satinlike sheen, and chylous effusions are milky white. A clear or slightly yellow pleural fluid is generally a transudate; however, exudates may also appear clear. Concomitant blood and pleural fluid protein and LDH determinations can help physicians make the distinction, as previously discussed. The more protein and cells, the deeper the color and the more turbid the fluid.

Although pleural fluid pH measurements have not been reported in series of empyema in pediatric patients, these measurements have been found to be valuable in the management of adults. Pleural fluid pH values of greater than 7.2 to 7.3 are generally found in sterile fluids not requiring drainage. In adults, a pleural fluid pH of less than 7.3 limits the differential diagnosis to empyema, malignancy, collagen vascular disease, tuberculosis, hemothorax, or esophageal rupture; a pH of less than 7.0 is seen only in empyema, collagen vascular disease, or esophageal rupture. One notable exception to the previous statement is empyema caused by *Proteus mirabilis,* which causes an elevated pleural fluid pH. It is also important to remember that pleural fluid pH will be lowered in the face of systemic acidosis. Therapeutically, in adults, a pleural fluid pH of less than 7.2 suggests that the effusion will not resolve spontaneously and will require chest tube drainage. Pleural fluid should be collected anaerobically in a heparinized syringe and transported to the laboratory on ice to ensure proper pH measurement.

A pleural fluid:serum glucose ratio less than 0.5 has a similar differential diagnosis as low pleural fluid pH. In animal studies, both leukocytes and bacteria have been shown to use glucose anaerobically, resulting in reduced glucose concentration, increased carbon dioxide and lactate levels, and decreased pH. Diseases known to depress pleural fluid glucose (less than 60 mg/100 mL) include infectious causes, collagen vascular diseases, malignancies, and esophageal rupture.

Most commonly performed, but least helpful, is the white blood cell count. Although counts are generally higher (greater than 10,000/mL) in children with purulent effusions and lower (less than 1,000/mL) in clear transudates, values overlap considerably. In large series of both adult and pediatric patients, pleural fluid white blood cell counts have not been helpful in narrowing the differential diagnosis or in determining the need for or duration of closed chest tube drainage.

Needle biopsy of the parietal pleura, although easily performed at the bedside, is not generally performed in the ED. Complications include pneumothorax, hemothorax, and rarely, spread of infection or tumor to the chest wall.

Obstructive Sleep Apnea

Background

Obstructive sleep apnea (OSA) is often discussed in adult medical journals but receives little attention in pediatric literature. Although the cause of this disorder is incompletely understood, OSA is most likely a multisystemic result of respiratory failure, involving airway structure and dynamics, autonomic nervous system function, the cardiovascular system, and other systems. Because of this systemic interaction, OSA could fall within the confines of several chapters within this text. It is reasonable to suggest that OSA in infants represents an example of a mixed apnea syndrome, with a significant central component. As experience with this disorder in older children increases, understanding of its cause and management should improve.

The sleep apnea syndrome is one with a rich history of description in both the medical and lay literature. The term *Pickwickian* was coined by William Osler in 1918, referring to obese, hypersomnolent patients. This association stems from the description of an incredibly fat boy with persistent somnolence in Charles Dickens' *Posthumous Papers of the Pickwick Club.*

Numerous other accounts of sleep apnea associated with obesity have since appeared in international literature. Individual descriptions of afflicted patients resulted in a profusion of named syndrome types. Confusion in nomenclature and incomplete understanding of the pathophysiology of these disorders have thwarted the accumulation of good epidemiologic data regarding this syndrome in older patients. Obesity-hyperventilation is a rarely reported syndrome in children. The recent literature contains few large reviews. Larger series of OSA patients have appeared.

OSA that fulfills the accepted criteria for apnea (cessation of air flow at the nose and mouth for 10 seconds or longer despite continued respiratory effort) can occur in children. The exact incidence is unknown, but it is likely that OSA in children is more common than previously suspected. The wide range and frequent absence of symptoms in the awake child can make the diagnosis of OSA in this age group difficult.

Pathophysiology

Three main types of apnea have been described: central, mixed, and obstructive. Apnea affecting infants is discussed in Chapter 10. In older patients, a central sleep apnea syndrome may develop into a mixed or obstructive syndrome during the natural evolution of the disorder.

As mentioned previously, the exact pathophysiology of OSA is unclear. Diaphragmatic electromyelographic (EMG) studies of obese patients have shown inhibition of activity during initial segments of apnea. In obstructive cases, increased diaphragmatic activity has been noted after one or two breaths. Partial obstruction also stimulates the autonomic and central nervous systems, resulting in disturbed sleep. Partial obstruction causes the diaphragm to work harder to maintain normal air exchange. In children, these increased inspiratory efforts may result in daytime complaints and nighttime symptoms.

Children with craniofacial anomalies are at higher risk for obstruction shortly after birth and can develop obstructive sleep disorders and OSA. In children whose craniofacial anomalies reduce the size of the nasal cavity, nasopharynx, or oropharynx, the normal pharyngeal tissues or minimal hyperplasia of the lymphoid tissue of Waldeyer ring can cause varying degrees of obstruction, including sleep apnea.

Other children with abnormal neuromuscular systems are also prone to obstruction and sleep apnea. In these children, the pharyngeal tissues from the nasopharynx to the hypopharynx may collapse during sleep. Children with neuromuscular disorders often have chronic pulmonary disease and impaired ability to clear pharyngeal secretions, which further complicates their evaluation and management.

Clinical Manifestations

Because a child's age will influence the interpretation of certain complaints, symptoms associated with different diseases can be difficult to quantify. Guilleminault et al. were able to review a large group of children diagnosed with OSA and report their presenting features. Interestingly, many of their referrals came from school teachers and other others who were not the parents charged with the children's daytime care. Most (73%) were noted to have abnormal daytime sleepiness. Other commonly associated behavioral abnormalities included hyperactivity, continuous fighting with peers, crying easily (especially in younger children), short attention span, and quick shifts from hyperactivity to excessive somnolence and withdrawal behavior.

Older children complained of sleepiness, tiredness, and fatigue. Decreased school performance, especially with regard to language acquisition, was seen in some children. One-fourth of patients reported morning headaches, and more than half demonstrated signs of failure to thrive. Severe symptoms included massive obesity in 11%, hypertension in 8%, and acute cardiac or cardiorespiratory failure in 17%.

Disordered sleep is the most common symptom of OSA. All children report continuous snoring, which is interspersed with pauses and snorts. More than 80% have disrupted nocturnal sleep (e.g., nightmares, night terrors, sleepwalking), and 90% sweat profusely during the night. Intermittent or nightly enuresis is also commonly noted (26%). Chronic nighttime cough may also be observed as a result of intermittent aspiration of small amounts of pharyngeal secretions and may aggravate other chronic conditions such as asthma.

Adult patients generally complain of daytime tiredness, fatigue, and sleepiness. Other common symptoms include deterioration of memory and judgment, nausea, headaches, mood swings, and neurotic behavior. Polycythemia, hypertension, cardiac arrhythmias, anoxic seizures, gastroesophageal reflux, and esophagitis are less common associated serious consequences. Two-thirds of one group of adult patients were reported to be overweight (more than 20% over estimates of ideal weight). This presentation is occasionally seen in children, but most are not obese. Because it is difficult for affected children to eat and breathe at the same time, they are often in the lower 25th percentile by weight and appear to have failure to thrive.

Management

Any child with a definite history of apnea must be managed with caution and concern. Hospitalization in a monitored setting is the rule. At the time of presentation, children might not appear in extremis, by definition, during nonwakeful periods. However, these children run the risk of repeated apnea and the related complication of subsequent hypoxemia. As always, acute obstruction should be managed according to Advanced Cardiac Life Support (ACLS) protocols. The determination of the exact cause of disease is of secondary importance.

In addition to a thorough general medical evaluation, children with OSA generally require otolaryngologic consultation for airway evaluation and, if needed, planning for relief of obstruction. Many children with OSA have preexisting anatomic airway abnormalities (e.g., facial dysmorphia syndromes or enlarged lymphoid tissue) that require the expertise of an otolaryngologist.

Polysomnography is an accepted method for evaluating apnea; however, it is inconvenient and expensive. This method of evaluation is generally reserved for the most severely obstructed patients. Roentgenograms of the neck taken with the child lying on his or her back can be helpful, but are not a definitive method of diagnosis. Both radiographs and physical examination of the pharynx have similar limitations in that the dynamics of the tissues at night cannot be observed. Standardized recordings of breathing sounds can also be taken either in the hospital or in the patient's home. These tapes can be evaluated for the quality of respiratory noise and presence of interrupted breathing. The lack of measurement of respiratory effort is usually not a problem because most older children with OSA have associated loud snoring respirations.

Numerous methods of management of OSA have been suggested and studied. Patients with obesity-hypoventilation syndrome (Pickwickian syndrome) may benefit from maintained weight reduction. Unfortunately, weight loss in these patients often is either difficult to achieve or only temporarily successful. For some such patients, nasally administered CPAP successfully alleviates hypoventilation. Trials of progesterone and protriptyline have also been reported to have some degree of success. Occasionally, patients require artificial airway placement and/or supplemental ventilation.

Patients with hyperplasia of the tonsils and adenoids as their only cause for obstruction will often have dramatic relief of symptoms following tonsillectomy and adenoidectomy. Patients with craniofacial anomalies and with neuromuscular disorders also usually have significant improvement after adenotonsillectomy. Other surgical procedures can be helpful in relieving obstruction in children with craniofacial anomalies or neuromuscular degenerative disorders.

Unfortunately, patients with obesity-hypoventilation syndrome often have multiple system dysfunction. By the time many patients present for treatment, they have significant airway and weight disorders, as well as coexistent motor, learning, and psychological dysfunction. Successful therapy depends on coordinated psychiatric, medical, and rehabilitative services.

In adults with primary snoring or OSA, oral appliances have been shown to improve snoring and reduce apnea and hypopnea. The appliances modify the upper airway by changing the posture of the mandible and tongue. Although appliances are generally tolerable, oral discomfort probably decreases compliance rates, which vary from 50 to 100%. Nevertheless, in adults, oral appliances seem to present a useful alternative to CPAP, especially for those patients who cannot tolerate CPAP or who are not candidates for tonsillectomy, adenoidectomy, craniofacial surgery, or tracheostomy.

Suggested Readings

ACUTE RESPIRATORY FAILURE

Centers for Disease Control and Prevention. US Health Statistics Web Site—Mortality Tables by ICD-9 code by age—1995.

Cherniack RM, Cherniack L. Respiration in Health and Disease. Philadelphia: WB Saunders, 1983.

Martin LD, Bratton SL, Walker K. Principles of respiratory support and mechanical ventilation. In: Rogers MC, ed. Textbook of Pediatric Critical Care. 3rd ed. Baltimore: Williams & Wilkins, 1996:265–330.

Mellins RB, Stripp B, Taussis LM, eds. Pediatric pulmonology in North America—coming of age. Am Rev Respir Dis 1986;134:849–853.

Murray JF. The Normal Lung. 3rd ed. Philadelphia: WB Saunders, 1986.

Shann F, Barker J, Poore P. Clinical signs that predict death in children with severe pneumonia. Pediatr Infect Dis J 1989;8:852–855.

BRONCHOPULMONARY DYSPLASIA

Farrell PM, Avery ME. State of the art. Am Rev Respir Dis 1975;111:657–688.

Goldson E. Bronchopulmonary dysplasia. Pediatr Ann 1990;19:13–18.

Groothuis JR, Gutierrez KM, Lauer BA. Respiratory syncytial virus infection in children with bronchopulmonary dysplasia. Pediatrics 1988;82:199–203.

Groothuis JR, Simoes EA, Hemming UG. RSV infection in preterm infants and the protective effect of RSV immune globulin (RSVIG). Pediatrics 1995;95:463.

Hazinski TA. Bronchopulmonary dysplasia. In: Chernick V, Boat TF, eds. Kendig's Disorders of the Respiratory Tract in Children. 6th ed. Philadelphia: WB Saunders 1998:364–387.

Kirkpatrick BV, Mueller DG. Bronchopulmonary dysplasia. In: Kendig E, Chernick V, eds. Disorders of the Respiratory Tract in Children. 4th ed. Philadelphia: WB Saunders, 1983:235–242.

Myers MG, McGuinness GA, Lachenbruch PA, et al. Respiratory illness in survivors of infant respiratory distress syndrome. Am Rev Respir Dis 1986;133:1011–1018.

Northway WJ Jr, Rosan RC, Porter DY. Pulmonary disease following respiratory therapy of hyaline membrane disease. N Engl J Med 1967;276:357–368.

O'Brodovich HM, Mellins RB. State of the art: bronchopulmonary dysplasia (unresolved neonatal acute lung injury). Am Rev Respir Dis 1985;132:694–709.

INTERSTITIAL PNEUMONIA

Bokulic RE, Hilman BC. Interstitial lung diseases in children. Pediatr Clin North Am 1994;41:543–567.

Carrington CB, Gaensler EA, Contu RE, et al. Natural history and treated course of usual and desquamative interstitial pneumonia. N Engl J Med 1978;298:801–809.

Fan LL, Lung MC, Wagener JS. The diagnostic value of bronchoalveolar lavage in immunocompetent children with chronic diffuse pulmonary infiltrates. Pediatr Pulmonol 1997;23:8–13.

Hauger SB, Powell KR. Infectious complications in children with HIV infection. Pediatr Ann 1990;19:421–433.

Jackson LK. Idiopathic pulmonary fibrosis. In: Fulmer JD, ed. Clinics in Chest Medicine: Interstitial Lung Diseases. Philadelphia: WB Saunders, 1982:579–592.

Rothenberg SS, Wagener JS, Chang JH, et al. The safety and efficacy of thoracoscopic lung biopsy for diagnosis and treatment in infants and children. J Pediatr 1996;31:100–103.

Wolff LJ, Bartlett MS, Baehner RL, et al. The causes of interstitial pneumonitis in immunocompromised children: an aggressive systematic approach to diagnosis. Pediatrics 1977;60:41–45.

ASPIRATION PNEUMONIA

Brook I, Finegold SM. Bacteriology of aspiration pneumonia in children. Pediatrics 1980;65:1115–1120.

Brook I. Aspiration pneumonia in institutionalized children. Clin Pediatr 1986;20:117.

Brook I. Percutaneous transtracheal aspiration in the diagnosis and treatment of aspiration pneumonia in children. J Pediatr 1986;96:1000.

Brook I. Treatment of aspiration of tracheostomy-associated pneumonia in neurologically impaired children: effect of antibioaucrobials effective against anaerobic bacteria. Int J Pediatr Otorhinolaryngol 1996;35:171–177.

Bynum LJ, Pierce AK. Pulmonary aspiration of gastric contents. Am Rev Respir Dis 1976;114:1129.

Downs JB, Chapman RL, Modell JH, et al. An evaluation of steroid therapy in aspiration pneumonitis. Anesthesiology 1974;40:129.

Hickling KG, Howard R. A retrospective survey of treatment and mortality in aspiration pneumonia. Intensive Care Med 1988;14:617–622.

Kirsch CM, Sanders A. Aspiration pneumonia, medical management. Otolaryngol Clin North Am 1988;21:677–689.

Murray HW. Antimicrobial therapy in pneumonia aspiration. Am J Med 1979;66:188.

Platxker ACG. Gastroesophageal reflux and respiratory syndromes. In: Chernick V, Boat TF, eds. Kendig's Disorders of the Respiratory Tract in Children. 6th ed. Philadelphia: WB Saunders, 1998:584–600.

Wynne JW, Modell JH. Respiratory aspiration of stomach contents. Ann Intern Med 1977;87:466.

PULMONARY EMBOLISM

Becker RC, Graor R, Holloway J. Pulmonary embolism: a review of 200 cases with emphasis on pathophysiology, diagnosis, and treatment. Cleve Clin Q 1984;51:519.

Bernstein D, Coupey S, Schonberg SK. Pulmonary embolism in adolescents. Am J Dis Child 1986;140:667.

Buck JR, Connors RH, Coon WW, et al. Pulmonary embolism in children. J Pediatr Surg 1981;16:385.

Goddard R, Scofield RH. Right pneumothorax with $S_1Q_3T_3$ electrocardiogram pattern usually associated with pulmonary embolus. Am J Emerg Med 1997;15:310–312.

Gouldin CW, DiGiulio GA, Gonzalez del Rey JA. Pulmonary embolism in children and adolescents. Ambul Child Health 1997;3:240.

McBride WJ, Gadowski GR, Keller MS, et al. Pulmonary embolism in pediatric trauma patients. J Trauma 1994;37:913–915.

Moser KM. Diagnosis and management of pulmonary embolism. Hosp Pract 1980;15(10):57.

Nguyen LT, Laberge J. Spontaneous deep vein thrombosis in childhood and adolescence. J Pediatr Surg 1986;21:640–643.

Nuss R, Hays T, Chudgar U, et al. Antiphospholipid antibodies and coagulation regulatory pattern in children with pulmonary embolism. J Pediatr Hematol Oncol 1997;19:202–207.

Sasahara AA, Sharma GVRK, Barsamian EM, et al. Pulmonary thromboembolism: diagnosis and treatment. JAMA 1983;249:2945.

PULMONARY EDEMA

Carter EP, Matthay M. Resolution of pulmonary edema: recent advances. In: George RB, Campbell GD, Jenkinson SG, Matthay RA, eds. Current pulmonary and critical care medicine. Vol. 17. St Louis: Mosby, 1996:351–376.

Hopkins RL, Levine, SD. Severe pulmonary edema in meningococcemia. Clin Pediatr (Phila) 1983;22:452.

Kanter RK, Watchko JF. Pulmonary edema associated with upper airway obstruction. Am J Dis Child 1984;138:356.

Mellins RB, Stalcup SA. Pulmonary edema. In: Kendig EL, Chernick V, eds. Disorders of the Respiratory Tract in Children. 4th ed. Philadelphia: WB Saunders, 1983.

Mulroy JJ, Mickell JJ, Tong TK, et al. Postictal pulmonary edema in children. Neurology 1985;35:403.

Robin ED, Coss CE, Zelis R. Pulmonary edema. N Engl J Med 1973;288:292.

Sarniak AP, Lieh-Lai M. Adult respiratory distress syndrome in children. In: Wilmott RW, ed. Pediatric Clinics of North America. Philadelphia: WB Saunders, 1994:337–363.

Scoggin CH, Myers TM, Reeves JT, et al. High-altitude pulmonary edema in the children and young adults of Leadville, Colorado. N Engl J Med 1977;297:1269.

Sofer S, Jacob B, Scharf SM. Pulmonary edema following relief of upper airway obstruction. Chest 1983;83:401.

Theodore J, Robin ED. Speculations on neurogenic pulmonary edema (NPE). Am Rev Respir Dis 1976;113:405.

PULMONARY HEMOSIDEROSIS

Boat TF. Pulmonary hemorrhage and hemoptysis. In: Chernick V, Boat TF, eds. Kendig's Disorders of the Respiratory Tract in Children. 6th ed. Philadelphia: WB Saunders, 1998:623–633.

Dearborn DG. Pulmonary hemorrhage in infants and children. Curr Opin Pediatr 1997;9:219–224.

Levy J, Wilmott RW. Pulmonary hemosiderosis. Pediatr Pulmonol 1986; 2:384–390.

PLEURITIS

Bechamps GJ, Lynn HB, Wenzl JE. Empyema in children: review of Mayo Clinic experience. Mayo Clin Proc 1970;45:43.

Blackmore CC, Black WC, Dallas RV, et al. Pleural fluid volume estimation: a chest radiograph prediction rule. Acad Radiol 1996;3:103–109.

Foglia RP, Randolph J. Current indications for decortication in the treatment of empyema in children. J Pediatr Surg 1987;22:28–32.

Freij BJ, Kusmiesz H, Nelson JD, et al. Parapneumonic effusions and empyema in hospitalized children: a retrospective review of 227 cases. Pediatr Infect Dis 1984;3:578.

Johnson RF, Dovnarsky JH. Pleural diseases. In: Fishman AP, ed. Pulmonary Diseases and Disorders. New York: McGraw-Hill, 1980.

Light RW. Pleural effusions. Med Clin North Am 1977;61:1339.

Light RW, MacGregor MC, Luchsinger PC, et al. Pleural effusions: the diagnostic separation of transudates and exudates. Ann Intern Med 1972;77:507.

Klein JS, Schultz S, Heffner JE. Interventional radiology of the chest: image-guided percutaneous drainage of pleural effusions, lung abscess, and pneumothorax. Am J Roentgenol 1995;164:581–588.

McLaughlin FJ, Goldmann DA, Rosenbaum DM, et al. Empyema in children: clinical course and long-term follow-up. Pediatrics 1984;73:587.

Pagtakhan RD, Chernick V. Pleurisy and empyema. In: Kendig EL, Chernick V, eds. Disorders of the Respiratory Tract in Children. 4th ed. Philadelphia: WB Saunders, 1983.

Sahn SA. The pathophysiology of pleural effusions. Ann Rev Med 1990; 41:7–13.

Swischuk LE. Emergency Radiology of the Acutely Ill or Injured Child. Baltimore: Williams & Wilkins, 1986.

Vives M, Porcel JM, de Vera MV, et al. A study of Light's criteria and possible modifications for distinguishing exudative from transudative pleural effusions. Chest 1996;109:1503–1507.

OBSTRUCTIVE SLEEP APNEA

Boxer GH, Bauer AM, Miller BD. Obesity-hypoventilation in childhood. J Am Acad Child Adolesc Psychiatry 1988;27:552–558.

Boxer GH, Miller BD. Treatment of a 7-year-old boy with obesity-hypoventilation (Pickwickian syndrome) on a psychosomatic inpatient unit. J Am Acad Child Adolesc Psychiatry 1987;26:798–805.

Guilleminault C. Obstructive sleep apnea: the clinical syndrome and historical perspective. Med Clin North Am 1985;69:1187–1203.

Guilleminault C, Korobkin R, Winkle R. A review of 50 children with obstructive sleep apnea syndrome. Lung 1981;159:275–287.

Potsic WP. Obstructive sleep apnea. In: Grundfast KM, ed. Pediatric Clinics of North America. Philadelphia: WB Saunders, 1989:1435–1442.

Schmidt-Nowara W, Lowe A, Wiegand L, et al. Oral appliances for the treatment of snoring and obstructive sleep apnea: a review. Sleep 1995; 18:501–510.

CHAPTER 96

Cystic Fibrosis

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Cystic fibrosis (CF) is the most common lethal inherited disease among Caucasians in the United States. CF is a generalized defect in all of the exocrine gland secretions. CF is inherited as an autosomal recessive condition. The CF gene and the most common mutation that causes CF have recently been identified. Most homozygotes for the disease have the “classical triad” of clinical findings: 1) chronic pulmonary disease, 2) malabsorption secondary to pancreatic insufficiency, and 3) elevated concentration of sweat electrolytes. However, it must be emphasized that the severity and the course of the disease vary greatly.

Whether CF is mild or severe, its course is generally a chronic progression of the pulmonary disease, and the severity usually correlates with the rate of progression. Many more CF patients are now surviving to adulthood and are leading active and productive lives. Several factors contribute to this improving survival rate, including more effective antibiotics, earlier diagnosis, and comprehensive care in CF centers. Another important factor in the improving outlook for CF patients is the prompt recognition and aggressive treatment of the serious, acute complications that can occur in this chronic disease.

Because CF affects the exocrine glands distributed throughout the body, it is understandable that such a wide variety of symptoms in several different organs can be associated with a single disease entity. It is impossible to discuss all features of the disease here, many of which are chronic rather than acute. However, several of the more common and severe symptoms of CF that are likely to be seen by a physician in an emergency department are listed in Table 96.1. A discussion of the clinical manifestations and treatment of these complications of CF form the basis of this section. If any of these conditions are present in a patient not previously diagnosed as having CF, the patient should be referred for diagnostic evaluation after treatment of the acute episode. The evaluation should include a sweat test performed using the quantitative pilocarpine iontophoresis method.

Meconium ileus	Pneumothorax
Rectal prolapse	Hemoptysis
Intestinal obstruction	Pulmonary exacerbation
Hypoelectrolytemia with metabolic	Cor pulmonale
alkalosis	Respiratory failure

Table 96.1. Common Manifestations of Cystic Fibrosis Requiring Emergency Interventions

PATHOPHYSIOLOGY

The exocrine glands, each of which performs a specialized function, are primarily affected in CF. Although the CF gene has been identified, the precise function of the gene product remains to be elucidated; therefore, it is not yet possible to describe precisely the pathogenesis of this complex disorder. Although it involves an enormous oversimplification, it is useful to consider the pathogenesis of symptoms in two large categories. First, viscous secretions result in obstructive phenomena in the respiratory and gastrointestinal tracts. Second, altered reabsorption results in electrolyte losses in the sweat glands.

The abnormally viscous mucous secretions and the chronic colonization of the respiratory tract with bacterial pathogens, predominantly *Staphylococcus aureus* and *Pseudomonas aeruginosa*, appear to be the major contributing factors in the progressive deterioration of pulmonary function that is characteristic of CF. The interplay of these two factors, mucous plugging and infection, produces a variable amount of hyperinflation, bronchiectasis, and atelectasis. Increasing ventilation-perfusion abnormalities and structural changes lead to chronic pulmonary insufficiency. Most CF patients

eventually die of respiratory failure complicated by cor pulmonale.

Pancreatic insufficiency occurs with the obstruction and dilation of pancreatic ducts and the production of viscous, low-volume, bicarbonate and enzyme-deficient pancreatic secretions. Abnormal intestinal mucins and biliary tract secretions have also been implicated in the intestinal malabsorption and obstruction seen in CF.

The high concentrations of sodium and chloride in the sweat of CF patients can lead to acute or chronic electrolyte depletion. The elevated sweat electrolytes are the most important criteria for establishing the diagnosis of CF.

Clinical Manifestations

Presentation

Patients who have CF but who have not yet been diagnosed with CF may present with a variety of chronic symptoms. Failure to thrive and a history of chronic respiratory and/or gastrointestinal symptoms are fairly typical. The respiratory symptoms may vary from a mild but persistent cough to recurrent pneumonia and atelectasis. Expiratory rhonchi and low-pitched wheezes are sometimes found on auscultation of the chest in CF patients. The atypical asthmatic who has digital clubbing, bronchiectasis, or a cough productive of purulent sputum may also have CF.

Frequent passage of pale, bulky, loose, and excessively foul-smelling stools is characteristic of CF. Patients with this presentation are often misdiagnosed as having chronic diarrhea or a milk allergy. The loose stools often prompt repeated formula changes for young children. Edema and hypoproteinemia may develop in children with CF and especially in those who are receiving a soy protein formula. In addition, a hemorrhagic diathesis resulting from vitamin K malabsorption has been reported.

In contrast to those patients with the more acute manifestations listed in Table 96.1, many patients with chronic symptoms will not require emergency treatment. However, proper management requires that they be referred for further evaluation, which should include a quantitative pilocarpine iontophoresis sweat test.

Meconium Ileus

CF often presents as intestinal obstruction secondary to meconium ileus in the neonatal period. A typical history is that after the first few feedings the infant develops abdominal distension and begins vomiting. The child usually has a history of passing little or no meconium stool. In addition to the obvious abdominal distension, peristaltic waves may be seen on the abdomen and a mass may be palpable. Three-view radiographic examination of the abdomen should be obtained promptly. The typical findings of uncomplicated meconium ileus include dilated loops of bowel and a bubbly, granular density in the lower abdomen. In many cases, air–fluid levels will not be seen (Fig. 96.1A). If associated signs of intestinal perforation are apparent, such as calcifications or free air in the abdomen, a laparotomy will be necessary. If no signs of perforation exist, a radiographic examination following contrast enema will typically show a microcolon of disuse and impacted meconium in the terminal ileum (Fig. 96.1B). Other abnormalities or complications such as volvulus can usually be seen on the barium enema. If complications exist or if the physician has some doubt about the diagnosis, laparotomy should be performed.

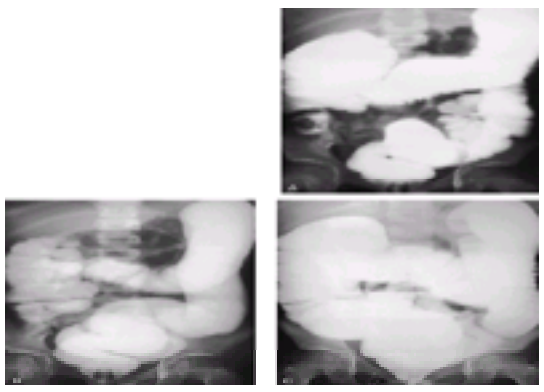

FIGURE 96.1. Distal intestinal obstruction syndrome (DIOS). **A.** Presenting Gastrografin enema of a child who had crampy abdominal pain and a right lower-quadrant mass. Fecal impaction with intussusception is demonstrated. **B.** Partial resolution of the obstruction after Gastrografin administration. **C.** Complete resolution of the intussusception and fecal impaction.

In cases of uncomplicated meconium ileus, an enema with diatrizoate methylglucamine (Gastrografin) can be used to clear the obstructing meconium, and surgery may not be necessary.

Rectal Prolapse

Rectal prolapse occurs most commonly in children less than 3 years old. Although several other conditions may cause a rectal prolapse, the association with CF is common and a sweat test should be performed on a child who has had rectal prolapse. In a child who is known to have CF, rectal prolapse usually results when pancreatic enzyme therapy has been inadequate. Although it may be frightening in appearance, the prolapse can easily be reduced by placing the infant in a comfortable position and using a lubricated glove for manual reduction. It is only in the unusual situation when an

intussusception is responsible for the prolapse that bowel strangulation may occur.

Intestinal Obstruction

Acute or chronic crampy abdominal pain is common in CF patients, and an associated fecal mass in the right lower quadrant is often present. Some patients with this history may present with signs and symptoms of intestinal obstruction, and roentgenograms of the abdomen may show dilated loops of bowel with air–fluid levels. Intestinal obstruction occurring beyond the neonatal period in patients with CF is often referred to as meconium ileus equivalent. It has been suggested that the abnormal intestinal mucus in CF patients causes a decreased motility that, combined with a decreased amount of abnormal pancreatic and biliary secretions, results in dry, puttylike stool that cannot pass from the terminal ileum to the cecum. In its mildest form, this situation may be responsible for intermittent abdominal pain. Eventually the fecal mass may cause an obstruction or serve as a leading edge for either an intussusception or a volvulus.

When the roentgenogram of the abdomen shows signs of obstruction, such as dilated loops of bowel and air–fluid levels, a barium or diatrizoate methylglucamine enema must be performed. If a nonreducible volvulus or intussusception is seen, emergency surgery is necessary. In some cases, an associated intussusception may be reduced by using diatrozoate methylglucamine as the contrast agent (Fig. 96.1A,Fig. 96.1B,Fig. 96.1C). If only a fecal mass is present without an associated volvulus or intussusception, medical management using diatrizoate methylglucamine and saline enemas usually results in dissolution of the impacted feces. Because diatrizoate methylglucamine has a very high osmolarity, the infant must be well hydrated before, during, and after the procedure. Fluid balance and serum electrolytes must be monitored closely. A useful technique is to instill a small amount of the diatrizoate methylglucamine as a bolus followed by the diluting water. This procedure often achieves a good result while using a lower total osmotic load. If some progress is made with the first diatrizoate methylglucamine enema, the procedure may be repeated. Pressure (other than hydrostatic) should not be exerted to instill the diatrizoate methylglucamine. External pressure to the infant's abdomen is also contraindicated. These procedures should be performed in consultation with a surgical team so that they may be prepared to intervene. In cases of fecal impaction without complete obstruction, enemas and oral administration of mineral oil and *N*-acetylcysteine (30 mL in 30 mL of cola) have been reported to be effective.

Hypoelectrolytemia and Metabolic Alkalosis

Especially during periods of hot weather, the increased loss of sodium and chloride in the sweat of patients with CF may lead to severe and symptomatic electrolyte depletion. Examples of the electrolyte abnormalities that were seen in two infants are shown in Table 96.2. The first patient was known to have CF. An intercurrent upper respiratory tract infection occurred during hot weather, and a decrease in oral intake was followed by profound lethargy. Several features of the electrolytes are characteristic of the abnormalities seen in CF. The extremely low chloride and elevated bicarbonate combined with a less severe hyponatremia probably reflect a renal compensation for the increased salt loss in the sweat of CF patients. The second patient presented with a history of an upper respiratory tract infection that progressed to bilateral upper lobe pneumonia and atelectasis during a period of warm weather. Again, an abrupt decrease in oral intake was followed by lethargy. In this second patient who was not previously diagnosed as having CF, the electrolytes provided an important clue for the subsequent diagnosis. In these patients, prompt fluid replacement with isotonic saline is critical; 20 to 30 mL/kg should be given within 15 minutes if signs of shock are present or within 1 hour in less severely ill patients. Potassium chloride should be administered as soon as urine output is established. However, the concentration of potassium should not exceed 40 mEq/L. Frequent determinations of serum electrolytes will be necessary to guide further therapy until correction is complete.

		Serum Electrolytes (mEq/L)				
Patient	Age (mo)	Na	K	Cl	CO_2	Serum pH
1	9	123	2.2	49	48	7.60
2	6	125	2.4	55	41	7.63

Table 96.2. Hypoelectrolytemia with Metabolic Alkalosis in Cystic Fibrosis Patients

Pneumothorax

Sudden onset of chest pain often referred to the shoulder and sometimes associated with the acute onset of increasing dyspnea and cyanosis is most likely the result of a pneumothorax in the CF patient. Rupture of a subpleural bleb introduces air into the pleural space. This complication is reported with frequency in older CF patients. It is important to realize that recurrences are very common and that tension pneumothorax has been reported in as many as 30% of these cases.

CF patients with a pneumothorax of larger than 10% of the area of the hemithorax should be treated with tube thoracostomy. This procedure should be performed promptly, but care should be taken to prepare the patient and surroundings properly and to consult an experienced physician, if one is available.

Needle aspiration of the pneumothorax should be avoided unless the patient's condition is rapidly deteriorating as the result of developing a tension pneumothorax (Fig. 96.2).

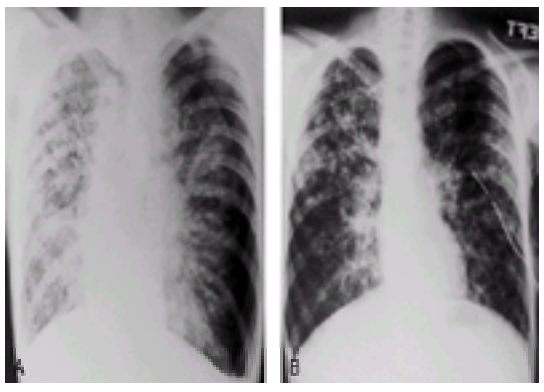

FIGURE 96.2. A. Chest roentgenogram showing a large left-sided pneumothroax with some shift of mediastinal structures of the right. **B.** Reexpansion of the left lung after tube thoracostomy and suction.

Hemoptysis

The expectoration of a small amount of blood, usually seen as bloodstreaking of the sputum, is a fairly common occurrence in CF patients. Although the first such episode may be very alarming to the patient and the parents, the patient's usual home care regimen does not need to be altered other than considering an appropriate course of antibiotic therapy to treat any intercurrent pulmonary infection.

Significant hemoptysis has been arbitrarily defined as the expectoration of at least 30 to 60 mL of fresh blood. The mechanism proposed to explain this event is the erosion of an area of local bronchial infection or bronchiectasis into a bronchial vessel. Hospitalization for observation is indicated for significant hemoptysis. Blood should be sent for a type and crossmatch in addition to hematocrit and prothrombin time determinations. Intravenous antibiotics against *Staphylococcus* and *Pseudomonas* are usually started. An intravenous line also provides an opportunity for rapid administration of saline or blood products if bleeding becomes more severe. If the patient is dyspneic, oxygen should be administered. If the prothrombin time is prolonged, vitamin K (5 mg initially) should be given. If bleeding persists, guidelines for replacement are essentially the same as for blood loss from other causes (see Chapter 59). Although it is less common, some patients with CF may have bleeding from esophageal varices secondary to advanced cirrhosis with portal hypertension. It is therefore important to establish whether the source of bleeding is from the respiratory or gastrointestinal tract. The treatment of bleeding esophageal varices is discussed in Chapter 30 and Chapter 93.

CF patients occasionally present with an episode of massive hemoptysis with volumes of blood loss ranging from 300 to 2500 mL. Massive hemoptysis represents a life-threatening situation, and in addition to instituting the measures already described, the skilled intervention of a team, including a bronchoscopist, anesthesiologist, and thoracic surgeon, may be necessary to maintain an airway and to locate and ligate the bleeding vessel. Bronchial artery embolization has been described for CF patients, and although potentially serious complications may result, this procedure may be valuable when conservative measures fail and surgery is not feasible.

Pulmonary Exacerbation

CF patients who experience an increase in respiratory symptoms such as cough and the rate and effort of breathing require careful evaluation. These symptoms often will occur after the onset of a mild upper respiratory tract infection. On physical examination, the patient will be tachypneic with intercostal retractions and may be cyanotic. Auscultation may reveal areas of coarse rales. A chest roentgenogram should be taken to determine whether pneumothorax, effusion, or local consolidation or atelectasis is present. However, in many cases, the roentgenogram will show only diffuse peribronchial thickening with a varying amount of fluffy infiltrates and hyperinflation. The roentgenogram is most helpful in assessing the degree of acute change if comparison can be made with previous roentgenograms and if medical personnel who are familiar with the patient's previous course can be contacted for advice. The establishment of the network of CF centers by the CF Foundation for the comprehensive care of CF patients has helped to ensure that such information will be available even on an emergency basis. If such guidance is not available and if lobar atelectasis, significant respiratory distress, or hypoxia (Pa O_2 less than 60 mm Hg) is present, the patient should be treated in a hospital setting with vigorous chest physiotherapy and antibiotics effective against *S. aureus* and *P. aeruginosa* (until results of sputum culture are available). Oxygen therapy should be guided by arterial blood gas determination or pulse oximetry.

Diffuse expiratory wheezing and prolonged expiration in a patient with CF suggest the possibility of coexisting asthma. A history of respiratory allergy with a good response to bronchodilators provides further support for this diagnosis. If these findings are present or if the patients show an improvement after a dose of epinephrine (1:1000, 0.01 mL/kg subcutaneously) or an inhaled bronchodilator (Albuterol 0.5%, 0.01–0.03 mL/kg 2 mL saline), therapy should be administered as outlined under Asthma in addition to treating for CF.

Cor Pulmonale

Patients with CF who have moderately severe pulmonary insufficiency and some degree of hypoxia will eventually develop right ventricular hypertrophy secondary to pulmonary hypertension. However, this condition is often not detected

by a standard electrocardiogram. Increased hypoxia during an exacerbation of pulmonary symptoms in such patients may precipitate an episode of congestive heart failure. In addition to cyanosis, tachypnea, and tachycardia, other associated signs are an enlarged, tender liver and in some patients a gallop rhythm, peripheral edema, and ascites. Most of these patients will have pronounced digital clubbing, which reflects the severity of their pulmonary disease. Rather than the elongated, narrow cardiac silhouette usually seen in the patient with CF, the chest roentgenogram will now show some cardiac enlargement with a prominence of the pulmonary vasculature. Oxygen and diuretics (furosemide 1 mg/kg given intravenously as an initial dose) have been most helpful in addition to starting treatment for the underlying pulmonary disease. Digitalis and pulmonary vasodilators have not been shown to be of proven benefit. However, many CF centers use digitalis during an acute episode of congestive failure and in selected CF patients (e.g., those with recurrent episodes of congestive heart failure and/or significant left ventricular dysfunction in addition to right ventricular dysfunction on echocardiography).

Some consideration must be given to the course of the patient's disease before the current episode when anticipating the response to therapy. The patient with a first episode of congestive heart failure precipitated by an acute exacerbation of the pulmonary disease may improve appreciably. However, in a patient in whom the congestive heart failure is superimposed on a course of inexorable deterioration in pulmonary status, a dramatic response is unlikely.

Respiratory Failure

When a CF patient presents with respiratory failure (i.e., hypercarbia—Pa CO_2less than 60 mm Hg) in addition to hypoxia, the management decisions become extremely difficult. CF patients in general do not respond as well to and have more complications from mechanical ventilation when compared with patients with other forms of chronic obstructive pulmonary disease.

If an acute episode such as viral pneumonia or status asthmaticus precipitates respiratory failure in a CF patient who had a history of good pulmonary function before the episode, mechanical ventilation should be considered. Factors in this decision include the patient's level of activity and pulmonary function before the episode, the course of the patient's disease, and the expectations of the patient and parents. Adequate, objective guidelines are not currently available, but one large retrospective study found that a history of hypercarbia indicated a poor prognosis. It seems reasonable that good pulmonary function before the acute episode provides an opportunity for a good result. However, when mechanical ventilation is used for a patient with CF, a skilled intensive care team must be prepared for a potentially difficult course.

When respiratory failure with increasing hypercarbia occurs in a CF patient after a course of progressive pulmonary insufficiency despite adequate medical therapy, mechanical ventilation is not indicated. However, consultation with the physicians providing long-term care for the patient is important before choosing this course. This consultation should include a determination of whether or not, and for how long, a patient has been listed for lung transplantation.

Suggested Readings

Davis PB, di Sant'Agnese PA. Assisted ventilation for patients with cystic fibrosis. JAMA 1978;239:1851–1854.

Del Pin CA, Czyrko C, Ziegler MZ, et al. Management and survival of meconium ileus: a 30-year review. Ann Surg 1992;215:179–285.

Fellows K, Khaw KT, Schuster S, et al. Bronchial artery embolization in cystic fibrosis: technique and long-term results. J Pediatr 1979;95: 959–963.

Fick RB, Stillwell PC. Controversies in the management of pulmonary disease due to cystic fibrosis. Chest 1989;95:1319–1327.

Kerem BS, Rommens JM, Buchanan JA, Markiewicz D, Cox TK, Chakravarti A. Identification of the cystic fibrosis gene: genetic analysis. Science 1989;245:1073–1080.

Kotloff RM, Zuckerman JB. Lung transplantation for cystic fibrosis: special considerations. Chest 1996;109:787–789.

McLaughlin FJ, Matthews WJ, Strieder DJ, et al. Pneumothorax in cystic fibrosis: management and outcome. J Pediatr 1982;100:863–869.

Ramsey BW. Management of pulmonary disease in patients with cystic fibrosis. N Engl J Med 1996;335:179–188.

Robinson C, Scanlin TF. Cystic fibrosis. In: Fishman AP, ed. Pulmonary Diseases and Disorders. 2nd ed. New York: McGraw-Hill, 1988:1273–1294.

Ruddy R, Anolik R, Scanlin TF. Hypoelectrolytemia as a presentation and complication of cystic fibrosis. Clin Pediatr 1982;21:36–38.

Stern RC, Borkat G, Hirschfeld SS, et al. Heart failure in cystic fibrosis. Am J Dis Child 1980;134:267–272.

Stern RC, Wood RE, Boat TF, et al. Treatment and prognosis of massive hemoptysis in cystic fibrosis. Am Rev Respir Dis 1978;117:825–828.

Welsh MJ, Tsui L-C, Boat TF, et al. Cystic fibrosis. In: Scriber CL, Beaudet AL, Sly WS, et al, eds. The Metabolic and Molecular Bases of Inherited Disease. 7th ed. New York: McGraw-Hill, 1995:3799–3876.

CHAPTER 97

Endocrine Emergencies

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Because the symptoms and signs of endocrinologic disease are associated with a wide range of nonendocrinologic diseases, many endocrine conditions are not recognized in the emergency department (ED). However, to achieve a favorable outcome, endocrinologic causes should be included in the differential diagnosis of ill children and appropriate testing should be performed. In many cases, the results of specific diagnostic tests will not be available on an emergency basis, and management will have to be initiated based on a probable diagnosis. In this chapter, emphasis is placed on the clinical and laboratory findings that are helpful in recognizing endocrinologic emergencies and in effectively treating those emergencies. Table 97.1 summarizes the major clinical features, recommended investigations, and treatments of pediatric endocrine emergencies.

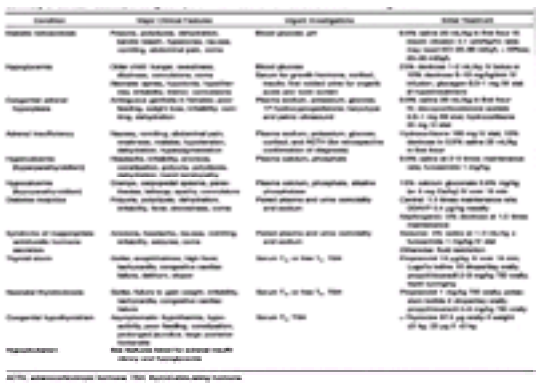

Table 97.1. Summary of Clinical Features, Investigations, and Initial Treatment of Pediatric Endocrine Emergencies

DIABETIC KETOACIDOSIS

Background

Severe ketoacidosis is a life-threatening complication of diabetes that is present in 20 to 40% of newly diagnosed juvenile-onset diabetic patients; it accounts for 65% of all admissions of diabetic patients less than 19 years old. The mortality rate for diabetic ketoacidosis (DKA) in children is less than 2% but still greater than ideal. Clinically, dehydration and acidosis are the serious immediate risks to the child. Characteristic biochemical findings include hyperglycemia and metabolic acidosis. DKA may be precipitated by acute infection, inadequate quantities of endogenous or exogenous insulin, or emotional factors. Recurrent or frequent episodes should lead to careful evaluation for this latter possibility. Nonketotic hyperosmolar coma, although rare in children, can be managed adequately using the principles outlined in the following sections.

Pathophysiology

Insulin deficiency is accompanied by elevations of the counterregulatory hormones, glucagon, catecholamines, corticosteroids, and growth hormone. These hormonal changes lead to hyperglycemia by reducing peripheral glucose use and increasing hepatic gluconeogenesis. Hyperglycemia promotes an osmotic diuresis and results in dehydration. Insulin deficiency also causes increased release of free fatty acids from adipose tissue; these fatty acids are subsequently converted into ketoacids in the liver. Ketoacids readily dissociate in the blood to produce free hydrogen

ions, and metabolic acidosis ensues. This reaction is partially compensated for by a respiratory alkalosis (hyperventilation), with a resultant lowering of P CO_2 and plasma bicarbonate (HCO^-_3).

Intracellular potassium is depleted because of transcellular shifts of this ion brought about by the acidosis and extracellular dehydration. Protein catabolism secondary to insulin deficiency causes a negative nitrogen balance and results in additional efflux of potassium from cells. The potassium is then lost in the urine during the osmotic diuresis. Volume depletion causes secondary hyperaldosteronism, which further promotes urinary potassium excretion. Thus, total body depletion of potassium occurs, although the plasma potassium concentration may not reflect the loss at the time of presentation.

Clinical Manifestations

In cases of new-onset diabetes, the child usually has a history of polyuria and polydipsia for a few days or weeks before the acute decompensation. A prolonged history of polyuria, or hyperglycemia without acidosis, should raise the possibility of type 2 diabetes, particularly in obese children from ethnic and racial minorities. Significant weight loss often occurs despite a vigorous appetite. In children known to have diabetes, the prodrome may be less than 24 hours and precipitated by an intercurrent illness, inappropriate sick-day management, or omission of insulin doses. Patients may complain of nausea, vomiting, and abdominal pain, and the parents may have noticed increasing listlessness. Fewer than 10% of children are in coma at the time of hospital admission, although a higher percentage have an altered state of consciousness. The history and physical examination usually suggest the diagnosis; however, particularly in the patient with new-onset diabetes, presenting clinical features can be misdiagnosed, especially in the infant or young child. For example, abdominal pain may be misinterpreted as appendicitis; hyperpnea may be mistaken as a sign of pneumonia or asthma; and polyuria may be incorrectly diagnosed as a urinary infection. Enuresis, polydipsia, and irritability are sometimes wrongly categorized as behavioral problems.

On physical examination, particular attention should be paid to the degree of dehydration, including skin turgor and dryness of mucous membranes. In severe cases, the child may exhibit signs of shock, including a thready pulse, cold extremities, and hypotension. The smell of ketones on the breath and the presence of deep sighing (Kussmaul) respirations reflect the ketoacidosis. The patient's consciousness level, which may range from full alertness to deep coma, should be noted. The child may have exquisite abdominal tenderness with guarding and rigidity, which can mimic an acute abdomen. The ears, throat, chest, and urine should be examined because infection is often a precipitating factor. The presence of hyperpigmentation (acanthosis nigricans) on the posterior neck should alert the clinician to the possibility of non–insulin-dependent diabetes.

Typical laboratory findings include a serum glucose greater than 200 mg/dL (usually 400 to 800 mg/dL), the presence of glucose and ketones in the urine, acidosis (venous pH less than 7.3 and HCO^-_3 less than 15 mEq/L), high or normal plasma potassium, and a slightly elevated blood urea nitrogen. Occasionally, DKA can occur with normoglycemia when persistent vomiting and decreased intake of carbohydrates are accompanied by continued administration of insulin. The serum sodium is usually low or in the low-normal range. Leukocytosis may be noted but does not necessarily signify an underlying infection. Hyperglycemia in the absence of acidosis should cause the clinician to consider additional possibilities (see subsequent section on Hyperglycemia).

Management

Careful clinical assessment and judgment is required. This section primarily focuses on the child who is significantly dehydrated, acidotic, and unable to take oral fluids because of vomiting or altered level of consciousness. Many cases of mild DKA can be managed with rehydration, either orally or intravenously, and with supplemental insulin either at home or in the ED. This possibility is addressed at the end of this section. For the severely dehydrated child, initial treatment is directed toward rapid expansion of intravascular volume and correction of the acidosis because these conditions are life-threatening. Subsequent treatment is directed at the normalization of all biochemical parameters by the use of insulin. Medical intervention carries significant risks of hypokalemia and cerebral edema (Table 97.2 and Table 97.3).

Table 97.2. Principles of Management of Diabetic Ketoacidosis

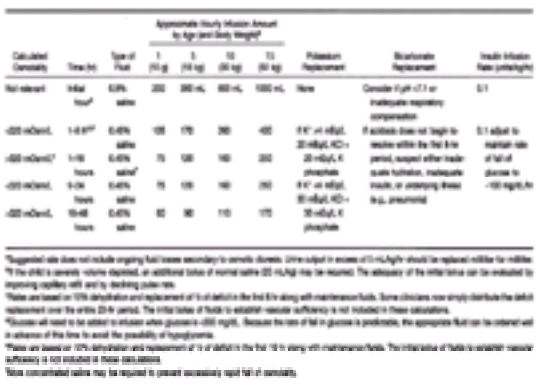

Table 97.3. Guide to Treatment of Severe Diabetic Ketoacidosis

Fluid and Electrolyte Replacement

Fluid replacement should be instituted promptly. In the first hour, isotonic (0.9%) saline should be infused intravenously at 20 mL/kg per hour to establish an adequate vascular volume and to improve tissue perfusion. This procedure may need to be repeated if the pulse rate and capillary refill rate do not decrease. Once adequate intravascular volume is established, the fluid deficit can be replaced over the next 24 to 48 hours, depending on the degree of hyperosmolality; however, once the child is able to drink, rehydration may occur enterally. It can be assumed that the fluid deficit is 10% of the body weight in children with DKA. Dehydration may actually be greater than that estimated on clinical appearance because of the hyperosmolar state. The volume of fluid given should replace the deficit (100 mL/kg), provide daily maintenance fluids (see Chapter 18 and Chapter 86), and replace ongoing urinary losses in excess of 5 mL/kg per hour (osmotic diuresis). For the child whose calculated osmolality is less than 320 mOsm/L, the deficit can be replaced over 24 hours, but a longer period (36 to 48 hours) should be used for children with higher osmolalities.

The Na^+ deficit is approximately 10 mEq/kg body weight and Na^+ maintenance is 3 mEq/100 per mL of maintenance fluid. From a practical point of view, half-normal (0.45%) saline can be started after the initial bolus of normal saline. The serum sodium should rise with initiation of therapy. If the initial serum sodium is less than 135 mEq/L, a falling serum sodium is an indication to use a more concentrated sodium stock and to increase monitoring, as well as to consider the possibility of the syndrome of inappropriate antidiuretic hormone.

All children with DKA are total body potassium depleted (5 mEq/kg body weight); therefore, potassium replacement is an important part of therapy. If the initial serum K^+ is greater than 4 mEq/L, 40 mEq/L of potassium is added to the infusion after vascular competency has been established and the child has urinated. Generally, K^+ is provided as potassium chloride (or acetate) and potassium phosphate in equal amounts. If the initial serum K^+ is less than 4 mEq/L, potassium replacement should be initiated promptly; doses of K^+ of 60 mEq/L or greater may be necessary. If the K^+ initial concentration is low, electrocardiographic (ECG) monitoring is indicated.

Phosphate depletion is almost universal in patients with DKA; however, the clinical significance of this reaction remains uncertain. As noted earlier, half of the K^+ replacement is with potassium phosphate.

Alkali

The most essential steps for the correction of acidosis are volume repletion and insulin. The use of sodium bicarbonate for correction of acidosis remains controversial because of the potential for paradoxical acidosis of the central nervous system (CNS) and resultant cerebral depression. Paradoxical acidosis occurs because administered HCO^-_3 combines with excess H^+ ions in the bloodstream to form H_2O and CO_2. Because the blood–brain barrier is relatively more permeable to CO_2 than to HCO^-_3, CO_2 accumulates in the CNS, resulting in further exacerbation of acidosis in this compartment, while acidosis is being corrected systemically. Most children will correct their acidosis during rehydration and initiation of insulin therapy without recourse to alkali. Bicarbonate therapy is generally reserved for children with an initial arterial pH of less than 7.1, and for those who are unable to compensate for their acidosis by hyperventilation. The adequacy of compensation can be evaluated if an PCO_2 and the serum $[HCO^-_3]$ are known. If the PCO_2 is in excess of $(1.5 \times HCO^-_3) + 8$, respiratory effort is inadequate for the degree of acidosis, and bicarbonate is required. The amount of bicarbonate needed may be calculated using the formula:

$$\text{Amount of bicarbonate (mEq)} = \text{Base deficit (mEq/L)} \times \text{Body weight (kg)} \times 0.6 \text{ (distribution factor for bicarbonate)}$$

Half of this amount is given intravenously over 2 hours. Biochemical studies are then repeated; the need for continued bicarbonate is reevaluated, and if necessary, a new dose of bicarbonate is calculated and administered. Alternatively, 1 to 2 mEq/kg may be given intravenously every hour until the pH is more than 7.25 and the HCO^-_3 is more than 15 mEq/L.

Insulin

Regular insulin is used for the treatment of ketoacidosis. Insulin is initially necessary to stop ongoing ketone body production, the primary cause of the acidosis. Insulin should be started at the same time as initial fluid expansion to correct the acidosis and may be either infused intravenously or injected intramuscularly at hourly intervals. Subcutaneous

injections of insulin should be avoided because of the uncertainties of absorption in a dehydrated patient. The starting dose of insulin for continuous infusion is 0.1 unit/kg per hour, which is normally infused by a regulated pump. The rate of insulin infusion should be adjusted to sustain a fall in the blood glucose of about 100 mg/dL per hour. Failure of the glucose to decrease in response to insulin suggests improper insulin preparation, inadequate hydration, or serious underlying disease (e.g., appendicitis with resultant significant increases in counterregulatory hormones). It is unnecessary to give an initial bolus of insulin. The dose for the hourly intramuscular injection is 0.25 unit/kg as a priming dose followed by 0.1 unit/kg per hour. Once the blood sugar is less than 300 mg/dL, glucose should be added to the intravenous (IV) fluids. As long as the child remains acidotic, insulin infusion should never be stopped; instead, the amount of glucose in the IV infusion should be increased and the insulin infusion adjusted to maintain the blood sugar between 100 and 200 mg/dL. When the child is able to eat and is no longer significantly acidotic, IV infusion of insulin can be discontinued. Because IV insulin is metabolized rapidly, subcutaneous insulin must be given promptly after the infusion is stopped. The initial dose of regular insulin should be about 0.25 unit/kg. If hourly intramuscular injections are used, they should be continued until the blood sugar is below 300 mg/dL and acidosis is correcting. By this time, perfusion is reestablished and subcutaneous insulin can be administered four times a day.

Monitoring

Close monitoring is mandatory, and a well-organized flow sheet ensures that all parameters are being observed. Admission to an intensive care unit is advisable if the patient is less than 1 year of age, has a Glasgow Coma Scale score of less than 12, has an initial calculated osmolality of more than 320 mOsm/L, has an initial [Na^+] of more than 145 mEq/L, or has an initial [K^+] of less than 4 mEq/L. The patient's blood pressure, pulse rate, respiratory rate, and the level of consciousness should be observed regularly. Careful neurologic examination, with particular attention to arousability and pupillary reactivity, should be performed frequently. The fluid input and output must be reviewed hourly to ensure that appropriate rehydration is occurring. The IV fluids should be checked frequently so that pump failure or fluid leakage into the subcutaneous tissues can be corrected quickly. In the severely ill child, an ECG monitor is advisable to detect arrhythmias associated with hyperkalemia or hypokalemia.

The serum glucose should be measured hourly until the blood glucose is stable and below 300 mg/dL and as long as the child is on an insulin infusion. Glucose measurement may be less frequent once the patient has been changed to subcutaneous insulin. Serum K^+ needs to be measured every 3 to 4 hours until the acidosis and hyperglycemia are normalized, or more frequently if hypokalemia is encountered or bicarbonate therapy is used. An arterial pH should be obtained before treatment and repeated if bicarbonate therapy is used or contemplated.

When the child is better hydrated and the acidosis resolves, mental alertness will improve and symptoms of nausea, vomiting, and abdominal pain should remit. If they do not resolve, an abdominal disorder should be considered. Some patients complain of blurred vision, which is caused by lens distortion resulting from fluid shifts of rehydration and correction of hyperglycemia. Twelve hours after initiation of treatment, most patients are able to tolerate oral fluids, at which point rehydration can be continued orally.

Mild Ketoacidosis

Some children with new-onset diabetes may also have hyperglycemia without ketoacidosis or with only mild acidosis. Generally, these patients are hospitalized for at least 12 to 24 hours to allow time to educate the family and stabilize the insulin dosage. These children require rehydration (as described in the next section) similar to patients with known diabetes and mild ketoacidosis. Insulin therapy can be initiated subcutaneously, at a total daily dose of 0.25 to 0.5 units/kg per day for the prepubertal child and 0.5 to 0.75 units/kg per day for the adolescent. Two-thirds of the total daily dose is administered in the morning, and one-third before dinner; two-thirds of the morning dose and one-half of the evening dose should be as an intermediate duration insulin (NPH, Lente).

Children with known diabetes often develop mild ketoacidosis during the course of intercurrent illness, especially gastroenteritis, or secondary to omission of insulin doses. Even the mildly dehydrated (5%) child with slight acidosis who presents to the ED benefits from a fluid bolus (20 mL/kg of normal saline); furthermore, this bolus will be given while awaiting laboratory test results. Once the laboratory results are available, the physician must decide whether to hospitalize the child, continue treatment in the ED, or send the child home. For purposes of definition, *mild DKA* is defined as a pH of more than 7.3, a bicarbonate of more than 15 mEq/L, and a calculated osmolality of less than 320 mOsm/L. Children who are significantly acidotic or hyperosmolar should be hospitalized and managed as outlined in the earlier section of this chapter. Several factors must be considered before sending a child home:

1. Is the child conscious and alert?
2. Can the child drink and retain oral fluids?
3. Can home glucose monitoring be done and are all related supplies available in the home?
4. Will the child have competent supervision at home?
5. Does the family have access to both a telephone and transportation?
6. Is there a physician available with whom the family can communicate by telephone?
7. Is the family comfortable with managing the mild acidosis at home?

If all of these questions can be answered in the affirmative, the child may be sent home.

Recommendations should be made to the family regarding fluid intake, insulin administration, and monitoring. Specific recommendations may vary with the age of the child and the experience of the family, but the following scheme may be helpful. Oral intake should be about the same as would be given intravenously to resolve the deficit and provide maintenance (e.g., the 10-year-old child would normally get about 260 mL/hour during the first 8 hours intravenously if he or she was hospitalized; therefore, the physician should suggest that the family try to get in about 8 ounces of liquid every hour for the next 6 to 8 hours). It is best if this liquid is taken in as sips. Additional insulin will be required. In the ED, two decisions will need to be made regarding insulin: First, how much should be given to the child before dismissal?

Usually, an amount equal to 10% of the child's usual daily dose will be adequate (e.g., a child normally takes 12 units R and 6 units N in the AM and 6 units R and 6 units N in the PM. The total daily dose is 30 units. Ten percent of 30 = 3 units. Therefore, 3 units of regular insulin would be given to the child before discharge). Second, how much should be given at home and with what frequency? Once home, the preceding 10% rule is generally applicable. If the blood glucose has not come down within 1 to 2 hours, consultation with the child's physician is recommended. If the blood glucose has decreased but is still more than 250 mg dL, a second dose of similar size is indicated. If a third home dose is contemplated, consultation with the child's physician is required. Many children have insulin adjustment algorithms for premeal adjustments from their endocrinologist. The family can begin using this algorithm once the child is able to return to a normal intake. Last, hourly monitoring of blood glucose, urine output, and ketones is recommended with the expectation that the blood glucose should decline, the urine output should fall, and the urine ketones should begin to clear.

HYPERGLYCEMIA

Background

According to new American Diabetes Association guidelines, a fasting glucose of greater than 125 mg/ dL or a random glucose greater than 199 mg/dL is highly suggestive of diabetes in an otherwise healthy person. These guidelines were developed primarily by specialists in adult diabetes and may not be completely applicable to the pediatric population, especially not infants and young children who are ill as well as distressed by the trauma of phlebotomy.

Therefore, it is essential that any elevated glucose level be evaluated within the context of the ill child and that other factors, such as simultaneous medication administration, be considered.

Pathophysiology

As noted in the previous section on diabetes and the following section on hypoglycemia, glucose homeostasis reflects the balance between glucose input (from gut absorption, hepatic glycogen breakdown, or gluconeogenesis) and disposal (via storage or oxidation). With the exception of gut absorption, this process is largely regulated by insulin, although counterregulatory hormones have a significant effect as well. Furthermore, tissue factors and medication also impact on the insulin effect.

Clinical Manifestations

Blood glucoses in the 200 to 300 mg/dL range rarely result in symptoms. This level of hyperglycemia may be accompanied by intermittently increased frequency of urination; however, parents are rarely aware of their child's frequency of urination once the child is toilet-trained unless the frequency becomes disruptive (e.g., the child begins having “accidents” at school). Children and adolescents have no sense of what is the normal frequency of urination, so they rarely complain, unless the frequent urination is accompanied by dysuria. Higher levels of glucose (greater than 300 mg/dL) may be associated with subtle clinical findings such as blurring of vision or dryness of oral membranes.

Significant hyperglycemia may occur without significant symptoms and can be tolerated for a prolonged period without clinical signs. In the ED, hyperglycemia is likely to be seen in several different situations. First, the child may be known to have diabetes and present with an intercurrent illness or traumatic injury. Both illness and injury result in increased counterregulatory hormones, which may lead to relative insulin resistance and hyperglycemia. The second presentation is the child for whom diabetes is suspected because of classical symptoms of polyuria, polydipsia, and polyphagia accompanied by weight loss. Almost half of children with new-onset diabetes mellitus present to their pediatrician or to the ED in this way. Third, some medical conditions are associated with persistent hyperglycemia, such as recurrent urinary tract infections and vaginal yeast infections. Furthermore, type 2 diabetes is increasingly being reported in minority adolescents; in many, hyperpigmentation of the posterior neck and axilla (acanthosis nigricans) may be noted. Fourth, a laboratory panel obtained for some other reason (e.g., abdominal pain) may reveal hyperglycemia.

Management

In the child with diabetes, unless the child is clinically dehydrated or is unable to take oral fluids, hyperglycemia is not a crisis. Oral fluids should be encouraged, and supplemental insulin may be required (usually 10% of the child's total daily dose, given as either regular insulin or the rapid-acting lispro insulin). Failure to respond to these simple measures, whether in the ED or at home, should lead to a consultation with the child's endocrinologist. If oral fluids must be restricted (e.g., a child with traumatic injury requiring surgery), IV fluids without glucose should be used and glucose should be monitored frequently. Supplemental insulin may be required, depending on when the child last received insulin and the response to simple hydration.

Clearly, the child with classic symptoms merits further evaluation to determine whether the child is dehydrated, hyperosmolar, or acidotic. If the child is simply hyperglycemic, hospitalization is often required for initiation of treatment and diabetes-related education; however, IV fluid resuscitation and intensive care are not indicated.

In children with signs or symptoms that may reflect hyperglycemia, a serum glucose should be obtained. If this test reveals hyperglycemia, as defined by the American Diabetes Association, the child should be referred for further evaluation and treatment.

Last, if hyperglycemia is a coincidental finding, the diagnosis requires thoughtful consideration. How traumatic was the blood draw? How upset was the child? What medications or IV fluids were given to the child just before the phlebotomy? What was the child drinking while waiting to see the physician? Are the symptoms in any way related to the hyperglycemia? Three simple evaluations are helpful in determining whether the hyperglycemia is circumstantial or suggestive of diabetes. Brief hyperglycemia resulting from a stress response to phlebotomy or secondary to oral intake

rarely results in significant glucosuria; therefore, a urine dip for glucose is often helpful. Second, in the absence of ongoing stress or input, glucose tends to fall over time. A fingerstick glucose is rarely stressful. Therefore, repeating a glucose measurement by fingerstick 1 to 2 hours after the original sample was sent is useful in separating disease from nondisease. Third, hyperglycemia secondary to these factors is usually mild (150 to 250 mg/dL). More significant hyperglycemia should raise the suspicion of diabetes or glucose intolerance.

HYPOGLYCEMIA

Background

Hypoglycemia is defined as serum glucose of less than 50 mg/L, regardless of whether symptoms are present. Hypoglycemia is a chemical finding that should lead to a diligent search for a cause. A differential diagnosis of hypoglycemia, as it may present in the ED, is provided in Table 97.4.

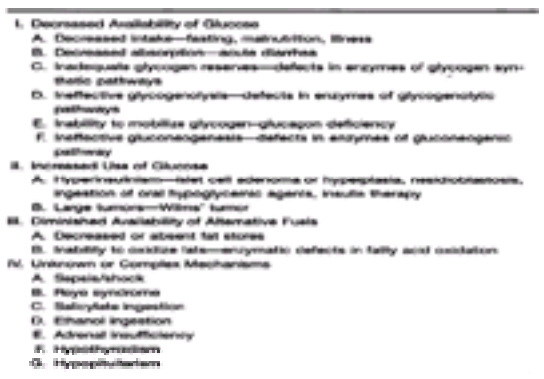
I. Decreased Availability of Glucose
A. Decreased intake—fasting, malnutrition, illness
B. Decreased absorption—acute diarrhea
C. Inadequate glycogen reserves—defects in enzymes of glycogen synthetic pathways
D. Ineffective glycogenolysis—defects in enzymes of glycogenolytic pathways
E. Inability to mobilize glycogen—glucagon deficiency
F. Ineffective gluconeogenesis—defects in enzymes of gluconeogenic pathway
II. Increased Use of Glucose
A. Hyperinsulinism—islet cell adenoma or hyperplasia, nesidioblastosis, ingestion of oral hypoglycemic agents, insulin therapy
B. Large tumors—Wilms' tumor
III. Diminished Availability of Alternative Fuels
A. Decreased or absent fat stores
B. Inability to oxidize fats—enzymatic defects in fatty acid oxidation
IV. Unknown or Complex Mechanisms
A. Sepsis/shock
B. Reye syndrome
C. Salicylate ingestion
D. Ethanol ingestion
E. Adrenal insufficiency
F. Hypothyroidism
G. Hypopituitarism

Table 97.4. Causes of Childhood Hypoglycemia

Hypoglycemia may be secondary to insulin therapy for diabetes. Excluding this category, almost all hypoglycemia in children occurs during periods of decreased or absent oral intake, often coupled with increased energy demand (e.g., viral gastroenteritis with fever). Postprandial hypoglycemia is unusual in children, except in those who have had prior gastrointestinal surgery.

Pathophysiology

Because glucose is necessary for cellular energy production in most human tissues, the maintenance of an adequate blood glucose is important for normal function. The serum glucose reflects a dynamic balance among glucose input from dietary sources, glycogenolysis and gluconeogenesis, and glucose use by muscle, heart, adipose tissue, brain, and blood elements.

The liver plays a unique role in glucose homeostasis because it stores glucose as glycogen. With fasting, this glycogen is degraded to glucose, which is released into the bloodstream. In addition, the liver synthesizes new glucose from glycerol, lactate, and certain amino acids. During fasting, lipolysis occurs and the resultant fatty acids are used for the production of both energy and ketones (acetoacetate and b-hydroxybutyrate) by the liver. The energy generated from the metabolism of fatty acids is essential to sustain maximal rates of gluconeogenesis and ureagenesis in the liver. The ketones are an important auxiliary fuel for most tissues, including the brain.

Muscle contains significant quantities of glycogen and protein. Under fasting conditions, the glycogen is degraded and used endogenously but is not released as free glucose into the bloodstream. Certain amino acids, particularly alanine and glycine, are released from the muscle and subsequently used by the liver for gluconeogenesis. Muscle derives an increasing proportion of its energy requirement from fatty acids as fasting proceeds.

Brain tissue is highly dependent on glucose for its energy requirements. Under certain circumstances, it can extract a limited proportion of its energy requirement from other substrates (e.g., glycerol, ketones, lactate), although this process requires a period of adaptation and does not negate the need for a constant supply of glucose.

Insulin is the primary hormone that regulates the blood glucose level. Insulin stimulates the uptake of glucose and amino acids into skeletal, cardiac, and adipose tissue and promotes glycogen and protein synthesis. It inhibits lipolysis and glycogenolysis. The net effect of insulin action is to accelerate the removal of glucose and gluconeogenic substrates from the bloodstream. Opposing or modulating the effects of insulin are cortisol, glucagon, epinephrine, and growth hormone. The effects of these hormones include inhibition of glucose uptake by muscle, mobilization of amino acids for gluconeogenesis, activation of lipolysis, inhibition of insulin secretion, and induction of gluconeogenic enzymes. The net effect is to increase the availability of gluconeogenic substrates to the liver, and to increase the accessibility and use of nonglucose fuels by other tissues.

Clinical Manifestations

Prompt recognition of hypoglycemia is important because brain damage may result if the hypoglycemia is prolonged or recurs frequently. The acutely ill child warrants a glucose determination if the level of consciousness is altered because hypoglycemia may accompany an illness that interferes with oral intake. Historical evidence may aid in establishing the cause of hypoglycemia. Because hypoglycemia in children occurs after a period of fasting, a careful chronology of dietary

intake during the preceding 24 hours should be obtained. The possibility of ingestion should be considered because ethanol, propranolol, and oral hypoglycemic agents are in common use.

The symptoms and signs of hypoglycemia are nonspecific and are often overlooked, especially in the infant and young child. The clinical findings of hypoglycemia reflect both the decreased availability of glucose to the CNS and the adrenergic stimulation caused by a decreasing or low blood sugar. Adrenergic symptoms and signs include palpitations, anxiety, tremulousness, hunger, and sweating. Irritability, headache, fatigue, confusion, seizure, and unconsciousness are neuroglycopenic symptoms. Any combination of these symptoms should lead to a consideration of hypoglycemia. Any child presenting with a seizure or unconsciousness should have a serum glucose determination.

Management

If hypoglycemia is suspected, blood should be obtained, if at all possible, before treatment. An extra tube (3 mL, red top) should be obtained and refrigerated until the laboratory glucose is known. Rapid screening should be performed using a portable glucose monitor while awaiting definitive laboratory results. Therapy should be instituted if this screen is suggestive of hypoglycemia. This method may lead to some overtreatment because the primary error with the chemically treated strips is underestimation of the serum glucose value; however, treatment holds minimal risk. It is preferable to overtreat than to allow a child to remain hypoglycemic until definitive laboratory results are available. If the laboratory glucose confirms that the blood glucose was less than 50 mg/dL, the reserved serum can be used for chemical (b-hydroxybutyrate, acetoacetate, free fatty acids, carnitine), toxicologic, and hormonal (insulin, growth hormone, cortisol) studies and may provide the correct diagnosis without extensive additional testing.

The first voided urine after the hypoglycemic episode should be saved for toxicologic and organic acid evaluation. In the ED, the urine can also be tested immediately for ketones. With hypoglycemia, ketones should be large. Failure to find large ketones in the presence of hypoglycemia strongly suggests either that fats are not being mobilized from adipose tissue, as might occur in hyperinsulinism, or that the fats cannot be used for ketone body formation, as might occur in enzymatic defects in fatty acid oxidation. The urine should be sent for organic acid analysis and toxicologic investigations. Both the urine and the serum results will be useful in determining the underlying cause of hypoglycemia.

The preferred treatment for hypoglycemia is 0.25 g of dextrose/kg body weight (2.5 mL/kg of 10% dextrose/kg, 1.0 mL/kg of 25% dextrose/kg) rapidly. The serum glucose should then be maintained by an infusion of dextrose at a rate of 6 to 8 mg/kg per minute. Generally, this goal can be accomplished by providing 10% dextrose at 1.5 times maintenance rates. Glucagon (1 mg intramuscularly) may be used to treat hypoglycemia that is known to be caused by hyperinsulinism but is not indicated as part of the routine therapy of hypoglycemia. Cortisol should not be used because it has minimal acute benefit and may delay identification of the cause of hypoglycemia.

The adequacy of therapy should be evaluated both chemically and clinically. The serum glucose should be monitored frequently until a stable level above 70 mg/dL is attained. Adrenergic symptoms should resolve quickly. The resolution of CNS symptoms may be prolonged, particularly if the child was initially seizing or unconscious. Seizures that do not respond to correction of hypoglycemia should be managed with appropriate anticonvulsants (see Chapter 83). The mild acidosis (pH 7.25 to 7.35) usually seen in hypoglycemia will correct without specific intervention. Marked acidosis (pH less than7.10) suggests shock or serious underlying disease and should be managed appropriately (see Chapter 3). Any child with documented hypoglycemia not secondary to insulin therapy should be hospitalized for careful monitoring and diagnostic testing.

HYPOPITUITARISM

Background

The term *hypopituitarism* generally applies to any condition in which more than a single pituitary hormonal deficiency is present. This condition may include deficiencies resulting from a lack of hypothalamic-releasing factors, as well as deficiencies of anterior and posterior pituitary hormones. Diabetes insipidus, the lack of antidiuretic hormone (ADH), may occur alone or in association with other hormonal defects and is discussed in a subsequent section.

Pathophysiology

Adrenocorticotropic hormone (ACTH) primarily affects adrenal glucocorticoid production; generally, it does not affect mineralocorticoid synthesis, which is primarily regulated by the renin–angiotensin system. A deficiency of ACTH production manifests as cortisol deficiency. Because cortisol plays a role as an insulin counterregulatory hormone, a lack of either ACTH or cortisol may result in hypoglycemia. Because the only identified role for thyroid-stimulating hormone (TSH) is the stimulation of thyroid hormone production, a deficiency of TSH is most likely to manifest as hypothyroidism. Luteinizing hormone (LH) and follicle-stimulating hormone (FSH) are involved in gonadal maturation and the regulation of gonadal functions. LH and FSH play an important role in testicular descent and penile growth in the fetus, as well as affecting the onset of puberty in the adolescent. The circulating levels of these two pituitary hormones are low in children and have no demonstrable function. Prolactin is primarily involved in the maintenance of lactation and is of minimal significance in childhood. Growth hormone is a principal regulator of linear growth and an important insulin counterregulatory hormone. The absence of growth hormone may be associated with hypoglycemia, particularly in infants and young children.

Clinical Manifestations

The symptoms and signs of hypopituitarism depend on which hormones are missing. Isolated growth hormone deficiency is most likely to present with poor linear growth, although occasionally, an infant or young child will present with hypoglycemia. The acute presentation of hypopituitarism is most likely to occur when the child is stressed by injury, illness, or fasting. The presentation may involve either an unusually rapid decompensation, reflecting the role of cortisol

in adaptation to stress, or as hypoglycemia, mirroring the role of both cortisol and growth hormone in opposing the effects of insulin. In the older child, no specific symptoms or signs indicate a lack of LH and FSH. An association between a lack of these hormones and anosmia has been noted (Kallmann's syndrome). In the adolescent, a deficiency of LH and FSH may be evidenced as pubertal delay. No specific signs or symptoms have been associated with a deficiency of prolactin in childhood.

In the neonatal male, hypopituitarism may be accompanied not only by hypoglycemia but also by micropenis (less than 2 cm, stretched length). This condition echoes the role of LH and FSH in stimulating testicular function in utero. Significant liver dysfunction in the neonatal period may be associated with congenital hypopituitarism. Hypopituitarism is seen with various midline structural anomalies, including optic nerve hypoplasia, cleft palate, absence of the septum pellucidum, and spina bifida.

In the older child, intracranial mass lesions, particularly with craniopharyngioma and other pituitary abnormalities, may cause hypopituitarism. The presence of visual field abnormalities may aid in localizing the site of the lesion. A history of severe head trauma, surgery for CNS tumors, or CNS irradiation should increase the suspicion of hypopituitarism.

Management

The child with hypopituitarism may require any or all of the following therapies. Adequate cortisol replacement is an absolute necessity in children with known or suspected secondary adrenal insufficiency (ACTH deficiency). Cortisol replacement under stress conditions (e.g., trauma, fever) should be the equivalent of 50 mg of hydrocortisone/m^2 per day (hydrocortisone semisuccinate, constant IV infusion; cortisone acetate 50 mg/m^2 intramuscularly every 24 hours; methylprednisolone 7.5 mg/m^2 per 24 hours every 8 hours). Because both cortisol and growth hormone are insulin counterregulatory hormones, children with hypopituitarism are prone to hypoglycemia. If oral intake is interrupted for prolonged periods, glucose should be supplied intravenously. Blood glucose should be monitored to ascertain the adequacy of therapy. Both adrenal insufficiency and diabetes insipidus can lead to fluid and electrolyte abnormalities; therefore, electrolytes should be determined at presentation and followed closely. Changes in IV therapy should be based on serum electrolytes. Judicious use of 1-desamino8- D-arginine vasopressin (DDAVP) may be helpful in managing diabetes insipidus, as outlined subsequently; however, this treatment is usually unnecessary at the time of acute presentation.

Although both thyroid hormone and sex hormone(s) may need to be replaced, this treatment is not required in the ED.

Because hypopituitarism often results from intracranial lesions, the demonstration of a pituitary or a hypothalamic mass, or a history of a significant cranial insult, should lead to a diligent search for hormonal deficits. Similarly, documented pituitary deficits should lead to a thorough radiologic investigation of the cranial cavity.

ACUTE ADRENAL INSUFFICIENCY

Background

Acute adrenal insufficiency occurs when the adrenal cortex fails to produce enough glucocorticoid and mineralocorticoid in response to stress. Patients at risk of this life-threatening event include individuals with primary adrenal disease and those who have adrenal insufficiency secondary to hypothalamic-pituitary suppression (Table 97.5). This emergency has become increasingly common with the widespread use of suppressive doses of corticosteroids in the treatment of chronic disease (e.g., nephrotic syndrome, acute lymphoblastic leukemia, asthma). Infection, trauma, or surgery in the susceptible patient generally precipitates the acute crisis. The diagnosis must be based primarily on clinical suspicion because prompt commencement of therapy is mandatory for survival and definitive diagnostic test results may not be available for weeks. Congenital adrenal hyperplasia is a unique form of adrenal insufficiency that is discussed in a subsequent section of this chapter.

I. Primary Adrenal Insufficiency
A. Congenital adrenal hyperplasia
B. Autoimmunity
C. Tuberculosis
D. Meningococcal septicemia
E. Adrenal hemorrhage
II. Secondary Adrenal Insufficiency
A. Suppression of adrenocorticotropic hormone by pharmacologic doses of glucocorticoid administration
B. Pituitary or hypothalamic tumors
C. Central nervous system surgery or irradiation
D. Structural abnormalities (septooptic dysplasia)
E. Congenital hypopituitarism

Table 97.5. Common Causes of Acute Adrenal Insufficiency in Children

Pathophysiology

Because the production of corticosteroids by the adrenal cortex is under pituitary and hypothalamic control, adrenal insufficiency can result from either an adrenal (primary) or hypothalamic–pituitary (secondary) disorder. Specific adrenal problems resulting in adrenal insufficiency include inborn errors of hormonal biosynthesis (discussed in the subsequent section on Congenital Adrenal Hyperplasia), autoimmune destructive processes, adrenoleukodystrophy, and adrenal

hemorrhage. Hypothalamic–pituitary causes include CNS tumors, trauma, and radiation therapy for a variety of neoplastic disorders. Exogenous administration of glucocorticoids also suppresses the adrenal–pituitary axis, an effect that often lasts well beyond the cessation of corticosteroid therapy.

Glucocorticoids are essential for withstanding stress; therefore, adrenal insufficiency is most likely to be manifested during an intercurrent infection or after trauma. Mineralocorticoids, especially aldosterone, play an important role in salt and water homeostasis by promoting salt resorption in the distal renal tubules and collecting ducts. Mineralocorticoid production is primarily regulated by the renin–angiotensin system; thus, adrenal insufficiency resulting from hypothalamic–pituitary causes is rarely associated with a lack of aldosterone. On the other hand, aldosterone deficiency is a common feature in primary adrenal insufficiency. Because of the nature of the pituitary–adrenal axis, primary adrenal insufficiency is accompanied by significantly elevated ACTH levels.

Clinical Manifestations

The historical information suggestive of adrenal insufficiency depends on the cause. Children with a primary adrenal defect are more likely to have had a gradual onset of symptoms, such as general malaise, anorexia, fatigue, and weight loss. Salt craving and postural hypotension may also have been noted. A child with secondary adrenal insufficiency is more likely to have a history of neurosurgical procedures, head trauma, CNS pathology, or chronic disease necessitating the prolonged use of glucocorticoids.

Findings on physical examination are more likely to be characteristic of the precipitating illness or trauma rather than specifically suggestive of adrenal insufficiency. Although a lack of glucocorticoid and aldosterone can be associated with hypotension and dehydration, a better clue to the possibility of adrenal insufficiency is inappropriately rapid decompensation in the face of metabolic stress. Hyperpigmentation may be present in primary adrenal insufficiency, especially of long duration.

Biochemical evidence suggestive of adrenal insufficiency includes hyponatremia, hyperkalemia, hypoglycemia, and hemoconcentration. Mild metabolic acidosis and hypercalcemia may be present. The definitive diagnosis depends on the demonstration of an inappropriately low level of cortisol in the serum. Blood should be obtained for the measurement of both cortisol and ACTH if the diagnosis is suspected, but results are unlikely to be available on an emergency basis.

Management

Treatment of adrenal crisis is based on rapid volume expansion and the administration of glucocorticoids. Immediate management consists of 50 to 100 mg of hydrocortisone intravenously. Subsequent management is hydrocortisone 50 mg/m^2 per 24 hours given continuously intravenously or methylprednisolone (Solu-Medrol) 7.5 mg/m^2 per 24 hours divided and administered every 8 hours intravenously. Volume expansion is accomplished with normal saline (20 mL/kg) in the first hour, followed by fluids appropriate for maintenance and replacement. Additional Na^+ may be needed in primary adrenal insufficiency because of ongoing urinary Na^+ losses. These fluids should contain 10% dextrose and should not contain potassium until the serum potassium is within the normal range. Mineralocorticoid therapy is rarely important in the acute phase, provided fluid therapy is adequate; however, patients with primary adrenal insufficiency may need replacement with a mineralocorticoid for long-term management. Specific therapy directed toward correction of the hyperkalemia is rarely required unless cardiac arrhythmias are present. Hypoglycemia is remedied by the use of dextrose and by the hyperglycemic effects of glucocorticoids. The precipitating factor, such as infection, also requires appropriate therapy.

Improvement in peripheral circulation and blood pressure should occur quickly. Dramatic improvement often occurs in all parameters within hours after the first dose of glucocorticoid. Because adrenal crisis is commonly brought on by another stress such as infection, the symptoms of malaise, anorexia, and lethargy may take longer to resolve. Once instituted, high-dose glucocorticoid therapy should be continued for 48 hours, and adequate hydration should be maintained either orally or intravenously. The patient known to be at risk for adrenal insufficiency should wear an identifying bracelet to alert ED personnel to this possibility.

CONGENITAL ADRENAL HYPERPLASIA

Background

Inborn errors of adrenal steroid biosynthesis are grouped under the term *congenital adrenal hyperplasia* (CAH). Two major modes of presentation occur in early infancy and require prompt diagnosis and treatment: acute salt-losing crisis and ambiguous genitalia (Table 97.6). CAH may also present in children as precocious virilization. This form of CAH warrants investigation, but it does not require emergency management. The most common form of CAH presenting in infancy is 21-hydroxylase deficiency, which is recessively inherited and accounts for 90% of all cases. Clinically apparent salt wasting develops in 30 to 70% of affected patients. In the United States, the incidence of 21-hydroxylase deficiency is about 1 in 15,000 live births.

	Clinical Features				
	Newborn with Sexual Ambiguity				
Enzyme Deficiency	Female	Male	Salt Wasting	Hypertension	Postnatal Virilization
21-Hydroxylase					
Non-salt wasting	+	0	0	0	+
Salt wasting	+	0	+	0	+
11-Hydroxylase	+	0	0	+	+
3β-Hydroxysteroid dehydrogenase	+	+	+	0	0
17α-Hydroxylase	0	+	0	+	0
Cholesterol desmolase	0	+	+	0	0
18-Hydroxylase	0	0	+	0	0
17β-Hydroxysteroid dehydrogenase	?	+	-	-	+

Table 97.6. Clinical and Laboratory Features of Various Forms of Congenital Adrenal Hyperplasia

Pathophysiology

The enzymes 21-hydroxylase, 11a-hydroxylase, 3b-hydroxysteroid dehydrogenase, and 20,22-desmolase are involved in the production of both cortisol and aldosterone (Table 97.6). Because the hypothalamic–pituitary axis is under feedback control by cortisol, the lack of production of this hormone caused by the enzyme deficiency results in a significant increase in ACTH. In turn, ACTH stimulates the adrenal to increase steroid hormone production. Because cortisol synthesis is impaired, the precursors of cortisol accumulate significantly. The symptoms and signs characteristic of each enzymatic deficiency reflect either the absence of cortisol or aldosterone or the accumulation of their precursors.

Impairment of mineralocorticoid synthesis by 21-hydroxylase, 3b-hydroxysteroid dehydrogenase, and 20,22-desmolase deficiency can result in salt wasting. Although 11a-hydroxylase deficiency also blocks aldosterone production, the immediate precursor to the block, desoxycorticosterone, has potent mineralocorticoid activity. Thus, instead of developing salt loss, patients with this enzyme defect often develop hypertension during childhood.

Androgenic compounds accumulate in 21-hydroxylase and 11a-hydroxylase deficiencies. Females with these defects are virilized in utero and are born with ambiguous genitalia; therefore, females are often identified in the newborn period. Some female infants are so virilized that they are mistaken as males with bilateral cryptorchidism. Males have normal genital development; therefore, the diagnosis is generally missed until they present with salt-wasting crisis during infancy or with evidence of precocious puberty during childhood. Deficiency of 3b-hydroxysteroid dehydrogenase leads to underproduction of testosterone. Boys with this deficiency are undervirilized because only weak androgens are produced, whereas girls are mildly virilized because of these weak androgens. Lack of cortisol renders the patient more susceptible to hypoglycemia and reduces the tolerance to severe stress, such as dehydration.

Clinical Manifestations

Initial evidence of CAH may be acquired at birth with the discovery of ambiguous genitalia, between 2 to 5 weeks of age when the baby presents with acute salt-losing crisis, or during childhood with the onset of precocious puberty. The affected child may come to the ED for any of these reasons. Two words of caution are in order regarding newborn screening for CAH. Although many states now screen newborns for CAH, the results may not be available for 3 to 4 weeks and the acute salt-losing crisis may occur before this time. Furthermore, the report of an abnormal test result may precipitate a visit to the ED: unless the child is ill, consultation with the pediatric endocrinologist is highly recommended.

The subsequent discussion deals primarily with the recognition and management of the acute salt-losing crisis, which is-life threatening. Salt wasting is present shortly after birth, but acute crisis usually does not occur until the second week of life. The appearance of symptoms can be insidious, with a history of poor feeding, lack of weight gain, lethargy, irritability, and vomiting. The nonspecificity of symptoms may lead to consideration of diagnoses far removed from CAH and delay initiation of treatment.

Examination of the child should include the vital signs and an assessment of the degree of dehydration. In severe cases, there may be shock and metabolic acidosis. The genitalia should be examined carefully because the degree of ambiguity of the genitalia varies considerably. Virilized females may have an enlarged clitoris and fusion of the labial folds. An undervirilized male may have a small phallus and/or hypospadias. The presence of gonads in the inguinal canals or labioscrotal fold is suggestive of a male karyotype. Hyperpigmentation of the labioscrotal folds and the nipples is occasionally present in the neonatal period; however, it is rarely prominent enough to alert the examiner to the possibility of CAH.

In the ED, the most urgent investigations are plasma electrolytes and blood glucose. The combination of hyperkalemia and hyponatremia is often the first clue to the diagnosis of CAH, especially in males. The plasma potassium is elevated, but in the presence of vomiting and diarrhea, the rise may be blunted. Levels between 6 and 12 mEq/L are commonly encountered, often without any clinical cardiac dysfunction or ECG changes. The plasma bicarbonate level is usually low, reflecting the metabolic acidosis that results from the retention of hydrogen ions in exchange for sodium loss. The blood glucose is usually normal; however, hypoglycemia may occur secondary to the lack of cortisol and the reduced caloric intake during the acute illness. Serum should be drawn for determination of an adrenal steroid profile to include 17-hydroxyprogesterone, dehydroepiandrosterone, androstenedione, and testosterone. Ideally, blood should be obtained for these tests before the administration of hydrocortisone. In the child in crisis, the diagnosis must be based on physical findings and electrolyte abnormalities, and treatment must be instituted before the definitive adrenal steroid profile is available.

Management

Fluid and Electrolyte Replacement

If the child is dehydrated, fluid replacement is urgent. Volume expansion should be effected by the rapid infusion of 20 mL/kg normal saline in the first hour or more rapidly, if needed. Because the dehydration in salt-losing CAH represents urinary losses of isosmotic fluid, replacement should consist of normal saline (0.9%). The volume to be replaced should constitute the child's daily requirements as well as the estimated fluid loss. Fluid input and output should be monitored carefully.

Mineralocorticoid Replacement

Principal management of the mineralocorticoid deficit is by the provision of sodium. In addition, hydrocortisone has some mineralocorticoid effect, particularly at high dosages. For long-term management, the child will require mineralocorticoid replacement (fludrocortisone 0.1 mg/day). Most infants also require oral Na^+ supplements for the first several months of life.

Glucocorticoid Replacement

Hydrocortisone (25 mg) should be given in an IV bolus, followed by hydrocortisone 50 mg/m^2 per 24 hours as a constant infusion. Alternatively, cortisone acetate 25 mg intramuscularly immediately, followed by 25 mg every 24 hours, may be used. Glucocorticoids can suppress ACTH and the precursor steroids production within a few hours of administration, thus making the diagnosis of an enzymatic deficiency more difficult. However, it is better to avoid the possibility of mortality by giving the steroid than to delay treatment for diagnostic purposes.

Correction of Hyperkalemia, Hypoglycemia, and Acidosis

Infants with CAH tolerate hyperkalemia far better than do other children and adults, with potassium levels as high as 12 mEq/L reported without clinical signs. Volume restoration with normal saline is the major and, usually, the only measure needed to lower the potassium. In the presence of arrhythmias, IV 10% calcium gluconate can be given for its membrane-stabilizing properties. Therapy with glucose and insulin is contraindicated because of the danger of precipitating hypoglycemia. If hypoglycemia is found at the time of presentation, it should be treated acutely by the administration of dextrose (0.25 g/kg) intravenously and by the subsequent inclusion of 10% dextrose in the infusate. Acidosis generally does not require specific treatment; however, the low serum bicarbonate may take days to fully correct.

PHEOCHROMOCYTOMA

Background

Pheochromocytomas are functional tumors that arise in chromaffin tissues. In most children, these tumors are in the adrenal medulla, but they may be found in aberrant tissue along the sympathetic chain. Less than 5% of all pheochromocytomas occur in children. They are twice as common in males as in females, with the incidence of malignancy estimated to be 2 to 4%. Most information on pheochromocytoma is derived from adult studies, especially regarding signs and symptoms. Few detailed studies are available on children.

Pathophysiology

Catecholamines are low-molecular-weight substances produced in the CNS, the sympathetic nerves, the adrenal medulla, and the extra-adrenal chromaffin cells. Catecholamines affect metabolic processes in most tissues of the body and have many effects, including accelerated heart rate, increased myocardial contraction, and increased peripheral vascular resistance. Excessive production of catecholamines by a pheochromocytoma results in intensification of the normal physiologic effects.

Clinical Manifestations

The detection of a pheochromocytoma requires expert clinical awareness. Most patients are symptomatic, but the symptoms are nonspecific and, in the child, are likely to be attributed to other disease entities. The symptoms and signs are related to the excess production of catecholamines and can be explained on the basis of the pharmacologic effects of these substances. The most common symptoms are headache, palpitations, and excessive or inappropriate sweating. The headache, characteristically, is pounding and may be severe. The palpitations may be accompanied by tachycardia. Almost all patients will have one of the three symptoms listed, and most will have at least two. Other symptoms may include nervousness, tremor, fatigue, chest or abdominal pains, and flushing.

The most useful screening tool for pheochromocytoma is the blood pressure cuff because most pheochromocytomas are associated with hypertension. Because this hypertension may be continuous or paroxysmal, frequent and repeated blood pressure determinations may be necessary. Hypertension is most likely to be found when the patient is symptomatic. A hypertensive patient who is asymptomatic is unlikely to have a pheochromocytoma. Paroxysmal symptoms and hypertension should lead to consideration of this diagnosis.

The diagnosis of a pheochromocytoma should also be considered in patients with malignant hypertension, in those who fail to respond or respond inappropriately to antihypertensive medications, and in those who develop hypertension during the induction of anesthesia or during surgery. Incidence of pheochromocytomas is increased among patients with

neurofibromatosis and with the multiple endocrine neoplasia syndromes types II and III.

Documentation of excess catecholamine in either the urine or serum confirms the diagnosis of pheochromocytoma. The most readily available and widely used test for this purpose is the measurement of urinary catecholamines or their metabolites (3-methoxy-4-hydroxymandelic acid and total metanephrines) in a 24-hour urine collection. The finding of significant elevations of these substances is adequate confirmatory data. Some false-negative results may occur using urinary catecholamines. When the degree of suspicion is high, repeated specimens may be needed. Plasma metanephrines and normetanephrines, when appropriately collected, have proven to be diagnostically useful.

Once the diagnosis is confirmed, anatomic localization is necessary using either computed tomography or nuclear magnetic resonance imaging. Occasionally, arteriography with selective sampling for epinephrine production is necessary for localization.

Management

Pheochromocytoma is cured by the surgical removal of the tumor. The focus of ED management should be on controlling hypertension and hypertensive crisis that may occur before the surgical procedure. a-Adrenergic blocking agents are useful in controlling hypertension and in minimizing blood pressure fluctuations during the surgical procedure.

Preferred drugs for controlling hypertension are phenoxybenzamine (Dibenzyline) and prazosin (Minipress). Dosage schedules and quantity must be tailored to the individual for adequate control of hypertension. Hypertensive crisis may be appropriately managed with IV phentolamine (Regitine 1 mg intravenously for children; 5 mg for adolescents) or sodium nitroprusside (0.5 to 8.0 µg/kg per minute).

DIABETES INSIPIDUS

Background

Diabetes insipidus (DI) is caused by an inability of the kidneys to concentrate urine and is characterized clinically by polyuria and polydipsia. Either a deficiency of ADH secretion from the hypothalamus and posterior pituitary gland or renal unresponsiveness to ADH can cause this disease (Table 97.7). Most central causes of DI in children are acquired and can present at any age. In contrast, the most common cause of nephrogenic DI in children is X-linked recessive and manifests in males during early infancy. Renal lesions associated with nephrogenic DI can present in later childhood.


I. Antidiuretic Hormone Deficiency
A. Head injury
B. Meningitis
C. Idiopathic
D. Suprasellar tumors and their treatment by surgery and/or radiotherapy
1. Craniopharyngioma
2. Optic nerve glioma
3. Dysgerminoma
E. Septooptic dysplasia
F. Association with midline cleft palate
G. Familial (dominant or sex-linked recessive)
H. Wolfram syndrome (diabetes insipidus, diabetes mellitus, optic atrophy, deafness)
I. Histocytosis X (Hand-Schüller-Christian disease)
II. Nephrogenic Diabetes Insipidus
A. Sex-linked recessive
B. Renal disease
1. Polycystic kidneys
2. Hydronephrosis
3. Chronic pyelonephritis
C. Hypercalcemia
D. Hypokalemia
E. Toxins:
1. Demeclocycline
2. Lithium
F. Sickle cell disease
G. Idiopathic

Table 97.7. Causes of Diabetes Insipidus in Children

Pathophysiology

ADH is synthesized in the supraoptic and paraventricular nuclei of the hypothalamus. It is transported along nerve axons to the posterior pituitary gland, where it is stored. ADH is released in response to increased plasma osmolality, hypernatremia, and decreased right atrial pressure secondary to hypovolemia. The distal convoluted tubules and the collecting ducts of the kidneys respond to ADH by increasing water reabsorption. Lack of ADH (central DI) can result from a wide variety of hypothalamic and pituitary lesions (Table 97.7). Conversely, in nephrogenic DI, ADH levels are normal or elevated because the defect resides in the renal collecting tubules, which are resistant to the action of ADH. In either case, failure of water reabsorption results in polyuria. A normal thirst mechanism contributes toward fluid balance by promoting adequate fluid intake; however, if this balance is not achieved, hypertonic dehydration ensues. If a hyperosmolar state develops abruptly, it may lead to dehydration of neural tissues, which can cause serious neurologic sequelae or result in death.

Clinical Manifestations

Urine excretion is increased in both volume and frequency in the child with DI. This condition may manifest as enuresis in the younger child. Provided the thirst mechanism is intact and fluids are accessible, the child can compensate for the water loss by drinking more. A history may be elicited of the child's awakening in the middle of the night to drink. If fluids are not available or if fluid intake is interrupted because of a viral illness, dehydration rapidly ensues. In the young infant who is not provided with adequate fluids and consequently is chronically dehydrated, the child may fail to thrive or may have a history of intermittent low-grade fevers. On the other hand, if the cries of the infant are interpreted as hunger rather than thirst, the infant with DI may be obese.

Physical examination may be normal, or signs of dehydration, such as dryness of mucous membranes, decreased skin turgor, sunken eyes, and in an infant, a depressed anterior fontanel, may be present. Because of the hyperosmolarity, the degree of dehydration may be underestimated on physical examination. Hypothalamic or pituitary lesions can lead to other endocrine abnormalities such as secondary hypothyroidism and growth failure. A craniopharyngioma or optic nerve glioma may affect the visual fields or cause raised intracranial pressure, which is indicated by papilledema.

DI is diagnosed by demonstrating that the kidneys fail to concentrate urine when fluid intake is restricted. This condition can be difficult to prove in children. Nonetheless, an adequate working diagnosis is usually obtained by finding an elevated serum osmolality (normal less than 290 mOsm/L) and an elevated serum [Na] (normal less than 145 mmol/L) in the presence of dilute urine (normal osmolality more than 150 mOsm/L). Blood glucose and serum creatinine levels are normal.

In many cases, the diagnosis can be ruled out by the demonstration of appropriately concentrated urine and normal serum osmolality on specimens obtained upon awakening. The definitive diagnosis is made by a formal water deprivation test. This test is performed electively in cases in which the diagnosis is uncertain and should never be performed if the child is already dehydrated. The measurement of ADH by radioimmunoassay is available but generally is not useful in the diagnosis of DI.

Management

In most cases, a diagnosis of DI is not known at the time of presentation; therefore, the acute management is directed toward correction of the dehydration and the hyperosmolar state. The treatment of DI is similar to that described for hypernatremic dehydration (see Chapter 18) with the notable addition that the fluid required for the replacement of urinary fluid losses will be far greater. In fact, the high urinary output, despite significant dehydration, often provides the first and most convincing evidence for DI. If the child is hypotensive or if the serum Na^+ is greater than 160 mmol/L, initial volume expansion is necessary, using 20 mL/kg normal saline during the first hour or more rapidly, if needed. Once an adequate intravascular volume has been achieved, further fluid replacement is accomplished slowly because overly rapid volume correction can cause cerebral edema, seizures, and death.

If the child is not hypotensive, or once the hypotension has been corrected, free water replacement is done over 48 hours. Calculations of appropriate fluids must include maintenance requirements, replacement needs, and ongoing urinary losses (see Chapter 18).

If DI is strongly suspected on the basis of discrepant serum and urine osmolality, DDAVP (5 to 20 µg intranasally or 0.2 to 0.4 µg/kg subcutaneously) may be a useful adjunct to IV fluid therapy. If DDAVP is not available or cannot be used for some reason, other antidiuretic agents are available (aqueous pitressin 1 to 5 units intramuscularly or 2 to 3 µU/kg per minute as a constant IV drip). DDAVP acts rapidly to promote tubular resorption of free H_2O; clinically, this reaction is apparent as decreased urinary output with increased osmolality within an hour of administration. Failure to respond to DDAVP suggests the possibility of tubular unresponsiveness to ADH (nephrogenic DI); however, more commonly, failure to respond results from improper administration of the medication or use of DDAVP that has lost its potency. Because of these factors, if cessation of diuresis is not noted within 2 hours of administration of the first dose, a second dose from a different bottle of DDAVP should be tried. The use of an ADH agonist generally simplifies management by reducing the quantity of fluid that must be infused; however, careful monitoring of input and output remains essential. Children who fail to respond to DDAVP are likely to have nephrogenic DI and must be acutely managed with fluid therapy alone. Paradoxically, the thiazide diuretics have proven to be useful in the chronic control of nephrogenic DI.

The child should be closely observed for changes in level of consciousness, pulse rate, and blood pressure. Fluid input and output should be meticulously monitored. Serum osmolality and $[Na^+]$ should be determined every 1 to 2 hours until the rate of their decline can be determined. Urine osmolality should be measured every 1 to 2 hours to determine the responsiveness of the renal tubule to DDAVP. Because large volumes of dextrose-containing fluids are used, the blood glucose should also be followed closely. If the blood glucose exceeds 160 mg/dL, the concentration of dextrose in the infusate should be decreased.

SYNDROME OF INAPPROPRIATE ANTIDIURETIC HORMONE SECRETION

Background

Excessive secretion of ADH accompanying normal or low plasma osmolality or [Na] is inappropriate because it further depresses the plasma osmolality and [Na]. Symptoms of excessive ADH secretion are not usually apparent until the plasma [Na] falls below about 120 mmol/L. The overall incidence of the syndrome of inappropriate antidiuretic hormone secretion (SIADH) in childhood is unknown, but it is common in certain disease states. More than 50% of children with bacterial meningitis, about 20% of patients on positive pressure ventilation, and about 70% of children with Rocky Mountain spotted fever develop SIADH.

Pathophysiology

ADH secretion is stimulated by hypertonicity of the fluid surrounding the hypothalamic osmoreceptors, by volume receptors in the left atrium, and by ill-defined nervous impulses from higher cortical centers. Disorders of the CNS (Table 97.8) may cause excessive ADH secretion by producing either a local disturbance of the hypothalamic osmoreceptors or some undetermined nervous stimuli. Many intrathoracic conditions are associated with SIADH, probably by stimulating the volume receptors in the left atrium. Physical and emotional stress and severe pain also cause ADH secretion. Excessive secretion of ADH leads to water retention by the collecting tubules of the kidneys, a mechanism mediated by cyclic adenosine monophosphate (cAMP). The retained water expands the intravascular compartment, dilutes all plasma

constituents, and lowers the plasma osmolality.

I. Disorders of Central Nervous System	III. Miscellaneous
A. Infection (meningitis, encephalitis)	A. Pain (e.g., after abdominal surgery)
B. Trauma, postneurosurgery	B. Severe hypothyroidism
C. Hypoxic insults, especially in the perinatal period	C. Congenital deficiency
D. Brain tumor	D. Tumors (e.g., neuroblastoma)
E. Intraventricular hemorrhage	E. Idiopathic deficiency
F. Guillain-Barré syndrome	IV. Drug-Induced
G. Psychosis	A. Increased antidiuretic hormone secretion
II. Intrathoracic Disorders	1. Vincristine
A. Infection (tuberculosis, pneumonia, empyema)	2. Cyclophosphamide
B. Positive-pressure ventilation	3. Carbamazepine
C. Asthma	4. Adenine arabinoside
D. Cystic fibrosis	5. Phenothiazines
E. Pneumothorax	6. Morphine
F. Patent ductus arteriosus ligation	B. Potentiation of antidiuretic hormone effect
	1. Acetaminophen
	2. Indomethacin

Table 97.8. Some Causes of Syndrome of Inappropriate Antidiuretic Hormone Secretion (SIADH) in Children

Clinical Manifestations

Most patients with SIADH are asymptomatic until the plasma [Na] falls below 120 mmol/L. Symptoms associated with hyponatremia range from anorexia, headache, nausea, vomiting, irritability, disorientation, and weakness to seizures and coma, leading ultimately to death. Absence of edema and dehydration are significant clinical findings.

Laboratory investigations for diagnostic purposes must include concomitant serum and urine samples (Table 97.9). Hyponatremia, hypoosmolality (serum), and low blood urea nitrogen will be present. In contrast, the urinary osmolality and [Na] are inappropriately elevated for the hypotonicity of the serum. Radioimmunoassay for ADH is now available and has been helpful in defining this syndrome; however, the results of this test are unlikely to be available on an emergency basis. The underlying cause of the syndrome should be investigated according to the physician's' clinical judgment. In the presence of hyperglycemia, hyperlipidemia, or hyperproteinemia, the serum sodium may be falsely low. Renal salt wasting, secondary to adrenal insufficiency, should be accompanied by hyperkalemia and dehydration. The urine osmolality in water intoxication states is usually low compared with that found in SIADH.

Hyponatremia, reduced serum osmolality
Urine osmolality that is inappropriately elevated (a urine osmolality <100 mOsm/kg usually excludes the diagnosis)
Urinary Na concentration that is excessive in comparison to the degree of hyponatremia (usually >18 mEq/L)
Normal renal, adrenal, and thyroid function
Absence of volume depletion

Table 97.9. Criteria for Diagnosis of Syndrome of Inappropriate Antidiuretic Hormone Secretion (SIADH)

Management

Severely Symptomatic Children

Patients with a persistent seizure attributable to severe hyponatremia and those who are lethargic or comatose need urgent treatment. Hypertonic (3%) saline is the preferred treatment. Infusing small amounts of 3% saline in the range of 3 mL/kg every 10 to 20 minutes until symptoms remit is probably the safest course of treatment. A single dose of furosemide (1 mg/kg) also can be administered intravenously. Close monitoring of fluid balance, plasma and urinary sodium, potassium, and osmolality is essential. Phenytoin (Dilantin) intravenously (5 to 10 mg/kg) inhibits ADH release and may be helpful in the patient with seizures secondary to CNS causes of SIADH. The underlying cause of SIADH, such as meningitis, should be treated when possible; successful treatment is usually accompanied by remission of inappropriate antidiuresis.

Asymptomatic or Mildly Symptomatic Children

Asymptomatic or mildly symptomatic patients are treated by rigorous fluid restriction. If the patient is not vascularly compromised, fluid input should be sharply limited, often below insensible loss, until the [Na] and osmolality begin to rise. If the initial [Na] is less than 125 mmol/L, all fluids must be withheld. Frequent measurements of plasma electrolytes and osmolality, as well as close monitoring of fluid input and output, are essential. As the serum [Na] rises and urine osmolality falls, the rate of fluid administration can be gradually increased. The child with chronic or recurrent episodes of SIADH may require treatment with demeclocycline 10 mg/kg. The underlying cause should be identified, treated, and eliminated, if possible.

HYPERPARATHYROIDISM

Background

Hyperparathyroidism is most commonly recognized during the third, fourth, and fifth decades of life. It is uncommon in children.

Pathophysiology

The parathyroid glands are derived from the third and fourth pharyngeal pouch and are usually embedded in the posterior aspect of the thyroid gland. Occasionally, a gland may be found in the anterior mediastinum. Parathyroid hormone (PTH) is the primary hormone produced by the parathyroid glands. The normal stimulus for PTH synthesis and release is a low calcium concentration. Prolonged hypocalcemia may lead to hypertrophy of the parathyroid glands. PTH acts on the kidney to decrease the excretion of calcium, magnesium, and hydrogen while increasing the excretion of phosphate, sodium, and bicarbonate. Many of the effects are mediated by cAMP, and an increased quantity of cAMP is present in the urine of patients with hyperparathyroidism. PTH also increases the formation of 1,25-dihydroxyvitamin D in the kidneys. PTH may increase intestinal absorption of calcium, although this effect is primarily mediated by 1,25-$(OH)_2D$. Both PTH and 1,25-$(OH)_2D$ affect bone. PTH acts on bone to increase the release of calcium by increasing the number and activity of the osteoblasts, whereas vitamin D decreases calcium use in bone formation by decreasing the number of osteoblasts. The net effect of the actions of PTH and vitamin D is to increase serum calcium by decreasing renal calcium excretion, decreasing new bone formation, increasing intestinal absorption of calcium, and increasing bone resorption.

Clinical Manifestations

Hyperparathyroidism has two common presentations in children. The first presentation is the critically ill infant who is found to have severe hypercalcemia during the course of diagnostic investigations. The serum calcium level may be extremely high. The second presentation is a child in the early to midteens with nonspecific symptoms including nausea, constipation, unexplained weight loss, personality changes, and headaches. Diffuse bone pain or renal colic may be reported, although these symptoms are less common in children than in adults.

The physical findings of hypercalcemia are hypotonia, weakness, and listlessness. Rarely, a palpable mass is located in the parathyroid region. Certain characteristic features have been associated with idiopathic hypercalcemia in infancy, including hypertelorism, broad forehead, epicanthal folds, prominent upper lip, an underdeveloped nasal bridge, and a small mandible. Not surprisingly, these same features have been noted in infants with hyperparathyroidism.

A family history may be helpful because hyperparathyroidism has been associated with both multiple endocrine neoplasia types I and II, which are inherited as autosomal-dominant conditions. Hyperparathyroidism also may occur in infants of hypoparathyroid mothers. Radiologic findings consistent with hyperparathyroidism include evidence of demineralization and bone resorption (Fig. 97.1 and Fig. 97.2). Osteitis fibrosa cystica, although highly suggestive of the diagnosis, is unusual in children.

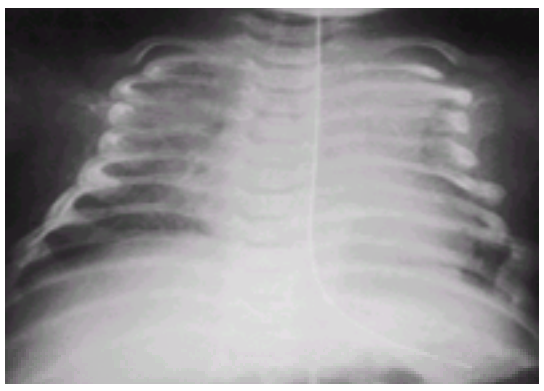

FIGURE 97.1. Primary hyperparathyroidism in a 3-day-old girl. Roentgenogram of the chest shows profound demineralization of the skeleton with loss of a well-defined cortical margin. Cystic changes in rib and subperiosteal bone resorption in humerus are seen. (Courtesy of Soroosh Mahboubi, MD, The Children's Hospital of Philadelphia.)

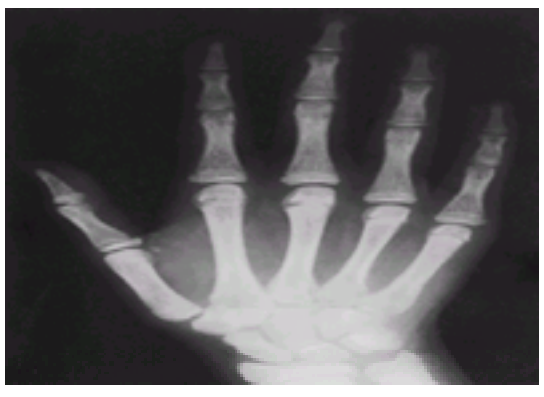

FIGURE 97.2. Secondary hyperparathyroidism in a 12-year-old girl with chronic pyelonephritis. There is moderate subperiosteal erosion on the radial side of the middle phalanges; note a lacy appearance of the periosteum and small-tuft

erosion. Subperiosteal bone resorption is the most significant radiologic finding in hyperparathyroidism; subperiosteal bone resorption and tuft erosion are seen in both primary and secondary hyperparathyroidism. (Courtesy of Soroosh Mahboubi, MD, The Children's Hospital of Philadelphia.)

The clinical laboratory is helpful in the recognition of hyperparathyroidism. Hypercalcemia is usually present but may be subtle or intermittent in mild cases. The serum inorganic phosphate level is usually low but may be normal, especially in patients with decreased renal function. Mild hyperchloremic acidosis may be present. Alkaline phosphates level and urinary hydroxyproline excretion may be elevated secondary to increased osteoclast activity. Because PTH causes a significant increase in cAMP in the kidney tubule, the presence of excess cAMP in the urine is strongly suggestive of excess PTH production. The determination of PTH levels is critical for diagnostic purposes, and elevated levels of PTH, when the patient is hypercalcemic, is a definitive laboratory finding.

Management

Acute management of hyperparathyroidism is essentially the same as management of hypercalcemia (see Chapter 86). The specific management of hyperparathyroidism depends on the level of calcium and on the presence of signs and symptoms. In the asymptomatic patient with a serum calcium of less than 12 mg/dL, careful follow-up with close attention to both bone mass and renal function is recommended. If the child is persistently hypercalcemic, parathyroid surgery is the preferred treatment. In the case of hyperplasia, the common reason for hyperparathyroidism in the infant, subtotal parathyroidectomy is indicated. If an adenoma is present, as is usually the case in the older child, simple removal of the involved parathyroid gland is adequate.

HYPOPARATHYROIDISM

Background

Hypoparathyroidism is rare in children. It may occur sporadically or be part of a familial syndrome consisting of combinations of several autoimmune diseases (e.g., Addison's disease, diabetes mellitus, lymphocytic thyroiditis, pernicious anemia, ovarian failure). Hypoparathyroidism is also associated with thymic aplasia and severe immunologic deficiencies (DiGeorge's syndrome). A transient form of hypoparathyroidism, lasting for as long as a year, has been reported in some infants. Hypoparathyroidism also may result from damage incurred during thyroid surgery or irradiation.

Pathophysiology

The basic actions of PTH are described in the preceding section on hyperparathyroidism. The lack of PTH, regardless of cause, has several deleterious effects on calcium homeostasis. Because PTH has significant effects on 1,25-$(OH)_2D_3$ formation, the absence of PTH is magnified by a consequent reduction in 1,25-$(OH)_2D_3$. The net effect of the lack of PTH (and decreased quantity of vitamin D) is a declining serum level of calcium, primarily caused by decreased intestinal absorption of calcium and decreased renal resorption of calcium.

Clinical Manifestations

The predominant historical features and clinical manifestations of hypoparathyroidism are the same as those of hypocalcemia (see Chapter 86). Unique historical information that may suggest the diagnosis of hypoparathyroidism includes other family members with autoimmune endocrine disease, recurrent episodes of serious infection in the affected child, and previous thyroid manipulations.

Most symptoms and signs of hypoparathyroidism are the same as those related to hypocalcemia. The particular symptoms and signs found depends on the age at onset of the disease, the chronicity of the disease, and the presence of other autoimmune or syndromic phenomena. Papilledema without hemorrhage may be seen during the initial examination and tends to resolve within several days after the initiation of therapy. Lenticular cataracts are common in hypoparathyroidism and are associated with long-standing hypocalcemia of any cause. Psychiatric and neurologic disorders occur in association with hypoparathyroidism. Subnormal intelligence occurs in about 20% of children with the idiopathic form of hypoparathyroidism, and the severity correlates closely with the period of untreated hypocalcemia. Dry, scaly skin is a common finding, as is patchy alopecia. Psoriasis or mucocutaneous candidiasis may be found on occasion. Unusually brittle fingernails and hair are often found. Hypoplasia of tooth enamel may be seen if hypoparathyroidism was present at the time of dental development. Intestinal malabsorption and steatorrhea have been reported in association with hypoparathyroidism.

In most cases, the diagnosis of hypoparathyroidism is first considered when low serum calcium is found. If an elevated phosphate accompanies low calcium, a low or normal serum alkaline phosphate, and normal blood urea nitrogen, hypoparathyroidism is a likely possibility. Finding a low or unmeasurable level of PTH in the presence of hypocalcemia and hyperphosphatemia makes the definitive diagnosis. Because PTH increases cAMP levels in the urine, the excreted amount of cAMP in the urine is low in patients with hypoparathyroidism and rises briskly with the administration of exogenous PTH. The presence of antibodies in other endocrine tissues or organs may help in delineating the cause of the hypoparathyroidism.

Management

The acute management of hypoparathyroidism is essentially the management of the hypocalcemia (see Chapter 86). Long-term management consists of treatment with vitamin D, usually with one of its more active analogs—1,25-$(OH)_2D_3$.

Supplemental oral calcium is almost always necessary. The goals of long-term therapy are to maintain the serum calcium in the lower range of normal and to avoid both vitamin D toxicity and hypercalcemia. Preparations of PTH are not available for the long-term management of hypoparathyroidism.

RICKETS

Background

Rickets describes a characteristic set of clinical features delineated centuries ago, which is now known to be predominantly caused by inadequate dietary vitamin D. With this awareness and the advent of vitamin D supplementation of foods, especially milk, the incidence of rickets has fallen significantly; however, rickets is still seen among certain ethnic groups, premature infants, children with severe malabsorption problems, and patients with serious renal disease.

Pathophysiology

Rickets is caused by the failure of mineralization of bone matrix in growing bone resulting from a lack of vitamin D. Consequently, unmineralized cartilage is excessive, and bone is soft. In addition to inadequate intake of vitamin D, the other causes of rickets are inability to form the active metabolite of vitamin D, excess phosphate excretion, and excess accumulation of acid.

Vitamin D may be obtained from dietary sources (especially animal fat) or synthesized from cholesterol via a complex pathway requiring the interaction of the precursor molecule with sunlight. Further hydroxylation of vitamin D in the liver (25-hydroxylation) and kidney (1-hydroxylation) leads to the formation of the active metabolite 1,25-dihydroxyvitamin D. Therefore, failure to form 1,25-$(OH)_2D_3$ may result from inadequate intake of vitamin D or insufficient exposure to sunlight. This is a particular problem among ethnic groups that eat small quantities of animal meat and that are extensively clothed when outdoors. Because vitamin D is fat soluble, any problem leading to prolonged fat malabsorption can result in rickets. Diseases affecting kidney or liver function may also lead to inadequate production of 1,25-$(OH)_2D_3$. An inherited deficiency of the 1-hydroxylase in the kidney (vitamin D–dependency rickets) is known. Certain drugs, such as phenobarbital and phenytoin, affect liver metabolism of vitamin D and can lead to rickets. Premature infants are particularly prone to vitamin D deficiency because of their minimal stores of vitamin D and their limited capacity for vitamin D synthesis.

Phosphate is a critical component of bone formation. Excess excretion of phosphate may lead to clinical rickets. Conditions that lead to excess phosphate excretion include primary hyperphosphaturia, Lowe's syndrome, and Fanconi's syndrome. Vitamin D–resistant rickets is a misnomer because the primary defect is in the renal tubular resorption of phosphate and not a resistance to vitamin D. Both an X-linked recessive and an autosomal-dominant form of phosphate wasting are known.

Rickets may also occur in conditions leading to chronic acidosis because bone is resorbed to buffer the acid load. This condition is seen in patients with distal renal tubular acidosis and may be partially responsible for the rachitic changes associated with Fanconi's syndrome.

Clinical Manifestations

Children with rickets may come to medical attention because of specific physical abnormalities (bowed legs), limb pain and swelling, seizures, failure to thrive (renal tubular acidosis), biochemical abnormalities (hypocalcemia), or radiographic findings (broadened, frayed metaphysis). A thorough social and dietary history is helpful in delineating the probable cause and in sparing the patient an extensive and expensive evaluation. A family history may be useful in identifying the 1-hydroxylase deficiency or renal phosphate wasting. If the child has previously been treated with vitamin D, the reported response to that treatment may be helpful in identifying the likely site of defect.

The clinical findings in rickets may vary considerably depending on the underlying disorder, the duration of the problem, and the child's age. Most features are related to skeletal deformity, skeletal pain, slippage of epiphyses, bony fractures, and growth disturbances. Muscular weakness, hypotonia, and lethargy are often noted. Failure of calcification affects those parts of the skeleton that are growing most rapidly or that are under stress. For example, the skull grows rapidly in the perinatal period; therefore, craniotabes is a manifestation of congenital rickets. On the other hand, the upper limbs and rib cage grow rapidly during the first year of life, and abnormalities at these sites are more common at this age (i.e., rachitic rosary, flaring of the wrist). Bowing of the legs is unlikely to be noted until the child is ambulatory. Dental eruption may be delayed, and enamel defects are common.

Radiography is the optimal way to confirm the clinical diagnosis because the radiologic features reflect the histopathology. Characteristic findings include widening and irregularity of the epiphyseal plates, cupped metaphyses, fractures, and bowing of the weight-bearing limbs (Fig. 97.3 and Fig. 97.4). The clinical laboratory is often helpful in correctly identifying the cause of rickets. Frank hypocalcemia (less than 7 mg/dL) is unusual in rickets. Calcium levels in the 7 to 9 mg/dL range are common and warrant careful attention because the initiation of vitamin D treatment increases bony deposition of calcium and may lead to a fall in serum calcium. Phosphate levels are often low. An amino aciduria is often present and may lead to some confusion of simple vitamin D deficiency with Fanconi's syndrome. Alkaline phosphatase levels are significantly increased, reflecting extremely active bony metabolism. Although PTH levels are elevated, the results of this test are unlikely to be available at the time initial clinical decisions are made. Chronic acidosis, liver disease, and renal disease should be ruled out.

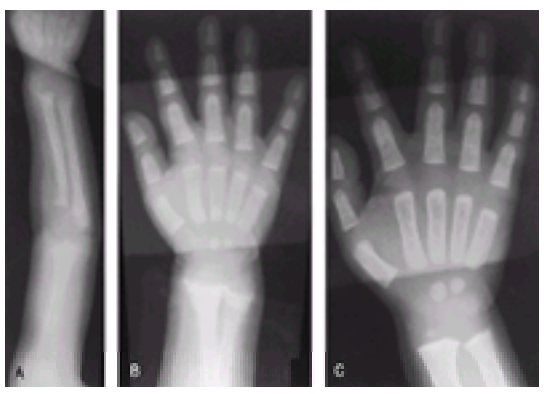

FIGURE 97.3. A. Rickets in an 11-month-old boy, breast-fed since birth. Roentgenogram of the upper extremity shows profound demineralization of the skeleton, with frayed, irregular cupping of the end of the metaphysis and poorly defined cortex. Note retardation of skeletal maturation. **B.** Same patient with some healing 4 weeks after supplemental vitamin D. Severe rachitic changes are noticeable. Periosteal cloaking, both of the metacarpals and of the radius and ulna, is evidence of healing. **C.** Complete healing of the rickets 8 months after treatment. Note the reappearance of the provisional zone of calcification. (Courtesy of Soroosh Mahboubi, MD, The Children's Hospital of Philadelphia.)

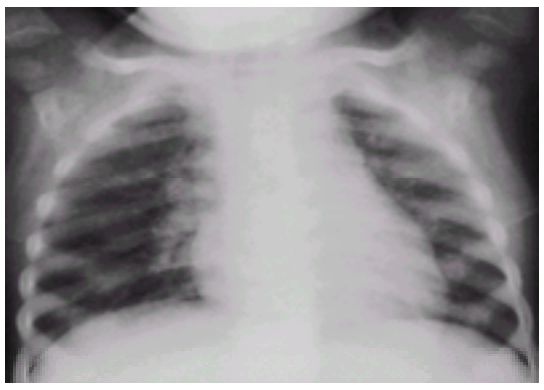

FIGURE 97.4. Rickets in an 11-month-old boy, breast-fed since birth. Roentgenogram of the chest shows demineralization of the skeleton with cupping of the distal end of ribs and humerus. (Courtesy of Soroosh Mahboubi, MD, The Children's Hospital of Philadelphia.)

Treatment

Treatment depends on the nature of the underlying disease. The response to treatment may be helpful in differentiating simple dietary vitamin D deficiency from more complex causes of rickets. In the absence of chronic disease, dietary rickets may be adequately treated with daily doses of 1000 to 2000 IU of vitamin D until healing occurs. Serum phosphate usually returns to normal within 1 to 2 weeks, and radiographic improvement is generally apparent by 2 weeks. Once healing is complete, the child should continue to be treated with 400 IU/day to prevent recurrence. If the initial serum calcium is borderline low or low, supplemental calcium should be initiated 48 hours before the institution of vitamin D, especially in the young child. Otherwise, the institution of vitamin D may cause a further decrease in serum calcium and elicit frank hypocalcemia. Children with symptomatic hypocalcemia or an initial serum calcium of less than 7 mg/dL on presentation warrant hospitalization and frequent calcium determinations. Failure to respond to vitamin D treatment suggests that the child has a more complex cause of rickets, and consultation with a pediatric nephrologist or endocrinologist is recommended.

THYROID STORM

Background

Thyroid storm, or thyrotoxic crisis, is a fulminating intensification of the hyperthyroid state. Because hyperthyroidism uncommonly occurs in children and because thyroid storm occurs in only 1% of patients with hyperthyroidism, thyroid storm is rare in children. Therefore, most information available on thyrotoxic crisis is derived from reports of this condition in adults. Precipitating factors include intercurrent infection, trauma, and subtotal thyroidectomy in an inadequately prepared patient. The mortality rate in adults may be as high as 20%; similar data are not available for children.

Pathophysiology

In thyroid storm, thyroid hormone is suddenly released into the circulation, which results in the uncoupling of oxidative phosphorylation and/or increased lipolysis, both of which contribute to excessive thermogenesis. Insensible fluid loss increases as a result of increased metabolism and sweating. Tachycardia is caused by both the hyperthermia and the direct action of thyroid hormones on the cardiac conduction system.

Clinical Manifestations

Almost all cases of thyroid storm occur in patients with known hyperthyroidism, although occasionally, a patient will present initially with thyroid storm. Most patients will have clinical findings characteristic of hyperthyroidism, including goiter (more than 95%), exophthalmos, tachycardia, bounding pulses, and systolic hypertension. Tremulousness,

restlessness, mania, delirium, or frankly psychotic behavior may be present. A primary feature that distinguishes thyroid storm from uncomplicated hyperthyroidism is the presence of high fever, often as high as 41°C (105.8°F). The marked increase in cardiac work load may result in congestive heart failure, in which case hypotension and pulmonary edema may be seen, rather than more classic hypertension.

Thyroid studies including serum thyroxine (T_4), triiodothyronine (T_3), T_3 resin uptake (T_3 RU), and TSH should be obtained. Many clinical laboratories can now perform T_4 assays on an emergency basis; however, in many cases, therapy must be initiated on the basis of clinical evidence. Furthermore, the T_4 and T_3 values seen in thyroid storm overlap with those found in frank hyperthyroidism without storm. Serum electrolytes should be obtained but are unlikely to reveal any characteristic abnormalities, except for evidence of modest dehydration. A chest radiograph and ECG are helpful in evaluating and following cardiac status as treatment is initiated.

Management

Initial treatment is directed toward lowering the metabolic rate and reducing the cardiac work load. Subsequent treatment is directed toward controlling thyroid hormone production. Because many of the hypermetabolic effects of hyperthyroidism are mediated by the adrenergic system, a blocker (propranolol starting at 10 µg/kg intravenously over 10 to 15 minutes) is useful in the acute management of thyroid storm. ECG monitoring for heart rate and arrhythmias is recommended. Because the metabolic rate is increased about 10% for every degree of body temperature above 36.5°C (97.7°F), lowering body temperature is an effective means of reducing the metabolic rate in the patient with thyrotoxicosis. Tepid sponging, use of a cooling blanket, and administration of acetaminophen can accomplish this task. Aspirin should not be used because it is a potential uncoupler of oxidative phosphorylation that may exacerbate the hypermetabolic state.

Treatment of the hyperthyroidism in thyroid storm is accomplished by the use of iodide and a thiourea derivative such as propylthiouracil. Iodide rapidly terminates thyroid hormone release; however, this effect is overcome after 3 to 5 days of iodide therapy. Lugol's iodide 5 drops once every 8 hours orally or sodium iodide 125 to 250 mg/day intravenously over 24 hours is the usual mode of iodide therapy. Propylthiouracil has at least two beneficial effects: 1) prevention of thyroid hormone synthesis in the thyroid gland and 2) inhibition of the peripheral conversion of thyroxine to its more active form, T_3. The dosage of propylthiouracil is 6 to 10 mg/kg per day, given orally every 6 to 8 hours. Its effects are minimally useful in acute management because the reduction in thyroid levels may take several days. Methimazole 0.6 to 0.7 mg/kg per day divided every 8 hours is an alternative to propylthiouracil, but its usefulness in thyroid storm is subject to the same limitations as propylthiouracil.

Adequate hydration is essential for effective treatment, and the estimate of fluid replacement should include a consideration of the significant increase in fluid requirements caused by fever and an accelerated metabolic rate. Glucocorticoids are useful in the acute situation because they appear to inhibit thyroid hormone release from the thyroid and decrease the peripheral conversion of T_4 to T_3. Dexamethasone (0.2 mg/kg) or hydrocortisone (5 mg/kg) can be given parenterally during the acute phase. Because intercurrent infection may be the precipitating factor, it should be searched for and treated appropriately. Broad-spectrum antibiotics should be considered while awaiting the results of cultures.

Improvement should be seen within a few hours after the initiation of treatment with propranolol, especially in terms of cardiovascular status. Full recovery and adequate control of the underlying thyroid disease will take several days to achieve. For the patient presenting with thyroid storm, serious consideration should be given to permanent treatment of the hyperthyroidism, either by surgery or radioiodide ablation.

NEONATAL THYROTOXICOSIS

Background

Neonatal thyrotoxicosis is a life-threatening condition that may not be correctly diagnosed in the newborn nursery and that may be discovered only when the child presents in extremis in the ED. Neonatal thyrotoxicosis is found in 1 to 5% of infants born to mothers with a history of hyperthyroidism; however, the maternal disease does not have to be active during the pregnancy.

Pathophysiology

Neonatal thyrotoxicosis is caused by excessive thyroid hormone produced by the neonatal thyroid that has been stimulated by maternal thyroid-stimulating antibodies present in the immunoglobulin G (IgG) fraction that have crossed the placenta. TSH, T_4, and T_3 do not cross the placenta in significant quantities. In most cases, the disease is self-limiting, and hyperthyroidism remits within about 6 weeks. Occasionally, the disease may run a protracted course and arise in the absence of maternal thyroid-stimulating antibodies.

Clinical Manifestations

Goiter and exophthalmos are almost always present; however, a goiter may be difficult to appreciate in a small infant with a short neck. The child usually fails to gain weight despite a ravenous appetite. The child may also be irritable and have tachycardia, as well as signs of congestive heart failure. Laboratory investigations should include estimations of serum T_4, T_3, and TSH, and thyroid-binding capacity. Increased concentration of T_4 in the presence of suppressed TSH levels is consistent with the diagnosis. If the mother is taking antithyroid medication, thyroid function tests on the infant may be unreliable in the first days of life because of suppression of the fetal thyroid by transplacental passage of maternal antithyroid medication. The bone age may be advanced. In most cases, treatment must be initiated on the basis of

historical and clinical findings.

For an infant who has an elevated level of T_4 but who has few, if any, symptoms or signs, consultation with a pediatric endocrinologist is strongly recommended. T_4 levels in all infants tend to be higher than those in older children because of increased TBG induced by maternal estrogen that crosses the placenta. Also, an elevated thyroxine may be seen with defects that alter the binding of T_4 to thyroid-binding globulin (TBG) or the end-organ sensitivity to T_4.

Management

Treatment is identical to that outlined for thyroid storm in older children. The duration of treatment is uncertain and should be based on serial thyroid function tests, especially TSH. It is anticipated that treatment need be continued only for 6 to 8 weeks in most cases because the causative agent is a subclass of IgG molecules with a serum half-life of about 2 weeks.

CONGENITAL HYPOTHYROIDISM

Background

Most westernized countries routinely screen infants in the first days of life for congenital hypothyroidism. The incidence of this problem is 1 in 3500 to 5000 live births. On occasion, notification of the parents by the screening program results in significant parental anxiety and leads to a visit to the ED. Emergency physicians should be knowledgeable about congenital hypothyroidism so that they can appropriately educate parents and initiate therapy. Acquired hypothyroidism rarely results in urgent clinical problems that lead to ED visits.

Pathophysiology

The causes of congenital hypothyroidism are numerous. About 20% of patients have ectopic glands, and another 50% have hypoplastic or aplastic thyroid glands. Other causes are less common and include dyshormonogenesis, maternal ingestion of antithyroid medication, hypothalamic–pituitary disorders, and defects in thyroglobulin metabolism. The dyshormonogenic disorders are inherited as autosomal-recessive conditions. Congenital thyroid deficiency may result in impaired neurologic development if not treated before 1 month of age.

Clinical Manifestations

Clinical symptoms and signs of congenital hypothyroidism may be subtle, especially during the first month of life. Severely affected infants may be relatively large at birth, have a large posterior fontanel, manifest hypothermia and hypoactivity, feed poorly, tend to become constipated, and have prolonged jaundice. An enlarged tongue, coarse facies, and a hoarse cry may also be noted but are unusual in the first weeks of life. An umbilical hernia may be present. If treatment is not started, the physical characteristics become more prominent as the child grows older. Thyroid function tests beyond the first 2 days of life are most useful diagnostically. The TSH level is elevated in primary hypothyroidism, and the T_4 level is low or normal for age. A thyroid scan (^{123}I) may be helpful in identifying the particular type of primary hypothyroidism, but treatment should not be delayed to obtain this study. A low T_4 level in the absence of elevated TSH level may result from a deficiency of thyroid-binding globulin, a pituitary deficiency of TSH, or prematurity.

Management

In term infants, treatment with l-thyroxine, 37.5 µg once daily should be instituted as soon as the relevant diagnostic tests are performed. This dosage can be adjusted to maintain a T_4 level between 10 and 14 µg/dL and a TSH value of less than 5.5 µU/mL after a few weeks of treatment. Both undertreatment and overtreatment must be avoided. Careful follow-up on a monthly basis during the first several months, preferably by a physician who is accustomed to dealing with congenital hypothyroidism, is strongly recommended.

Suggested Readings

DIABETIC KETOACIDOSIS

Burghen GA, Etteldorf JN, Fisher JN, et al. Comparison of high-dose and low-dose insulin by continuous intravenous infusion in the treatment of diabetic ketoacidosis in children. Diabetes Care 1980;3:15–20.

Butkiewicz EK, Liebson CL, Obrien PC, et al. Insulin therapy for diabetic ketoacidosis: bolus injection versus continuous insulin infusion. Diabetes Care 1995;18:1187–1190.

Finberg L. Why do patients with diabetic ketoacidosis have cerebral swelling, and why does treatment sometimes make it worse? Arch Pediatr Adol Med 1996;150:785–786.

Hale PM, Rezvani I, Bruanstein AW, et al. Factors predicting cerebral edema in young children with diabetic ketoacidosis and new onset type 1 diabetes. Acta Paediatr 1997;86:626–631.

Jacobson AD, Hauser ST, Willett J, et al. Consequences of irregular versus continuous follow-up in children and adolescents with insulin dependent diabetes mellitus. J Pediatr 1997;131:727–733.

Klekamp J, Churchwell KB. Diabetic ketoacidosis in children: initial clinical assessment and treatment. Pediatr Ann 1996;25:387–393.

Krane EJ, Rockoff MA, Wallman JK, et al. Subclinical brain swelling in children during treatment of diabetic ketoacidosis. N Engl J Med 1985;312:1147–1150.

Lindsey R, Bolte RG. The use of insulin bolus in low-dose insulin infusion for pediatric diabetic ketoacidosis. Pediatr Emerg Care 1989;5:77–79.

Mel JM, Werther GA. Incidence and outcome of diabetic cerebral oedema in childhood: are there predictors? J Pediatr Chi Health 1995;31:17–20.

Morris AD, Boyle DIR, McMahon AG, et al. Adherence to insulin treatment, glycaemic control and ketoacidosis in insulin dependent diabetes mellitus. Lancet 1997;350:1505–1510.

Rosenbloom AL, Hamas R. Diabetic ketoacidosis (DKA): treatment guidelines. Clin Pediatr 1996;35:261–266.

Thompson CJ, Cummings F, Chalmers J, et al. Abnormal insulin treatment behavior: a major cause of ketoacidosis in the young adult. Diabet Med 1995;12:429–432.

HYPOGLYCEMIA

Aynsley-Green A. Hypoglycemia in infants and children. Clin Endocr Metab 1982;11:159–193.

Haymond MW. Hypoglycemia in infants and children. Endocrinol Metab Clin North Am 1989;18:211–252.

LaFranchi S. Hypoglycemia of infancy and childhood. Pediatr Clin North Am 1987;34:961–982.

Pagliara AS, Karl IE, Haymond M, et al. Hypoglycemia in infancy and childhood. Parts I and II. J Pediatr 1973;82:365-379, 558–577.

Stanley CA, Baker L. Hyperinsulinism in infants and children: diagnosis and therapy. Adv Pediatr 1976;23:315–355.

HYPOPITUITARISM

DeVile CJ, Stanhope R. Hydrocortisone replacement therapy in children and adolescents with hypopituitarism. Clin Endocrinol 1997;47:37–41.

Lovinger RD, Kaplan SL, Grumbach MM. Congenital hypopituitarism associated with neonatal hypoglycemia and microphallus: four cases secondary to hypothalamic hormone deficiencies. J Pediatr 1975;87:1171–1181.

Rogol AD. Hypopituitarism. Curr Ther Endocrinol Metabol 1994;5:26–29.

Stahnke N, Koehn H. Replacement therapy in hypothalamus-pituitary insufficiency: management in the adolescent. Horm Res 1990;4(Suppl 33):38–44.

Willnow S, Kiess W, Butenandt O, et al. Endocrine disorders in septo-optic dysplasia: evaluation and follow-up of 18 patients. Eur J Pediatr 1996;155:179–184.

ACUTE ADRENAL INSUFFICIENCY

August GP. Treatment of adrenocortical insufficiency. Pediatr Rev 1997;18:59–62.

New MI. Replacement doses of glucocorticoids. J Pediatr 1991;119:161.

Urban MD, Kogut MD. Adrenocortical insufficiency in the child. Curr Ther Endocrinol Metab 1994;5:131–135.

CONGENITAL ADRENAL HYPERPLASIA

Azziz R, Dewailly D, Owerbach D. Nonclassical adrenal hyperplasia: current concepts. J Clin Endocrinol Metab 1994;78:810–815.

Cutler GG Jr, Laue L. Congenital adrenal hyperplasia due to 21-hydroxylase deficiency. N Engl J Med 1990;323:1906–1913.

Miller WL. Congenital adrenal hyperplasia. Endocrinol Metab Clin North Am 1991;20:721–749.

Newfield RS, New MI. 21-hydroxylase deficiency. Ann NY Acad Sci 1997;816:219–229.

Pang S. Congenital adrenal hyperplasia. Endocrinol Metab Clin North Am 1997;26:853–891.

PHEOCHROMOCYTOMA

Caty MG, Coran AG, Geagen M, et al. Current diagnosis and treatment of pheochromocytoma in children. Arch Surg 1990;125:978–981.

Ein SH, Pullerits J, Crighton R, et al. Pediatric pheochromocytoma: a 36-year review. Pediatr Surg Int 1997;12:595–598.

Fonkalsrud EW. Pheochromocytoma in childhood. Prog Pediatr Surg 1991;26:103–111.

Lenders JWM, Keiser HR, Goldstein DS, et al. Plasma metanephrines in the diagnosis of pheochromocytoma. Ann Intern Med 1995;123:101–109.

Werbel SS, Ober KP. Pheochromocytoma: update on diagnosis, localization and management. Med Clin North Am 1995;79:131–153.

DIABETES INSIPIDUS

Buonocore CM, Robinson AG. The diagnosis and management of diabetes insipidus during medical emergencies. Endocrinol Metab Clin North Am 1993;22:411–413.

Harris AS. Clinical experience with desmopressin: efficacy and safety in central diabetes insipidus and other conditions. J Pediatr 1989;144:711–718.

Knoers N, Monnens LA. Nephrogenic diabetes insipidus: clinical symptoms, pathogenesis, genetics and treatment. Pediatr Nephrol 1992;6:476–482.

Lee YJ, Huang FY, Shen EY, et al. Neurogenic diabetes insipidus in children with hypoxic encephalopathy: six new cases and a review of the literature. Eur J Pediatr 1996;155:245–248.

Niaudet P, Dechaux M, Trivin C, et al. Nephrogenic diabetes insipidus: clinical and pathophysiologic aspects. Adv Nephrol Necker Hosp

1984;13:247–260.

Ober KP. Endocrine crisis: diabetes insipidus. Crit Care Clin 1991;7:109–125.

Seckl JR, Dunger DB. Diabetes insipidus: current treatment recommendations. Drugs 1992;44:216–224.

SYNDROME OF INAPPROPRIATE ANTIDIURETIC HORMONE SECRETION

Ganong CA, Kappy MS. Cerebral salt wasting in children: the need for recognition and treatment. Am J Dis Child 1993;147:167–169.

Kaplan SL, Feigin RD. Syndromes of inappropriate secretion of antidiuretic hormone in children. Adv Pediatr 1980;27:247–274.

Kappy MS, Ganong CA. Cerebral salt wasting in children: the role of atrial natriuretic factor. Adv Pediatr 1996;43:271–308.

Robinson AG. Disorders of antidiuretic hormone secretions. Clin Endocrinol Metab 1985;14:55–88.

HYPERPARATHYROIDISM

Lawson ML, Miller SF, Ellis G, et al. Primary hyperparathyroidism in a paediatric hospital. QJM 1996;89:921–932.

Matsuo M, Okita K, Takemine H, et al. Neonatal primary hyperparathyroidism in familial hypocalciuric hypercalcemia. Am J Dis Child 1982;136:728–731.

Ross AJ, Cooper A, Attie MF, et al. Primary hyperparathyroidism in infancy. J Pediatr Surg 1986;21:493–499.

HYPOPARATHYROIDISM

Rosen JF, Fleischman AR, Finberg L, et al. 1,25-Dihydroxycholecalciferol: its use in the long-term management of idiopathic hypoparathyroidism in children. J Clin Endocrinol Metab 1977;45:457–468.

Winter WE, Silverstein JH, Maclaren NK, et al. Autosomal-dominant hypoparathyroidism with variable, age-dependent severity. J Pediatr 1983; 103:387–390.

RICKETS

Chesney RW, Mazess RB, Rose P, et al. Long-term influence of calcitriol and supplemental phosphate in X-linked hypophosphatemic rickets. Pediatrics 1983;71:559–567.

Dwyer JT, Dietz WH, Hass G, et al. Risk of nutritional rickets among vegetarian children. Am J Dis Child 1979;133:134–140.

Harrison HE, Harrison HC. Rickets then and now. J Pediatr 1971;87: 1141–1149.

Lovinger RD. Rickets. Pediatrics 1980;66:359–365.

Swischuk LE, Hayden CK Jr. Rickets: a roentgenographic scheme for diagnosis. Pediatr Radiol 1979;8:203–208.

THYROID STORM

Foley TP Jr. Thyrotoxicosis in childhood. Pediatr Ann 1992;21:43–49.

Foley TP Jr, Charron M. Radioiodine treatment of juvenile Graves disease. Exp Clin Endocrinol Diab 1997;105(Suppl 4):61–65.

Hung W. Graves' disease in children. Curr Ther Endocrinol Metab 1997; 6:77–81.

NEONATAL THYROTOXICOSIS

Dirmikis SM, Munro DS. Placental transmission of thyroid-stimulating immunoglobulins. Br Med J 1975;2:665–666.

Foley TP Jr. Maternally transferred thyroid disease in the infant: recognition and treatment. Adv Exp Med Biol 1991;299:209–226.

Singer J. Neonatal thyrotoxicosis. J Pediatr 1977;91:749–750.

CONGENITAL HYPOTHYROIDISM

Barsano CP, DeGroot LJ. Dyshormonogenetic goitre. Clin Endocrinol Metab 1979;8:145–165.

Burrow GN, Dussault JH. Neonatal Thyroid Screening. New York: Raven, 1980.

Fisher DA. Effectiveness of newborn screening programs for congenital hypothyroidism. Pediatr Clin North Am 1987;34:881–890.

Fisher DA, Klein AH. Thyroid development and disorders of thyroid function in the newborn. N Engl J Med 1981;304:702–712.

New England Congenital Hypothyroidism Collaborative. Characteristics of infantile hypothyroidism discovered on neonatal screening. J Pediatr 1984;104:539–544.

CHAPTER 98

Metabolic Emergencies (Inborn Errors of Metabolism)

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The inborn errors of metabolism comprise a group of clinically diverse disorders that usually involve a deficiency of an enzyme or transport system. Consequently, one or more metabolites accumulate in body fluids, often to a concentration that causes serious toxicity of the central nervous system. This chapter focuses on those disorders in which coma and other neurologic signs are major clinical findings. A summary of these diseases, together with suggestions for initial evaluation and treatment, is given in Table 98.1.

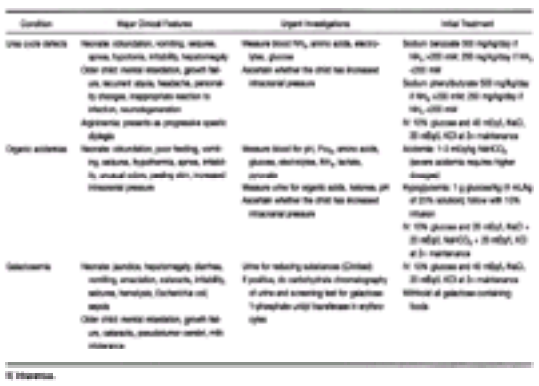

Table 98.2. General Features of Inborn Errors of Metabolism

Failure to make an early diagnosis, when appropriate treatment often can be administered, may lead to irreparable brain injury. Physicians therefore should consider the inborn errors of metabolism in the differential diagnosis of the patient with unexplained neurologic findings. The alert caregiver should request appropriate laboratory tests when indicated by the history and physical findings. The latter are summarized in Table 98.2.

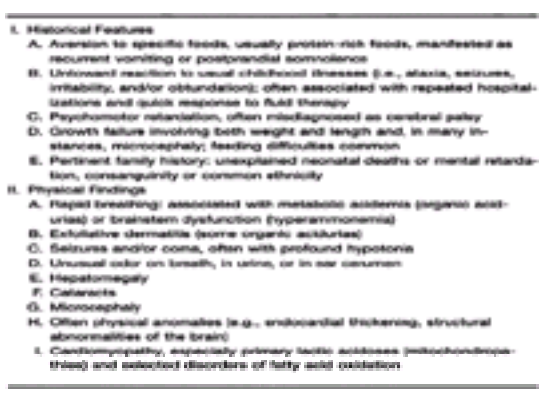

Table 98.1. Summary of Clinical Features, Investigations, and Initial Treatment of Inborn Errors of Metabolism

A careful history is essential. Is the disease episodic? Has an acute attack of obtundation, seizures, and acidosis been preceded by a high intake of protein or an acute infection? Even otherwise trivial infections may precipitate a fulminant syndrome. Parents sometimes report that their child has been admitted repeatedly to the hospital, only to recover after administration of intravenous fluids and glucose. Such a history, especially if coupled with psychomotor retardation and/or growth failure, should always prompt consideration of an inherited metabolic defect.

Many discrete metabolic diseases have been identified. The spectrum of clinical manifestations is broad, ranging from clinically silent biochemical derangements that are detected through a routine screening program to potentially fatal disturbances (e.g., the urea cycle defects, which are characterized by a dramatic clinical presentation entailing sudden

neurologic decompensation).

Metabolic diseases are relatively rare, often occurring perhaps only once in 100,000 or 200,000 live births. A precise estimate is elusive because some cases remain undetected. In addition, the classical phenocopy of a given biochemical defect often is not the only mode of clinical presentation. The literature now abounds with descriptions of variant forms that manifest a later age of onset and a clinical course that is milder than that of the more complete metabolic lesion; the latter usually is described first because of the more dramatic clinical findings that are associated with a pervasive enzymatic deficiency.

UREA CYCLE DEFECTS

Background

All cells produce ammonia as an end-product of nitrogen metabolism. Because high ammonia concentrations are toxic to the brain, various biochemical strategies have evolved for the efficient disposal of this metabolite. Many animals, including humans, convert ammonia to urea via the urea cycle, as shown in Figure 98.1. Waste nitrogen is thereby eliminated as urea, which is excreted into the urine. The most vivid example of the importance of the urea cycle to metabolism is the hyperammonemia that occurs in hepatic failure or a congenital deficiency of one of the urea cycle enzymes (Table 98.3).

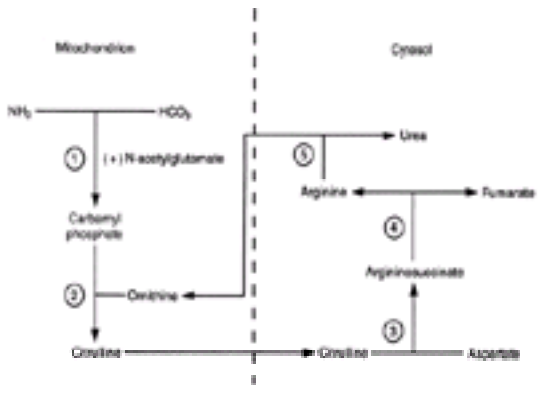

FIGURE 98.1. The urea cycle. Enzymes: *1,* carbamyl phosphate synthetase; *2,* ornithine transcarbamylase; *3,* argininosuccinate synthetase; *4,* argininosuccinate lyase; *5,* arginases. The symbol + indicates that *N*-acetylglutamate is a positive effector for carbamyl phosphate synthetase.

I. Primary Urea Cycle Enzyme Deficiencies	II. Hepatic Amino Acid Transport Defects
A. Carbamyl phosphate synthetase deficiency	A. Lysinuric protein intolerance
B. Ornithine transcarbamylase deficiency	B. Homocitrullinuria, hyperornithinemia, and hyperammonemia
C. Citrullinemia	
D. Argininosuccinic aciduria	
E. Argininemia	

Table 98.3. Inherited Causes of Hyperammonemia

Pathophysiology

The relationship between elevated ammonia levels and impaired neurologic function remains poorly understood. A derangement of energy metabolism may be involved because adenosine triphosphate (ATP) levels can be low, particularly in the brainstem reticular-activating system. Energy metabolism may be compromised by interference with the malate-aspartate shuttle, which is responsible for the transport of reducing equivalents from cytoplasm to mitochondria, thereby permitting the storage of energy released from glycolysis. Other evidence links ammonia toxicity with a derangement of neuronal membrane ion transport, especially chloride conductance. Recently, elevated concentrations of brain glutamine, which is formed from ammonia and accumulates in hyperammonemic conditions, has been shown to lead to a loss in the integrity of the blood–brain barrier and to swelling of brain cells. Ammonia toxicity also may impair the metabolism of brain glutamate, the major excitatory neurotransmitter, as well as the activity of glutamate receptors.

Clinical Manifestations

Signs and Symptoms

A variety of clinical presentations may accompany hyperammonemia. The severity of the clinical syndrome usually reflects the completeness of the inherited enzymatic defect, with milder syndromes being associated with a relative

preservation of enzyme activity.

Neonatal Catastrophe

The infant appears well at birth, although the ammonia level may be high in umbilical blood. The disorder usually manifests during the first few days of life, after the baby ingests protein in infant formula or breast milk. Vomiting is a common early finding, followed by poor feeding, a loss of muscle tone, and a progressive diminution of consciousness culminating in frank coma. Convulsions occur in many cases. Hepatomegaly without liver failure is often observed. If timely treatment is not initiated, ventilatory failure secondary to brainstem involvement develops by the end of the first or second week. Death is common, and the prognosis is poor for complete neurologic recovery among surviving babies. The duration of coma, rather than the severity of the hyperammonemia, best predicts outcome.

Recurrent Coma in the Older Infant and Child with Retarded Psychomotor Development

Urea cycle defects may be the true cause of developmental delay in the older infant or child who is thought to have cerebral palsy. Careful questioning of the parents may elicit a history of vomiting and/or lethargy after ingestion of protein-rich foods. Indeed, many patients intuitively avoid meat, chicken, fish, and so on. The marginal dietary intake may result in growth failure.

An important clinical feature is that minor infections may precipitate hyperammonemia. The reason for this relationship is that infections may evoke a protein catabolic response that leads to increased ammonia production. Affected youngsters may be admitted repeatedly to the hospital before the metabolic defect is appreciated.

Recurrent Vomiting and Ataxia

Frank psychomotor retardation does not occur, but physical development is poor because of inadequate dietary intake. Vomiting, dizziness, ataxia, and obtundation may result in repeated hospitalizations. Patients may be diagnosed incorrectly as having vestibular dysfunction, migraine, or even epilepsy. School performance may be poor. Parents may report that the child often seems listless and has a brief attention span. An aversion to foods rich in protein is common. The acute symptoms improve after treatment with parenteral fluids, but symptoms reappear after a few weeks or months.

Progressive Neurologic Deterioration

These patients seem normal for years, after which intellectual performance and overall neurologic function decline progressively. The pathophysiology associated with an extended period of normal development and subsequent decompensation is unknown.

Laboratory Findings

Blood ammonia levels are usually elevated in the acutely ill child. Infants with near complete enzymatic defects may have levels greater than 1000 µg/dL (normal is less than 50 µg/dL). Lesser increases are often seen in the older infant or child, even when acute symptoms are present. Conversely, a patient occasionally will seem asymptomatic even though the blood ammonia is 100 to 150 µg/dL. The reason for the absence of a strict correlation between the blood [NH_3] and the clinical findings remains uncertain. One possibility is that ammonia is not the only toxin. Thus, some evidence points to a toxic role for glutamine, which the brain forms in large amounts during hyperammonemia. Symptoms may also reflect not only the magnitude of the hyperammonemia but also the rate of rise of the ammonia concentration. The brain of the chronically hyperammonemic child may have adjusted to the metabolic perturbation. An acute neurologic syndrome occurs when the level increases abruptly.

The blood urea may be low, as would be anticipated in a disorder of ureagenesis, but a normal blood urea does not exclude a congenital hyperammonemia syndrome, especially if the child is dehydrated from vomiting and renal urea excretion is consequently diminished. Furthermore, in patients with an incomplete enzymatic defect, enough urea may be produced to bring this metabolite into the low-normal range.

The blood electrolytes, including pH, are usually normal. Although ammonia is a base, the amounts that accumulate are not large enough to cause metabolic alkalosis. Conversely, metabolic acidosis does not occur unless the patient experiences cardiorespiratory arrest. The latter point deserves emphasis; clinicians often erroneously assume that the absence of acidosis is evidence against the existence of an inborn error of metabolism.

The routine urinalysis is normal. However, careful microscopic examination of the urine in the patient with ornithine transcarbamylase (OTC) deficiency, the most common urea cycle defect, may reveal the presence of orotic acid crystals. These long, spindle-shaped crystals are excreted in excess because carbamyl phosphate, a substrate for the OTC reaction, is produced in the liver mitochondria but is not converted to citrulline (Fig. 98.1). The excess carbamyl phosphate spills over into the cytoplasm, where it is used to make orotic acid, a precursor to pyrimidines. Orotic aciduria in OTC deficiency is of diagnostic importance. The peripheral blood count is nondiagnostic.

The cerebrospinal fluid (CSF), glucose, and protein levels are normal unless intraventricular hemorrhage has occurred. Both brain and pulmonary hemorrhage can occur, especially in the fulminant hyperammonemia syndrome encountered in the neonate. The CSF ammonia is elevated. The amino acid concentrations of the CSF will reflect the underlying urea cycle defect. Glutamine is often increased, particularly when the ammonia level is high. Other CSF amino acids (e.g., citrulline, argininosuccinate, arginine) will be elevated depending on the underlying enzymatic defect.

Extreme hyperammonemia may be accompanied by elevation of the blood transaminases and other enzymes marking liver dysfunction. Indeed, the urea cycle defects have been confused with acquired syndromes of hyperammonemia and

liver failure (e.g., Reye syndrome). Differentiation between these disorders may require microscopic examination of a liver biopsy to detect the characteristic microvesicular fat deposition of Reye syndrome.

The blood amino acids are important to diagnosis. As noted in Figure 98.1, an enzymatic defect of one of the three final steps in the urea cycle should produce elevations in blood citrulline, argininosuccinic acid, or arginine, respectively. An increased concentration of one of these amino acids in a hyperammonemic child constitutes a presumptive diagnosis of a specific urea cycle defect. If none of these three amino acids is increased in blood but levels of glutamine and alanine, the primary nitrogen carriers, are increased, an enzymatic block may exist at the level of either carbamyl phosphate synthetase (CPS) or OTC. Quantitation of urine orotic acid, which is high in OTC deficiency and normal or low in CPS deficiency, often differentiates between these two disorders. OTC deficiency is far more common than CPS deficiency.

Definitive diagnosis requires enzymatic assay and/ or analysis of deoxyribonucleic acid (DNA). If a patient dies in the emergency department (ED), the physician should try to secure appropriate postmortem samples of tissue, blood, and urine. Identification of the enzymatic lesion is important for genetic counseling. Liver tissue should be obtained quickly and kept frozen, if possible at –80°C (–176°F), until assay of the urea cycle enzymes can be undertaken.

Management (See Table 98.1)

Blood ammonia must be lowered as quickly as possible because the long-term prognosis depends on the severity and duration of hyperammonemia. Effective management should include a double-pronged attack: 1) minimization of ammonia production and 2) facilitation of ammonia removal. The latter goal has been abetted in recent years by the development of new antidotes to ammonia toxicity and by the increasing use of dialysis in the pediatric population.

Parenteral fluids should be administered to stimulate urine flow and renal ammonia excretion. The infusion of glucose augments tissue anabolism by stimulating secretion of insulin, which promotes protein synthesis and minimizes protein catabolism. The administration of 10% glucose and 45 mEq/L sodium chloride at twice the maintenance fluid rate should be adequate. Brain swelling sometimes occurs, and fluid administration should be adjusted accordingly.

Unless the patient has acidemia, bicarbonate should not be given because alkalinization of the blood favors conversion of NH_4^+ to NH_3, which more readily traverses the blood–brain barrier than does the ionized species. Similarly, maintenance of a relatively low urine pH favors excretion of NH_4.

Dialysis and exchange transfusion have been used to remove ammonia, with the former technique being more effective. Blood ammonia can be reduced dramatically by peritoneal dialysis, hemodialysis, or continuous arteriovenous hemofiltration. This treatment is particularly effective in the neonate with severe hyperammonemia (greater than 500 µg/dL), in whom dialysis for 2 to 3 days lowers blood [NH_3]. The potential risks of short-term dialysis are outweighed by the risks of prolonged hyperammonemia. Clearly, dialysis should only be attempted at centers with the requisite expertise.

Highly effective antidotes to ammonia toxicity have been developed in recent years. These antidotes are compounds that react with amino acids to form acyl conjugates that are rapidly excreted into the urine, thereby providing a mechanism, in addition to urea synthesis, by which excess nitrogen is eliminated.

One such agent is sodium benzoate. As illustrated in Figure 98.2, benzoate and glycine are converted to hippurate, which is excreted readily in the urine. Because glycine nitrogen is derived directly from ammonia, excretion of the latter as hippurate is an effective detoxification mechanism. This treatment has been particularly useful for the management of congenital defects in either of the first two steps of the urea cycle (i.e., carbamyl phosphate synthetase deficiency or ornithine transcarbamylase deficiency). In the ED, it is preferable to administer benzoate (250 to 500 mg/kg per day) as a parenteral (10%) solution. Toxicity associated with benzoate treatment is relatively low.

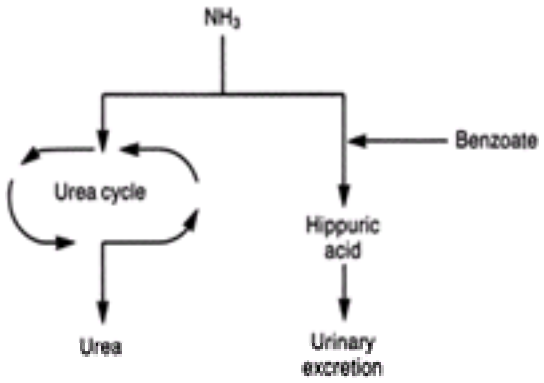

FIGURE 98.2. The benzoate effect.

Benzoate treatment can be supplemented with enteral (oral or nasogastric) sodium phenylbutyrate (250 to 500 mg/kg per day), which the liver converts to phenylacetate. The latter is then conjugated with the glutamine to form phenylacetylglutamine, which is cleared quickly by the kidney. Because glutamine contains 2 atoms of nitrogen, in contrast to the single atom present in glycine, therapy with phenylbutyrate is even more efficient than benzoate treatment in promoting the elimination of waste nitrogen.

For defects involving either of the next two steps of the urea cycle (i.e., citrullinemia or argininosuccinicaciduria),

treatment with arginine (1 to 3 mmol/kg per day) is indicated. Arginine therapy, which can be administered parenterally, reduces blood ammonia by a variety of mechanisms. First, the arginine is cleaved in the liver to urea and ornithine, the latter being a positive effector for the synthesis of *N*-acetylglutamate, which stimulates activity of carbamyl phosphate synthetase, the first step of the urea cycle (Fig. 98.1). In addition, because the patient with either citrullinemia or argininosuccinicaciduria has a relative inability to produce arginine via the urea cycle, overall protein synthesis is diminished. Administration of arginine corrects this deficiency and thereby favors protein anabolism and minimizes catabolism and concomitant production of ammonia. Finally, arginine treatment promotes formation of either citrulline or argininosuccinic acid, both of which are excreted in large amounts, carrying with them NH_3 on an equimolar basis.

If parenteral solutions are unavailable, benzoate, phenylbutyrate, and arginine can be administered by a nasogastric tube. A 10% solution in water is usually convenient. A therapeutic effect, that is, a lowering of blood ammonia, should be noted in about 2 hours.

In addition to the treatments already described, gut sterilization may minimize ammonia formation by enteric urea-splitting bacteria. Absorption of ammonia from the gut can be lowered by giving lactulose (5 mL for infants, 10 to 20 mL for children), a nonabsorbable carbohydrate that acidifies the colon and thereby favors formation of NH_4^+ rather than the more readily absorbed NH_3.

The mainstay of long-term therapy is a low-protein diet. During the first year of life, 1.5 to 2 g/kg per day of protein are necessary to satisfy the growth requirement. Thereafter, an allotment of approximately 1.0 g/kg per day should be sufficient. In most patients, the combination of a low-protein diet and supplemental sodium phenylbutyrate and/or arginine is compatible with reasonable control.

ORGANIC ACIDURIAS

Background

All cells oxidize carbohydrates, fats, and amino acids to yield carbon dioxide, water, and in the case of the amino acids, ammonia. This metabolic interconversion does not occur in a single step, however. Instead, carbohydrates, fats, and amino acids are first converted to metabolic intermediates, many of which are organic acids, as illustrated in Figure 98.3.

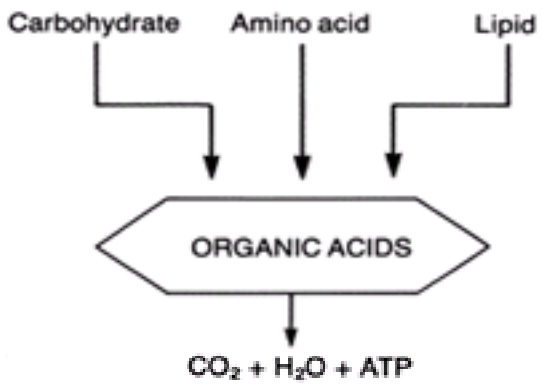

FIGURE 98.3. Formation of organic acids. *ATP,* adenosine triphosphate.

The term *organic aciduria* refers to a class of diseases characterized by excessive accumulation of one or more organic acids in body fluids. This reaction may occur as a secondary phenomenon, as in the lactic acidosis consequent to tissue hypoxia. In this case, lactate cannot be converted to pyruvate because of an increased cytoplasmic NADH:NAD ratio in hypoxic tissue. Another common cause of secondary organic aciduria is the ketoacidosis of diabetes mellitus, in which a lack of insulin leads to increased production of acetoacetate and 3-hydroxybutyrate from fatty acids.

This section focuses on the primary organic acidurias, that is, a group of inherited disorders that are usually caused by the congenital deficiency of an enzyme mediating the metabolism of a particular organic acid. As a result, the acid accumulates, often to toxic levels. As noted in Table 98.4, the number of disorders is large because of the diverse metabolic pathways involved in the oxidation of carbohydrates, fats, and amino acids. The associated clinical presentations are also varied, although sufficient features are shared to warrant consideration as a single group.

I. Disorders of Amino Acid Metabolism
A. Maple syrup urine disease
B. Isovaleric acidemia
C. Propionic acidemia
D. Methylmalonic acidemia
E. β-Ketothiolase deficiency
F. β-Methylcrotonic aciduria
G. Holocarboxylase synthetase deficiency
H. Biotinidase deficiency
I. Glutaric aciduria (types I and II)
J. β-Methylglutaconic aciduria
II. Disorders of Carbohydrate and Energy Metabolism
A. Primary lactic acidosis
1. Pyruvate dehydrogenase deficiency
2. Pyruvate carboxylase deficiency
3. Inherited defects of the tricarboxylic acid cycle
4. Disorders of the electron transport chain
B. Glyceric aciduria
III. Disorders of Fatty Acid Oxidation
A. Medium-chain acyl-CoA dehydrogenase deficiency
B. Long-chain acyl-CoA dehydrogenase deficiency
C. Very-long-chain acyl-CoA dehydrogenase deficiency
D. Carnitine palmitoyltransferase II deficiency
E. Type II glutaric aciduria
F. 3-Hydroxy-acyl-CoA dehydrogenase deficiency
G. Primary carnitine deficiency

Table 98.4. Inherited Organic Acidurias

The occurrence of individual disorders is relatively low, with an incidence of perhaps 1 in 100,000 to 200,000 live births. The collective incidence, of course, is much higher. As awareness of these disorders has increased and the technology necessary for diagnosis has become more available, many more cases have been detected.

Pathophysiology

The central nervous system is usually prominently involved. Untoward concentrations of certain compounds (e.g., methylmalonate) have deleterious effects on cerebral oxidative metabolism. These compounds may also inhibit important hepatic metabolic pathways, such as ureagenesis and gluconeogenesis, thereby causing hyperammonemia or hypoglycemia. Disorders of lactate oxidation (e.g., pyruvate dehydrogenase deficiency, electron transport defects) are usually damaging to the brain; the degree of damage depends on the severity of the block of oxidation of lactate and pyruvate derived from glucose via glycolysis.

Acidosis is a common, although not invariant, feature of the organic acidurias. On rare occasions, the acidosis is the consequence of impaired renal bicarbonate reabsorption and consequent proximal renal tubular acidosis.

A more common kind of acidosis, especially during acute illness, is associated with an anionic gap. This abnormality develops when the positive charges (cations) in the blood (Na^+ plus K^+) significantly exceed the negative charges (anions) (Cl^- plus HCO_3^-). Because blood is electrically neutral, the sum of the positive and negative charges must be equal. As shown in Figure 98.4, the anions of blood are primarily chloride and bicarbonate, with lesser contributions from sulfate, phosphate, and negatively charged proteins. Few negative charges are contributed by the organic acids, which are ordinarily oxidized completely to carbon dioxide and water (Fig. 98.3). However, a congenital deficiency of an enzyme mediating the breakdown of a particular organic acid will increase the contributions such acids make to the anions of the blood.

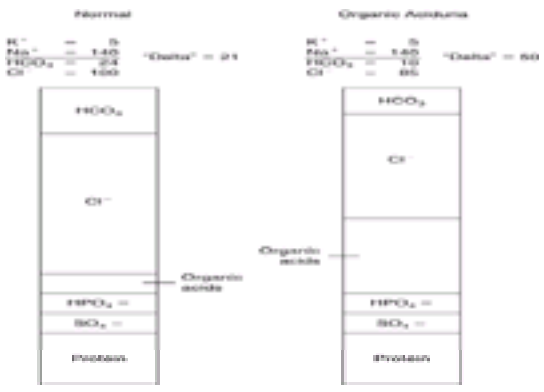

FIGURE 98.4. The anionic gap.

The clinician may infer the presence of a large organic acid concentration by inspection of the routine blood electrolytes. As noted in Figure 98.4, if the total of cationic charges is 145 mEq/L and the sum of chloride plus bicarbonate is only 95 mEq/L, an anionic gap of some 50 mEq/L of negative charges compared with a normal anionic gap of only 15 to 25 mEq/L clearly exists. The unidentified negative charges could have been derived from phosphate or even sulfate rather than from the carboxylate anions, but discriminating clinically among these possibilities is usually not difficult.

In addition to the central nervous system, the toxicity of organic acid accumulation extends to the liver and bone marrow. Hepatic gluconeogenesis and ureagenesis are often impaired. The precise mechanisms are poorly defined, although a likely factor is a depletion of free intracellular coenzyme A, which is consumed rapidly by esterification with the accumulated organic acid. Because these metabolic processes, especially ureagenesis, depend on an adequate supply of coenzyme A, hyperammonemia may supervene.

The bone marrow toxicity reflects an inhibition in the maturation of leukocyte and platelet precursors rather than an inhibition of stem cell formation. Neutropenia and thrombocytopenia result and may cause confusion of the organic acidurias with septicemia.

Clinical Manifestations

The clinical presentations of the organic acidurias resemble those of the urea cycle defects. Indeed, the two groups of diseases can be confused, particularly because hyperammonemia can occur in several of the organic acidurias. However, certain clinical findings should suggest a primary disorder of organic acid metabolism.

Acute Catastrophe in the Newborn Period

This clinical presentation has been noted in association with almost all the disorders listed in Table 98.4. The infant seems well for the first few days or weeks of life, after which he or she becomes increasingly apathetic and irritable. Progression to frank coma can occur abruptly. Symptoms generally do not become apparent until after institution of the first feedings. Intrauterine development is usually normal because organic acids can be metabolized by the mother. Vomiting, mild hepatomegaly, hypothermia, hypotonia, and convulsions occur often. An unusual symptom may be a

bizarre odor in the urine, breath, perspiration, saliva, or ear cerumen. This finding is attributable to the accumulation of organic acid. The repertoire of possible odors is varied, ranging from the sweet smell of branched-chain keto acids in maple syrup urine disease to the “sweaty socks” scent of isovaleric acid encountered in isovaleric acidemia (Table 98.5).

Disorder	Odor
Maple syrup urine disease	Burnt sugar or maple syrup
Isovaleric acidemia	Sweaty socks or cheeselike
Methylmalonic acidemia	Fruity (from ketosis), ammoniacal
Phenylketonuria	Mouse urine, musty
Tyrosinemia	Cabbagelike
Methionine malabsorption	Malt or hops
3-Methylcrotonic aciduria	Tomcat urine
3-Hydroxy-3-methylglutaric aciduria	Tomcat urine
Propionic acidemia	Fruity (ketosis), ammoniacal

Table 98.5. Odors Associated with Organic Acidurias

The long-term outlook is related to the time elapsed before diagnosis. Treatment (see the following) should be started early to realize maximum benefit.

Recurrent Coma in a Child with Mental Retardation and Growth Failure

A subset of patients come to attention because of delayed psychomotor and physical development. Cerebral palsy may have been misdiagnosed. An otherwise trivial infection may cause sudden clinical decompensation, including vomiting, obtundation, and frank coma. Parents often report an intolerance to protein-rich foods and a history of recurrent vomiting. Physical examination discloses a comatose, hyperventilating child whose weight and height are often below average. Both urine and breath should be scrutinized for unusual odors. Convulsions and focal neurologic findings may be present. Several children have had severe choreoathetosis, reflecting basal ganglia involvement. In others, particularly children with the primary lactic acidoses, spinocerebellar degeneration has been noted.

Older Infant or Child with Acute Decompensation and Liver Failure

These children may have mild to moderate mental retardation and/or growth failure. Some may be asymptomatic until severe vomiting and dehydration develop in association with a stress, usually a viral illness. The progression to altered consciousness and coma may be very rapid. A careful history often reveals that these children have been hospitalized previously with vomiting and loss of consciousness. The family history also is important: a similar presentation in siblings suggests an inborn error of metabolism.

A subset of these children have defects of fatty acid oxidation, such as medium-chain acyl-CoA dehydrogenase deficiency or primary carnitine deficiency. The primary clinical findings are coma, hypoglycemia, hyperammonemia, and hepatomegaly. Muscle weakness and cardiomyopathy may occur. Analysis of the urine organic acids usually discloses a typical pattern of metabolite excretion.

Laboratory Findings

Metabolic acidosis may occur during the acute illness when an anionic gap develops. The alkali requirement may be very high, perhaps 10 to 15 mEq/kg per day of HCO^{-}_{3}, to maintain the blood bicarbonate in the low-normal range.

Patients may occasionally demonstrate hyperchloremic metabolic acidosis, particularly if they are dehydrated from vomiting and/or diarrhea. In rare instances, hyperchloremic metabolic acidosis is also seen in patients with primary lactic acidosis or methylmalonic aciduria and secondary renal tubular acidosis.

Ketonuria is common and probably reflects an inhibition of ketone body oxidation by accumulated organic acids. Significant ketonuria is unusual in newborn infants; its presence should alert the clinician to the likelihood that the baby may have an inherited disorder of organic acid metabolism.

Hypoglycemia is common. In some disorders (e.g., maple syrup urine disease, methylmalonic acidemia), the cause is a primary failure of hepatic gluconeogenesis. In certain disorders of fatty acid oxidation (e.g., medium-chain acyl-CoA dehydrogenase deficiency), the hypoglycemia reflects a rate of glucose consumption that exceeds the rate of glucose production.

Hyperammonemia occurs because high levels of organic acids may inhibit ureagenesis. The hyperammonemia sometimes leads to confusion of the primary organic acidurias with the urea cycle defects. Careful inspection of the concentrations of the blood amino acids and urine organic acids should permit discrimination between these two categories of disease.

Neutropenia and thrombocytopenia are common findings. The occurrence of these derangements in the neonate often suggests the diagnosis of septicemia. Infants may be erroneously treated with antibiotics instead of a specific therapy for organic acidurias (see the following).

The plasma lactate and pyruvate levels are usually elevated in children with primary lactic acidosis, caused either by a primary defect of pyruvate oxidation (e.g., pyruvate dehydrogenase deficiency) or by a defect in the respiratory chain (e.g., cytochrome oxidase deficiency). Special care should be exercised in obtaining blood to measure the lactate level, which is often artifactually increased by extended application of a tourniquet. The lactate concentration may also be artifactually raised by the metabolism of erythrocytes if a delay occurs between blood sampling and analysis. Therefore, laboratory guidelines for blood collection should be followed carefully. In a minority of patients, the plasma lactate level is normal, and the diagnosis of primary lactic acidosis requires quantitation of the CSF lactate level.

Analysis of urine organic acids usually points to a specific syndrome. Organic acid excretion is commonly abnormal even when the patient is clinically stable. However, in children with partial enzymatic defects (i.e., those who have some residual capacity to oxidize the organic acid in question), the biochemical abnormality may be detectable only during episodes of clinical decompensation. Therefore, repeated testing is indicated for children at risk in whom initial testing proves negative. Such testing requires gas–liquid chromatography of the urinary organic acids. Because urine is an exceedingly complex mixture, normally containing many hundreds of different compounds, organic acid analysis should be undertaken in laboratories with the requisite expertise, including gas chromatography–mass spectrometry, to identify with certainty the many peaks in the chromatographic effluent.

The plasma aminogram is not necessarily diagnostic, but it provides useful information. Thus, significant elevations of blood glycine are common in methylmalonic aciduria and propionic acidemia, in which the accumulated organic acid inhibits hepatic oxidation of glycine. The levels of leucine, isoleucine, and valine are extremely high in patients with maple syrup urine disease who lack the ability to metabolize these branched-chain amino acids. The blood alanine is high in children with primary lactic acidosis because alanine is formed rapidly from pyruvate via transamination. Patients with primary lactic acidosis also may have increased levels of proline, reflecting an inhibition of proline oxidase activity by high levels of lactate.

The emergency physician will often lack access to specialized laboratory tests such as gas–liquid chromatography. An alternative approach to diagnosis is the use of simple liquid screening tests that can be performed in any ED. Thus, the ferric chloride reaction is grayish green in the urine of children with maple syrup urine disease. The 2,4-dinitrophenol reaction is positive (i.e., a yellowish precipitate forms), in the same disorder. The para-nitroaniline reaction gives a bright emerald green reaction in the urine of patients with methylmalonic aciduria.

Management

The treatment of a child with primary organic aciduria should be directed to three primary goals: 1) correction of abnormalities (e.g., acidosis) that are consequences of the primary metabolic insult; 2) reduction of organic acid production; and 3) if possible, enhancement of organic acid disposal.

The acidosis may be severe. Indeed, when a patient needs high dosages of alkali to control metabolic acidemia, primary organic aciduria must be considered among the diagnostic possibilities.

The rationale for bicarbonate therapy is twofold. First, acidosis impairs normal metabolism and physiology. In addition, a higher pH favors ionization of the carboxylate anion (COO^- versus COOH). The ionized species is more efficiently excreted into urine. Thus, the urine pH should be kept at 6 or higher. The initial dose of bicarbonate should be 1 to 2 mEq/kg, or based on arterial blood gases. A dosage of 1 to 3 mEq/kg per day should be adequate for most children after recovery from acute decompensation. A notable exception is the rare patient whose organic acidemia also causes renal tubular acidosis (e.g., a child with methylmalonic aciduria) and some of the primary lactic acidoses. In the latter group, extremely high dosages of bicarbonate (8 to 13 mEq/kg per day) are sometimes needed.

In addition to metabolic acidosis, hypoglycemia is common during the acute presentation. The hypoglycemia, which may be severe in maple syrup urine disease and methylmalonic aciduria, is caused by defective gluconeogenesis (see previous section). Therefore, clinicians should carefully monitor the blood glucose. Emergency treatment consists of intravenous administrations of 1 g (4 mL)/kg of 25% glucose followed by a 10% glucose infusion.

The administration of intravenous glucose also alleviates the ketoacidemia, which may be severe. This reaction occurs because glucose infusions favor the secretion of insulin, which inhibits the production of ketone bodies from fatty acids.

Organic acid production must be minimized by avoiding the administration of compounds that are precursors to the organic acid in question. Most clinically relevant organic acids are derived from amino acids; therefore, protein intake should be restricted. In the ED, the patient should receive adequate glucose (i.e., a 10% glucose infusion) to provide a source of calories and to minimize endogenous protein catabolism. Such treatment also helps correct the hyperammonemia that often accompanies these disorders (see previous section).

The organic acidurias are best conceptualized as toxicity syndromes, and a specific antidote is often available, as indicated in Table 98.6. Two kinds of antidotes have been developed: 1) a particular vitamin that, at a high dosage, stimulates the congenitally defective metabolic pathway, thereby allowing increased oxidation of the accumulated organic acid, and 2) a compound that reacts with the organic acid to form a conjugate that is excreted more efficiently into the urine. An example of the latter approach is the use of glycine to treat children with isovaleric acidemia. The hepatic enzyme glycine *N*-acylase mediates the formation of isovalerylglycine from glycine and isovalerate. Glycine therapy can safely reduce the concentration of isovaleric acid in many patients.

Thiamine (25–100 mg/day, orally or parenterally)
Maple syrup urine disease
Primary lactic acidosis with megaloblastic anemia
Biotin (10–40 mg/day, orally or parenterally)
Holocarboxylase synthetase deficiency
Biotinidase deficiency
Vitamin B_{12} (2 mg intramuscularly)
Methylmalonic acidemia with homocystinuria
Folic acid (1–5 mg/day, orally or parenterally)
Methylmalonic acidemia with homocystinuria
Lipoic acid (25–50 mg/kg/day, orally)
Primary lactic acidosis secondary to lipoamide dehydrogenase deficiency
Vitamin C (50 mg/kg/day) and vitamin K (0.6 mg/kg/day as menadione)
Primary lactic acidosis secondary to electron transport defect
Glycine (250 mg/kg/day, orally or parenterally)
Isovaleric acidemia
Carnitine (50 mg/kg/day, orally)
Carnitine deficiency
Dichloroacetate (experimental, dose not established)
Primary lactic acidosis

Table 98.6. Specific Treatments for Organic Acidurias

Another therapeutic approach is the administration of L-carnitine. The blood concentration of carnitine, a cofactor for the oxidation of long-chain fatty acids, is significantly diminished in several organic acidurias, perhaps because of increased excretion as carnitine esters. Exogenous carnitine may favor esterification and thereby alleviate the toxicity associated with high free organic acid concentrations.

Detoxification of the child with isovaleric acidemia and other organic acidurias often requires 1 to 2 weeks because much acid is probably sequestered in adipose tissue, from which it is released only slowly. Therefore, initiation of glycine or carnitine treatment is not imperative during the relatively brief interlude most patients spend in the ED.

The organic acidurias that can be managed with either vitamins or conjugating agents are described in Table 98.6.

Recent evidence indicates that dialysis, which efficiently removes ammonia from the body (see previous section), can dramatically reduce organic acid levels in the acutely affected child. Peritoneal dialysis, hemodialysis, and continuous arteriovenous hemofiltration have been used, primarily in the neonate with a fulminant syndrome. The risks of performing such a procedure for 2 to 3 days appear to be less than those of prolonged exposure of the brain to elevated levels of a potentially toxic organic acid. Obviously, dialysis should only be performed by a staff with the necessary expertise.

GALACTOSEMIA

Background and Pathophysiology

Approximately 15% of the calories of human milk are derived from galactose, which together with glucose constitutes the disaccharide lactose. To use this energy, the infant must convert galactose to glucose, which is then oxidized, first via the glycolytic pathway and then in the tricarboxylic acid cycle. The conversion of galactose to glucose is mediated through the uridine nucleotide pathway, as noted in Figure 98.5.

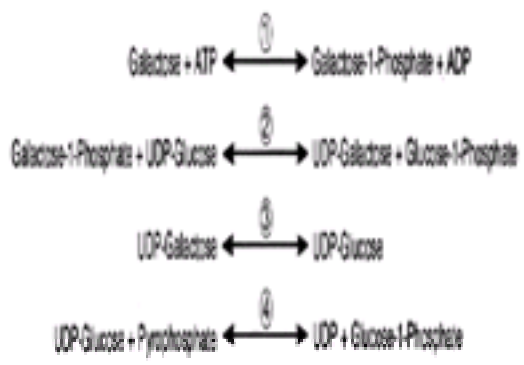

FIGURE 98.5. The uridine nucleotide pathway. *1,* galactokinase; *2,* galactose-1-phosphate uridyltransferase; *3,* epimerase; *4,* UDP-glucose pyrophosphorylase.

Galactosemia is caused by a failure to metabolize galactose to glucose. In most cases, the disease is associated with an inherited deficiency of galactose-1-phosphate uridyltransferase (Fig. 98.5). Rarely, inherited defects of galactokinase or of uridine diphosphate (UDP)-galactose 4-epimerase cause the galactosemia syndrome.

The accumulation of galactose in the body fluids of the transferase-deficient patient affects primarily the brain, liver, eyes, and kidneys. The toxic factor appears to be galactose-1-phosphate, not galactose. Early diagnosis and treatment can attenuate severity of the clinical syndrome.

Clinical Manifestations

Failure to thrive is characteristic and may be the reason for referral. Most affected children also have gastrointestinal problems, particularly vomiting and diarrhea, a few days after exposure to dietary lactose. By the end of the first week of life, the classic patient shows signs of liver disease. Hyperbilirubinemia, which may be intense, is caused by both hepatic dysfunction and hemolysis. Cataracts are present in almost all babies at birth, although a slitlamp examination may be

necessary to visualize an embryonic cataract. Neurologic deterioration with coma ensues in severe cases. The risk of infection is also increased, and death from *Escherichia coli* septicemia is not unusual, often as the initial manifestation of the disorder.

Mental retardation may occur in patients who survive the neonatal period. Indeed, recent data indicate a high incidence of learning disabilities even in children who are treated with a galactose-free diet from an early age. This correlation may reflect either subtle intrauterine injury or an autointoxication syndrome caused by the endogenous production of galactose, which probably continues throughout the life span.

If the toxicity syndrome is not alleviated with a low-galactose diet, patients may develop renal tubular dysfunction with hyperchloremic acidosis and generalized aminoaciduria. Albuminuria is common. Progressive hepatic failure is encountered. Hypergonadotropic hypogonadism is common even in treated female galactosemics. Gonadal function in male patients appears to be normal.

It is important to emphasize that the severity of transferase-deficiency galactosemia varies in different racial groups. Thus, African-American patients tend to have more residual erythrocyte transferase activity than do white patients. The African-American child's tolerance for milk may be correspondingly greater, and the clinical presentation may be delayed until the child is several years of age, at which time hepatomegaly, growth failure, cataracts, and mental retardation become evident. Such patients often receive partial treatment once parents appreciate that the administration of milk evokes vomiting.

Laboratory Findings

A presumptive diagnosis is made by the presence of galactosuria in a symptomatic individual. This diagnosis is accomplished most simply by testing the urine for the presence of a reducing agent. Clinitest tablets are commonly used for this purpose. If a reducing substance is present, the urine should be tested with a paper strip impregnated with glucose oxidase to rule out glycosuria.

Precise quantitation of galactose in the urine or blood requires either enzymatic assay or isolation of the galactose with gas–liquid chromatography.

Most patients have abnormal liver function tests. Jaundice is common, especially in babies who initially develop indirect hyperbilirubinemia followed by direct hyperbilirubinemia 1 to 2 weeks later.

Urine may disclose bicarbonaturia and generalized aminoaciduria. The most important clinical counterpart of the renal tubular dysfunction is hyperchloremic metabolic acidosis.

In some patients, particularly during the newborn period, hemolysis may be prominent and may be so severe that it is confused with erythroblastosis. The hemolysis is a contributing factor to the hyperbilirubinemia.

Definitive diagnosis requires demonstration of the enzyme deficiency in the patient's cells. Erythrocytes are usually used, although the defect is probably present in all tissues. Much simpler, semiquantitative enzyme assays have been developed for screening purposes. These tests can be performed on filter paper impregnated with the patient's blood.

Management

Complete exclusion of dietary galactose is the mainstay of treatment. In the newborn period, this treatment is accomplished by feeding the child a lactose-free formula, such as Nutramigen or Pregestimil. In later life, when patients are able to eat a more diverse diet, parents must avoid all foods that may contain galactose, however trivial the amount.

An important adjunct in monitoring the scrupulousness of diet therapy are erythrocyte levels of galactose-1-phosphate. The goal of diet therapy should be to maintain erythrocyte galactose-1-phosphate levels as close to normal as possible (less than 1 mg/dL erythrocyte volume).

The emergency physician can best aid the galactosemic patient by being mindful of this diagnosis when managing the newborn with a suggestive clinical pattern. Liver failure and electrolyte abnormalities consequent to the renal tubulopathy are of special concern during the acute illness. Because defective bactericidal activity seems to be an intrinsic feature of the syndrome, appropriate cultures should be obtained and antibiotic therapy instituted quickly.

Various agents (e.g., menthol, progesterone) have been used to promote galactose oxidation in humans. Unfortunately, none has proved to be of real clinical benefit. Experimental evidence suggests that high doses of folate can stimulate galactose oxidation in a rat, and this approach may hold promise for some patients.

Suggested Readings

Batshaw ML, Thomas GH, Brusilow SW. New approaches to the diagnosis and treatment of inborn errors of urea synthesis. Pediatrics 1981;68:290.

Chalmers RA, Lawson AM. Organic Acids in Man. London: Chapman and Hall, 1982.

Cohn RM, Roth KS. Metabolic Disease: A Guide to Early Recognition. Philadelphia: WB Saunders, 1983.

Goodman SI, Markey SP, eds. Diagnosis of Organic Acidemias by Gas Chromatography–Mass Spectrometry. New York: Alan R Liss, 1981.

Jouvet P, Poggi F, Rabier D, et al. Continuous venovenous haemodiafiltration in the acute phase of neonatal maple syrup urine disease. J Inherit

Metab Dis 1997;20:463.

Leonard JV. The management and outcome of propionic and methylmalonic acidaemia. J Inherit Metab Dis 1995;18:430.

Scriver CR, Beaudet AL, Sly WS, Valle D, eds. The Metabolic Basis of Inherited Disease. New York: McGraw-Hill, 1989.

CHAPTER 99

Dermatology

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ATOPIC DERMATITIS

Background

The definition of atopic dermatitis is confusing. Many alternative terms are used to describe skin inflammation (dermatitis) that is chronic and relapsing. Although the eruption may have a variable appearance (erythema, edema, papules, vesicles, serious discharge, and crusting), its constant feature is unrelenting pruritus. The eruption often has a characteristic distribution, depending on age (Fig. 99.1), and often occurs in allergic (atopic) individuals or those with a family history of allergies (e.g., hayfever, asthma, allergic rhinitis). Although many theories relating to cause exist (e.g., genetic, physiologic, pharmacologic, immunologic), the data are conflicting.

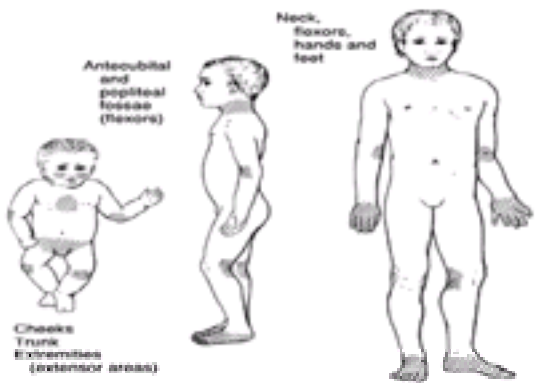

FIGURE 99.1. Distribution of atopic eczema at various ages.

Atopic dermatitis affects about 3% of children, beginning at 1 to 2 months of age. Of children who acquire atopic dermatitis, 60% will do so by the end of their first year of life, 90% by 5 years, 95% by 10 years, and 99% between 10 and 20 years of age. The course of an individual case is difficult to predict, but only 30% of those who develop the problem during the first year continue to have the disease during childhood. Of all children who have atopic dermatitis

during childhood, 90% will be clear by the time they reach adolescence.

Pathophysiology

No single theory explains the initiation and progression of atopic dermatitis. Some evidence suggests that a combination of factors including altered physiologic, pharmacologic, and immunologic mechanisms, is involved in the exaggerated reactivity of the skin.

Various studies have found that patients with atopic dermatitis have dry skin. This reaction may be caused by increased transepidermal water loss and/or decreased quantities of sebaceous gland-derived lipids at the skin surface. Patients also have an increased sweating response to Mecholyl. If this drug is injected into the skin of an individual with atopic dermatitis, it causes blanching rather than the usual erythema. Simple scratching of the skin in an atopic will induce white dermatographism. Finally, the b-adrenergic blockade therapy hypothesizes that reduced function of the b-adrenergic system leads to decreased production of cyclic adenosine monophosphate (cAMP). This reaction results in increased release of the pharmacologic mediators, such as histamines, producing pruritus and inflammation of the skin.

Atopic dermatitis patients have immune system dysregulation, which includes increased production of immunoglobulin E (IgE) by B cells, elevated prostaglandin E_2, abnormal lymphokine secretion profiles, and abnormalities of Langerhans cells. How these changes produce what is called atopic eczema is slowly becoming clearer. cAMP phosphodiesterase is abnormally increased. This response lowers cAMP, producing increased release of histamine, prostaglandin E_2, and other cytokines. This reaction leads to decreases in cell-mediated interferon-g responses and increases in interleukin-4 and interleukin-5 responses. The resulting inflammatory mediators arising from these complex interactions trigger the itch– scratch cycle.

Clinical Manifestations

The patient's age often determines the distribution and appearance of the skin lesions. During infancy, the itch–scratch cycle, which usually begins at 2 to 3 months of age, produces the erythematous, exudative lesions that appear on the cheeks and extensor surfaces. At times, the process becomes generalized. Near the age of 2 years, the more characteristic flexural involvement occurs. Also indicative of atopic dermatitis are 1) varying sized patches of hypopigmentation, especially prominent on the cheeks (pityriasis alba) (Fig. 99.2); 2) patchy or diffuse, fine papules (follicular accentuation) (Fig. 99.3); 3) scaling in the scalp with or without hair loss; and 4) hyperlinear palms and soles (Fig. 99.4), which may show desquamation. Involvement of the feet in such a manner often leads to the misdiagnosis of tinea pedis, which occurs less often in the pediatric population before adolescence. During adolescence, the distribution remains the same; however, a greater incidence of involvement of the face, neck, posterior auricular areas, and the hands and feet occurs. The major physical findings of chronicity, hyperpigmentation, and lichenification are often present (Fig. 99.5).

FIGURE 99.2. Postinflammatory hypopigmentation occurring in a child with atopic dermatitis (pityriasis alba).

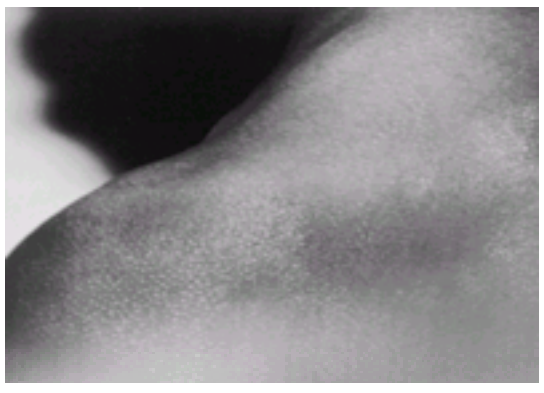

FIGURE 99.3. Follicular accentuation in a patient with atopic eczema.

FIGURE 99.4. A patient with atopic eczema who has hyperlinearity of the soles.

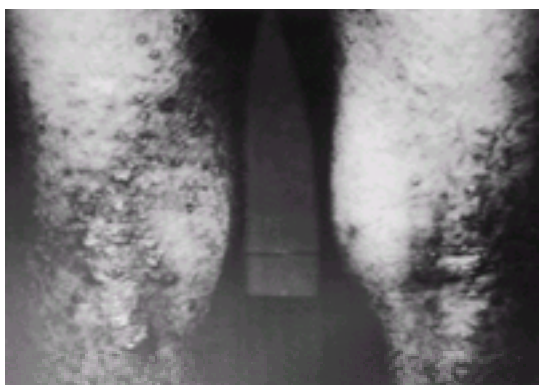

FIGURE 99.5. Chronic changes of hyperpigmentation and lichenification often seen in atopic eczema.

The diagnosis of atopic dermatitis is based on the presence of pruritus, typical morphology, and distribution, as well as a tendency toward chronically relapsing dermatitis. Other possible features are listed in Table 99.1. Unfortunately, laboratory tests are not helpful in the diagnosis of this disorder. Although eosinophilia and elevated serum IgE levels are present, they are not specific for this condition.

Major	Ichthyosis
Typical morphology and distribution	Tendency toward nonspecific hand and foot dermatitis (pseudotinea pedis)
Pruritus	
Chronically relapsing course	Tendency toward repeated cutaneous infections
Personal or family history of atopic disease	White dermographism
Additional Features	Elevated serum immunoglobulin E
Xerosis	*Minor*
Hyperlinear palms and soles	Cataracts
Follicular accentuation	Keratoconus
Pityriasis alba	Dennie-Morgan (infraorbital) fold
Scaling of the scalp	

Table 99.1. Diagnostic Features of Atopic Dermatitis

Differential Diagnosis

Atopic and seborrheic dermatitis may be difficult to differentiate when first appearing in a 1- to 2-month-old infant. Both conditions may cause scaling in the scalp or diaper dermatitis. Clues pointing to seborrheic dermatitis include involvement of the flexural and intertriginous areas in the infant; a salmon-colored eruption with greasy, yellow scaling; and the lack of pruritus. Therapeutic clues include a rapid response to antiseborrheic shampoos and steroids in seborrheic scaling of the scalp. Atopic dermatitis, on the other hand, is often worsened with antiseborrheic shampoos and responds slowly to topical steroids.

Nummular eczema is an eruption that differs from atopic eczema in that the lesions are circular, erythematous, scaling, crusted patches or plaques. The lesions begin as papules and vesicles that spread and coalesce, forming the typical coin-shaped patches. Pruritus is variable. Affected patients do not usually have an atopic background, and IgE levels are generally not elevated. This disorder may be a manifestation of dry skin and, in fact, is aggravated by overwashing, harsh soaps, low temperatures, and low humidity. Decreased bathing, use of mild soaps, and topical steroids are generally helpful.

Xerosis, or dry skin, is a condition that is commonly seen in patients who bathe frequently and use harsh soaps. Low temperatures and humidity will exacerbate this disease. Therefore, it is more common during winter months. The rash is pruritic and appears as rough, red, dry, scaling skin. It is similar in appearance to chapped hands and cheeks seen in cold weather. Decreased bathing, use of mild soaps, and lubrication of the skin are helpful.

Many immune and metabolic disorders are also associated with a rash that is similar in appearance to atopic dermatitis. These disorders are listed in Table 99.2.

Metabolic Disorders	Immunologic Disorders
Phenylketonuria	Ataxia-telangiectasia
Acrodermatitis enteropathica	Letterer-Siwe disease
Histidinemia	Wiskott-Aldrich syndrome
Gluten-sensitive enteropathy	X-linked agammaglobulinemia
Hartnup's syndrome	Hyper immunoglobulin E syndrome
Hurler's syndrome	Selective immunoglobulin A deficiency
	Severe combined immunodeficiency

Table 99.2. Immune and Metabolic Disorders Causing Rash That Resembles Atopic Dermatitis

Complications

Infection of the existing dermatosis is the principle complication in atopic dermatitis. Colonization and infection with *Staphylococcus aureus* is common among atopic children and may account for flare-ups or failure to respond to therapy. Group A b-hemolytic streptococci are also cultured from many individuals with secondarily infected skin.

Viral skin infections also occur more often in patients with atopic dermatitis. Whether this infection is directly correlated to the impaired cellular immunity problem these patients have has not been proven. Eczema vaccinatum, once a dreaded and often fatal complication, rarely occurs now that routine smallpox vaccination has been discontinued. The common causes for what is termed Kaposi's varicelliform eruption are mainly herpes simplex virus (Fig. 99.6) (eczema herpeticum) or, on occasion, coxsackievirus infection. Groups of umbilicated vesicles or areas of increased crusting and ulceration should be cultured for herpes simplex. A diagnostic procedure that may yield a quick answer to the presence of herpes simplex is the Tzanck test (Fig. 99.7). Material from the base of a freshly opened vesicle is scraped for a Giemsa stain. Multinucleated giant cells and balloon cells indicate the presence of herpes simplex. A much more sensitive and specific test is the rapid direct immunofluorescent test (DIF) described in Chapter 67. This virus can also cause localized flare-ups of eczema without dissemination. Leyden described culturally proven recurrent local attacks of herpes simplex virus appearing as discrete punched-out ulcerations. Varicella virus lesions also tend to concentrate in areas of inflamed skin as a result of leakage of virons through dilated vessels. The viruses that cause molluscum contagiosum and warts also infect individuals with atopic dermatitis more often than the average patient (Fig. 99.8).

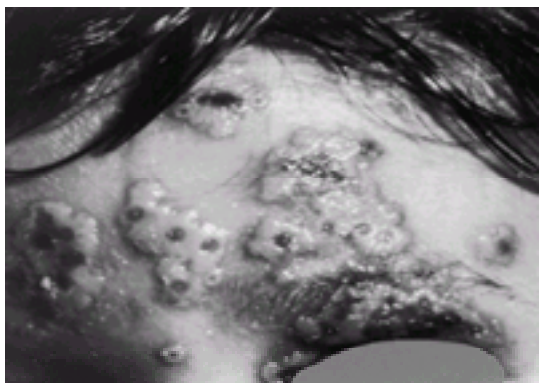

FIGURE 99.6. Eczema herpeticum.

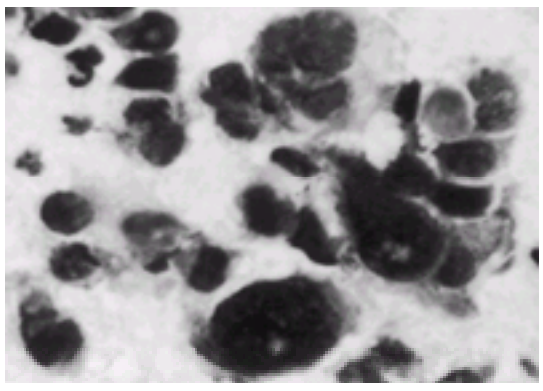

FIGURE 99.7. Positive Tzanck test demonstrating multinuclear giant cells.

FIGURE 99.8. An atopic eczema patient whose face is covered with molluscum contagiosum.

Management (Table 99.3)

Reduction of Pruritus		*Reduction of Inflammation*
Mild soaps	skin care	Skin care
Infrequent washing		Topical steroids (high potency)
Skin lubrication		Systemic steroids (rarely necessary)
Topical steroids (high potency)		*Control of Infection*
Systemic steroids (rarely necessary)		Penicillinase-resistant antibiotics
Antihistamines (children >4 yr old)		

Table 99.3. Acute Treatment of Atopic Dermatitis

Skin tests and hyposensitization are of little value and are rarely indicated. Dietary restrictions may be helpful in certain patients but are difficult to maintain. The four main objectives in the treatment of uncomplicated atopic dermatitis are 1) reduction of pruritus, 2) reduction of inflammation, 3) protection of the skin from unknown irritants, and 4) removal of known irritants. Reduction of pruritus can be accomplished in numerous ways. Most important of these methods is limitation of bathing (at times, to only once per week) and the use of a mild soap (e.g., Dove, Tone, Caress). Lubrication of the skin with Nivea cream, Eucerin cream, or Moisturel (which contains no lanolin or perfumes) ameliorates dryness, which may be a factor in producing pruritus. Antihistamines can be helpful, although, during infancy, the necessity for soporific doses results in their being less helpful therapeutically. Old standbys include diphenhydramine hydrochloride (5 mg/kg per day in three to four divided doses) and hydroxyzine (2 mg/kg per day in three to four divided doses). Newer preparations include topical doxepin and oral cetirizine. Control of inflammation is accomplished with the use of topical steroids (Table 99.4). During the acute phase, potent steroids should be used to bring the situation under control. (At times, systemic steroids are used to bring an acute flare-up under control. Fortunately, this measure is rarely necessary.) Once control is achieved, steroids of mild potency should be used and applied less often. (Note: Steroids should not be used on the face for prolonged periods.) Maintenance with the least potent steroid, applied as infrequently as possible, is advisable. However, continued therapy is usually necessary. After control has been maintained for a fairly prolonged period, an attempt to discontinue steroid therapy can be made. Protection of the skin against unknown irritants is best done by covering it. Long-sleeve polo shirts and leotards are helpful in preventing dust and pollens from coming into contact with the skin. Removal of known irritants is achieved by 1) environmental control (i.e., no stuffed toys, wool clothing or blankets, or pets); 2) avoidance of harsh soaps; and 3) keeping fingernails short. At times, hospitalization for control is advisable and certainly a more desirable alternative than systemic steroids.

Mild	
Hydrocortisone 1%	
Moderate	
Aristocort ointment 0.1% (triamcinolone acetonide)	
Cordran ointment 0.05% (flurandrenolide)	
Synalar cream 0.025%	(fluocinolone acetonide)
Synalar ointment 0.025%	
Valisone cream 0.1% (betamethasone valerate)	
Potent	
Diprosone ointment 0.05% (betamethasone dipropionate)	
Florone ointment 0.05% (diflorasone diacetate)	
Lidex cream or ointment 0.05% (fluocinonide)	
Topicort ointment 0.25% (desoximetasone)	

[a]All the synthetic preparations are fluorinated. Hydrocortisone is not.

Table 99.4. Potency of Topical Steroids [a]

Appropriate antibiotics are important in the treatment of secondary bacterial infections. A child who is not toxic can often be treated orally in the home setting. Because penicillin-resistant staphylococcal organisms are commonly involved,

antibiotics such as erythromycin (40 mg/kg per day) or dicloxacillin (50 mg/kg per day) provide suitable coverage. These antibiotics also treat group A b-hemolytic streptococci that may be present. When a child is toxic, intravenous (IV) therapy is advisable; therefore, hospitalization is necessary. Again, penicillinase-resistant antistaphylococcal drugs should be given.

Eczema herpeticum that is localized and that has not produced toxicity in a child can be treated symptomatically and will usually clear in 2 to 3 weeks. With severe infection, especially in young infants, more aggressive therapy may be necessary. A daily dosage of 700 mg/m^2 per 24 hours of acyclovir, given in a 1-hour IV infusion every 8 hours, is advised. More localized primary or secondary infections can be treated orally at a dosage of 15 to 30 mg/kg per 24 hours in four to five divided doses for 10 days.

Studies evaluating therapy with medications such as Tacrolimus ointment, oral cyclosporine, interferon gamma, as well as many others, are under way.

SEBORRHEIC DERMATITIS

Background

Seborrheic dermatitis is the term given to the salmon-colored patches with yellow, greasy scales occurring primarily in the so-called seborrheic areas (face, postauricular area, scalp, axilla, groin, presternal area). During childhood, seborrheic dermatitis is seen in infants or adolescents. Its onset occurs during the first 3 months of life and generally disappears shortly thereafter, only then reappearing in adolescence.

Pathophysiology

Although sebaceous gland dysfunction is often cited as a cause, the definite cause of this disorder has not been established. In fact, surface fat levels are normal in seborrheic dermatitis, but their ratio is altered. The presence of seborrheic dermatitis correlates strongly to concomitant emotional stress or neurologic disorders.

Clinical Manifestations

The two common locations of skin involvement during infancy are the scalp (“cradle cap”), as shown in the infant with seborrheic dermatitis in Figure 99.9, and diaper area. Most commonly, yellow, greasy scales are found over the anterior fontanel. Scaling is concentrated in this location because of the fear some mothers have about rubbing or scrubbing over the fontanel. Many times the scaling is limited to this area; however, occasionally, the scaling is spread to the forehead, eyebrows, nose, ears, and neck. The intertriginous and flexural areas may also become involved. This reaction is especially seen in the diaper area (see Fig. 99.15). The child is not irritable, and pruritus does not seem to be present. The prognosis for clearing is excellent, and resolution usually occurs within several weeks to months.

FIGURE 99.9. Seborrheic dermatitis.

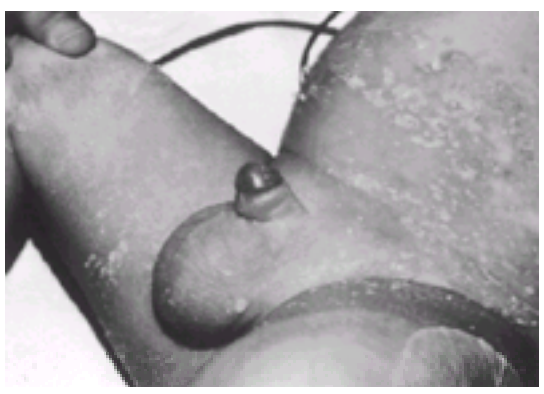

FIGURE 99.15. Infant with seborrheic diaper dermatitis.

Between the periods of infancy and adolescence, scaling of the scalp usually indicates causes other than seborrheic dermatitis (atopic dermatitis or tinea capitis). In fact, true seborrheic dandruff does not appear until puberty, when

excessive production of sebum occurs. Most commonly, scaling before puberty and after infancy indicates the presence of atopic dermatitis or tinea capitis (especially Trichophyton tonsurans). Differentiation is aided by clinical appearance, cultures, and response to therapy. Atopic dermatitis is often worsened with harsh shampoos and responds slowly to topical steroids. The diagnosis of tinea capitis is best made with cultures. If steroids are used in the presence of tinea capitis, scaling of the scalp often increases secondary to suppressed local immunity of the skin and increased growth of the fungus. Seborrheic dermatitis of the scalp during the adolescent period is similar in nature to the condition in adults. Scaling in the scalp appears, and the seborrheic areas are variably involved. Erythema and scaling occur between the eyebrows, on the eyelid margins, and in the nasolabial creases, sideburns, beards, mustache, posterior auricular areas, and aural canals. Rarely, the patient may develop a secondary infection with monilia or bacteria.

Management

Seborrheic dermatitis of the scalp responds readily to antiseborrheic shampoos (i.e., selenium sulfide) and topical steroids such as fluocinolone acetonide or betamethasone valerate (Table 99.4). In infants, loosening of the scales with a soft toothbrush or fine-toothed comb before shampooing often hastens clearing of the cradle cap. Topical steroids are effective in the treatment of seborrheic dermatitis. Because hairy locations are commonly involved, steroid preparations in the form of lotions or gels are advisable. The strength of the steroid and the frequency of application is determined by the response to therapy. Steroids should not be used for prolonged periods on the face because of potential damage to the skin in that area. Secondary infection with bacteria can be treated with appropriate antibiotics. If *Candida albicans* secondarily invades the lesions, topical clotrimazole cream, applied twice a day, is useful.

ALLERGIC CONTACT DERMATITIS

Background

Allergic contact dermatitis is a cell-mediated reaction to antigenic material in contact with the surface of the skin. The incidence in children is about 1.5%, a considerably lower value than that given for adults. Children less than 1 year of age rarely respond to contactants and, until nearly 3 years of age, have a reduced incidence of contact dermatitis.

Pathogenesis

An allergen penetrates the stratum corneum (facilitated by trauma at times) and combines with a carrier protein to form the foreign substance that is responsible for initiating the sensitization process. This complex is carried via lymphatics to the regional lymph nodes where processing by macrophages occurs. Recognition by T lymphocytes follows; these cells then leave the node, enter the bloodstream, and migrate into the skin. When the antigen again comes in contact with the skin, sensitized T lymphocytes combine with the specific foreign material and release inflammatory lymphokines. The characteristic dermatitis occurs 6 to 18 hours later.

Clinical Manifestations

The acute onset of linear or geometric areas of erythema, edema, eczematization, and papulovesiculation usually indicates the presence of allergic contact dermatitis. Because skin involvement is limited to areas of contact, the distribution, pattern, and shape of the dermatitis provide important clues for the clinician (Table 99.5). Therefore, a round lesion on the back of the wrist would incriminate a wristwatch; a linear pattern encircling the waist points to the rubber in the waistband of a garment; linear lesions on exposed portions of the body indicate brushing against the leaves of a poison ivy plant (Fig. 99.10); and extensive involvement of exposed areas of skin suggests an airborne allergen, as with ragweed or vaporized oil transmitted in the smoke of burning poison ivy (Fig. 99.11). Generally, the scalp, palms, and soles are less permeable to allergens and therefore are less often involved. Involvement of oral mucous membranes is uncommon. As previously mentioned, trauma or nonspecific factors such as pressure, heat, and perspiration may predispose the skin to allergic contact dermatitis.

Head and Neck
Scalp—hair dye, hair spray, shampoo
Ear canal—neomycin
Forehead—hat band
Eyelids—nailpolish, volatile gases, false eyelash cement, mascara, eye shadow/cosmetics
Perioral—dentrifices, bubble gum, chewing gum
Ears—earrings, perfume
Trunk
Axilla—deodorant, clothing dye
Breasts—metal, elastic in bra
Arms
Wrist—cosmetic jewelry (nickel), leather (p-phenylenediamine, chrome)
Abdomen
Waistline—rubber dermatitis from elastic in pants, jockstrap (lower)
Lower extremities
Feet—shoe dermatitis

Table 99.5. Regional Predilection of Various Substances That Cause Contact Dermatitis

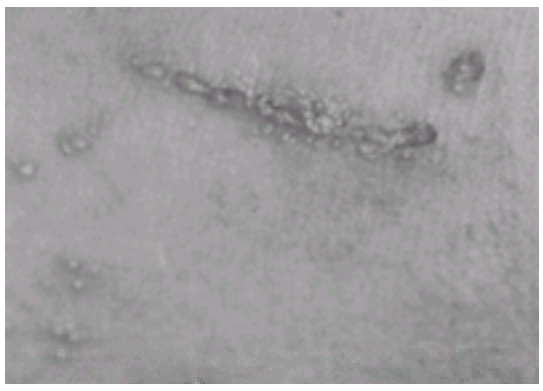

FIGURE 99.10. Typical linear pattern after exposure to the poison ivy plant.

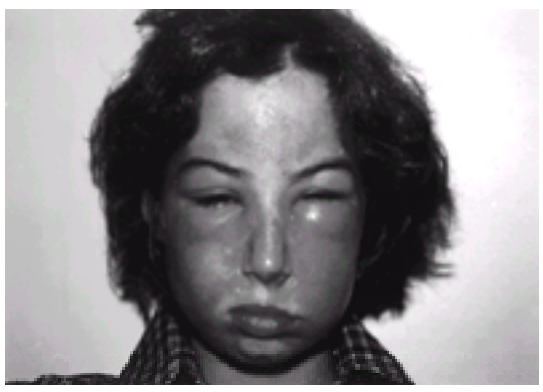

FIGURE 99.11. Facial edema and inflammation in response to exposure to airborne contact allergen (e.g., vaporized oil in smoke of burned poison ivy plants). *Please see the color-tip insert (Color Plate 99.11).*

Fisher states that the most common causes of contact dermatitis in order of frequency are rhus (poison ivy, oak, sumac), *p*-phenylenediamine, nickel, rubber compounds, and the dichromates. Although these conclusions are based on an adult sample, they generally apply to children, except for the substance *p*-phenylenediamine, which is found in hair dyes and is probably at the bottom of the list.

Rhus (Poison Ivy, Oak, Sumac)

Rhus dermatitis is the most common allergen involved in the production of contact dermatitis. The poison ivy plant (Fig. 99.12A) occurs in all parts of the United States as a shrub or vine, often on trees or fences. Poison oak (Fig. 99.12B), an upright shrub, appears only on the west coast. Poison sumac (Fig. 99.12C) grows as a shrub or tree east of the Mississippi. Seventy percent of the population will become sensitized if exposed to the oleoresin, known as urushiol, contained on the leaf, stem, or root of the plant. The active ingredient in this oil is pentadecylcatechol. The oil can be carried on clothing and pets or by the wind.

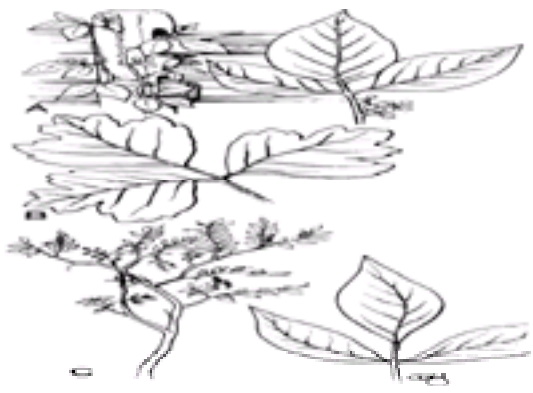

FIGURE 99.12. A. Poison ivy *(Rhus radicans).* **B.** Poison oak *(Rhus toxicodendron).* **C.** Poison sumac *(Rhus vernix).*

Each plant produces an identical redundant eruption. From the time of exposure, the average time to the appearance of the rash is 48 hours. At that time, onset of pruritus, inflammation, and grouped or linear papulovesicles or bullae occurs. With severe exposure, the face and eyelids become uniformly edematous. The eruption can last from 1 to 3 weeks.

Avoidance of exposure is the best prophylaxis in treatment. Topical barrier agents such as the product that is used by forest rangers have been developed to prevent rhus dermatitis in high-risk individuals. However, at times, protection is impossible. Once an individual is exposed, contaminated clothing should be removed and laundered, and the body should be bathed with any soap as soon as possible, preferably within 5 to 10 minutes. Once the oil has been removed, spread does not occur, even from vesicular fluid. Although sequential outbreaks on various parts of the body suggest spread, lesions appearing later in time indicate initial exposure to a lesser dose of the offending oil.

Antipruritic lotions such as calamine are useful. Topical steroids are minimally effective, and topical antihistamines and anesthetics should be avoided because they can be sensitizers. Antihistamines can be helpful. With generalized reactions, oral prednisone 1 to 2 mg/kg once daily for 1 week than tapered over the next week is advisable.

Nickel Contact Dermatitis

Nickel dermatitis is seen commonly in the female in response to nickel-containing jewelry. Earlobes are commonly involved because of the popularity of pierced ears. Most articles of jewelry contain nickel, including those made of gold and silver. Any person wearing these items is at risk. Perspiration begins the process by leaching the nickel from the jewelry around the neck, wrist, or the fingers. Treatment consists of removing the offending object, avoiding further contact with nickel-containing jewelry, and applying topical steroids to the affected areas of skin.

Shoe Contact Dermatitis

Erythema, blistering, weeping, crusting, or lichenification of the dorsal aspects of the toes and instep of the feet, with sparing of the interdigital webs, suggest shoe contact dermatitis. The responsible antigens are usually the rubber, glues, dyes, and tanning agents used in making the shoes. Although not often recognized, the problem is common. Children who sweat freely are more likely to be affected because of the leaching of allergens from the shoes onto the skin. Secondary infection is common, and an “id” reaction, similar to that seen in tinea pedis, can cause involvement of the hands and other areas of the skin distant from the primary site.

Patch testing kits for shoe components are available to determine specific sensitizing substances. This testing should be done only after the skin problem is brought under control, and probably by a dermatologist. Control is achieved by avoiding shoes when possible, treating secondary infection with appropriate antibiotics, and using topical steroids. All antihistamines are helpful for reducing pruritus. An id reaction consisting of huge bullae on the hands and feet can occur. The child is often unable to walk. Hospitalization and the use of systemic steroids (see previous section regarding Rhus) are necessary.

Cosmetics

Many practicing physicians do not know that nail lacquers (containing sulfonamides and formaldehyde resins) or nail hardeners (containing formaldehyde) are a common cause of allergic reactions on the skin of the eyelids. The skin in this area is thin and permeable. Simply rubbing the eyes with fingernails that have polish on them can induce the problem. *p*-Phenylenediamine, which is contained in hair dyes, will also cause eczematous eruptions of the scalp and face.

Management

Elimination and avoidance of the causal antigen is the most effective preventive and therapeutic measure. Topical steroids and antihistamines help with the inflammation and pruritus.

With localized involvement, moderate to high potency topical steroids can be helpful in reducing symptoms while the dermatitis clears. With generalized skin involvement, oral steroids are effective at a dosage of 1 to 2 mg/kg per day over 7 to 10 days then tapered over the next 7 days (rebound less likely). Patch testing should not be done during an acute episode because contact with the allergen may cause worsening of the rash.

DIAPER DERMATITIS

Background

Diaper dermatitis is a general term used to describe skin abnormalities beneath the diaper secondary to a variety of causes. The problem is common in children 2 years of age or younger who require the use of a diaper. It generally disappears after toilet training.

Pathophysiology

The pathogenesis of the problem is multifactorial (Fig. 99.13) and not clearly defined. The possibilities include the concentration of bacteria or fungi, the action of organisms on the urine, and moisture itself.

FIGURE 99.13. Proposed sequence of events producing skin inflammation in the diaper area.

No firm proof exists that bacteria play a major role. However, bacterial overgrowth does occur on moist skin with increasing time. Bacteria have been implicated in liberating ammonia from urine and raising urine pH. The rise in pH increases the activity of fecal proteases and lipases, which can damage skin. Bile salts can potentiate this damage.

C. albicans is found on the skin in 40% of infants with active diaper dermatitis within 72 hours of the appearance of the rash. Because studies show that this organism is present in less than 10% of infants without diaper dermatitis, *C. albicans* may be playing a significant role. Sources of *C. albicans* include the gastrointestinal (GI) tract and secondary implantation from a mother with candidal vaginitis.

Chronic exposure of the skin to moisture, especially under occlusion by the diaper, leads to maceration and alteration of the epidermal barrier with overgrowth of bacteria and *C. albicans.* If one major instigating factor exists, the effect of chronic exposure to moisture is critical to the development of diaper dermatitis.

Another consideration is the predisposition of certain individuals to react more easily and negatively to varying irritants. Generally, infants with an atopic or seborrheic background are at greater risk for the development and persistence of diaper dermatitis.

Clinical Manifestations

Differentiation of the various types of diaper dermatitis is difficult. Clues from the history and physical examination are necessary when characterizing the cause of this problem. The different types of diaper rashes include occlusion dermatitis, atopic dermatitis, seborrheic dermatitis, moniliasis, and mixed or not diagnosable rash.

Occlusion Dermatitis (Fig. 99.14)

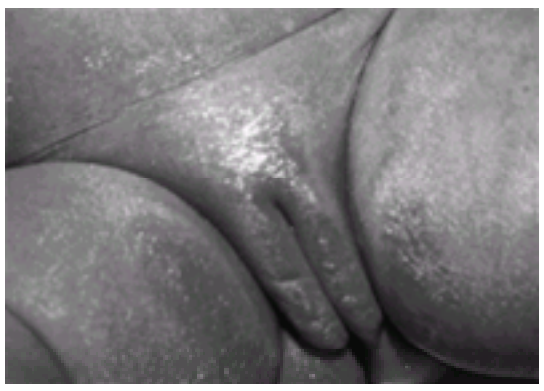

FIGURE 99.14. Infant with occlusion diaper dermatitis. *Please see the color-tip insert (Color Plate 99.14).*

Occlusion dermatitis contains two components. The first, friction, occurs mainly on those portions of the diaper area where contact with the diaper is greatest (inner thighs, lower abdomen, and prominent surfaces of the genitalia and buttocks). The rash waxes and wanes and often has a shiny, glazed surface appearance. Occasionally, papules are associated with the rash. The second component, trapped moisture, causes the erythema and maceration that occurs in the intertriginous parts of the diaper area (inguinal, genital, intergluteal, and folds of the thighs). This type of problem is often associated with and precipitated by tightly applied diapers, commercial plastic diapers (especially those made with elastic edges to prevent leakage around the thighs), and rubber pants placed over cloth diapers. Such coverings increase friction and prevent the evaporation of moisture.

Atopic Dermatitis

The appearance of the rash in the diaper area is not different from occlusion dermatitis. It is, however, more chronic and difficult to treat. Examination may disclose lesions on other body surfaces (cheeks, antecubital and popliteal spaces) typical of atopic involvement, and a family history of atopy often exists.

Seborrheic Dermatitis (Fig. 99.15)

Generally, the rash has an erythematous, salmon-colored base that is covered with yellow, greasy scaling. Similar involvement of other seborrheic locations such as the scalp, postauricular area, or other flexures helps to establish the diagnosis. At times, a family history of seborrheic dermatitis exists.

Moniliasis

Moniliasis is the most characteristic of the diaper rashes. The skin in the diaper area has clusters of erythematous papules and pustules that go on to coalesce into an intensely red confluent rash with sharp borders. Beyond these borders are satellite papules and pustules. At times, the infant has concomitant oral thrush. When the problem is chronic and recurrent, seeding from the GI tract or from a mother with monilial vaginitis should be considered.

On rare occasions an id reaction occurs (Fig. 99.16). Beside the primary monilial diaper rash lies an antigenic dissemination with involvement of the intertriginous areas as well as scattered small patches or plaques of scaling

erythema on other parts of the skin surface. Generally, *C. albicans* cannot be cultured from these plaques.

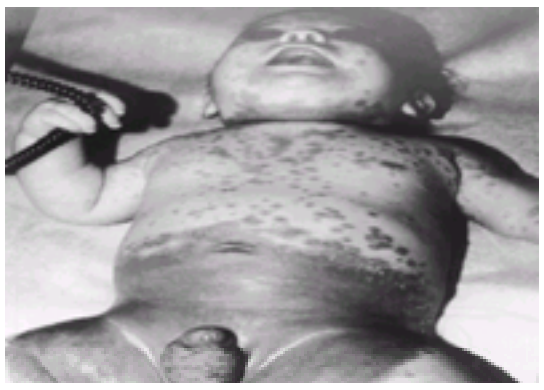

FIGURE 99.16. Infant with monilial diaper dermatitis with id reaction.

Mixed or Not Diagnosable Rash

Mixtures of the above categories of diaper dermatitis are often found on infants. A diagnosis is often difficult to make. Secondary invasion with *C. albicans* is common as mentioned. The potential for secondary bacterial infection exists. If blistering occurs, *S. aureus* infection should be considered.

Management

Treatment is determined by the cause of the dermatitis. In general, proper skin care, which includes decreased frequency of washing, use of mild soaps, and keeping the diaper off as much as possible, will help resolve diaper dermatitis resulting from any cause. With occlusive dermatitis, avoidance of tightly fitting diapers, plastic-covered paper diapers, and rubber pants is important. When atopic dermatitis is present, the use of topical steroids is necessary. It is important to avoid fluorinated or other potent steroids in the diaper area because occlusion by the diaper enhances the steroid effect and is more likely to produce skin atrophy and striae. The newer antifungal–steroid combinations should also be avoided for these same reasons. Therefore, 1% hydrocortisone cream no more than twice daily over a short period is recommended. Hydrocortisone (1%) is also effective for seborrheic diaper dermatitis and can be used intermittently.

With monilial diaper dermatitis, the use of preparations such as econazole twice a day is effective. If thrush is also present, oral nystatin 200,000 units (2 mL) four times a day for 7 days, is advisable. This medication will also be useful if the infant is seeding *C. albicans* from the GI tract onto the skin of the diaper area. Because another potential source of *C. albicans* is from a vaginal infection in the infant's mother, the mother should be questioned for this problem; if a vaginal discharge is present, it should be checked by a gynecologist and treated appropriately. Patients with id reactions, as described before, require oral nystatin, econazole on the diaper and intertriginous areas, and 1% hydrocortisone applied to the plaques. Resolution usually takes 7 to 10 days. Mycolog II cream is recommended by many physicians for monilial diaper rashes; however, the clinician should be cognizant that this preparation contains a fluorinated steroid. Secondarily infected dermatitis, such as bullous impetigo, should be treated with the appropriate systemic antibiotics.

Whether traditional diaper creams and ointments are effective is still unproven. Their ability to provide an effective barrier that reduces irritation remains to be established.

DRUG REACTIONS IN THE SKIN

Background

When a drug is being taken by a child, any reaction of the skin that is not expected should be considered a drug reaction. Hospitalized patients are more likely (30%) to have a reaction to a drug because of multiple exposures. Approximately 2 to 3% of these inpatients have cutaneous reactions. The rate of adverse effects depends on the particular drug. Arndt et al. showed that reactions occur at the rate of 59 per 1000 drug courses for trimethoprim–sulfamethoxazole, whereas with the use of chloralhydrate, only 0.2 reactions per 1000 courses occurred. When considering all drugs, they found reactions at a rate of 3 per 1000 courses of therapy. Penicillins, sulfonamides, and blood products were responsible for most reactions.

When considering penicillin and its derivatives alone, allergies to these substances affect 1 to 10% of the population. Fatal anaphylaxis occurs in approximately 2 of 100,000 patients taking penicillin. Penicillin reactions appear less often in children, as is the case with any drug reaction. An increased risk for the development of anaphylactic reactions to penicillin is present in atopic individuals.

Pathophysiology

The pathogenesis of such reactions can be on an immune or nonimmune basis. When occurring on an immune basis, any of the four types of immunologic mechanisms (IgE-mediated, immune complex, cytotoxic, cell-mediated) can be involved. However, reactions also occur on the basis of overdosage, specific toxicity, common side effects of a particular drug, and unusual drug interactions. Pathogenic mechanisms in specific situations often cannot be identified.

Clinical Manifestations

The appearance of drug reactions is nonspecific and may mimic almost any known dermatosis. Therefore, the clinician cannot make a diagnosis of a drug reaction based on the appearance of the rash alone because the skin can react only in a limited number of ways to many different stimuli. However, certain patterns should raise suspicion of the presence of an adverse reaction.

Laboratory analysis is usually not helpful in the diagnosis of drug eruptions. Peripheral eosinophilia, thought to occur commonly with adverse drug reactions, is actually uncommon. Skin biopsy can be helpful.

Specific Reaction Patterns

Urticaria. Urticaria constitute the most common expression of drug sensitivity. Most commonly, reactions occur within 1 week of drug exposure. When an individual is on multiple agents and has a reaction, the clinician should suspect those agents that were most recently introduced or those medications that are known to be commonly associated with drug reactions (Table 99.6) When questioning for the use of medications, it is important to ask not only about prescription items, but also about over-the-counter preparations. Occasionally, patients who use aspirin, acetaminophen, laxatives, or ear, nose, or eye drops do not consider these substances to be medications or drugs.

Trimethoprim-sulfamethoxazole	Cephalosporins
Ampicillin	Dipyrone
Semisynthetic penicillins (carbenicillin, cloxacillin, dicloxacillin, methicillin, nafcillin, oxacillin)	Nitrazepam
	Barbiturates
Sulfisoxazole	Nitrofurantoin
Penicillin G	Glutethimide
Gentamicin	Indomethacin

Table 99.6. Drugs Most Commonly Associated with Allergic Skin Reactions

Maculopapular Eruptions Similar to Those of a Viral Exanthem. Maculopapular eruptions are the second most common of all drug-induced rashes and may be caused by many different agents. These eruptions are symmetric and consist of erythematous macules and papules with areas of confluence. Variable involvement of the palms, soles, and mucous membranes, as well as purpura may occur. The presence and severity of pruritus is variable. Ampicillin is a medication often associated with this type of skin reaction, particularly in patients with infectious mononucleosis.

Erythema Multiforme. Erythema multiforme is an acute and often recurrent inflammatory syndrome often secondary to drugs (e.g., penicillins, sulfonamides, hydantoins, barbiturates) or infections. Recent observations suggest that a significant portion of idiopathic erythema multiforme cases may be caused by herpes simplex virus. The skin findings include macules, papules, vesicles, and pathognomonic target or iris lesions (Fig. 99.17) that tend to be more or less symmetrically distributed. Bullous lesions may also be present. In the more severe cases, constitutional symptoms occur; when mucous membranes are involved, the term *Stevens-Johnson syndrome* is used (Fig. 99.18 and Fig. 99.19). Erythema multiforme consists of two lesion types: macular-urticarial and vesicular-bullous. There is a predilection for the backs of the hands, palms, soles, and extensor surfaces of the limbs. The lesions may begin at these sites and then spread diffusely or they may begin generalized. In 25% of the patients, the mucous membranes are involved and, in fact, can be the sole site of involvement. The usual sites of mucous membrane involvement are the lips, buccal mucosa, palate, conjunctivae, urethra, and vagina. With severe involvement, the pharyngeal, tracheobronchial, and esophageal mucous membranes are also affected. Less common sites are the anal and nasal mucosa. When the eyes are involved, there may be simple conjunctivitis, severe keratitis, or panophthalmitis. These changes may lead to blindness in 3 to 10% of these patients. Therefore, close attention to involvement of the eyes is necessary. Lesions may continue to erupt in crops for as long as 2 to 3 weeks. Death occurs in 3 to 15% of patients.

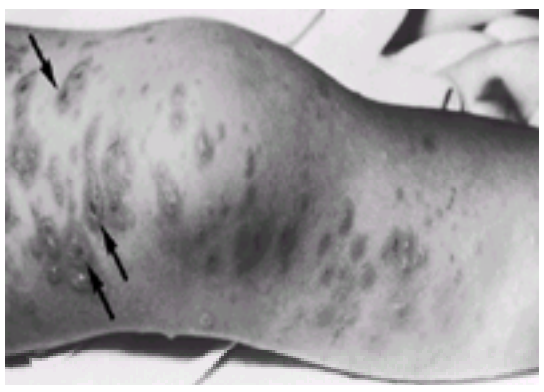

FIGURE 99.17. Iris or target lesions pathognomonic of erythema multiforme.

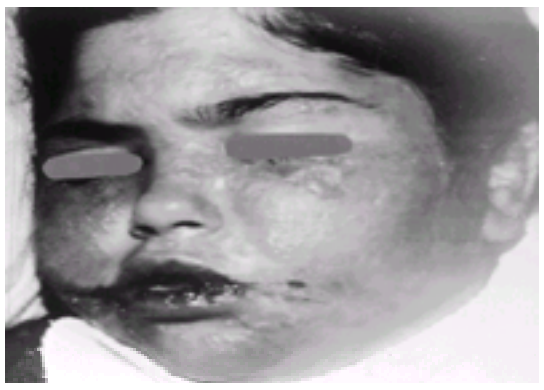

FIGURE 99.18. Adolescent with Stevens-Johnson syndrome secondary to sulfonamides. Note involvement of mucous membranes of the mouth.

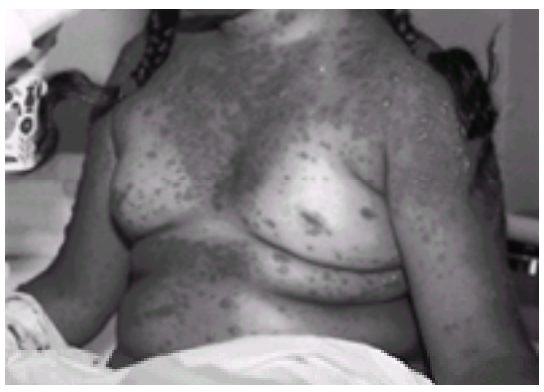

FIGURE 99.19. Same child as seen in Figure 99.18. Note photo distribution of lesions. *Please see the color-tip insert (Color Plate 99.19).*

Vasculitis. The classic lesions of vasculitis are palpable purpura. Although these lesions are characteristic, vasculitis may be manifest by erythematous macules, papules, urticaria, and hemorrhagic vesicles and bullae (Fig. 99.20). The diagnosis is made by a skin biopsy, which shows leukocytoclasis, endothelial cell necrosis, and destruction of dermal vessels.

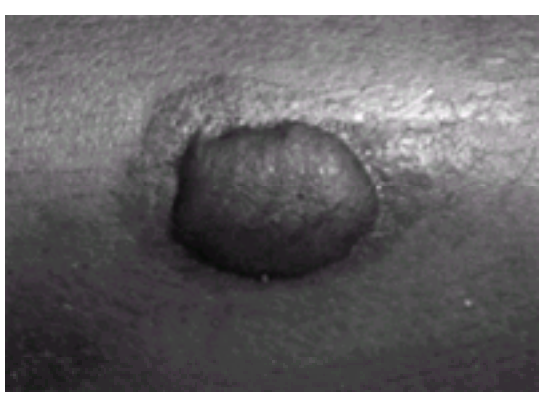

FIGURE 99.20. Hemorrhagic bulla in patient with vasculitis. *Please see the color-tip insert (Color Plate 99.20).*

Erythema Nodosum. The lesions of erythema nodosum appear as deep, tender, erythematous nodules or plaques of the extensor surfaces of the extremities (Fig. 99.21). They are thought to be hypersensitivity phenomena secondary to infections (e.g., streptococcal pharyngitis, tuberculosis, coccidioidomycosis, histoplasmosis), inflammatory bowel disease, sarcoidosis, malignancies, and occasionally, drugs. The exact immunologic mechanism has not been clarified.

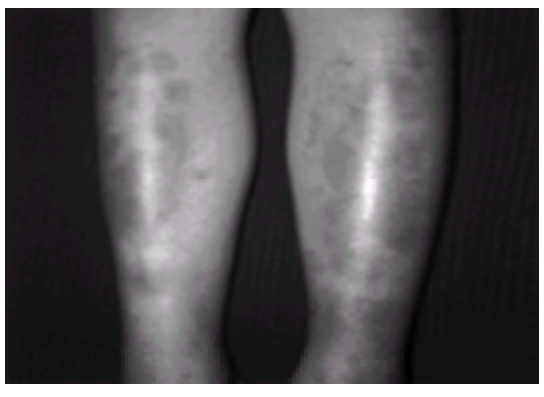

FIGURE 99.21. Extensor surface involved with lesions of erythema nodosum. *Please see the color-tip insert (Color Plate*

99.21).

Photosensitive Cutaneous Eruption. When a drug causes an exaggeration of the sunburn response, a phototoxic eruption should be considered. However, photoallergic eruptions usually do not occur on first exposure to a medication because immunologic induction must first occur. Because a hypersensitivity reaction is also involved, the eruption, although concentrated most heavily on sun-exposed areas, can also occur on non–sun-exposed areas. Tetracycline and sulfonamides can be involved in this reaction.

Toxic Epidermal Necrolysis. Drug-induced toxic epidermal necrolysis (TEN) must be differentiated from an illness caused by a circulating staphylococcal exotoxin. If a child who has TEN has been taking drugs long term or shortly before onset of the rash, is over 6 years of age, or has a mixed rash (i.e., areas with the appearance of erythema multiforme as well as toxic epidermal necrolysis), a biopsy must be performed to distinguish between the two disorders. With drug-induced TEN, dermal–epidermal separation is visible on histologic examination. If epidermolytic toxin has been released by staphylococci, epidermal cleavage occurs in the granular layer (see Fig. 99.24). With extensive exfoliation of skin, fluid and electrolyte disturbances may occur, and the potential for bacterial sepsis is present.

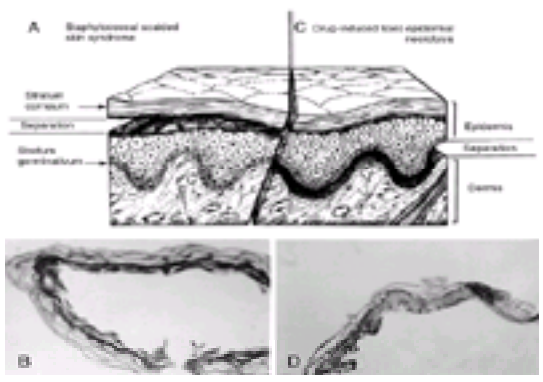

FIGURE 99.24. A. and **B.** Pathology in staphylococcal scalded skin syndrome (SSSS). **C.** and **D.** Pathology in drug-induced toxic epidermal necrolysis.

Fixed Drug Eruption. Fixed drug eruption refers to a localized round or oval dermatitis that tends to recur at the same location each time there is exposure to the offending drug. The lesions are generally erythematous and may or may not contain vesicles. They disappear over 7 to 10 days after cessation of the drug, leaving various shades of postinflammatory hyperpigmentation in their place. The discoloration may persist for months or years. Initially, lesions are solitary but then can become multiple; they often involve the palms, soles, glans penis, and lips.

Management

Vital to the management of any suspected drug reaction is the identification and removal of the offending drug. Pruritus can be controlled with antihistamines, and open lesions are responsive to compressing with Burow's solution and topical silver sulfadiazine. When extensive exfoliation occurs, attention to fluid and electrolyte balance and secondary infection is essential. Any patient with mucous membrane involvement should have an ophthalmologic examination to rule out the presence of corneal involvement. Hospitalization should be considered in any patient who has severe involvement of the skin, is toxic, or has extensive exfoliation.

The literature suggests that steroid therapy of Stevens-Johnson syndrome and drug-induced toxic epidermal necrolysis is of no value, will prolong hospital stays, and may in fact be harmful. Steroids (i.e., an equivalent of prednisone 1 to 2 mg/kg per day) must be started within the first 2 days of the eruption to be effective. Progression of the reaction after 5 days of steroid therapy indicates that the medication is ineffective and should be discontinued. If skin denudation is greater than 20% of the child's body surface area steroid therapy should be avoided. If denudation progresses to greater than 25% of body surface area the child should be transferred to a burn unit.

STAPHYLOCOCCAL SCALDED SKIN SYNDROME

Background

The term *toxic epidermal necrolysis* is often used indiscriminately. Because the gross dermatologic changes are the same despite different causes, this term should be used to indicate only the visible changes. In this way, the physician will approach patients with an open mind and be more likely to consider the alternative possibilities: although rare, a drug-induced TEN in children and bacteria-induced TEN in adults. *Staphylococcus*-induced disease, or the staphylococcal scalded skin syndrome (SSSS), is presented in this section. Included under the heading of SSSS are bullous impetigo (staphylococcal pustulosis), in newborns, scarlatiniform rashes induced by *S. aureus,* and the generalized exfoliative syndrome caused by *S. aureus* seen in newborns (Ritter's disease) or in children (Lyell's syndrome).

Pathophysiology

The mechanism of these reactions was described by Mellish and Glasgow. They injected coagulase-positive phage group II staphylococci into newborn mice, producing erythema and a positive Nikolsky sign (denudation of skin with gentle rubbing) in 12 to 16 hours, followed by bullae and extensive exfoliation in 16 to 20 hours. Since that time, phage group I and III staphylococci have also been implicated. The disease is believed to occur primarily in children because they lack antibodies against the organism and are unable to metabolize and excrete the toxin as well as adults.

Clinical Manifestations

The illness begins with malaise, fever, and irritability. The irritability is often caused by significant tenderness of the skin when touched. Mothers will relate that their infant does not want to be held and cries when handled. A “sunburn” erythema follows, which first begins and is most intense around the neck, the intertriginous areas, and periorifically (especially the eyes and mouth). The erythema spreads to varying portions of the skin surface, and the child may be very toxic. With mild involvement of the skin, superficial desquamation (flaking) then follows similar to the reaction that occurs after an ordinary sunburn (Fig. 99.22). With severe involvement, large sheets of skin shear away, leaving a denuded, oozing surface similar to the reaction that occurs after a burn (Fig. 99.23). The skin can often be rubbed off (Nikolsky's sign). Vesicles, pustules, and bullae can also occur during the exfoliative phase. Often, a purulent discharge emits from the eyes, but no conjunctival injection is present. Mucous membranes are not involved. Most children do well, and clearing of the skin occurs in 12 to 14 days, leaving no residua.

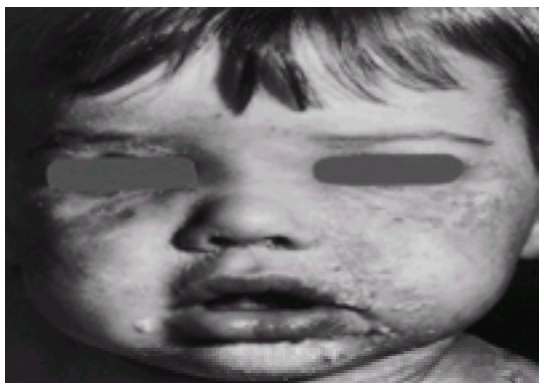

FIGURE 99.22. Desquamation of the skin of the face in the staphylococcal scalded skin syndrome (SSSS).

FIGURE 99.23. Denudation of skin of nose in child with staphylococcal scalded skin syndrome (SSSS).

The complete blood count (CBC) and urinalysis are not helpful in the evaluation of such children. Although blood cultures should be done, they are usually negative, as are cultures of intact vesicles or bullae. At times, *S. aureus* can be grown from exfoliating skin, the umbilicus, circumcision wounds, throat, eyes, ears, nose, or rectum. Histologic examination of the skin distinguishes between changes caused by staphylococci or a drug. In SSSS, skin clippings or a punch biopsy show separation of the superficial layer of the epidermis subcorneally (Fig. 99.24A and Fig. 99.24B). Patients who have drug-induced TEN will have dermal–epidermal separation (Fig. 99.24C and Fig. 99.24CD). Children taking medications long term or shortly before the eruption of the rash, children older than 6 years of age, or children with a mixed rash (i.e., areas of TEN and erythema multiforme) should have a skin biopsy taken for differentiation.

Causes other than *S. aureus* or drugs may produce a similar clinical picture. These conditions include certain fumigants, lymphomas, aspergillosis, irradiation, and graft-versus-host reaction. More recently, the toxic shock syndrome has been described (Table 99.7).

Fever	Sterile pyuria
Toxic epidermal necrolysislike rash	Elevated bilirubin and enzymes
Desquamation (after 10 days)	Low platelets
Hypotension	Disorientation or alteration in consciousness
Vomiting/diarrhea	
Hyperemia of the mucous membranes	

Table 99.7. Toxic Shock Syndrome

Management

Most of the time, SSSS is a self-limited disorder. Antibiotics probably ameliorate the course of the disease, but steroids have no beneficial effects. In fact, steroids may exacerbate the dermatitis by increasing the ability of the organisms to proliferate and produce greater amounts of epidermolytic toxin.

Neonates and children less than 1 year of age should be admitted to the hospital and started on IV antistaphylococcal antibiotics (cefazolin, oxacillin) after blood cultures are obtained. In addition, any older child who is toxic or who has severe skin involvement with significant denudation should be admitted. Close attention should be paid to the child's state of hydration and electrolyte imbalances when a significant amount of skin is lost. Secondary infection, similar to a patient with a major burn, is an important consideration.

Older children with mild involvement limited to dry desquamation, who are not toxic, can be managed on an outpatient basis. These children can be started on oral dicloxacillin or erythromycin (depending on local sensitivities) or cephalexin and followed closely. Skin care is nonspecific unless extensive denudation occurs, then the use of Silvadene cream is warranted.

BITES AND INFESTATIONS (SEE ALSO CHAPTER 91)

Children are often bitten by insects (especially mosquitoes and fleas) and at times are infested by parasites. The papules, urticaria, blisters, and hemorrhagic lesions produced are commonly misdiagnosed. The season of the year, area of the country, grouping and appearance (central punctum) of the lesions, and distribution on exposed surfaces provide the clues necessary for diagnosis.

Mosquitoes and Fleas

Mosquitoes are probably the most common cause of insect bites in children, followed closely by fleas (Fig. 99.25). Mosquito bites are generally limited to the warm months of the year. On the other hand, flea bites, which predominate from spring to fall, can also occur during the winter months as a result of cats and dogs living indoors. At times, flea bites occur without an animal living in the household. Generally, the clinician should ask for a history of visits to a household that has pets or whether the patient's family has recently moved into a home in which the prior owners had pets. In the latter situation, fleas can live in carpeting for a long time.

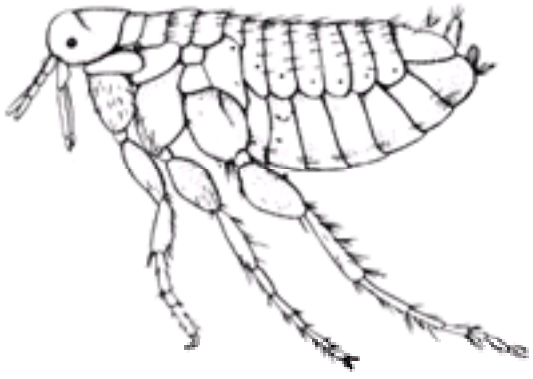

FIGURE 99.25. Flea *(Pulex irritans).*

The distribution of lesions is a valuable clue in making the diagnosis of mosquito or flea bites. Insect bites generally involve the exposed surfaces of the head, face, and extremities. The lesions are usually urticarial wheals that occur in groups or along a line on which the insect was crawling. On occasion, both mosquito bites and flea bites can cause blistering lesions. These lesions are not caused by secondary infection but rather by a violent immune response to the bite. Certainly, excoriation with resulting secondary infection with *S. aureus* or group A streptococci can complicate a simple bite.

A recurrent papular eruption called papular urticaria can occur in young children who become sensitized to insect bites.

Although the lesions tend to occur on exposed parts of the body, with sensitization they may appear at sites distant from the primary bite.

Unfortunately, no specific treatment exists for insect bites. Antihistamines, calamine lotion, or topical steroids have a limited or temporary effect. Prevention by the prophylactic use of insect repellents offers the best solution. Obviously, elimination of the biting insects by treatment of the homes with insecticides or treatment of the infested animals is important.

Tick Bites

Tick bites usually cause only local reactions. Rarely, they are associated with significant systemic illness, including Rocky Mountain spotted fever, tick paralysis, and Lyme arthritis.

When ticks are removed, it is important not to leave fragments of the mouth parts in the skin or to introduce body fluids containing infectious organisms. Various methods have been recommended for removal of ticks from the skin. The only safe method is to use a blunt curved forceps, tweezers, or fingers protected by rubber gloves. The tick is grasped close to the skin surface and pulled upward with a steady even force. The tick must not be squeezed, crushed, or punctured. If mouth parts are left in the skin, they should be removed.

Spider Bites

Loxosceles reclusus, or the brown recluse (Fig. 99.26), found most commonly in the south central United States, is responsible for most skin reactions caused by the bite of a spider. This spider is small, the body being only 8 to 10 mm long, and bears a violin-shaped band over the dorsal cephalothorax. The venom contains necrotizing, hemolytic, and spreading factors. The initial symptoms include mild stinging and/or pruritus. A hemorrhagic blister then appears, which can develop into a gangrenous eschar. Severe bites can cause a generalized erythematous macular eruption, nausea, vomiting, chills, malaise, muscle aches, and hemolysis. Treatment includes oral steroids within 6 to 12 hours after the bite, antibiotics to prevent secondary infection, and surgical removal of the necrotic area to prevent spread of the toxin. An antivenom has been developed at the Vanderbilt School of Medicine.

FIGURE 99.26. Brown recluse spider *(Loxosceles reclusus).*

Scabies Infestation

The cardinal symptom of any infestation with scabies is pruritus. Infants and children excoriate themselves to the point of bleeding. Two clues should be considered when attempting to make this diagnosis: 1) distribution (concentration on the hands, feet, and folds of the body, especially the finger webs) and 2) involvement of other family members. It is important not only to ask other family members if they have pruritus but also to examine their skin. In contrast to adults, infants may develop blisters and also exhibit lesions on the head and face.

The diagnosis is made by scraping involved skin and looking for mites under the ×-10 microscope objective (Fig. 99.27). The materials necessary to examine the skin for scabies are a glass slide, immersion oil, and a No. 15 scalpel blade. A drop of immersion oil should be placed on the glass slide and the No. 15 blade edge dipped into this oil. The best areas to scrape are the interdigital webs of the hands or tiny linear lesions that represent burrows caused by the mite. A recently described procedure involves the use of fountain pen ink applied to the suspected area of skin. After the surface ink is cleaned away, the mite burrows can be identified by means of the ink that has trickled into them.

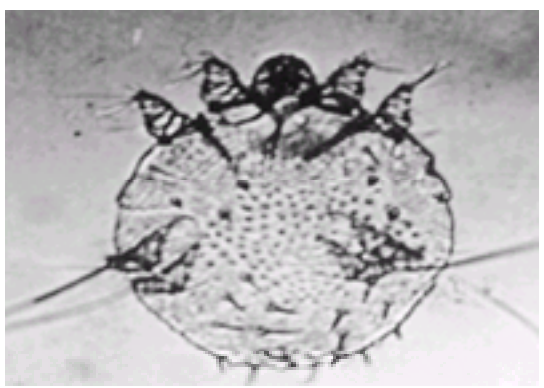

FIGURE 99.27. Mite that causes scabies *(Sarcoptes scabiei).*

Once an infestation occurs, it usually takes 1 month for sensitization and pruritus to develop. The introduction of 5% permethrin cream (Elimite) has obviated the need for lindane and its potential risks. This cream should be applied from head to toe and left on for 8 to 14 hours. The preparation is then washed off with soap and water. Its safety for use in pregnant females has not been proven. All family members and close contacts (e.g., baby-sitters, grandparents) should be treated simultaneously.

Louse Infestation

Three forms of lice infest humans: 1) the head louse, 2) the body louse, and 3) the pubic or crab louse (Fig. 99.28). The major louse infestation in children involves the scalp and causes pruritus. The female attaches her eggs to the hair shaft. The egg then hatches, leaving behind numerous nits (Fig. 99.29) that resemble dandruff. Secondary infection can occur from vigorous scratching. Body lice generally reside in the seams of clothing and lay their eggs there. They go to the body to feed, particularly the interscapular, shoulder, and waist areas. Red pruritic puncta that become papular and wheal-like then occur. Pubic lice occur in the genital area, lower abdomen, axillae, and eyelashes. Transmission is usually venereal. Blue macules (maculae caeruleae) that are 3 to 15 mm in diameter can be seen on the thighs, abdomen, or thorax of infested persons. These macules are secondary to bites.

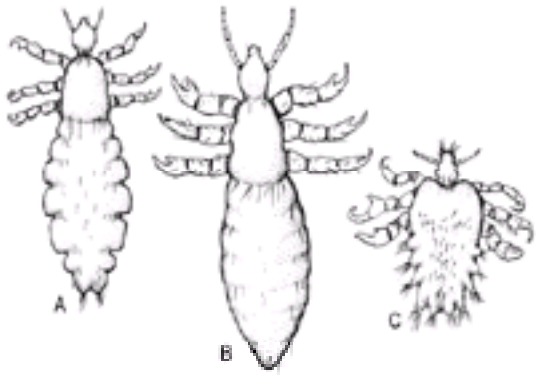

FIGURE 99.28. A. Head louse (*Pediculus humanus var.Capitis*). **B.** Body louse (*Pediculus humanus var. Corporis*). **C.** Pubic louse (*Phthirus pubis*).

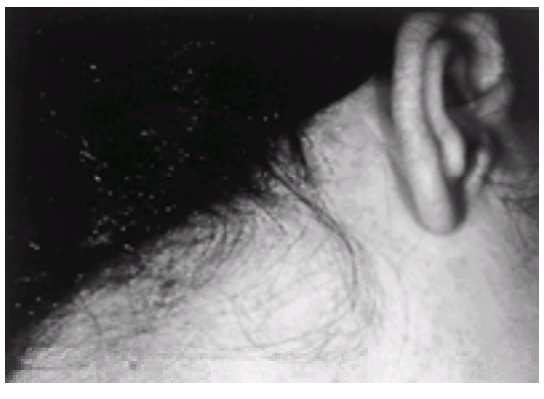

FIGURE 99.29. Nits in the hair of a child with head lice.

Because the body louse resides in clothing, therapy consists mainly of disinfecting the clothing with steam under pressure. Pediculosis capitis is most effectively treated with 1% permethrin creme rinse (Nix). The patient's hair should be shampooed, rinsed, and toweled dry. Enough permethrin to saturate the hair and scalp is applied. The medication is washed out after 10 minutes. Pediculosis pubis is best treated with the same preparation. Any nits are removed with a fine-toothed comb. The safest treatment for lice in the eyelashes is the application of white petrolatum twice daily for 8 days. The lice stick to the Vaseline, cannot feed, and die. Another less safe therapy is physostigmine ophthalmic ointment.

SUPERFICIAL FUNGAL INFECTIONS OF THE SKIN

Tinea Corporis

Tinea corporis (Fig. 99.30) is characterized by one or more sharply circumscribed scaly patches. The center of the circular patch generally clears as the leading edge spreads out. The leading edge may be composed of papules, vesicles, or pustules. The lesions are most commonly confused with nummular eczema. The diagnosis can be made by scraping the active outer rim of papules and examining the scales with a potassium hydroxide (KOH) preparation under the microscope (Fig. 99.31). These lesions do not fluoresce under the Wood's light. The most common offending fungi

are Trichophyton tonsurans and Microsporum canis. Treatment with topical antifungal agents such as clotrimazole, miconazole, or econazole produces clearing in 7 to 10 days. Therapy should be maintained for 2 weeks. If improvement does not occur, treatment with griseofulvin (15 mg/kg per day in two divided doses) will usually resolve the problem.

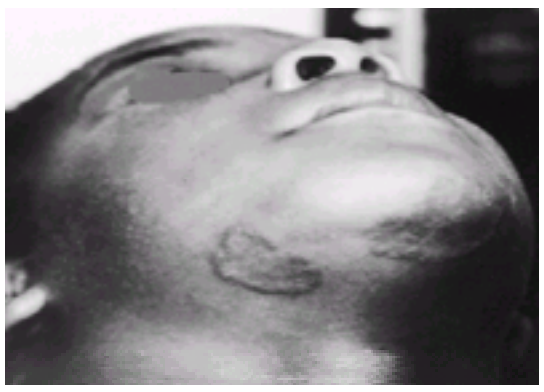

FIGURE 99.30. Child with tinea corporis.

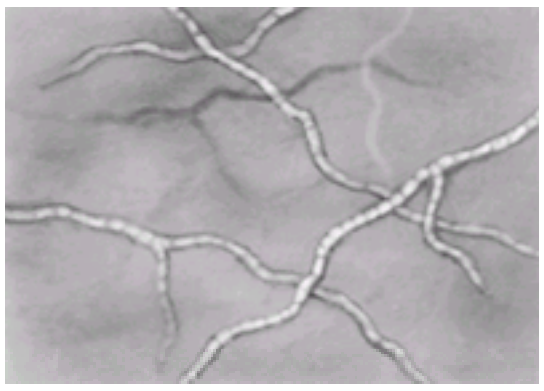

FIGURE 99.31. Characterization of a positive potassium hydroxide (KOH) preparation demonstrating branching hyphae running across the microscope field.

Tinea Capitis

Although tinea capitis was commonly caused by the *Microsporum* species in the past, it usually results now from infection by *Trichophyton tonsurans.* The two forms have different clinical appearances. The *Microsporum* species (Table 99.8) generally causes round patches of scaling alopecia (Fig. 99.32). Illumination of a lesion with a Wood's lamp gives a blue–green fluorescence. Kerion formation can occur as a swollen, boggy abscess. The *Trichophyton* species (Table 99.9) usually causes scattered alopecia, not always oval or rounded; the alopecia is irregular in outline with indistinct margins. Normal hairs grow within the patches of alopecia. At times, the hairs break off at the surface of the scalp, leaving a “black dot” appearance (Fig. 99.33). Diffuse scaling may simulate dandruff, and although minimal hair loss is present, it is not perceived. Wood's light examination of the lesion does not produce fluorescence. The organism can cause a folliculitis suppuration and kerion formation (Fig. 99.34). Diagnosis is made by culturing the affected scalp area (Fig. 99.35). The clinician should consider the presence of tinea capitis when a nonresponsive seborrheic or atopic dermatitis of the scalp is present, black dots are seen, or increased scaling follows the use of topical steroids. With the use of dermatophyte test media (DTM), a color change occurs in the media (yellow to red) in the presence of a growing dermatophyte. If a kerion is present, the swelling (allergic reaction to the fungus) can be controlled by a combination of prednisone and griseofulvin.

Round patches of scaling alopecia
Fluoresce blue-green
Kerion formation

Table 99.8. Tinea Capitis *Microsporum* Species

Partial scattered alopecia—not always oval or rounded	Black dots, diffuse scaling ("dandruff")
Alopecia irregular in outline with indistinct margins	Nonfluorescent
Normal hairs growing within patch of alopecia	Folliculitis, suppuration, kerion formation

Table 99.9. Tinea Capitis *Trichophyton* Species

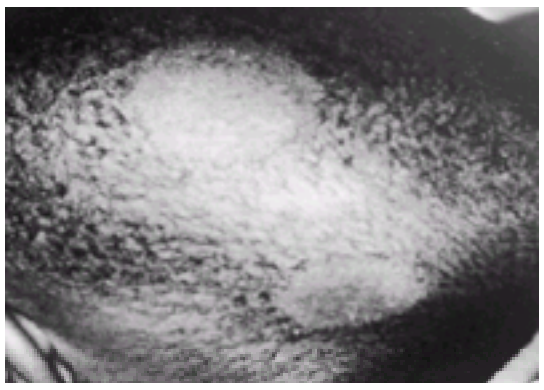

FIGURE 99.32. Tinea capitis secondary to infection with *Microsporum audouinii.*

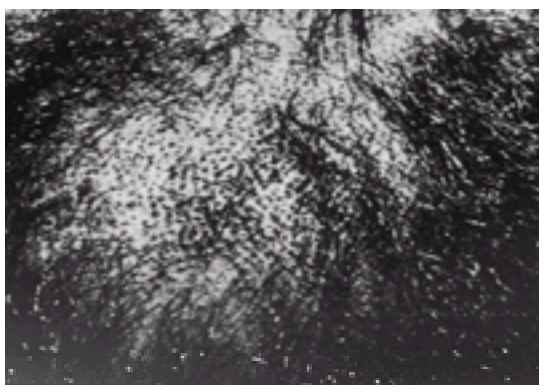

FIGURE 99.33. "Black-dot" appearance of scalp infection with *Trichophyton tonsurans.*

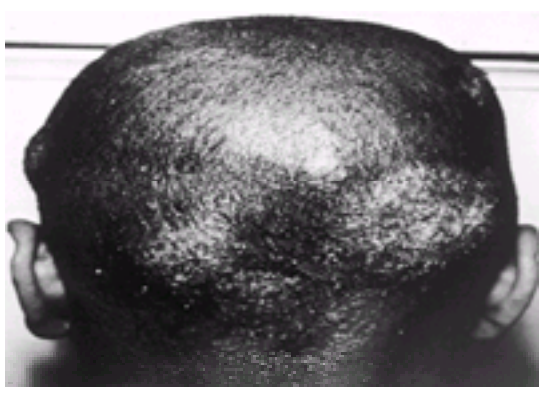

FIGURE 99.34. Patient with tinea capitis and multiple kerions.

FIGURE 99.35. Toothbrush implants of scalp brushing that are growing fungus.

In the differential diagnosis of patchy hair loss, as is seen in tinea capitis, the clinician should consider alopecia areata (Fig. 99.36). However, with alopecia areata, no inflammation or scaling of the scalp occurs. *Trichotillomania,* the term given to the habit children develop of rubbing, twirling, or playing with their hair to the point that the hair breaks and is lost in irregular patches, should also be considered. Traction alopecia occurs with certain hair styles. Hair is lost at the margins of the hairline with the ponytail style or frequent use of hair rollers. Tight braiding or corn-rowing can cause hair loss on any area of the scalp. At times, papules or pustules occur where the skin has been disrupted by the traction. Infants who are left on their backs for long periods may lose hair at the occiput from the constant friction in that area.

FIGURE 99.36. Child with alopecia areata.

Treatment for tinea capitis consists of orally administered griseofulvin 15 to20 mg/kg per day in two divided doses with a glass of milk for 6 weeks. Adjunctive therapy includes the use of 2.5% selenium sulfide shampoo twice weekly. With the use of this shampoo, shedding of spores is decreased within 1 to 2 weeks. Two newer oral medications, itraconazole and terbinafine, are currently under study and may eventually replace griseofulvin as the preferred medication.

Tinea Cruris

Tinea cruris begins as a small, red, scaling rash in the groin that spreads peripherally and clears centrally. The edges are sharply marginated and scalloped, extending down the thighs. Generally, the scrotum is not noticeably involved. This fungal infection is most common in semitropical regions where heat and high humidity are prevalent. Tight-fitting clothes also contribute to the problem by preventing evaporation. Other conditions to consider are seborrheic dermatitis (which usually can be differentiated by involvement of other areas of the body such as the ears, scalp, and eyelids), intertrigo (generally secondary to friction and maceration), contact dermatitis, candidiasis (which usually involves the inner thigh and causes the scrotum to appear bright red), and erythrasma. The clinician should always check the feet to make sure there is not fungal involvement in that area as well. In general, this condition affects only postpubertal children. Diagnosis is made by KOH preparation. Nonspecific measures for treatment include loose-fitting clothing, reducing the amount of perspiration by using dusting powders, and decreasing intake of caffeine-containing foods. Clotrimazole, miconazole, tolnaftate, and econazole are useful as topical antifungal agents. Oral griseofulvin may be needed in severe cases.

Tinea Pedis

Tinea pedis is generally caused by *Trichophyton rubrum* or *T. mentagrophytes.* It occurs most commonly in postpubertal children. The cracking and peeling of the skin suggestive of tinea pedis in prepubertal children more often indicates the presence of atopic eczema or hyperhidrosis. Tinea pedis is a penalty of civilization in that it occurs only in those individuals who wear shoes. KOH preparation will demonstrate hyphae, especially when samples are taken from between the fourth and fifth interspaces of the toes. Clinically, the skin has a dry, white, hazy appearance and is often pruritic. When secondary bacterial infection is present, an odor occurs. At times, an inflammatory type of lesion (caused by *T. mentagrophytes*) causes blistering. The presence of an id reaction indicates dissemination of antigen to other parts of the body, especially the hands.

The differential diagnosis of tinea pedis includes simple maceration, contact dermatitis, and atopic eczema. Treatment consists of drying the feet thoroughly after washing; wearing dry, clean socks; avoiding caffeine-containing foods to decrease sweating; keeping shoes off as much as possible; and walking barefoot or in sandals. Topical antifungal agents (see the previous section on Tinea Cruris) and/or oral griseofulvin are used to treat this condition. Newer medications (see the previous section on tinea capitis) are on the horizon.

Tinea Versicolor

Tinea versicolor refers to a superficial infection of the skin caused by *Pityrosporum orbiculare,* which produces color changes of the skin, hypopigmentation, hyperpigmentation, and redness (Fig. 99.37). Wood's light examination usually shows yellowish brown fluorescence. Because moisture promotes growth of the organism, exacerbations occur in warm weather or in athletes who sweat excessively. The infection is difficult to eradicate and recurs frequently. A KOH

preparation shows large clusters of spores and short, stubby hyphae, often called meatballs and spaghetti.

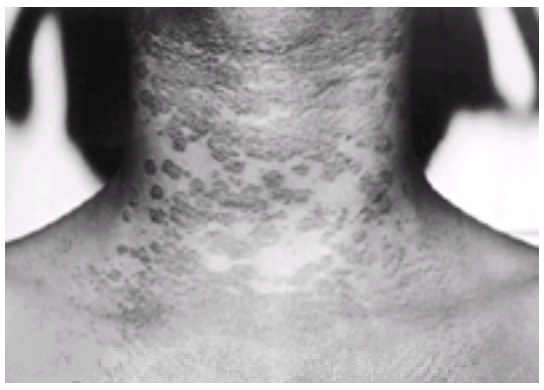

FIGURE 99.37. Adolescent with tinea versicolor.

Treatment consists of lathering the entire body with selenium sulfide shampoo (2.5% concentration) after wetting the skin surface in a shower. The lather is left on for 20 minutes and is then showered off. This procedure is carried out multiple times during the first week, with decreasing frequency over the ensuing weeks. Maintenance therapy is advisable because of the high incidence of recurrence. Localized areas of involvement can be treated with topical antifungal agents (e.g., econazole, ketoconazole topically). Adolescents can be treated with 400 mg of ketoconazole initially and then 200 mg at monthly intervals during the warm summer months or during a sports season when the child sweats frequently.

PYOGENIC GRANULOMAS

Pyogenic granulomas (Fig. 99.38) are vascular nodules that develop rapidly at the site of an injury such as a cut, scratch, insect bite, or burn. The histologic picture is that of proliferating capillaries in a loose stroma. Although this lesion was previously believed to be caused by infection of a small wound, the definite cause has not been established. Pyogenic granulomas occur commonly in children and young adults, usually on the fingers, face, hands, and forearms.

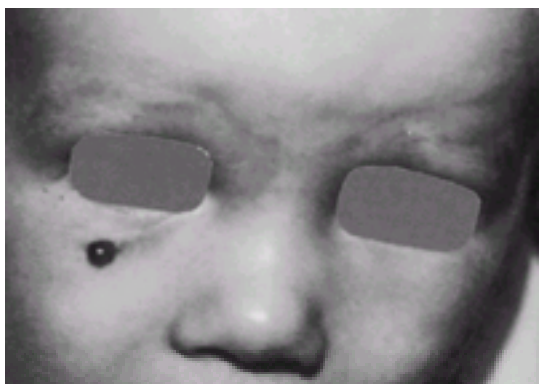

FIGURE 99.38. Pyogenic granuloma on the cheek of a child.

Clinically, the lesions are bright red to reddish brown or blue–black. The vascular nodules are pedunculated, ranging from 0.5 to 2 cm in size. Their surfaces are glistening, or raspberrylike, often becoming eroded and crusted. They bleed easily. Generally, they are asymptomatic. Because spontaneous disappearance is rare, the lesions must be removed by excision, electrosurgery, or cryosurgery.

URTICARIA

Background

Urticaria as a symptom complex is often encountered in the pediatric population, occurring in 2 to 3% of all children. In most cases, no cause is identified. A small number of cases are caused by allergic reactions from the ingestion of drugs or foods (e.g., nuts, eggs, shellfish, strawberries). Urticaria also follow viral (e.g., Epstein-Barr virus, hepatitis), bacterial (streptococcal), or parasitic infections. Physical factors, including dermographism, cholinergic stimulation (induced by heat, exercise, and emotional tension), cold (acquired and familial), and solar exposure, can induce urticaria. Finally, urticaria may be caused by factors producing a vasculitis and substances causing degranulation of mast cells (radiocontrast material). Episodes of urticaria that last less than 6 weeks are termed transient or acute. The most common causes of urticaria are infection, insect bites, drugs, and foods. Chronic urticaria are defined as those that last more than 6 weeks. No cause is found in 90% of children. These cases include the physical urticarias or urticarial vasculitis.

Pathophysiology

The lesion itself follows vasodilation and leakage of fluid and red blood cells from involved vessels. The vascular damage can be caused by mediators such as histamine complement and immune complexes. IgE can attach to and cause

degranulation of mast cells in sensitized individuals with resulting histamine release. Urticarias are usually acute and transient, but at times become chronic and recurrent.

Clinical Manifestations

The typical urticarial lesions are familiar to all physicians. They can be localized or generalized (involving the entire body). At times, the lesions are giant with serpiginous borders. Individual wheals rarely last more than 12 to 24 hours. Most commonly, the lesions appear in one area for 20 minutes to 3 hours, disappear, and then reappear in another location. The total duration of an episode is usually 24 to 48 hours; however, the course can last 3 to 6 weeks.

Management

Acute relief can be accomplished with subcutaneous epinephrine (1:1000) 0.01 mL/kg and intramuscular diphenhydramine 1 mg/kg. Prolonged sympathetic effect can be maintained with Sus-Phrine 0.005 mL/kg. Oral antihistamines are useful for maintenance therapy for transient urticaria. Hydroxyzine hydrochloride (2 mg/kg per day in three to four divided doses) or diphenhydramine hydrochloride (5 mg/kg per day in three to four divided doses) should be prescribed for at least 10 days. Newer long-acting antihistamines include terfenadine (dose varies by age) or loratadine (10 mg once per day).

PITYRIASIS ROSEA

Pityriasis rosea can occur in all age groups but is seen predominantly after 10 years of age and only rarely under 5 years of age. The cause is unknown; however, a viral cause is suspected. Less than 5% of cases occur in multiple family members. In 80% of children, a large, oval, solitary lesion known as the herald patch appears on the trunk (Fig. 99.39) before the eruption of subsequent lesions. Individual lesions are oval and slightly raised, pink to brown, with peripheral scaling. Because the lesions follow the cleavage lines (Fig. 99.40) of the skin, the backs of patients have a “Christmas tree” appearance. Generally, the face, the scalp, and distal extremities are spared. On occasion, an inverse distribution occurs (lesions on the face and extremities with truncal sparing). The rash is pruritic early in the course but then becomes asymptomatic. It lasts 4 to 8 weeks.

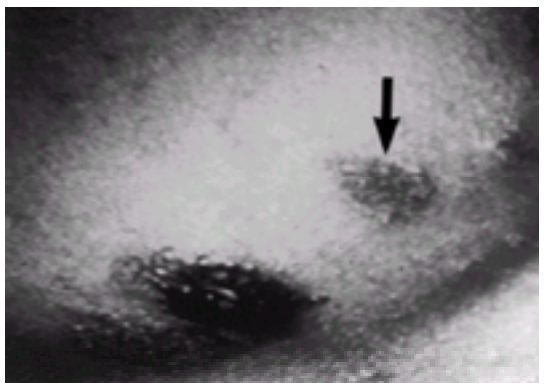

FIGURE 99.39. Herald patch *(arrow)* in adolescent with pityriasis rosea.

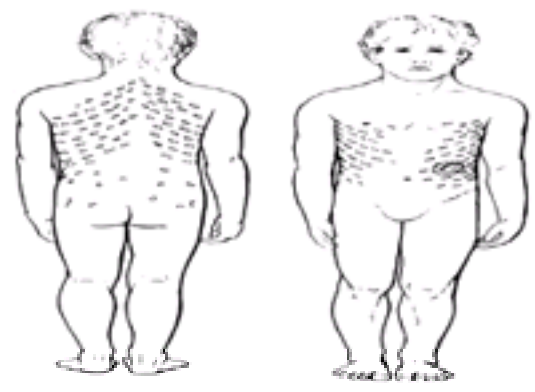

FIGURE 99.40. Typical distribution of pityriasis rosea.

The herald patch can be mistaken for tinea corporis, but a KOH preparation eliminates that possibility. When pityriasis rosea appears in adolescence, it must be differentiated from secondary syphilis. Clinical clues are helpful (Table 99.10), but serologic testing is necessary.

	Pityriasis Rosea	Syphilis
Herald patch	+	-
Ovals follow dermatomes	+	-
Lymphadenopathy	-	+
Mucous membrane lesion	-	+

Table 99.10. Differential Diagnosis

Treatment is symptomatic. Antihistamines and topical emollients can help the pruritus.

PANNICULITIS

Erythema Nodosum

Erythema nodosum seems to be a hypersensitivity reaction to infection (streptococci, tuberculosis, coccidioidomycosis, histoplasmosis), inflammatory bowel disease, sarcoidosis, and drugs. The exact immunologic mechanism has not been clarified. The entity occurs predominantly in adolescents during the spring and fall. Females are affected more often than males.

The lesions of erythema nodosum appear as deep, tender, erythematous nodules or plaques on the extensor surfaces of the extremities (Fig. 99.21). The sedimentation rate is generally elevated and usually returns to normal with disappearance of the eruption unless an underlying disease is present. The reaction usually lasts 3 to 6 weeks. Treatment should be directed toward the cause when and if established; otherwise, it is symptomatic (aspirin and antihistamines). Hospitalization is unnecessary. Corticosteroids should not be used except in severe cases after an underlying infection has been ruled out.

Cold Panniculitis

Cold panniculitis is secondary to cold injury to fat. During the cold of winter, infants and some older children develop red, indurated nodules and plaques on exposed skin, especially the face. The subcutaneous fat in infants and some children solidifies more readily at a higher temperature than that of an adult because of the relatively greater concentration of saturated fats. Infants who hold ice popsicles in their mouths are also susceptible to this phenomenon (Fig. 99.41). The lesions gradually soften and return to normal over 1 or more weeks. Treatment is unnecessary.

FIGURE 99.41. Infant with popsicle panniculitis of the cheek. *Please see the color-tip insert (Color Plate 99.41).*

WARTS AND MOLLUSCUM CONTAGIOSUM

Warts

Warts affect 7 to 10% of the population and are one of the most common dermatologic problems encountered in pediatrics. The peak incidence is during adolescence. Sixty-five percent of common warts disappear spontaneously within 2 years, and 40% of plantar warts disappear within 6 months in prepubertal children. However, immunosuppressed patients may have extensive spread of the lesions.

The common wart resembles a tiny cauliflower. The shape of the wart varies with its location on the skin. They may be long and slender (filiform) on the face and neck or flat (verruca plana) on the face, arms, and knees. When located on the soles, they are called plantar warts, and when in the anogenital area, they are referred to as condyloma acuminata.

The tendency for recurrence of warts makes the treatment of this condition frustrating. Because most warts disappear spontaneously with time, procedures that are least traumatic for the child should be attempted first (Table 99.11). The

simple, nontraumatic method of airtight occlusion with plain adhesive tape for 1 month has been shown to be successful on many occasions. Topical application of salicylic acid in flexible collodion (Duofilm; Table 99.12) is good for home use, as are some of the over-the-counter preparations (e.g., Compound W). When simple methods are unsuccessful, touching the warts with liquid nitrogen for 20 to 30 seconds or surgical removal can be attempted. Both procedures are painful.

Decrease irritation—cover with tape (1–2 months)
Over-the-counter preparations such as Compound W (1 month)
Salicylic in collodion (Duofilm) (1 month)
Refer to dermatologist

Table 99.11. Management of Warts

1. Soak wart for 5 min.	5. Cover with tape.
2. Dry.	6. Repeat twice a day.
3. Surround with petroleum jelly.	7. Pare dead skin.
4. Apply Duofilm (let dry for few minutes).	

Table 99.12. Use of Duofilm

Plantar warts can be treated with 40% salicylic acid plaster. Circular pieces, slightly larger then the plantar wart, are cut from a sheet of this material. They are placed on the wart and kept in place continuously with adhesive tape for 1 to 2 weeks. At that point, the dead tissue is carefully pared away. Treatment is continued until normal skin is seen in place of the wart.

Anogenital warts are treated with 20% podophyllin. This medication is carefully applied to the wart only and washed off in 4 to 6 hours (see Chapter 94). Severe burning of the skin occurs if the material is not completely removed. Child abuse should be considered in any child with genital warts.

Molluscum Contagiosum

The lesion, produced by the common poxvirus, is a papule with a white center (Fig. 99.42). It occurs at any age during childhood. It is more common in swimmers and wrestlers. Patients with atopic eczema are especially susceptible. Most lesions resolve in 6 to 9 months, but some may persist for more than 3 years. Spread is by autoinoculation.

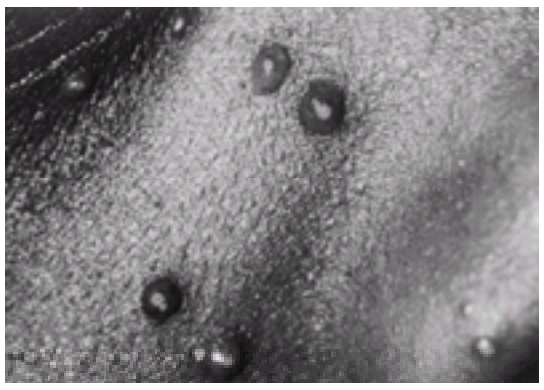

FIGURE 99.42. Molluscum contagiosum. Papules with white centers that contain the virus.

Lesions can be single or numerous and favor intertriginous areas such as the groin. They are usually 2 to 5 mm in diameter, but several can coalesce and form a lesion 1.5 cm in diameter. They may become inflamed, which sometimes heralds spontaneous disappearance. At times, an eczematous reaction occurs around some lesions, and they can

become secondarily infected.

Treatment should be gentle. Removal of the white core will cure the lesion. This treatment can be performed by applying eutectic mixture of local anasthetics (EMLA) cream under occlusion to the lesion 1 to 2 hours before treatment. This procedure will anesthetize the area and allow the physician to prick the skin open over the core with a 26-gauge needle, and squeeze the core out with a comedome extractor. Multiple light touches with liquid nitrogen can also be effective. With widespread lesions, nonpainful procedures are preferable. Application of 0.1% retinoic acid one to two times daily may induce enough inflammation to hasten the host's immune response or cause extrusion of the central core. Multiple painful treatments will cause great fear in the patient and make future visits to a physician difficult for the family.

CONGENITAL HERPES SIMPLEX VIRUS

Congenital herpes simplex virus (HSV) infection encompasses a broad clinical spectrum ranging from localized cutaneous and mucosal lesions to life-threatening central nervous system and internal organ involvement. Studies have shown that the prevalence of HSV-2 infection has increased by 30% from 1976 to 1994. The greatest increases occurred in white teenagers and white women in their twenties. The risk of acquiring HSV during pregnancy was 2% in susceptible women. The risk of acquiring the infection is similar in each trimester. Less than 10% of those who were seropositive gave a history indicating genital herpes infection. Other patients noted nonspecific genital urinary symptoms such as dysuria, leukorrhea, hematuria, and pelvic pain. If seroconversion occurs in the mother before delivery, the risk to the infant is small. However, if a primary infection occurs shortly before labor, neonatal HSV infection can occur in up to 50% of newborns. Whereas most infections are transmitted by direct contact with the infected birth canal during the second stage of labor, some evidence supports transplacental infection of the fetus.

The incubation period of congenital HSV infections ranges from 2 to 30 days after exposure. Lesions present at birth or shortly thereafter have been explained by transplacental passage of the virus. The clinical manifestations of congenital HSV infection are diverse, but more than 50% of infected neonates present with external involvement. In vertex deliveries, the scalp is a common site for the vesicles. Conversely, infants delivered by breech often develop lesions of the buttocks and perianal area initially. The lesions are not unlike those seen in older children or adults in that they are grouped tense vesicles arising on an erythematous base. However, the infection may present on the skin as individual vesicles, pustules, bullae, or denuded skin. Unfortunately, when infants have disease limited to the integument, HSV infection is often not considered as a possibility. Instead, these children are treated for “impetigo.” The correct diagnosis may not be considered until constitutional symptoms such as fever, hypothermia, poor feeding, irritability, lethargy, and vomiting have appeared. By then, dissemination of the disease has occurred.

Diagnosis of congenital HSV infection should be suspected in any infant less than 1 month of age who has a vesicular eruption on an erythematous base. A Tzanck preparation of the base of an unbroken vesicle is an easy and rapid diagnostic tool to aid in the recognition of this potentially lethal disease (Fig. 99.7). Giemsa, Papanicolaou, or hematoxylin-eosin stains of the smeared preparation of vesicles infected with HSV will reveal multinucleated giant cells, intranuclear inclusions, ballooning degeneration, or margination of nuclear chromatin. Rapid slide tests, using monoclonal antibodies, are also available for rapid diagnosis. Viral culture still remains the gold standard for proving that HSV is present.

The differential diagnosis of vesiculopustular lesions in the newborn includes bullous impetigo, congenital cutaneous candidiasis, congenital syphilis, neonatal pustulomelanosis, and cytomegalovirus infection. Differentiation of these entities requires the use of Gram stains, KOH scrapings, serologic studies for syphilis, and appropriate cultures. Although many serologic tests are currently available to detect the presence of HSV antibodies, none of these studies is valuable in arriving at an early diagnosis of congenital infection. Direct culture of the herpes virus from a lesional vesicle takes 24 to 48 hours for identification.

All infants with suspected congenital HSV should be treated with IV acyclovir, which is the preferred drug. Acyclovir is should be given at a dosage of 30 mg/kg per day, divided every 8 hours (10 mg/kg per dose), in a 1- to 2-hour IV infusion.

DISORDERS OF PIGMENTATION

Hypopigmentation

A dominant form of partial albinism occurs, in which localized areas of skin and hair are devoid of pigment. Ocular albinism is also seen. Two syndromes with albinism are Waardenberg's syndrome (white forelock, heterochromia of the iris, sensorineural hearing loss) and Chediak-Higashi syndrome (immunodeficiency, leukocytes with giant granules).

Loss of pigmentation can be a result of absence of melanocytes as in vitiligo and halo nevi. Vitiligo is a symmetric, patchy loss of pigmentation. Hair located in areas of vitiligo is often white. Vitiligo can be associated with alopecia areata, pernicious anemia, Addison's disease, hypothyroidism, diabetes mellitus, hypoparathyroidism, and other endocrine disorders. Vitiligo and some of the diseases associated with it may be autoimmune disorders. Antibodies directed against melanocytes have been detected.

Suppression of melanocytic pigment production can cause loss of pigmentation as in postinflammatory hypopigmentation. An example of this condition is the white patch of hypopigmentation and scaling often seen on the face, trunk, or extremities of children with atopic eczema. The ash-leaf macule is a flat, hypopigmented (whitish) spot that is present in more than 90% of patients with tuberous sclerosis. In white patients, they are more easily seen by shining a Wood's lamp on the skin.

Hyperpigmentation

Diffuse hyperpigmentation is associated with Addison's disease, acromegaly, and hemochromatosis.

Pigment deep in the dermis appears gray or blue at the surface of the skin. Mongolian blue spots are an example. The nevus of Ota is dermal pigment in the distribution of the ophthalmic branch of the fifth nerve; this pigmentation can involve the sclera and palate as well.

Certain syndromes, including neurofibromatosis, are associated with pigmented skin lesions. Patients with this disease have café-au-lait spots, which are flat, nonpalpable, coffee-colored lesions of varying size and shape. When six or more lesions are present, greater than 0.5 cm in size, neurofibromatosis should be considered. The Peutz-Jeghers syndrome is a dominantly inherited condition that includes frecklelike lesions of the lips, nose, buccal mucosa, fingertips, and subungual areas associated with polyps in the small intestine, stomach, or colon. Melena and intussusception are the chief complications that may develop, usually in the second decade of life. Albright's syndrome should be suspected when unilateral café-au-lait spots with irregular borders occur in the lumbosacral area. Included in this syndrome are bony abnormalities and precocious puberty.

Single or multiple red–brown papules or nodules occurring on the extremities or face of children may have a confusing tissue structure. Although the lesions are benign, they have commonly been misdiagnosed as malignant melanoma. Therefore, the name *benign juvenile melanoma* has been assigned to this condition. Other names include *Spitz tumors* or *spindle-cell epithelioid nevi.* True malignant melanomas are rare in children. They usually arise from congenital pigmented nevi.

Suggested Readings

GENERAL

Fitzpatrick TB. Dermatology in General Medicine. 4th ed. New York: McGraw-Hill, 1993.

Hurwitz S. Clinical Pediatric Dermatology. 2nd ed. Philadelphia: WB Saunders, 1993.

Schachner LA, Hansen RC. Pediatric Dermatology. 2nd ed. New York: Churchill Livingston, 1995.

Solomon LM, Esterly NB. Neonatal dermatology. In: Major Problems in Clinical Pediatrics. Philadelphia: WB Saunders, 1973.

Weston WL, Lane AT. Color Textbook of Pediatric Dermatology. 2nd ed. St Louis: Mosby, 1996.

ATOPIC DERMATITIS

Hanifin JM. Atopic dermatitis in infants and children. Pediatr Clin North Am 1991;38:763.

Leyden JJ, Baker DA. Localized herpes simplex infections in atopic dermatitis. Arch Dermatol 1979;115:311.

Rothe MJ, Grant-Kels JM. Atopic dermatitis: an update. J Am Acad Dermatol 1996;35:1.

Sampson HA, Scanlon SM. Natural history of food hypersensitivity in children with atopic dermatitis. J Pediatr 1989;115:23.

ALLERGIC CONTACT DERMATITIS

Epstein WL. Topical prevention of poison ivy/oak dermatitis. Arch Dermatol 1989;125:499.

Gaul LE. Development of allergic nickel dermatitis from earrings. JAMA 1967;200:176.

Weston WL, Weston JA. Allergic contact dermatitis in children. Am J Dis Child 1984;138:932.

DIAPER DERMATITIS

Berg RW, Buckingham KW, Stewart RL. Etiologic factors in diaper dermatitis: the role of urine. Pediatr Dermatol 1986;3:102.

Buckingham KW, Berg RW. Etiologic factors in diaper dermatitis: the role of feces. Pediatr Dermatol 1986;3:107.

Honig PJ, Gribetz B, Leyden JL, et al. Amoxicillin and diaper dermatitis. J Am Acad Dermatol 1988;19:275.

Leyden JJ, Katz S, Stewart R, et al. Urinary ammonia and ammonia-producing microorganisms in infants with and without diaper dermatitis. Arch Dermatol 1977;113:1678.

Weston WL, Lane AT, Weston JA. Diaper dermatitis: current concepts. Pediatrics 1980;66:532.

DRUG REACTIONS

Esterly NB. Corticosteroids for erythema multiforme? Pediatr Dermatol 1989;6:229.

Kramer MS, Hutchinson TA, Flegel KM, et al. Adverse drug reactions in general pediatric outpatients. J Pediatr 1985;106:305.

Orton PW, Huff JC, Tonnesen JG, et al. Detection of herpes simplex viral antigen in skin lesions of erythema multiforme. Ann Intern Med 1984;101:48.

Prendeville JS, Hebert AA, Greenwald MJ, et al. Management of Stevens-Johnson syndrome and toxic epidermal necrolysis in children. J Pediatr 1989;115:881.

STAPHYLOCOCCAL SCALDED SKIN SYNDROME

Amon RB, Diamond RL. Toxic epidermal necrolysis: rapid differentiation between staphylococcal and drug-induced disease. Arch Dermatol

1975;111:1433.

Melish ME, Glasgow LA. The staphylococcal scalded-skin syndrome: development of an experimental model. N Engl J Med 1970;282:1114.

Murono K, Fujita K, Yoshioka H. Microbiologic characteristics of exfoliative toxin-producing staphylococcus aureus. Pediatr Infect Dis J 1988;7:313.

BITES AND INFESTATIONS

Davies HD, Sakuls P, Keystone JS. Creeping eruption. Arch Dermatol 1993;129:588.

Hershey FB, Aulenbacher CE. Surgical treatment of brown spider bites. Ann Surg 1969;170:300.

Meinking TL, Taplin D, Hermida JL, et al. The treatment of scabies with Ivermectin. N Engl J Med 1995;333:26.

Needham GR. Evaluation of five popular methods for tick removal. Pediatrics 1985;75:997.

Paller AS. Scabies in infants and small children. Semin Dermatol 1993;12:3.

Taplin D, Meinking BA, Porcelain SL, et al. Permethrin 5% dermal cream: a new treatment for scabies. J Am Acad Dermatol 1986;15:995.

FUNGAL INFECTIONS

Gupta AK, Sauder DN, Shear NH. Antifungal agents: an overview. Part I, Part II. J Am Acad Dermatol 1994;30:677,911.

Guzzo C, Honig PJ, Rabinowitz LG. Fungal infections from head to toe. Patient Care 1987;21:62.

Honig PJ, Caputo GL, Leyden JJ, et al. Microbiology of kerions. J Pediatr 1993;123:422.

Honig PJ, Caputo GL, Leyden JJ, et al. Treatment of kerions. Pediatr Dermatol 1994;11:69.

URTICARIA

Ellis CN. Current management of urticaria and angioedema: proceedings of a round table. J Am Acad Dermatol 1991;25(Suppl):Part 2.

Ghosh S, Kanwar AJ, Kaur A. Urticaria in children. Pediatr Dermatol 1993;10:107.

Schuller DE, Elvey SM. Acute urticaria associated with streptococcal infection. Pediatrics 1980;65:592.

CONGENITAL HERPES SIMPLEX

Brown ZA, Selke S, Zeh J, et al. The acquisition of herpes simplex virus during pregnancy. N Engl J Med 1997;15:337.

Brown ZA, Vontver LA, Benedetti J, et al. Effects on infants of a first episode of genital herpes during pregnancy. N Engl J Med 1987;317:1246.

Fleming DT, McQuillan, Johnson RE, et al. Herpes simplex virus type 2 in the United States, 1976 to 1994. N Engl J Med 1997;337:1105.

Goodyear HM. Rapid diagnosis of cutaneous herpes simplex infections using specific monoclonal antibodies. Clin Exp Dermatol 1994;19:294.

PITYRIASIS ROSEA

Parsons JM. Pityriasis rosea update: 1986. J Am Acad Dermatol 1986;15:159.

CHAPTER 100

Oncologic Emergencies

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Each year only 1 child in 10,000 develops cancer. Yet, next to accidents, cancer remains the leading cause of death in children 100older than 1 year of age. Fortunately, with carefully planned therapy, more than half the children diagnosed with cancer can now be expected to become healthy, productive members of society.

Not so much because cancer is potentially fatal, but because it is potentially curable, pediatric oncology concerns all physicians who see children. The pediatrician or the emergency physician must first consider cancer as a diagnosis and must decide which child needs a complete blood count (CBC), radiograph, or biopsy. Children who develop cancer complain of the same things that bother most children: fatigue, fever, headache, or pain. After a history, physical examination, and appropriate laboratory tests, the emergency physician may already have sufficient information to make a presumptive diagnosis of cancer. His or her role in the management of the child may consist of sharing this information with the family and then referring the child to a specialist who is qualified to confirm the diagnosis and undertake the care of pediatric cancer. On some occasions, however, the emergency physician must begin supportive therapy that is potentially lifesaving or care for a child who has a complication of cancer therapy.

This chapter is divided into two sections. The first section describes the presentation and initial care of children with cancer. Leukemia is described first and in greatest detail because it is often diagnosed in the emergency department (ED); the lymphomas and histiocytic diseases are discussed separately because they may present in a variety of locations. Soft tissue and bone sarcomas are discussed separately for the same reason. Other solid tumors, including central nervous system (CNS) tumors, are presented by location because signs, symptoms, and potential emergencies are related to the surrounding anatomy.

Common problems of children undergoing cancer therapy or with progressive tumor are discussed in the second section. During treatment, the most common problems relate to 1) the risk of hemorrhage, anemia, and infection as a result of bone marrow suppression; 2) metabolic derangement caused by tumor lysis or the side effects of chemotherapy; 3) the compression of a vital organ by progressive tumor growth; or 4) damage to a vital organ as a result of cancer therapy.

Pain is associated with cancer at diagnosis, at the time of painful procedures, and when the disease has failed to respond to therapy. The pain of cancer is often severe and can qualify as an emergency. This topic is discussed briefly in the second section (see also Chapter 4).

DIAGNOSIS AND INITIAL CARE OF PATIENTS WITH CANCER

The early symptoms and signs of cancer can be indistinguishable from the initial manifestations of more common childhood illnesses. Table 100.1 reviews common symptoms and signs associated with pediatric malignancies. The history and physical examination often suffice to rule out cancer. In general, any mass in the abdomen of a child should be considered possibly malignant. Multiple cytopenias must be considered a sign of malignancy until a bone marrow examination proves otherwise or until other evidence establishes the cause. The primary care or emergency physician should remember that, except in the rare circumstance when the need for emergency care is clear, the diagnostic workup of a malignancy should be completed in a pediatric oncology center where all tests required by modern treatment protocols are available. Tissue for diagnosis should be obtained at the institution that will undertake the child's care. Today, for almost all diseases, viable tissue is essential for categorizing diseases and determining treatment; histologic slides and cell blocks may not be sufficient.

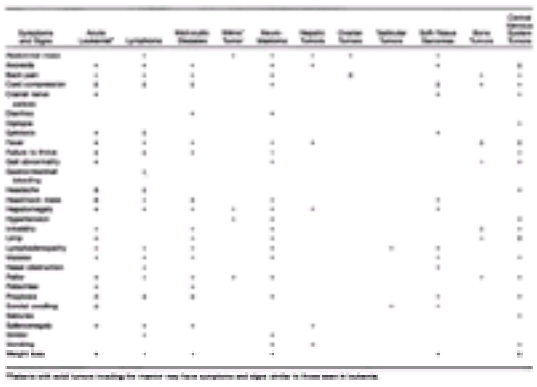

Table 100.1. Common Presenting Symptoms and Signs of Pediatric Malignancies

Leukemia

Background

Definition

In leukemia, excessive numbers of immature malignant white blood cells (WBCs), “leukemic blasts,” replace normal marrow hematopoietic cells. Leukemic blasts leave the marrow, circulate in the peripheral blood, and infiltrate lymph nodes, liver, spleen, meninges, and soft tissues. Leukemia manifests as bone marrow failure and by infiltration into extramedullary sites.

Classification

Leukemia is a group of diseases. The various types of leukemia are classified according to whether the disease is acute or chronic and according to the morphology of the leukemic blasts. Acute leukemia is characterized by the rapid onset of bone marrow failure. Untreated, the disease is fatal within a few weeks to several months from the onset of symptoms. Most childhood leukemia is acute. Chronic leukemia, in contrast, evolves over months to years; initially, peripheral blood granulocytes of all stages of maturation increase, but ultimately, blasts predominate (“blast crisis”) and marrow failure ensues.

Morphologic classification of leukemia defines two discrete types according to the appearance of the blasts: 1) lymphoblastic (lymphoid, lymphocytic, lymphatic) and 2) myelogenous (myeloid, myelocytic, myeloblastic, acute nonlymphoblastic). The distinction is based on the superficial resemblance of the lymphoid blasts to immature or transformed lymphocytes and of the myeloid blasts to normal myeloblastic, promyelocytic, or monocytic precursors. These two types of leukemia are further classified by immunologic surface markers and chromosomal abnormalities. A small percentage of leukemic blasts have features of both types. Unlike normal myeloid and lymphoid precursors, the leukemic blasts do not have the ability to mature and differentiate *in vivo,* do not obey the signals that regulate proliferation of normal cells, and make no contribution to the body's immune system.

The distinction between acute myelogenous and lymphoblastic leukemia is important: childhood acute lymphocytic leukemia (ALL) responds to different therapy and has a substantially better prognosis than acute myelogenous leukemia (AML). About 75% of childhood leukemia is lymphoblastic.

Epidemiology

Leukemia is the most common childhood malignancy in the United States. The yearly incidence is about 4 cases per 100,000 children. The peak incidence is between 3 and 6 years.

Physical agents (e.g., ionizing radiation) and chemical agents (e.g., benzene) can cause leukemia in humans, and certain DNA viruses and RNA retroviruses are associated with leukemia in laboratory animals, but the cause of most childhood leukemia is unknown. Certain groups are at especially high risk of developing leukemia: 1) an identical twin of an infant with leukemia; 2) children with Down's syndrome, Bloom's syndrome, ataxia telangiectasia, or neurofibromatosis type 1; or 3) rarely, children who have previously received radiation and chemotherapy for another tumor. When

discussing the possibility of a child having leukemia, the emergency physician should reassure the parents that the cause is unknown in most cases, that the parents could not have prevented its occurrence, and that the disease is treatable.

Clinical Manifestations and Pathophysiology

The symptoms of acute leukemia are the symptoms of marrow failure and tissue infiltration. Marrow failure leads to anemia, thrombocytopenia, and leukopenia. Anemia causes pallor, fatigue, and irritability. Thrombocytopenia causes spontaneous bruising, epistaxis, and oozing from gums. Patients with myelogenous leukemia, especially hypergranular promyelocytic leukemia, are prone to CNS hemorrhage.

Infiltration of the marrow with leukemic cells can cause bone pain. Joint pain and dactylitis also occur, but their cause is obscure. Some children have a limp because of bone or joint pain. Leukemic infiltration of the reticuloendothelial system can cause adenopathy, hepatosplenomegaly, and mediastinal mass, especially in T-cell leukemia. In myelogenous leukemia, gingival hypertrophy is seen, and in monocytic leukemia, subcutaneous and soft tissue collections of leukemic blasts, called chloromas, are not uncommon. Leukemic cells invading the CNS can be clinically silent or can produce a meningitic syndrome or spinal cord compression. Extreme leukocytosis can also cause CNS symptoms. A high WBC count, CNS disease, and the presence of a mediastinal mass are unfavorable prognostic signs.

Table 100.2 shows the clinical and laboratory presentation of 1637 children with ALL. Although most patients have some hematologic abnormalities, many do not show such profound leukocytosis that a diagnosis of leukemia is certain from the peripheral count. Strikingly abnormal blood counts can also be seen in aplastic anemia, infectious mononucleosis, cytomegalovirus (CMV) infection, pertussis or parapertussis infection, chronic myeloid leukemia, and leukemoid reactions to bacterial or viral infections. Isolated thrombocytopenia is unusual in leukemia and is usually indicative of immune thrombocytopenic purpura (ITP). Anemia, reticulocytopenia, and neutropenia may be seen in transient erythroblastopenia of childhood (TEC). However, the neutropenia is usually mild in TEC and the WBC morphology is normal (see Chapter 87 for a discussion of ITP and TEC).

Clinical		Laboratory	
Characteristic	Percentage	Characteristic	Percentage
Age (yr)		Hemoglobin (g/100 mL)	
< 2	9	< 7	44
2–6	61	7–10	36
7–10	15	> 10	20
> 10	15	WBC × 10^3/mm^3	
Sex (male)	58	< 5	30
Race (white)	85	5–10	24
Adenopathy	61	10–20	15
Hepatomegaly	65	20–50	14
Splenomegaly	57	50–200	12
		> 200	5
		Platelets × 10^3/mm^3	
		< 50	50
		50–150	32
		> 150	18
		Mediastinal mass	5

Modified with permission from Hammond D, et al. Publication of Children's Cancer Study Group (USC Comprehensive Cancer Center, Los Angeles), 1980.
WBC, white blood cell.

Table 100.2. Acute Lymphocytic Leukemia: Presentation of 1637 Children from 1972 to 1977, Children's Cancer Study Group

Although an experienced hematologist can usually differentiate blasts from atypical mononuclear cells on the peripheral smear, the diagnosis of leukemia is made by marrow aspiration or biopsy. The leukemic marrow is hypercellular and contains a monotonous population of blasts. Normal precursors are rare or absent. Performing the marrow examination in an ED is not advisable because diagnosis and current management of acute leukemia require karyotyping, immunophenotyping of surface markers, and special histochemical stains; these studies need to be arranged by the physician who will assume responsibility for the management of the patient.

Management

In many instances, confirmation of the diagnosis of leukemia is not made in the ED. However, when leukemia is the probable diagnosis or the only tenable diagnosis (e.g., WBC count of 100,000/mm ^{3} with a predominance of blasts), the emergency physician or pediatrician should discuss the possibility that a child's disease is leukemia. This practice is accepted provided the physician has laboratory evidence to support the diagnosis, can take time to explain the reasons for concern, and can reassure the family that leukemia is no longer an untreatable disease. After the physician has begun appropriate supportive care, he or she should arrange admission to the hospital or referral of the child to a pediatric oncologist. Referral to a pediatric oncologist is important because survival is generally better when a child is treated at a pediatric cancer center or is managed by a specialist according to a current protocol.

Specific treatment of leukemia with chemotherapy should not be undertaken in the ED. Immediate antileukemic therapy in a child with a large tumor burden can precipitate massive tumor lysis, hyperkalemia, hypocalcemia, renal failure, and possibly death (see the following section on metabolic complications). Definitive therapy requires medical and emotional support and use of chemotherapeutic protocols designed for specific subsets of leukemia. The goals of therapy are to achieve a marrow “remission” (i.e., a normal bone marrow within 1 or 2 months of intensive “induction” therapy) and then to maintain the remission. Induction is followed by a period of prevention or treatment of occult CNS disease and then maintenance therapy. Treatment is changed if the disease recurs in the marrow or appears in the CNS or testes. Treatment is stopped after months or years of continuous remission.

Table 100.3 outlines the initial care of patients with acute leukemia. Fortunately, most patients require only general supportive care. However, some patients require immediate intervention for specific life-threatening problems. All

patients and their families require emotional support and sensitivity to deal with the fear and disorientation they feel when confronted with the diagnosis of cancer.

Table 100.3. Emergency Department Care of the Patient with Probable or Certain Acute Leukemia

Hematologic Complications

The emergency physician should take special care to inform the blood bank that a patient may have acute leukemia. Current blood bank practices for the patient with acute leukemia include 1) initial complete red blood cell (RBC) antigen typing to facilitate future crossmatches if the patient develops anti-RBC antibodies; 2) the use of CMV-negative products for potential bone marrow transplant patients, all patients with myeloid leukemia, and CMV-seronegative patients with lymphoblastic leukemia; 3) the use of in-line leukocyte-depletion filters in many types of acute leukemia; and 4) irradiation to 1500 cGy or greater of all blood products used in oncology patients. Some or all of these practices are followed in major pediatric oncology centers, but transfusion of an acutely ill patient should not be delayed to follow these recommendations.

Anemia. At the time of diagnosis, most children with leukemia are anemic. If the hemoglobin is less than 8 g/dL, administration of packed blood cells is advisable (Table 100.3) because the child is unlikely to have the ability to produce erythrocytes for several weeks; however, if the child is not having symptoms or showing signs of severe anemia, transfusion does not need to take place in the ED. If, in the absence of hemorrhage, the child has profound anemia (i.e., hemoglobin, 1 to 4 g/dL), transfusions at the usual rate can precipitate heart failure. Blood should be replaced slowly, at 3 to 5 mL/kg over 4 hours, and supplemental oxygen should be given to enhance oxygen delivery to tissues. Furosemide (1 mg/kg) can help avoid fluid overload or heart failure; however, if the WBC count is greater than 100,000/mm ^{3}, diuretics should be withheld because intravascular dehydration encourages sludging and thrombosis. Exchange transfusion has been advocated by some physicians in these exceptional circumstances. When hemorrhage is the cause of a low hemoglobin, transfusion therapy should be carried out quickly to replace losses.

Hemorrhage. Hemorrhage is the second most common cause of death in leukemia. Spontaneous bleeding can occur at platelet counts less than 10,000 to 20,000/mm^3, although spontaneous bruising is seen at higher levels in some patients. Other factors contributing to a bleeding tendency include 1) infection with associated disseminated intravascular coagulation (DIC); 2) consumptive coagulopathy in acute promyelocytic leukemia, monoblastic leukemia, and T-cell ALL; 3) antibiotic therapy, which causes hypoprothrombinemia; and 4) aspirin, which interferes with platelet function.

In most newly diagnosed leukemic children, bleeding problems can be controlled with local measures alone (i.e., pressure and topical thrombin for epistaxis), or in conjunction with platelet transfusions (0.2 unit/kg platelets). Epistaxis is sometimes a serious problem that may last for hours and may fail to respond to pressure. If local measures (see pp. 1177–1178 and Chapter 23) and platelets fail, packing is a necessary but uncomfortable therapy. Sedation and analgesia may be helpful in controlling the anxiety associated with severe bleeding.

In some patients with AML, especially those with hypergranular promyelocytic leukemia and some with monoblastic leukemia, a bleeding diathesis may occur at presentation or upon initiation of therapy. Bleeding is generally refractory to platelet transfusion. Patients show prolonged prothrombin and partial thromboplastin times, elevated fibrin split products, and drastically shortened fibrinogen half-life. The most common form of bleeding in this situation is in the CNS; it may be fatal in the first few days of illness. Fresh-frozen plasma (10 mL/kg) and cryoprecipitate can help maintain levels of fibrinogen and clotting factors. Platelet transfusions (0.2 unit/kg) are given to correct thrombocytopenia. Although no controlled study has been conducted to demonstrate the benefit of prophylactic heparin, heparinization (loading dose, 50 units/kg; then 5 to 10 units/kg per hour to maintain the partial thromboplastin time at approximately 1.5 times normal) is often given to patients who show no improvement with aggressive blood product support.

Extreme Leukocytosis. Extreme leukocytosis with a WBC count of greater than 200,000/mm^3 occurs at diagnosis in about 5% of patients with acute leukemia and in those with chronic myelogenous leukemia (CML). In acute leukemia, extreme leukocytosis predisposes patients to early bleeding and thrombosis in the CNS. It has been suggested that patients with WBC counts greater than 200,000/mm^3 should receive prompt cranial irradiation or leukocytopheresis, but these therapies are unproven. Patients with CML may also have the problem of sludging and thrombosis seen in AML when they have a predominance of blasts or when their WBC counts are greater than 200,000/mm ^{3}. Hydration and alkalinization often result in a substantially lower WBC count (Table 100.3).

Leukopenia. *Leukopenia* is defined as a WBC count of less than 3000/mm^3 and *neutropenia,* in children, as less than 500 neutrophils/mm^3. Leukopenia per se is not life-threatening, but it can predispose patients to life-threatening

infections (see the following).

Infectious Complications

Infection remains the leading cause of death in acute leukemia. The patient with acute leukemia has quantitative and qualitative cellular immune dysfunction. Leukopenia and neutropenia are common at diagnosis. Even if the absolute neutrophil count is more than 500/mm^3, as may be the case in a patient with a WBC count of 50,000/mm^3 and 1% neutrophils, defective chemoattraction and killing have been documented in patients with acute leukemia. Therefore, any patient with newly diagnosed leukemia, especially those with AML, should be considered at increased risk for infection. In the ED, handwashing by staff, strict asepsis for drawing blood and performing other procedures, and protection of the patient from airborne infections (e.g., measles and varicella) must be enforced.

Although fever is a symptom of leukemia in 25% of patients, more often it indicates infection. In the leukemic patient, significant fever is defined by three temperature elevations of 38.0°C (100.4°F) or higher over 4 hours or a single temperature higher than 38.4°C (101.1°F). There are exceptions to the association between fever and infection. A leukemic patient in septic shock may have no fever or may be hypothermic.

Management of fever begins with a thorough physical examination to search for localizing signs of infection. Even an apparently minor swelling or a tear in the skin or a mucosal surface can be a source of disseminated infection. Blood and urine cultures should be obtained. All leukemic patients should have baseline chest radiograph films taken. Thereafter, chest radiograph is needed only if the child has respiratory symptoms or signs. Bacterial meningitis is rare in leukemia, but if symptoms of meningitis are found, spinal fluid should be sent for bacterial, fungal, and viral culture and for cytology to rule out CNS leukemia.

After the appropriate cultures are obtained, intravenous broad-spectrum antibiotics should be started in a patient with leukemia and fever. The choice of antibiotic combinations is reviewed in Figure 100.7. Most patients do not have invasive fungal disease. However, oral candidiasis sometimes occurs in a patient who has been treated with antibiotics before the diagnosis of leukemia. Thrush should be treated with oral Mycostatin, fluconazole, or clotrimazole troches.

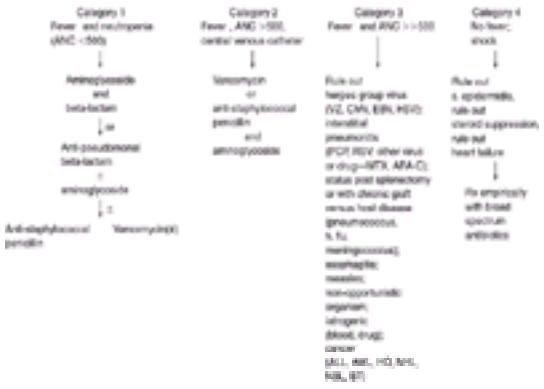

FIGURE 100.7. Initial management of the child with cancer and presumed infection. *ANC,* absolute neutrophil count; *VZ,* varicella-zoster; *CMV,* cytomegalovirus; *EBV,* Epstein-Barr virus; *HSV,* herpes simplex virus; *PCP, Pneumocystis carinii* pneumonia; *RSV,* respiratory syncytial virus; *MTX,* methotrexate;*ARA-C,* cytarabine; *ALL,* acute lymphoblastic leukemia; *AML,* acute myelogenous leukemia; *HD,* Hodgkin's disease; *NHL,* non-Hodgkin's lymphoma;*NBL,* neuroblastoma; *BT,* brain tumor. [a]Fever: temperature 38°C × 3 or >38.4°C × 1. [b]Use of vancomycin or anti-staphylococcal penicillin varies according to prevalence and resistance patterns of institutional Gram-positive bacteria. NB: Aminoglycoside alone is not adequate Gram-negative coverage in the neutropenic patient.

Infection can cause septic shock in a patient with leukemia. Treatment consists of aggressive measures to maintain intravascular volume and blood pressure (see Chapter 3) and broad-spectrum antibiotics. Because the patient with leukemia is commonly anemic at diagnosis, bolus infusions of packed RBCs can be useful when treating shock.

Metabolic Complications

Uric Acid Nephropathy. Some patients with acute leukemia have an elevated serum uric acid caused by spontaneous breakdown of leukemic cells. The excess uric acid precipitates in the renal tubules. Antileukemic therapy accelerates breakdown of leukemic cells; more urates are formed and renal failure can occur. To prevent urate nephropathy, all children with leukemia should receive the xanthine oxidase inhibitor allopurinol (150 mg/d orally in three daily doses for those 6 years old or younger; 300 mg/d orally in three daily doses for those older than 6 years) for at least 24 hours before starting therapy. Hydration at the rate of twice maintenance and alkalinization of urine to pH between 6.5 and 7.5 with sodium bicarbonate (40 mEq/L) facilitates dissolution of uric acid crystals. Once the serum uric acid level is normal and the child is adequately hydrated and producing a dilute urine, specific antileukemic therapy may begin. Bicarbonate is stopped at this time to avoid development of hypoxanthine nephropathy.

Hypercalcemia. Hypercalcemia occurs in about 2% of patients with acute leukemia. Hypercalcemia is caused by destruction of bone by malignant cells, by ectopic production of parathormone by the leukemic cells themselves, or by elevated levels of peripheral plasma prostaglandin E_2, vitamin D–like substances, or osteoclast-activating factors. The serum calcium can reach levels high enough to cause anorexia, nausea, vomiting, constipation, lethargy, confusion, coma, tachycardia or bradycardia, and renal failure. The ultimate therapy of leukemic hypercalcemia is treatment of the leukemia. Interim supportive management consists of hydration with normal saline (200 mL/m^2 per hour), followed by diuresis with furosemide (1 to 2 mg/kg intravenously every 4 to 6 hours). Monitoring of the cardiovascular status is

essential. For indications for corticosteroids, calcitonin, gallium nitrate, or dialysis in symptomatic hypercalcemia (usually greater than 12 to 15 mg/dL), see Chapter 86. Phosphorus has no role in the emergency treatment of the patient who has leukemia and an acute elevation of serum calcium. Malignant hypercalcemia may require management in an intensive care setting.

Tumor Lysis Syndrome. In a patient with a large tumor burden, especially patients with leukemia and massive organomegaly or with high WBC counts, or in patients with non-Hodgkin's lymphoma, antineoplastic therapy can cause a potentially fatal tumor lysis syndrome that consists of a rapid rise in serum potassium and phosphorus, a precipitous fall in serum calcium, and elevations of the serum uric acid, blood urea nitrogen (BUN), and creatinine. These abnormalities may occur despite appropriate hydration and allopurinol therapy. Hyperkalemia, the most dangerous abnormality, demands prompt treatment with Kayexalate, insulin and glucose infusion, or dialysis (see Chapter 86).

Other Complications

Spinal Cord Compression. Spinal cord compression occurs in leukemia because of epidural or subarachnoid collections of leukemic cells. Symptoms include radicular pain, back pain, difficulty with urination, paresis, and paralysis. Physical examination can be unremarkable or may show percussion tenderness over spinous processes, weakness, hyperactive (or later, absent) deep tendon reflexes, absent superficial reflexes, inability to walk on the toes or heels, and a sensory level. Radiographs may show a collapsed vertebral body, but they are often normal. Magnetic resonance imaging (MRI) with and without gadolinium confirms the diagnosis; therapy consists of immediate corticosteroid administration (dexamethasone 0.25 to 0.5 mg/kg every 6 hours), prompt irradiation, or both. Leukemia and lymphoma of the spinal cord respond to steroids or radiation therapy and do not require laminectomy.

Central Nervous System Leukemia. Leukemia can present or relapse in the CNS as diffuse subarachnoid disease or as localized deposits of cells. Symptoms include headache, stiff neck, malaise, cranial nerve palsy, and rarely, fever. Diagnosis is made by finding greater than 5 leukemic blasts/mm^3 in the spinal fluid. Spinal fluid should be examined by cytospin preparation. Treatment consists of intrathecal chemotherapy and craniospinal irradiation, as well as reinstitution of systemic chemotherapy in the case of CNS relapse.

Testicular Leukemia. Clinically apparent testicular disease is rarely present at diagnosis, but the first site of relapse may be the testes. A painless, hard swelling is seen in one or both testes. Treatment consists of reinduction therapy and irradiation. Testicular relapse may be a harbinger of marrow relapse.

Non-Hodgkin's Lymphomas

Background

Definition

Non-Hodgkin's lymphomas (NHL) are malignant tumors that originate in lymphatic tissues: lymph nodes, Waldeyer's ring, the appendix, mesentery, Peyer's patches, or rarely, the spleen. Sometimes NHL arises in extralymphatic sites such as bone, ovaries, or the nervous system. In contrast to NHL in adults, which often remains indolent and localized for years, NHL in children has a tendency to proliferate rapidly and spread outside the primary lymphatic area in weeks or months. Often, it metastasizes to bone marrow, the CNS, or both. When extensive marrow replacement occurs, the distinction between NHL and acute leukemia is difficult.

Classification

No disease classification is more controversial than that of NHL. The NHLs are classified according to histology, cytology, and immunology. The types of lymphoma seen in children are almost always poorly differentiated and diffuse (as opposed to the nodular NHL common in adults), and they efface entirely the architecture of the tissue of origin. In children, the malignant cells are often B lymphoblasts or T lymphoblasts (lymphoblastic NHL), undifferentiated stem cells, Burkitt-type B-cells, or histiocytelike cells (nonlymphoblastic NHL). NHLs are also classified according to stage. Stage reflects the tumor burden and extent of spread. A small tumor burden that is confined to the tissue or region of origin usually confers an excellent prognosis. However, most pediatric NHL is widely disseminated.

Epidemiology

NHL usually occurs before puberty or in early adolescence. The incidence in childhood is 0.5 per 100,000. There is a male predominance of 1.4:1. Patients with immunodeficiency diseases have an exceptionally high incidence of NHL. The inherited immunodeficiency states associated with NHL include severe combined immunodeficiency, variable immunodeficiency, X-linked hypogammaglobulinemia, IgA deficiency, IgM deficiency, X-linked lymphoproliferative syndrome (Duncan's disease), ataxia telangiectasia, and Wiskott-Aldrich syndrome. In addition, NHL occurs with increased frequency in patients with acquired disorders of immunity, including rheumatoid arthritis, systemic lupus erythematosus, sarcoidosis, celiac disease, and acquired immune deficiency syndrome (AIDS). NHL develops with increased frequency in renal and cardiac transplant patients, in patients receiving long-term immunosuppressive therapy, and in patients with Hodgkin's disease treated with radiation and chemotherapy.

Clinical Manifestations and Pathophysiology

Localized NHL

About 25% of childhood NHL presents as localized disease. Presentations include swelling in the cervical or inguinal nodes, nasopharyngeal masses, mediastinal masses, and bone pain and swelling. Primary disease in the abdomen may

be confused with acute appendicitis or intussusception. Gastrointestinal (GI) hemorrhage can result from tumor infiltration of the bowel and may be the presenting symptom.

B-Lymphocyte NHL (American Burkitt's Lymphoma)

American Burkitt's lymphoma usually causes generalized abdominal enlargement, pain, nausea, vomiting, constipation, and in about half of patients, malignant ascites. In these circumstances, the primary tumor is either in the ovaries or retroperitoneal nodes, but kidneys and sometimes bowel are involved. Jaw tumors occur in only 20% of American cases and cervical node primary tumors in about 15%.

Burkitt tumor cells proliferate exceedingly rapidly. Tumors grow quickly and tend to extend locally over several days to weeks, causing ascites, pleural effusion, and renal failure. They often metastasize to the marrow and meninges. Untreated, American Burkitt's lymphoma is rapidly fatal, usually because of renal failure.

Mediastinal NHL

Mediastinal NHL occurs most often in males in late childhood or early adolescence and, untreated, metastasizes to marrow, nervous system, and testes. The symptoms and signs of mediastinal NHL may be those of leukemia or other thoracic tumors. Mediastinal NHL sometimes causes life-threatening superior vena cava (SVC) syndrome or tracheal compression (see the following section about thoracic tumors).

Management

Localized NHL

When localized NHL is suspected, the node or mass should be biopsied promptly and ideally at an institution that has the facilities to provide phenotyping and the necessary genetic studies on the malignant cells. Open biopsy is mandatory to evaluate node architecture. Before biopsy, it is advisable to obtain a chest radiograph and blood count to ensure that no large mediastinal mass or unsuspected leukemic process is present. When localized NHL is treated with surgery and chemotherapy, with or without irradiation, the prognosis is excellent. Complete surgical resection should not be attempted when extensive disease is present.

Burkitt's Lymphoma with Extensive Abdominal Involvement

When the primary tumor is abdominal, the disease usually constitutes a medical emergency because the tumor grows day by day and rapidly encroaches on vital organs, especially the kidneys. Spontaneous tumor lysis causes metabolic abnormalities, discussed in the previous section about tumor lysis syndrome.

The role of the emergency physician who suspects intra-abdominal Burkitt's lymphoma is to ascertain the hematologic and metabolic status of the child with the following studies: CBC, BUN, creatinine, uric acid, electrolytes, calcium, phosphorus, and liver function tests. Hydration (3000 mL/m^2 per day of D5W/0.25 NS), alkalinization, and allopurinol (150 mg/d orally in three daily doses for those 6 years old or younger; 300 mg/d orally in three daily doses for those older than 6 years) should be started immediately. If renal failure is present, determining whether it is from uric acid nephropathy, tumor infiltration, or obstructive uropathy is necessary. An ultrasound examination can detect infiltration or compression by tumor. Catheterization may be necessary to establish whether the kidneys are forming urine and to rule out bladder obstruction. Treatment of renal failure and secondary hypertension varies with the cause (see Chapter 88). Antineoplastic therapy should be directed by a pediatric oncologist.

Mediastinal Lymphoma

When the chest radiograph reveals a mediastinal mass and no obvious congenital abnormality or infectious cause is apparent, cancer must be ruled out. Sometimes it is possible to diagnose NHL by the combination of an abnormal blood count or bone marrow examination and a mediastinal mass. Other times, biopsy of a peripheral node or the mass itself is necessary. Often, open biopsy is a straightforward procedure, but it can be life-threatening. For a discussion of mediastinal masses, see the following section about thoracic tumors. Mediastinal NHL is responsive to combinations of chemotherapy and irradiation and most patients are now long-term survivors.

Hodgkin's Disease

Background

Definition

Like NHL, Hodgkin's disease is a malignancy of the lymphatic tissues. It usually begins in a cervical node and spreads in a predictable and orderly sequence from one lymph node region to the next; untreated, it progresses to involve organs outside the lymph nodes. In contrast to NHL, spread to the marrow or CNS is rare. The malignant cell in Hodgkin's disease is the Reed-Sternberg cell, which is believed to originate from either a B lymphocyte or a histiocyte precursor.

Classification

Hodgkin's disease is classified according to the extent of disease at presentation, the stage, and the histology of the involved lymph node. Unlike most other malignancies, Hodgkin's disease is multicellular (i.e., more than one cell type is present, and the interaction of these cells comprises the malignant process). The accepted histologic classification is a modification of the Rye, New York, classification based on the number of lymphocytes, the number of “reactive” cells, the

number of Reed-Sternberg cells, and the amount of fibrosis. The four histologic types are 1) lymphocyte predominant, 2) nodular sclerosing, 3) mixed cellularity, and 4) lymphocytic depletion. Two-thirds of pediatric patients have the nodular sclerosing type.

Epidemiology

The incidence of Hodgkin's disease is similar to that of NHL, 0.6 per 100,000 per year. Although Hodgkin's disease has been reported in infants, it is rare in children less than 5 years of age; there is one peak in adolescence and a later peak in the sixth decade. In the United States, Hodgkin's disease tends to occur more often in the higher socioeconomic classes. The cause is unknown.

Clinical Presentation and Pathophysiology

Hodgkin's disease most often presents as a mass in the neck; about 4% of patients have masses in the groin. The mass may have been present for days, months, or years. Some patients complain of adenopathy localized to a few lymph node regions; others note generalized adenopathy. Anterior mediastinal masses occur in 50% of patients. Hodgkin's disease rarely occurs in Peyer's patches, Waldeyer's ring, or epitrochlear nodes. Large mediastinal Hodgkin's tumors can cause the same cardiovascular or respiratory symptoms as mediastinal NHL (see the following section about thoracic tumors). Most Hodgkin's disease patients are well, but 30% have fever, involuntary weight loss, or both. At diagnosis, the fever may be low or high grade. The Pel-Epstein fever, a high, debilitating fever followed by a drenching sweat, is usually a sign of advanced disease. Incapacitating pruritus, seen in about 15% of adults, is uncommon in children.

Patients with Hodgkin's disease manifest defects of cellular immunity early in the course of their disease. B-lymphocyte function remains relatively intact, but almost all patients have impaired T cell–mediated immunity that results in increased susceptibility to fungal, viral, and certain bacterial infections. Complete anergy to recall antigens does not usually occur until the disease is advanced.

Hodgkin's disease is classified by stage according to the extent of disease and the presence of symptoms: stage I, one lymph node region; stage II, two lymph node regions on the same side of the diaphragm; stage III, lymph node involvement (including spleen) on both sides of the diaphragm; stage IV, lymph node disease plus lung, liver, marrow, or bone involvement. “A” patients have no symptoms; “B” patients have fever and weight loss. The clinical stage is that which is determined by physical examination and radiograph studies, including lymphangiogram, retroperitoneal computed tomography (CT) scan, or both. The pathologic stage refers to the stage as determined by laparotomy with splenectomy and surgical sampling of abdominal lymph nodes, liver, and bone marrow.

Management

The first step in treating Hodgkin's disease is to obtain a tissue biopsy using the same guidelines and precautions as for NHL. The second step is to determine the stage of the disease by either clinical or pathologic staging. In the past, most patients with Hodgkin's disease underwent pathologic staging, including splenectomy. Those with stages IA to IIIA were treated with large fields of irradiation, and those with more advanced disease were treated with chemotherapy combinations (MOPP: nitrogen mustard, vincristine (oncavin), prednisone, and procarbazine; AVBD: adriamycin, vinblastine, bleomycin, and 5-(3, 3-dimethyltriazeno)imidazole-4-carboxamide [DTIC]; or other combinations). With the advance of these therapies, most Hodgkin's disease patients can be cured. However, splenectomy carries a high risk of hyperacute pneumococcal infection in children with Hodgkin's disease. Furthermore, irradiation causes major growth disturbances in children less than 18 years old and some risk of secondary sarcomas 10 to 20 years after treatment. On the other hand, chemotherapy and large-field irradiation predispose to the development of leukemia and NHL in about 5% of patients. Awareness of these complications is leading many pediatric oncologists to investigate staging without laparotomy and to use different combinations of chemotherapy and lower dosages and smaller fields of irradiation.

The issues regarding biopsy and SVC syndrome in patients with Hodgkin's disease are presented in the following section on thoracic tumors. Infections and other special problems seen during therapy for Hodgkin's disease are discussed in the following section about complications of therapy and progressive disease.

Histiocytic Diseases

Background

Histiocytic diseases include a heterogeneous group of benign and malignant disorders. With some of these diseases, determining whether the histiocytes are benign or malignant is difficult because they may appear histologically benign and yet metastasize; conversely, they can appear histologically malignant and behave with relative benignity.

Clinical Manifestations and Pathophysiology

Histiocytic diseases are best classified according to their disease functions into lipid storage diseases, reactive diseases, and neoplastic diseases. Among the reactive disorders are those diseases formerly called histiocytosis X and hemophagocytic lymphohistiocytosis. Some reactive disorders tend to behave in a malignant fashion. Among the neoplastic group, acute monocytic leukemia and malignant histiocytosis are seen in children and adolescents. Three of the disorders, which behave in a malignant fashion regardless of cause or disease function, are discussed: 1) the spectrum of eosinophilic granulomas, 2) familial and nonfamilial hemophagocytic lymphohistiocytosis, and 3) malignant histiocytosis. Monocytic leukemia is considered a form of AML (see previous section about leukemia).

Eosinophilic Granuloma Syndromes

A solitary eosinophilic granuloma presents as a painless or mildly painful swelling in the skull, long bones, ribs, pelvis, or vertebra. Radiograph films show a lytic lesion with well-defined borders. Multiple eosinophilic granulomas can arise over weeks, months, or years. They can be confined to the long bones or may be distributed in the skull in a manner that causes proptosis, diabetes insipidus, cholesteatomas, and loss of teeth (Hand-Schüller-Christian disease). Rare cases of paraplegia caused by vertebral lesions have been described.

Widespread eosinophilic granulomas of bone and soft tissue in an infant constitute Letterer-Siwe disease. Symptoms and signs include failure to thrive, diarrhea, chronic seborrhea, chronic otitis media and otorrhea, purpuric rash, generalized adenopathy, and hepatosplenomegaly. The chest radiograph may show interstitial pneumonitis. The blood count may be normal, but marrow involvement or massive splenomegaly can lead to pancytopenia.

Familial and Nonfamilial Erythrophagocytic (Hemophagocytic) Lymphohistiocytosis

Familial erythrophagocytic lymphohistiocytosis is an autosomal-recessive systemic disease presenting in early infancy with fever, vomiting, anorexia, irritability, and occasionally (25%) with seizures, cranial nerve abnormalities, and aseptic meningitis. Nonfamilial forms have been reported, and some cases have followed infection with Epstein-Barr virus (EBV) or similar viruses. Generalized adenopathy and hepatosplenomegaly are common, and a purpuric rash sometimes occurs. Laboratory abnormalities include hypofibrinogenemia, hypertriglyceridemia, elevated liver enzymes, and hypogammaglobulinemia or hypergammaglobulinemia. Biopsy of involved tissues or bone marrow aspiration in advanced disease shows diffuse infiltration by morphologically benign histiocytes and prominent erythrophagocytosis. Thymic and lymphoid tissue is depleted.

Malignant Histiocytosis (Histiocytic Medullary Reticulosis)

Malignant histiocytosis presents with fever, adenopathy, hepatosplenomegaly, pancytopenia, or leukocytosis. The mean age at presentation is 31 years but the age range is from 1 to 71 years. Biopsy of involved tissues reveals infiltration with malignant histiocytes of all stages of maturation. Infiltrates are prominent in the medullary zone of lymph nodes.

Management

The role of the emergency physician in the management of the histiocytic disorders is to suspect them, recognize their presentations, refer those patients who require chemotherapy or irradiation, and handle the complications of the diseases and therapy. Eosinophilic granulomas can cause diabetes insipidus (see Chapter 86 and Chapter 97), pathologic fracture, or rarely, paraplegia. Letterer-Siwe disease can be suspected from the clinical findings but must be differentiated from severe combined immunodeficiency. The complications of pancytopenia are the same as in leukemia. Massive organomegaly can cause respiratory embarrassment. Irradiation and chemotherapy usually restore adequate ventilation.

Specific therapy depends on the extent of the disease. Isolated eosinophilic granulomas are cured with surgical curettage. If the lesion is inaccessible and in a critical area (e.g., pituitary fossa), low-dose irradiation can be used. The disease often arrests spontaneously with prolonged observation. Disseminated histiocytosis requires multiagent chemotherapy under the direction of a pediatric oncologist.

Familial and nonfamilial erythrophagocytic lymphohistiocytosis and malignant histiocytosis are usually fatal, but remissions have been achieved using multiagent chemotherapy and bone marrow transplantation. Plasmapheresis and immunotherapy have been used to achieve remission in several patients with erythrophagocytic lymphohistiocytosis. The major complications are marrow failure that predispose the patient to infection and hemorrhage and respiratory failure as a result of massive hepatosplenomegaly and ascites.

Thoracic Tumors

Anterior Mediastinum

Hodgkin's disease and NHL are the most common malignant tumors of the anterior mediastinum. Other tumors include thymoma, thymic cyst, thymic hyperplasia, teratoma, ectopic thyroid, thyroid carcinoma, sarcomas, neuroblastoma (rarely), lymphangiomas, and inflammatory processes such as sarcoid. Half of anterior mediastinal masses are benign. However, almost all large tumors and masses that cause compromise to the great vessels or cause pleural effusions are malignant.

Large tumors in the anterior mediastinum can cause tracheal narrowing, SVC syndrome, or both. Tracheal compression causes tracheal deviation, stridor, cough, dyspnea, and orthopnea. Compression of the vena cava by tumor may cause headache, dyspnea, orthopnea, syncope, or cardiovascular collapse. Physical examination may be unremarkable or may show venous distension, plethora, cyanosis, and edema anatomically confined to the head, neck, thorax, and arms. When the arm is involved, the veins will remain full when the arm is raised. Conjunctival and retinal vessels may be engorged. Children with SVC syndrome are often anxious and diaphoretic and will resist efforts to place them in a supine position. When these findings are present, narcotics, sedatives, and any drugs that interfere with venous return are contraindicated. Chest radiograph reveals a mass in the anterior mediastinum (Fig. 100.1). If a tumor is ausing SVC syndrome in a child or adolescent, the diagnosis is almost certain to be NHL or Hodgkin's disease.

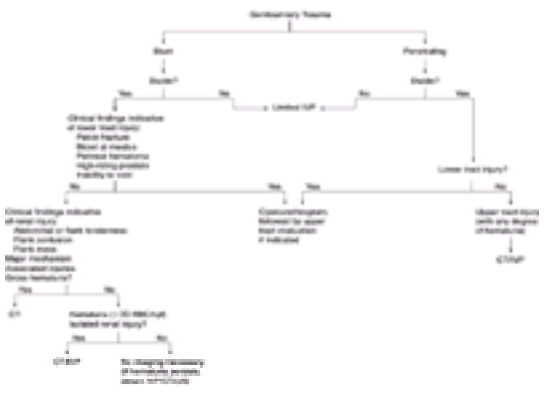

FIGURE 100.1. Management guidelines for children with an anterior mediastinal mass who are at risk for superior vena cava syndrome. (Modified with permission from Kelly KM, Lange B. Oncologic emergencies. Pediatr Clin North Am 1997;44:809–831.)

SVC syndrome is life-threatening. It may be ill-advised to try to obtain a histologic diagnosis under general anesthesia because these patients tolerate procedures poorly. Even placing the patient in the CT scanner can cause cardiorespiratory decompensation. If both arms are involved, intravenous infusions should be started in the feet, provided that venous return is good in the lower extremities. Infusions in an affected arm can cause respiratory distress, thrombosis, or phlebitis. In the unstable patient with SVC syndrome, empiric therapy with radiation (50 to 100 cGy to the midplane for 2 or 3 days), corticosteroids (hydrocortisone 2 mg/kg every 6 hours), or both may alleviate symptoms. Either treatment may make later interpretation of histology difficult; however, studies of tissue obtained a day or two after initiation of emergent treatment have documented that the initial clinical impression of the treating oncologist is usually correct. If the patient can tolerate procedures, effusions should be drained, a bone marrow aspirate should be taken as early as possible (to rule out NHL), and a biopsy should be performed on an accessible peripheral node under local anesthesia.

Middle Mediastinum

Most tumors of the middle mediastinum are benign. Malignant tumors include Hodgkin's disease of the hilar nodes or pericardium, NHL or metastatic tumors such as neuroblastomas (less commonly), and sarcomas (rarely). Many large anterior mediastinal tumors extend into the middle mediastinum. Rarely, these large tumors may cause cardiovascular compromise by compression of the vagus nerve, coronary vessels, or pericardial invasion. Spontaneous syncope or syncope with a Valsalva maneuver are clues to vagal involvement. Similar to the rare patients with SVC syndrome, patients with a history of syncope are poor surgical risks. They require hospitalization and supportive care.

Posterior Mediastinum

Most tumors in the posterior mediastinum are of neurogenic origin: neuroblastoma, neurofibrosarcoma, ganglioneuroblastoma, ganglioneuroma, and neurofibroma (Fig. 100.2). Only the first two tumor types have malignant potential. The more slowly growing tumors are often detected on chest films taken for other purposes. Larger, rapidly growing tumors can cause pain and neurologic deficits.

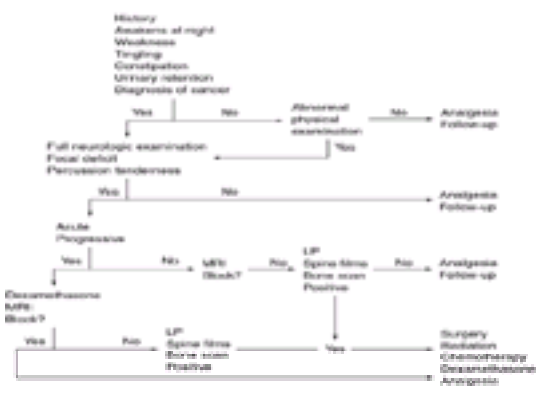

FIGURE 100.2. Approach to back pain in children at risk for spinal cord compression. (Modified with permission from Kelly KM, Lange B. Oncologic emergencies. Pediatr Clin North Am 1997;44:809–831.)

Pulmonary Parenchyma

Nodules (“coin” lesions or “cannonball” lesions) in a child with a solid tumor are usually metastases. In the everely compromised host, they may be fungal legions. Rarely, bacteria such as staphylococci may cause coin lesions; in this case, the patient is usually acutely ill.

Abdominal, Genitourinary, and Retroperitoneal Tumors

Intra-abdominal tumors manifest themselves as discrete masses or as generalized abdominal enlargement. Wilms' tumor and neuroblastoma are the most common intra-abdominal malignant tumors in young children. Wilms' tumor presents as a large flank mass in a well child, and abdominal neuroblastoma, following the newborn period or infancy, as a midline or

flank mass in an ill child. Hepatic tumors are sometimes associated with systemic illness. Pelvic sarcomas, teratomas, neuroblastomas, or lymphomas may first cause neurologic deficits or abdominal pain. (Pelvic sarcomas are discussed in the following section about soft-tissue sarcomas.)

For most of these tumors, staging usually depends on the amount of tumor that has been surgically removed. Stage I tumors are those that have been completely removed. If microscopic or gross residual tumor or spread to regional nodes occurs, the tumors are stage II or stage III. The most advanced stage (IV) is defined by the presence of distant metastases. Each tumor category also has a classification based on histology, which is prognostically important.

Wilms' Tumor

Background

Definition

Wilms' tumor (nephroblastoma), the most common intrarenal malignancy of childhood, is an embryonal neoplasm of mixed tissue structure. About 5% of Wilms' tumors are bilateral.

Classification

The accepted classification of Wilms' tumor is based on extent of disease after surgery and on tumor histology. The grouping system of The National Wilms' Tumor Study Group (NWTS), a multi-institutional cooperative, is as follows: a stage 1 tumor has been resected entirely, stage 2 has microscopic residual, stage 3 has gross residual, stage 4 has metastatic disease, and stage 5 has bilateral tumors. The most favorable histology has a high degree of differentiation of the epithelial elements. Histologically unfavorable tumors are sarcomatous, anaplastic, clear cell, or rhabdoid.

An intrarenal tumor that is clinically similar in presentation to Wilms' tumor but that is morphologically distinct is congenital mesoblastic nephroma. Mesoblastic nephroma occurs in children younger than 1 year of age, is composed primarily of mesenchymally derived cells, and lacks the malignant elements of Wilms' tumor. It is treated by nephrectomy alone.

Epidemiology

Wilms' tumor occurs in 0.78 per 100,000 children less than 15 years of age. In children less than 5 years old, it is the fourth most common malignancy; 65% of children presenting with Wilms' tumor are younger than 5 years of age.

A significant association exists between Wilms' tumor and congenital anomalies, including aniridia, hemihypertrophy, genitourinary anomalies, hemangiomas, hamartomas, cardiac anomalies, and Beckwith-Wiedemann syndrome. There are reports of familial cases of Wilms' tumor, as well as reports of families in which some of the members had the congenital anomalies previously listed and other members had Wilms' tumor.

Clinical Manifestations and Pathophysiology

Most patients with Wilms' tumor have an abdominal mass that is discovered accidentally by the parents while bathing or clothing a child or by a physician during a routine physical examination. The mass is deep in the flank and can be either firm or soft. Abdominal pain, fever, anorexia, malaise, vomiting, and weight loss are rare presenting complaints. Gross hematuria occurs in less than 25% of patients. Some institutions report hypertension in up to 15% of patients. Hypertension is thought to result from increased renin secretion secondary to compression of the renal artery. By a related mechanism, polycythemia occasionally occurs in patients with Wilms' tumor. More often, anemia results from bleeding into the tumor. When performing a physical examination, the physician should be particularly attentive to the size of the liver and spleen, the site and size of the mass, the patient's blood pressure, and the presence of the previously listed congenital anomalies. Laboratory evaluation should include CBC, BUN, and creatinine. Abdominal radiograph (kidney, ureter, bladder [KUB]) may reveal a mass displacing bowel. Intravenous pyelogram (IVP) demonstrates an intrarenal mass distorting the calyces. Ultrasound, when available, is the preferred imaging technique for initial localization of these tumors (Fig. 100.3).

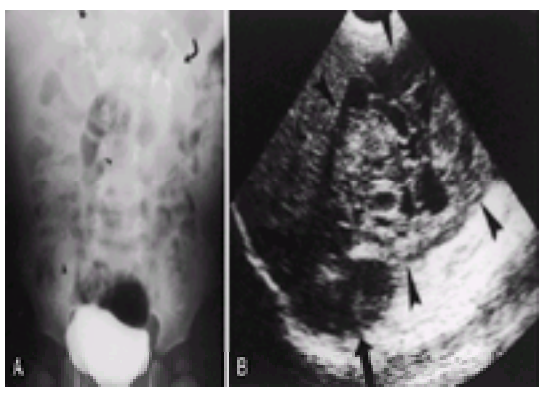

FIGURE 100.3. A. Wilms' tumor. Intravenous pyelogram demonstrates a normal left kidney *(curved arrow).* There is upward displacement and splaying of the right renal collecting system *(arrow)* by a large nonopacified mass *(arrowhead).* **B.** Wilms' tumor. Right parasagittal sonogram shows the liver *(L),* the right upper renal pole *(arrows),* and a large complex mass *(arrowheads)* arising from the midpole of the right kidney. The hypoechoic areas represented necrosis within the tumor. (Courtesy of Dr. Henrietta Kotlus Rosenberg, Director, Division of Ultrasound, Children's Hospital of

Philadelphia.)

Management

Children with a presumed Wilms' tumor should be referred to a surgeon experienced in the diagnosis and treatment of this tumor. Definitive therapy for localized disease with favorable histology consists of complete resection and chemotherapy. Surgery, chemotherapy, and irradiation are used in patients with advanced disease or unfavorable histology. The chemotherapeutic agents with proven efficacy are vincristine, actinomycin D, and doxorubicin.

Wilms' tumor exemplifies the advances made in pediatric oncology with multimodal therapy: disease-free survival of all patients approaches 90%. Even patients with metastatic disease, bilateral tumors, or unfavorable histology can be treated successfully.

Delayed complications from the combined use of radiation and chemotherapy can occur months or years after treatment. They include inflammation and then fibrosis of lung, liver, or kidneys. Postradiation hepatitis is often associated with thrombocytopenia. The complications can be either debilitating or fatal; fortunately, they are rare.

Neuroblastoma

Background

Definition

Neuroblastoma is a malignant tumor that arises from the sympathetic tissue in the adrenal medulla or in the sympathetic chain along the craniospinal axis in the neck, in the posterior mediastinum (see previous section on thoracic tumors), intra-abdominally, or in the pelvis. The tumor is composed of small, round cells with scant neurofibrillar cytoplasm. The cells may form rosettes. Neuroblastoma commonly metastasizes to bone, bone marrow, liver, skin, lymph nodes, and uncommonly to lungs or to the CNS.

Classification

Several staging systems have been in use for neuroblastoma. To provide a uniform nomenclature, the International Committee for Neuroblastoma Staging has proposed a staging system based on extent of disease and the completeness of surgical resection. Recently, factors other than stage have been shown to be important for predicting outcome. These factors include the Shimada-Chatten histology, the child's age, serum ferritin, amplification of the *MYCN* oncogene in tumor tissue, and the DNA content of the tumor cells.

Epidemiology

After CNS tumors, neuroblastoma is the most common solid tumor in childhood. About 50% occur before the age of 2 years and about 80% are found in children less than 5 years old. Neuroblastoma occurs in approximately 1 per 100,000 children less than age 16 each year.

Clinical Manifestations and Pathophysiology

About two-thirds of patients have widespread metastases. Symptoms and signs include irritability, anorexia, weight loss, pallor, and subcutaneous nodules. Adrenal neuroblastoma as well as nonadrenal intra-abdominal tumors can cause an abdominal mass that is palpable on routine physical examination. Involvement of the cervical sympathetic ganglia can cause Horner's syndrome or hoarseness by compressing the recurrent laryngeal nerve. Neuroblastoma arising from the sympathetic ganglia and extending into and out of the intravertebral foramina (“dumbbell tumor”) can cause symptoms and signs of compression of the spinal cord. Skeletal lesions lead to bone pain or pathologic fractures. Periorbital metastases cause proptosis and periorbital ecchymosis (“raccoon eyes”). Patients with black eyes can be mistaken for attered children (Fig. 100.4). Rare presentations include opsoclonus-myoclonus, hypertension, tachycardia, skin flush, and chronic diarrhea from excess catecholamine secretion.

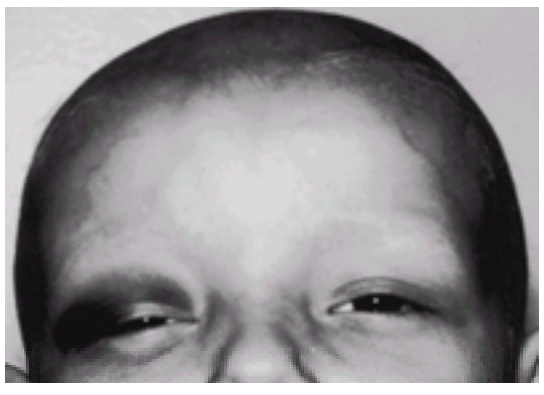

FIGURE 100.4. Neuroblastoma with “black eyes” that may be mistaken for child abuse. (Courtesy of Dr. Audrey Evans, Department of Pediatrics, Children's Hospital of Philadelphia.)

An intriguing characteristic of neuroblastoma is its ability to undergo spontaneous regression or to undergo maturation to more benign lesions, ganglioneuroblastoma, and ganglioneuroma. Whether all ganglioneuromas were once neuroblastoma is unclear, but it is generally agreed that they have the same cellular origin.

Stage 4S (formerly IVS of the Evans classification) encompasses a unique group of patients with metastatic disease: generally well-appearing infants whose primary tumor is small, with metastases limited to liver, skin, and marrow. Most of these patients have spontaneous regression of tumor, even when the tumor burden is massive. Infants with stage 4S appear to have a tumor that is biologically distinct from the tumor of older patients with stage 4 disease metastatic to bone and bone marrow. Patients with stage 4S present in the first few months of life with gross hepatomegaly or skin nodules. The primary tumor may not be obvious. The emergency physician who notes massive hepatomegaly in an infant who otherwise looks healthy should consider stage 4S neuroblastoma in the differential diagnosis.

Management

Evaluation of patients with suspected neuroblastoma should include a CBC, ultrasound, chest radiograph, skeletal survey, bone scan, and bone marrow aspiration and biopsy. Radiographs of painful areas in bones may show symmetric lytic lesions of the metaphyses of the long bones. Cortical destruction is seen in flat bones. Skull films may show increased soft tissue over the involved orbit in a child with proptosis. Abdominal radiographs reveal a retroperitoneal mass displacing normal structures. Adrenal primaries commonly have fine calcifications. Ultrasound can show a suprarenal mass that displaces the kidney down and away from its normal axis. All patients with suspected neuroblastoma should have quantitative evaluation of serum ferritin and urinary catecholamines including homovanillic acid (HVA) and vanillylmandelic acid (VMA). Evaluation requires several days and is most efficiently performed at a pediatric oncology center. The primary tumor is examined for the amount of stroma and number of mitoses, and viable tissue is assayed for amplification of *MYCN*, a cellular oncogene of prognostic importance in this tumor.

Treatment of neuroblastoma is directed toward both the tumor itself and complications of the tumor. The choice of treatment is based on several prognostic factors: age, stage, initial ferritin, histology, and *MYCN* oncogene amplification. Most low-stage tumors are associated with a good prognosis and are treated with surgical excision. Patients with poor-prognosis disease are treated with combined modalities, including bone marrow transplantation. Radiation may be used to reduce tumor bulk and for palliation, especially to treat painful bone lesions. Chemotherapeutic agents used to treat advanced neuroblastoma include vincristine, cyclophosphamide, doxorubicin, DTIC, VP-16, and cisplatin. Even though most poor-prognosis patients initially respond to therapy, many eventually die of recurrent or progressive disease.

Emergency physicians need to recognize special conditions in neuroblastoma.

1. Massive hepatomegaly in a neonate or infant can cause hepatic failure or respiratory embarrassment. These children need to be admitted for supportive care.
2. Dumbbell tumors can cause cord compression. These children need immediate evaluation. The acute onset of paraplegia in a child less than 5 years of age should be considered neuroblastoma unless another cause has been proven. Treatment may consist of chemotherapy, laminectomy, surgical removal, and irradiation.
3. Rarely, airway obstruction can result from large mediastinal tumors.
4. Extensive marrow disease can cause pancytopenia. Children with neuroblastoma and fever and neutropenia need to be managed similarly to children with leukemia.
5. Opsoclonus-myoclonus (dancing eyes, dancing feet) can resemble a toxic drug reaction or neuromuscular disease. This disorder is usually seen in children with limited surgically resectable disease; however, surgery may not cure the neurologic disorder.

Hepatic Tumors

Background

Definition

The two major primary liver tumors in children are hepatoblastoma and hepatocellular carcinoma. iver tumors originate most commonly in the right lobe of the liver. The common sites of metastases are lung and intra-abdominal lymph nodes and viscera. Bone and bone marrow spread is unusual.

Epidemiology

The incidence of primary hepatic tumors in children is 0.2 per 100,000 each year. Hepatoblastoma usually occurs before the age of 3 years. Incidence of hepatocellular carcinoma peaks around 4 years of age and again around 15 years.

A strong association exists among the appearance of cirrhosis, prior hepatitis B virus infection, and the development of hepatocellular carcinoma, although the association in children is not as strong as it is in adults. Metabolic diseases that cause cirrhosis (von Gierke's disease, Niemann-Pick disease, galactosemia, and chronic hereditary tyrosinuria) can progress to hepatic cancer. Patients who have received anabolic steroid therapy are at risk for developing benign hepatic neoplasms, and a few cases of hepatocellular carcinoma after androgen therapy or long-term hyperalimentation have been reported.

Clinical Manifestations and Pathophysiology

The most common presentation of a hepatic tumor is an enlarging abdomen, an abdominal mass, and symptoms and

signs resulting from the mass—anorexia, weight loss, vomiting, pain, fever, diarrhea, and irritability. Jaundice is uncommon, but pallor as a result of chronic disease is seen. Virilization, which may result from production of chorionic gonadotropin, is rare.

Management

The evaluation of patients with suspected liver tumors includes roentgenographic examination of the abdomen, which may show an enlarged liver and occasionally hepatic calcifications; a chest roentgenogram should be taken to look for metastases. CT scan shows lucent areas with increased vascularity after contrast. Initial laboratory tests should include a CBC and liver function tests (although they are often normal) and serum a-fetoprotein. Further evaluation necessitates admission to the hospital.

Tumors of the Female Genitalia

Background

Definition

Tumors of the female genitalia include primary ovarian tumors and vulvar, vaginal, and intrauterine tumors. Most ovarian tumors are cystic and histologically benign. Malignant ovarian tumors are solid, and in children, they are usually derived from the germ cells. Malignant tumors of the vulva are usually sarcomas; vaginal tumors are either sarcoma botryoides in younger girls or adenocarcinomas in adolescents.

Classification

The ovarian tumors seen in children are dysgerminomas, embryonal carcinomas, endodermal sinus tumors, teratomas, choriocarcinomas, and mixed germ-cell tumors. The staging system is based on the extent of tumor spread.

Epidemiology

Malignant ovarian tumors are uncommon, accounting for only about 1% of malignant neoplasms in girls less than 17 years old. Incidence increases with age, with approximately 20% occurring between 0 and 4 years, 30% between 5 and 9, and 50% between 10 and 14. Menarchial hormonal factors may play a role in development of ovarian tumors.

Female offspring of mothers who received diethylstilbestrol (DES) during pregnancy are at risk for developing vaginal adenocarcinoma.

Presentation and Pathophysiology

Ovarian tumors are usually large and may be mistaken for pregnancy. They can cause painful ovarian torsion or an acute abdomen, especially if hemorrhage or rupture of a cyst occurs; right-sided ovarian tumors may mimic acute appendicitis.

Stromal tumors may produce hormones, causing precocious puberty, vaginal bleeding (granulosa cell tumors), or masculinization (arrhenoblastoma). Ovarian tumors spread by direct extension along the adnexal structures to lymph nodes and the peritoneal surface of the bladder, uterus, sigmoid colon, liver, diaphragm, and small intestine. Rarely, these tumors metastasize to lung, liver, and bone. Ascites and pleural effusion are seen with fibroid tumors (Meigs' syndrome); however, fibroids are rare in children and adolescents.

Vaginal sarcomas may present as exophytic masses. Vaginal adenocarcinoma is detected by a routine Papanicolaou smear or by the presence of a mass or bleeding.

Management

Girls with suspected genital neoplasms need a thorough physical examination, including careful inspection of the external genitalia, bimanual pelvic examination, and evaluation of pubertal status. This evaluation should be performed by a physician experienced in pediatric and adolescent gynecology. Abdominal radiograph may reveal displacement of bowel, calcifications, teeth or other aberrant tissue structures, and ascites, but abdominal and pelvic ultrasound is the preferred imaging technique. A chest radiograph should be taken to look for metastatic disease. Laboratory examinations for ovarian tumors should include measurement of a-fetoprotein, which is produced by embryonal carcinoma and endodermal sinus tumor, and human chorionic gonadotropin, produced by embryonal carcinoma and choriocarcinoma.

Tumors of the Male Genitalia

Background

Tumors of the male genitalia include testicular, paratesticular, and prostatic tumors. Germ-cell tumors comprise about 75% of the primary testicular tumors. Leukemia can metastasize to the testes. Paratesticular and prostatic tumors are usually rhabdomyosarcomas.

A relatively high incidence of germ-cell testicular tumors occurs in infancy. The incidence decreases after age 4 years and begins to rise again in puberty, with a continued increase into early adulthood. A cryptorchid testis has a 30 to 50 times greater risk of developing a neoplasm; 10 to 20% of all testicular cancers arise from a cryptorchid testis. Sarcomas of the male genitalia occur mostly between the ages of 2 and 10 years.

Clinical Manifestations

Most testicular tumors present as slowly growing scrotal masses. Occasionally, a patient may have pain. About 25% of the patients have either an associated hydrocele or an inguinal hernia. Testicular and paratesticular tumors most commonly metastasize to lungs or spread to regional lymph nodes.

Management

Patients with solid testicular or paratesticular masses should be referred to a urologic surgeon with experience in removing pediatric neoplasms. For germ-cell tumors, treatment usually consists of orchiectomy with high ligation of the spermatic cord. Sarcomas require chemotherapy and irradiation in addition to surgery.

Tumors of the Head, Neck, and Extremities

Tumors of the head, neck, and extremities include those that present as masses in the brain (brain tumors) or near the brain (parameningeal tumors), in the eye (retinoblastoma), and in the neck or extremities. Most malignant tumors of the nasopharynx, middle ear, or behind the eye are rhabdomyosarcomas. In the neck, lymphomas are most common; sarcomas, neuroblastomas, thyroid carcinomas, and metastatic tumors also occur. Soft-tissue sarcomas and bone sarcomas are the most common malignant tumors of the extremities.

Retinoblastoma

Background

Retinoblastoma is the most common primary intraocular malignancy in children. Retinoblastoma arises from the nuclear layer of the retina and is of neurologic origin. Most cases of retinoblastoma are cured by enucleation. Retinoblastoma occasionally metastasizes hematogenously to bone marrow, bones, lymph nodes, and liver. It can also spread by direct extension via the optic nerve to the meninges and into the spinal fluid. The staging system for retinoblastoma is based on size, number, and location of lesions in the retina and vitreous.

Retinoblastoma occurs in 1 in 23,000 births. Most patients are diagnosed by 2 years of age. Two patterns of retinoblastoma occur: 1) a hereditary form that is autosomal dominant with about 90% penetrance, and 2) a sporadic form. Approximately 35 to 45% of retinoblastomas are hereditary. Bilateral cases are almost always hereditary. Patients with retinoblastoma are at risk for developing osteosarcoma or pineal tumors (i.e., trilateral retinoblastoma) later in life.

Clinical Manifestations

Two-thirds of children with retinoblastoma have a white pupil (leukocoria or “cat's eye”), often detected by the parents (Fig. 100.5). The white pupil is actually the tumor itself, as seen through the vitreous. Other symptoms and signs include strabismus (see Chapter 25); a unilateral, fixed, and dilated pupil (see Chapter 26); decreased visual acuity; a red, painful eye (see Chapter 24) caused by glaucoma; spontaneous hyphema; proptosis; and heterochromia iridis. In families with known hereditary forms of retinoblastoma, most tumors are detected while they are asymptomatic because of careful and frequent ophthalmologic examinations.

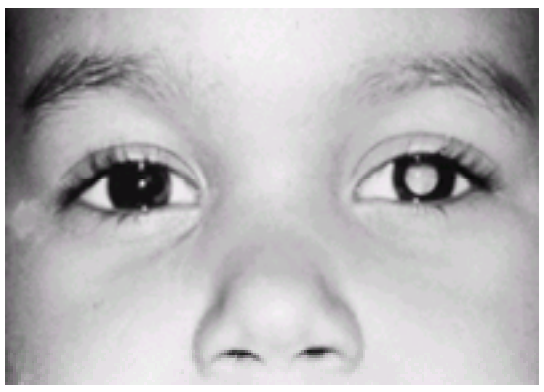

FIGURE 100.5. Leukocoria in a child with retinoblastoma of the left eye. (Courtesy of Dr. Anna Meadows, Department of Pediatrics, Children's Hospital of Philadelphia.)

Management

All children who present with a white pupil or with suspected retinoblastoma need a thorough ocular examination under general anesthesia by an experienced ophthalmologist. The ophthalmologist must be able to differentiate other causes of leukocoria (nematode endophthalmitis, persistent hyperplastic primary vitreous, Coats' disease, coloboma, idiopathic retinal detachment, and congenital cataract) from a neoplasm. Treatment is usually enucleation; more ecently, insertion of radioactive plaques or use of laser in small tumors has been under investigation. Chemotherapy is reserved for metastatic disease or for bilateral disease.

Thyroid Cancer

Background

Thyroid cancers are carcinomas, usually arising in the thyroid gland and less commonly in ectopic thyroid tissue. Thyroid cancer is divided into four histologic subtypes: papillary, follicular, medullary, and anaplastic. Papillary carcinoma accounts for approximately 70% of cancers, and follicular carcinoma comprises 20%.

Thyroid cancer is uncommon in children. Exact figures regarding the incidence in childhood are difficult to obtain. The disease is approximately twice as common in females as in males.

A well-documented association exists between previous low-dose head and neck irradiation and the development of thyroid cancer. This association was discovered when the histories of many patients with thyroid cancer revealed that they had received irradiation for conditions such as acne, tonsil enlargement, or thymic enlargement. All persons with previous neck irradiation should have careful neck examinations every 1 to 2 years, and all palpable nodules should be evaluated.

Clinical Manifestation and Pathophysiology

Thyroid cancer usually presents as one or more firm, painless nodules in the neck. Occasionally, patients will have symptoms of dysphagia or hoarseness. Thyroid cancer commonly spreads to cervical lymph nodes and less commonly to lungs and bones. Local invasion of the trachea, larynx, or esophagus can also occur. Any patient suspected of having a thyroid nodule should have a radioisotope study of the thyroid with either iodine-123 or technetium-99 pertechnetate. Thyroid scan usually reveals a “cold” nodule, although “hot” nodules can also be malignant. The evaluation should also include radiographs of the chest and neck, radionuclide bone scans, and measurement of triiodothyronine (T_3), thyroxine (T_4), and thyroid-stimulating hormone (TSH).

Soft-Tissue Sarcomas

Background

Definition

Soft-tissue sarcomas are derived from mesenchymal tissue: muscle, tendon, nerve, fat, and endothelium. Rhabdomyosarcoma arises from muscle and is the most common member of this group in children. Less common are Ewing's sarcoma (PNET), fibrosarcoma, neurofibrosarcoma, synovial sarcoma, mesenchymoma, malignant fibrous histiocytoma, and leiomyosarcoma.

Classification

Soft-tissue sarcomas are classified according to the apparent tissue of origin. Rhabdomyosarcoma is further subdivided according to histologic subtype (embryonal, alveolar, or pleomorphic) and the extent of disease after surgery.

Epidemiology

Soft-tissue sarcomas can occur at birth. Their incidence peaks at 2 to 6 years of age. Sarcoma botryoides of the bladder, prostate, or vagina tend to occur in infants; paratesticular sarcomas occur in childhood or adolescence. Soft-tissue sarcomas occur at an annual rate of 0.84 per 100,000 in white children and 0.39 per 100,000 in African-American children. They account for approximately 6% of all neoplasms in children.

Clinical Presentation and Pathophysiology

A sarcoma usually presents as a lump in the soft-tissues. The lump may be painless or painful. Specific symptoms depend on the location of the tumor.

Orbital tumors cause rapidly developing unilateral proptosis. Nasopharyngeal tumors can present with recurrent epistaxis, chronic sinusitis, chronic nasal obstruction, or dysphagia. Middle ear rhabdomyosarcoma can cause a chronic otitis media, ear pain, cranial nerve palsies, and rarely, sarcomatous meningitis. Neck tumors can cause dysphagia, hoarseness, or simply a painless mass in the posterior cervical triangle. Tumors of the bladder or prostate lead to acute urinary tract obstruction or constipation. Sarcoma botryoides of the vagina present with bleeding or as a polypoid mass protruding from the introitus. Extremity sarcomas usually appear as painless masses.

Management

The role of the emergency physician is to include cancer in the differential diagnosis of masses in the soft tissues of children and to refer children for prompt biopsy of the mass when a benign cause has been ruled out. Definite therapy consists of surgery, chemotherapy, and usually radiotherapy.

Bone Tumors

Background

Definition

A variety of neoplasms, many of which are benign, occur in bones. The malignant tumors are sarcomas arising from the cells of the cortical or cancellous bone (osteogenic sarcoma), cartilaginous bone (chondrosarcoma), periosteum (periosteal sarcoma), or reticuloendothelial cells of the marrow (Ewing's tumor).

Classification

Malignant bone tumors are classified according to the cell of origin. The two malignant bone tumors that are of practical importance are osteogenic sarcoma and Ewing's tumor.

Epidemiology

Osteogenic sarcoma and Ewing's tumor are more common in adolescents than in children. However, both tumors have been described in infants. Sixty percent of osteogenic sarcomas occur just above or below the knee. Both osteogenic sarcomas and Ewing's tumor show a male:female predominance of 1.6:1. The incidence of osteogenic sarcoma in the United States is approximately 0.5 per 100,000 children and that of Ewing's tumor is 0.1 per 100,000. Osteogenic sarcoma shows no racial predilection; Ewing's tumor, in contrast, is exceedingly rare in African-Americans.

Considerable epidemiologic data have accumulated to show that ionizing radiation causes osteogenic sarcoma. Despite the extensive epidemiologic data linking radiation and osteogenic sarcoma, most adolescents with osteosarcoma have had no excessive exposure to sources of radiation. Likewise, the cause of Ewing's tumor is unknown.

Clinical Presentation and Pathophysiology

Patients with malignant bone tumors complain of pain or a painful lump. The pain commonly occurs after a well-remembered episode of trauma and has often been present for weeks to months. Traumatic hemorrhage into the tumor may call attention to a lesion. Often, the pain is more intense at night and may awaken the patient. Patients with osteogenic sarcoma are almost always well. About one-third of patients with Ewing's tumor have a history of systemic symptoms, including fever, weight loss, anorexia, and malaise.

When the tumor is in an extremity, physical examination may reveal a hard, tender mass. Large tumors may feel warm and may have obviously increased vascularity. Disuse muscular atrophy may occur in the affected limb.

Trunk lesions, especially those of the pelvis, are difficult to diagnose. Often the mass is buried in gluteal tissue and cannot be appreciated on routine physical examination or on routine radiographs. A meticulous neurologic examination with particular attention to bladder and bowel function and the lower extremities is mandatory.

Diagnosis of a malignant bone tumor requires a radiograph and a biopsy of the lesion. Radiographs will reveal changes associated with a destructive process in bone. Early changes include loss of soft-tissue fat planes and periosteal elevation (Codman's triangle). Later, an “onionskin” periosteal reaction caused by repetitive episodes of the lesion pushing out the periosteum and the periosteum responding by laying down calcium is visible. Further along in disease development, normal trabeculation disappears, and areas of lysis are seen. No defined sclerotic margin is visible around the area of destruction, and the tumor's precise limits are impossible to determine. Eventually, the tumor breaks through the cortex, weakening bone and predisposing the patient to pathologic fracture. The “sunburst” phenomenon of osteosarcoma occurs as the tumor blood vessels grow perpendicularly to the shaft of the bone and malignant osteoblasts lay down bone along the vessels (Fig. 100.6). Almost all osteosarcomas are seen on plain radiographs. Some Ewing's tumors may not be obvious; if the history and physical examination suggest that trauma alone cannot account for the pain, a bone scan is the next radiologic study to order. Sometimes, pelvic Ewing's tumors may be difficult to see on plain radiographs; CT scan is helpful in delineating the full extent of these tumors. In osteosarcoma and Ewing's tumor, MRI may help delineate the extent of disease, but it is rarely needed to make the initial diagnosis. Other studies that may be useful are a CBC, sedimentation rate, serum lactate dehydrogenase (LDH) in Ewing's tumors, and serum alkaline phosphatase in osteosarcoma. If a presumed malignant tumor is seen, a plain chest radiograph should be taken to determine whether obvious metastases are present.

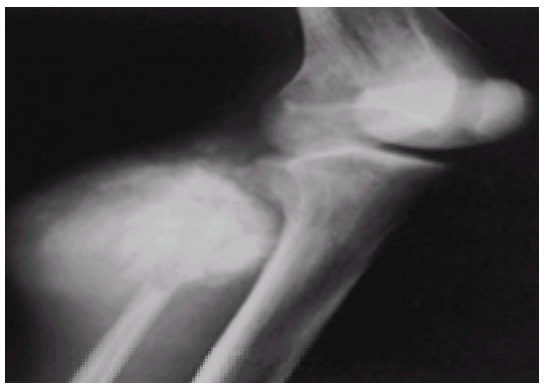

FIGURE 100.6. Osteosarcoma in the fibula in a 14-year-old girl. Radiograph shows lytic bone destruction, malignant new bone formation, cortical breakthrough, and massive extraosseous tumor.

Management

If the clinical findings suggest osteomyelitis, blood culture and culture from the lesion are necessary. If the clinical presentation is more suggestive of tumor, a biopsy is indicated. Ideally, biopsy of a Ewing's tumor or osteosarcoma should be performed by an orthopedic surgeon who has expertise in the management of eoplastic bone lesions. A large lesion with a thin cortex in a weight-bearing bone may require immediate immobilization to prevent pathologic fracture.

Before 1970, patients with osteogenic sarcoma or Ewing's tumor succumbed to pulmonary metastases within months or years. Since the early 1970s, adjuvant chemotherapy has been combined with surgery (amputation or en bloc resection for osteogenic sarcoma) or high-dose irradiation (Ewing's tumor). Agents used to treat osteogenic sarcoma include vincristine, high-dose methotrexate with leucovorin rescue, Adriamycin, and cisplatin. Agents used to treat Ewing's tumor include vincristine, Adriamycin, cyclophosphamide, actinomycin D, VP-16, and ifosfamide. Current projected survival rate for patients with these tumors is roughly 50%.

The major complication of osteogenic sarcoma therapy that may bring a patient to the attention of an emergency facility is methotrexate toxicity. Patients treated with high-dose methotrexate can develop life-threatening mucositis, renal compromise, or cerebral vascular changes.

Rarely, patients with osteosarcoma develop pneumothoraces. This condition is usually an indication that a subpleural metastasis has broken through the pleura. No immediate intervention is warranted unless the pneumothorax causes respiratory distress.

Patients with Ewing's tumor who have received irradiation and chemotherapy are prone to develop pathologic fractures. These fractures may occur silently or may cause pain and dislocation. Treatment consists of external immobilization. Many fractures eventually require internal fixation.

Central Nervous System Tumors

Background

Definition

Primary CNS tumors, whether histologically benign or malignant, arise in the brain, meninges, or spinal cord. This group of diseases includes many histologic variants, but only the relatively common primary neoplasms are discussed. In the past, metastatic brain tumors were relatively uncommon in children, but as chemotherapy changes the natural history of some tumors, cerebral or epidural metastases of some tumors (rhabdomyosarcoma, Ewing's sarcoma, osteogenic sarcoma, and lymphomas) are becoming more common.

Classification

Several classification systems have been used for characterization of brain tumors, based on histology, benignancy or malignancy, and location. For the clinician, a more useful classification is one based on the location of the tumors because lesions at specific locations produce corresponding specific neurologic deficits. In contrast to adult brain tumors, most pediatric brain tumors are infratentorial. Histologically, the most common brain tumors are gliomas. Astrocytomas, a subset of gliomas, comprise about one-third of all brain tumors. Astrocytomas are graded according to the degree of malignancy: grade 1 astrocytoma is histologically benign and grade 4, glioblastoma multiforme, is the most malignant. More than half of astrocytomas are infratentorial, predominantly cerebellar in location. Brainstem gliomas are usually regarded as a separate subset of astrocytomas. Astrocytomas and ependymomas are the common low-grade spinal cord gliomas.

Primitive neuroectodermal tumors (PNETs), the most common of which was formerly called medulloblastoma, are the second most common histologic types of brain tumor. They mostly occur in the cerebellum, arising from germinative cells of the external granular layer, and grow predominantly from the vermis.

Epidemiology

Brain tumors occur in 4 of 100,000 children yearly and are the most common group of solid tumors in children. They can occur at any age, but their incidence peaks from ages 5 to 10 years. Gliomas are more common in children less than 5 years old, and PNETs are most common around age 8 years, with a striking male predominance of 4:1. Certain constitutional disorders are associated with an increased risk of development of brain tumors. Most notable is neurofibromatosis type 1 (NF-1) in which optic pathway, hypothalamic, and brainstem gliomas are common; in addition, MRI screening programs have identified benign, sometimes transient unidentified bright objects (UBOs) in young children with NF-1. Many brain tumors in patients with NF-1 do not require treatment. Patients with NF-2 may develop acoustic neuromas and meningiomas. Other associations of brain tumors with constitutional disorders are tuberous sclerosis with subependymal giant cell astrocytoma, von Hippel-Lindau disease with retinal or brainstem hemangioblastomas as well as renal neoplasms, and nevoid basal cell carcinoma syndrome with PNETs.

Clinical Presentation and Pathophysiology

Most brain tumors cause symptoms and signs of increased intracranial pressure (see Chapter 13, Chapter 78, Chapter 83, and Chapter 125). In fact, brain tumors are now a relatively common cause of increased intracranial pressure because the incidence of other causes, such as brain abscess, is decreasing. The symptoms of increased intracranial pressure vary with age. Infants show personality changes, vomiting, lethargy, loss of previously acquired motor skills, seizures, and symptoms of obstructive hydrocephalus. In older children, headache is the most common complaint. Early in the course, headaches are usually intermittent. A history of headache on arising in the morning should alert the

emergency physician to the possibility of a CNS tumor. Morning vomiting, with or without nausea, is also a common presenting symptom. Other early anifestations include diplopia, ataxia, hemiparesis, speech disturbance, stiff neck, dizziness, lethargy, and coma.

Specific tumors cause specific focal neurologic changes. Brainstem gliomas cause cranial nerve deficits usually in the order of VI, VII, IX, and X. Facial palsies and dysphagia are presenting symptoms; ataxia and hemiparesis follow. Localizing symptoms often occur before increased intracranial pressure. PNETs and cerebellar astrocytomas, because of their midline location, cause truncal ataxis and an unbalanced reeling gait. If PNETs “drop” metastases to the cord, cord compression may occur (see p. 1183 and Chapter 83). Cerebellar astrocytomas that occupy one hemisphere cause ipsilateral hypotonia and a tendency to fall to the side of the lesion. Herniation of a cerebellar tonsil causes head tilt and neck stiffness. Tumors near the third ventricle (craniopharyngiomas, germinomas, optic gliomas, and hypothalamic and pituitary tumors) cause visual impairment, increased intracranial pressure, and hydrocephalus. Tumors of the chiasmatic/hypothalamic region may produce the diencephalic syndrome (failure to thrive, wasting, and inappropriate alertness) in infants and young children. However, these tumors may cause major deficits in visual acuity and reduction in visual fields that are difficult to detect in young children. Pineal tumors obstruct the aqueduct of Sylvius, producing increased intracranial pressure and Parinaud's syndrome (upward gaze paralysis, convergence nystagmus, and decreased pupillary response). Cerebral astrocytomas, ependymomas, and oligodendrogliomas cause seizures and hemiparesis.

Spinal cord tumors cause pain at the site of the tumor and neurologic deficits. The tumor is sometimes localized by tenderness of the overlying vertebral segments. Depressed motor function, sensory deficit, hyperreflexia below the lesion, and a Babinski sign may be present. Later, paralysis, areflexia, and loss of bladder and bowel control may occur.

Management

The long-term management of CNS tumors requires a team approach that includes the participation of a pediatric neurologist, neurosurgeon, oncologist, radiation therapist, and psychosocial support staff. The role of the emergency physician in the management of CNS tumors is one of recognizing CNS lesions, performing the initial diagnostic procedures to establish the presence of the lesion, and stabilizing the patient who has a life-threatening increase in intracranial pressure or spinal cord compression. When an intracranial lesion is suspected, a complete neurologic evaluation, including an eye examination and developmental assessment, is mandatory. CT scanning is the most rapid way to establish the presence of a mass lesion. The mass will show enhancement with the use of contrast material. Definitive localization of intracranial and spinal cord tumors is now accomplished using MRI. Myelography and angiography are reserved for those patients in whom the neurosurgeon needs further information for planning the surgical approach.

Lumbar puncture is rarely useful in the initial evaluation of a child with a CNS tumor. If focal neurologic signs or signs of increased intracranial pressure are present, a CT scan should precede any attempt to perform a lumbar puncture. If an immediate lumbar puncture is deemed necessary to rule out meningitis, the protocol outlined in Chapter 12 should be followed.

Increased intracranial pressure may be treated with the administration of dexamethasone (0.5 to 1.0 mg/kg per day in four divided doses) and diuretics such as mannitol (1 to 2 g/kg over 30 to 60 minutes). Depending on the severity of the neurologic findings, intubation and hyperventilation may be indicated. (See Chapter 13 and Chapter 83 for additional discussions of increased intracranial pressure.)

The patient should be examined carefully for signs of spinal cord compression. Tumors that cause spinal cord compression, whether intrinsic to the cord, such as astrocytomas, ependymomas, or chordomas, or extrinsic to the cord, such as metastatic PNETs, lymphomas, or sarcomas, demand immediate attention by a pediatric neurosurgeon and oncologist. (See the following section on cord compression, p. 1183.)

Definitive diagnosis of a CNS tumor should be made by biopsy. Surgical resection is becoming increasingly important because surgery in the hands of experienced pediatric neurosurgeons appears to improve the prognosis for CNS tumors. An operative procedure that removes as much of the tumor as possible is a goal that must be weighed against the risk of neurologic deficits. Tumors that are deep in the dominant hemisphere or in the brainstem are not usually amenable to aggressive surgical resection.

Definitive therapy for CNS tumors depends on the patient's age, the location of the tumor, tumor histology, and the extent of the surgical resection. Radiation therapy (4500 to 6500 cGy) has improved survival in most children with unresectable CNS tumors, especially PNETs, ependymomas, and low-grade astrocytomas. However, the dosages used may be devastating to children less than 5 years of age, and chemotherapeutic regimens are now being used in this group of patients. Adjunctive chemotherapy and bone marrow transplantation in patients with CNS tumors are under investigation. Many patients are now receiving multimodal therapy with surgery, radiation therapy, and multiagent chemotherapy. Chemotherapeutic agents used for CNS tumors include lomustine (CCNU), carmustine (BCNU), cyclophosphamide, vincristine, cisplatin, carboplatin, ifosfamide, prednisone, VP-16, procarbazine, and thiotepa.

COMPLICATIONS OF THERAPY AND PROGRESSIVE DISEASE

Children with cancer have many problems that are no different from those of normal children. However, the ffects of their disease and of the surgery, chemotherapy, and radiation used to treat their disease create problems unique to these patients. This section reviews the most common problems that are unique to children with cancer. These problems generally arise from 1) bone marrow suppression, 2) metabolic derangements caused by the primary disease or chemotherapy, and 3) major organ damage caused by treatment or progressive disease.

The evaluation of a patient with cancer who presents to the ED begins with establishing the patient's history. Table 100.4

lists the basic data needed to evaluate a cancer patient. Knowledge of the most recent course of therapy is important because it establishes the potential for bone marrow suppression at the time of the visit.

I. Primary diagnosis	IV. Central venous access device
A. Disease	A. Type
B. Primary and metastatic sites	B. Previous infections
C. Date of diagnosis	V. Current medications
D. Status of disease (remission, relapse, completed therapy)	A. Corticosteroids
II. Surgical history	B. Chemotherapy
A. Date	C. Trimethoprim-sulfamethoxazole prophylaxis
B. Extent of resection	D. Growth factors (e.g., g-CSF, erythropoeitin)
III. Last treatment	VI. How is the child acting?
A. Chemotherapy	VII. Has the problem occurred before?
1. Drugs	VIII. What does the parent believe to be the problem or the cause of the problem?
2. Date	
B. Radiation therapy	
1. Dosage	
2. Fields	
3. Date	

Table 100.4. Basic Historical Data Needed for Evaluation of a Cancer Patient in the Emergency Department

In general, the first few months of cancer therapy are intensive. During this time, oncologists are particularly cautious about the risk of infection associated with bone marrow suppression. Early in the course of therapy a patient may be admitted to the hospital for observation on the basis of ill appearance alone, even if the neutrophil count is adequate and the patient is afebrile.

The parents of children with cancer are often uncomfortable in the ED because of the absence of doctors and nurses with whom they are familiar, the perceived risk that their child will be exposed to communicable diseases, and the need to recite their child's complex history. However, the parents of oncology patients usually acquire a great deal of sophistication about their child's illness and the symptoms that their child manifests when ill. Cancer chemotherapy can cause unusual side effects and can mask some of the symptoms and signs of illness; therefore, soliciting the parents' impression of their child's problem is important. The emergency physician must also recognize the parents' concerns that an acute problem may represent relapse of their child's cancer.

Except for patients who are still on mask precautions after bone marrow transplant, oncology patients do not need to be isolated in private rooms. However, they do need separation from patients who have contagious illnesses, especially varicella or measles infection. Strict handwashing by all personnel is mandatory.

Hematologic Complications

Anemia

Anemia is common in children being treated for cancer and is usually well tolerated. Most often, anemia is a result of the suppression of normal hematopoiesis by chemotherapy. Other causes, such as nutritional deficiencies, chronic blood loss, infection, and hemolysis, should be ruled out if the anemia is problematic. After induction therapy, routine transfusion for a hemoglobin in the range of 7 to 10 g/dL is not advisable unless the anemia is causing symptoms or blood loss is continuous. If the child is a potential candidate for bone marrow transplant, blood products from family members should not be used.

Guidelines for the use of packed RBCs are reviewed on p. 1160 and in Table 100.3. If there is a question about the need for CMV-negative blood, the child's oncologist or the blood bank director should be consulted. RBC products should be irradiated to 1500 cGy before transfusion. Leukodepletion filters should be used. It is useful to write “oncology patient” on blood bank requests so that the product can be properly prepared.

Some cancer patients may receive recombinant human erythropoietin to ameliorate the effects of chemotherapy-induced anemia. A patient who receives erythropoietin should not receive a packed RBC transfusion without his or her oncologist being consulted.

Hemorrhage

After infection, hemorrhage is the second most common serious complication of cancer therapy. Often the hemorrhage is minor (mild epistaxis, gum bleeding), but when the platelet count is below 10,000 to 20,000/mm ^{3}, spontaneous intracranial hemorrhage or GI bleeding becomes more likely. See the previous section on hemorrhage in the discussion of leukemia and Table 100.3 for treatment guidelines.

Two special circumstances require a high index of suspicion of hemorrhage. Patients who have had a resection of a CNS tumor are at particular risk for bleeding into the tumor bed. Also, many cancer patients taking high dosages of corticosteroids must be considered at increased risk of GI hemorrhage.

For epistaxis or bleeding of the oral mucosa, local measures to control the bleeding should be tried before blood products are used. Local pressure should be applied to the bleeding site or to the nasal bridge; the use of 2 to 3 drops of phenylephrine (0.25 or 0.5%) is ften helpful in epistaxis. One or two vials of topical thrombin (1000 units/vial, as the powder) may be applied after the large unstable clot is removed. Desmopressin (DDAVP, 10 µg/m ^{2} intravenously) has been used successfully in thrombocytopenic patients with mucosal bleeding that is unresponsive to local measures and

platelet transfusion.

Leukocytosis/Leukopenia

Leukocytosis is discussed in the previous section on leukemia. Infection in the leukopenic patient is discussed in the following section.

Infectious Complications

Despite advances in supportive care, infection remains a major cause of morbidity and mortality in cancer patients. As chemotherapy regimens have become more aggressive, periods of neutropenia and mucositis have become more severe and prolonged. The disruption of mechanical barriers to infection and the quantitative neutrophil defect place the cancer patient at increased risk of opportunistic infection. Indeed, even a minor infection can overwhelm the compromised immune system of a cancer patient. (See infectious complications in the previous section on leukemia, p. 1162, for a discussion of problems seen in patients with newly diagnosed leukemia.) A general approach to infection in the child with cancer is presented in Figure 100.7.

Febrile Neutropenic Patient

The most important variable in determining the outcome of infection is the degree of neutropenia caused by the disease itself and by many cytotoxic agents. In adults, *neutropenia* is defined as fewer than 1000 neutrophils/mm^3. In practice, children tolerate neutropenia better than adults, and most can remain free of infection with counts of more than 500 neutrophils/mm^3. However, with fewer than 500 neutrophils/mm^3, the risk of infection increases; at less than 200/mm^3, it becomes more likely. If the neutrophil count is on its way down from 200 to 0, the risk of infection becomes even greater. Duration of neutropenia is also significant. Infection is less likely if neutropenia represents a transient dip in the neutrophil count after a large dose of chemotherapy, and recovery is likely within a few days. If infection occurs in this setting, the patient's own defenses can usually be of aid in time. If, on the other hand, the neutropenia is of several weeks' duration, overwhelming infection is more of a threat.

Management of fever and presumed infection begins with a careful history. This historical information is important because the neutropenia and cytotoxic therapy can mask signs of illness and cause a physician who does not know the child well to underestimate the potential seriousness of the illness.

A thorough physical examination sometimes uncovers the source of fever. In addition to examining the common sites of infection in children (the buccal mucosa, perianal area, and nail beds), muscles, joints, bones, and former sites of intravenous infusions should be evaluated. In a patient undergoing treatment for cancer, just as in the patient with newly diagnosed leukemia, an apparently minor swelling or a tear in a mucosal surface or the skin can be a source of disseminated infection. Perirectal abscesses, commonly seen in patients with AML, may cause pain and discoloration but no edema. Seemingly insignificant paronychia may be the source of significant bacteremia. Blood and urine cultures should be obtained and a chest radiograph should be taken in patients with respiratory symptoms, but a lumbar puncture is not indicated unless signs of meningitis are present; for unknown reasons, infectious meningitis is rare in children with cancer.

The accepted practice in treating a febrile neutropenic patient is to begin empiric antibiotic therapy with combinations of intravenous broad-spectrum antibiotics. The choice of antibiotic combination depends on the prevailing patterns of infection and antimicrobial resistance in the area. First-line combinations currently in use are listed in Figure 100.7. These choices are based on experience; most infections in neutropenic patients arise from the patient's endogenous flora and are caused by invasion of skin (*Staphylococcus aureus* and *Staphylococcus epidermidis*) or intestinal *flora (Escherichia coli, Pseudomonas, Klebsiella, Enterococcus, Serratia). S. epidermidis* is a common cause of sepsis in children with indwelling Broviac or Hickman catheters. Antibiotics should be started if the child appears toxic or the neutrophil count is less than 500/mm^3. When possible, the primary physician should be contacted for advice.

Most patients with cancer do not present to the ED with invasive fungal disease unless they are in relapse or have been recently discharged from the hospital after a protracted course of antibiotics or hyperalimentation. Corticosteroids and indwelling catheters also predispose cancer patients to fungal disease. Fungal overgrowth can present as esophagitis, cellulitis, pneumonitis, sinusitis, meningitis, urinary tract infection, septicemia, or fever of unknown origin. Mucositis and esophagitis are discussed in the following sections about GI and genitourinary complications and pneumonitis is discussed in the following section about interstitial pneumonitis. In general, invasive fungal infections must be treated with intravenous amphotericin B. However, initiation of therapy with amphotericin B is not usually indicated in the ED.

Bacterial, viral, or fungal infections can cause septic shock in the neutropenic patient. Treatment consists of aggressive measures to maintain intravascular volume and blood pressure (see Chapter 3), broad-spectrum antibiotic therapy appropriate for neutropenic patients, and hematologic support with red cells, platelets, and plasma. Stress doses of corticosteroids should be given to patients who have been on adrenal-suppressive doses of prednisone or dexamethasone for more than 10 days.

Patients with Central Venous Catheters

Many children with cancer have central venous catheters. These catheters may be of the Hickman/Broviac type, which have an external portion, or of the subcutaneous port type, which are completely internal. Patients with these devices are at increased risk of three types of infection: 1) bacteremia or sepsis related to an indwelling catheter, particularly with *S. epidermidis, Streptococcus viridans,* and Gram-negative enteric organisms; 2) local infection along the tunnel tract from the central vessel to the exit or port site or surrounding the port, usually with *S. aureus* or *S. epidermidis;* and 3) infection

at the exit site of externalized catheters.

Patients with central venous catheters and significant fever (38°C [100.4°F] × 3 or 38.4°C ([101.1°F] or higher × 1) require admission and intravenous antibiotics, after blood cultures are obtained. Blood cultures must be drawn from each lumen of the catheter. The choice of antibiotics depends on the patient's absolute neutrophil count (Fig. 100.7, Category 1 and Category 2). Fever, headache, nausea and vomiting, or chills that occur within 20 to 45 minutes after a central venous catheter is flushed indicate bacteremia related to catheter infection. The patient with erythema, swelling, or tenderness along the tunnel tract or around the port should be treated with intravenous antibiotics (Fig. 100.7, Category 1 or 2) even if fever is not present, and a pediatric surgeon familiar with central venous access devices should examine the patient. Catheters may become obstructed by blood clot formation. This may act as a nodus for infection.

Patients with erythema around the exit site of an externalized catheter, no fever, and an absolute neutrophil count of more than 500/mm^3 may be treated with conservative measures. Dressing changes should be increased to once or twice per day; mupirocin ointment may be added to the dressing change routine. During warm weather, fungal dermatitis under the catheter dressing is common. Frequent dressing changes and the use of clotrimazole cream twice daily will usually result in rapid resolution of this problem.

Interstitial Pneumonitis

Interstitial pneumonitis in the oncology patient may be caused by a wide range of infectious agents or may be a result of chemotherapy or radiation therapy. Therefore, it occurs in patients with different primary diagnoses and in different clinical settings. A patient often presents with interstitial pneumonitis when the eutrophil count is normal and when the primary disease is under control.

Before the advent of effective prophylaxis with trimethoprim–sulfamethoxazole (TMP-SMZ given at a dosage of 2.5 to 5 mg/kg per dose of TMP twice daily on 2 to 3 consecutive days per week), *Pneumocystis carinii* pneumonia (PCP) presented a special problem for patients with ALL. PCP was the most common cause of death in ALL patients in remission before the use of TMP-SMZ. PCP has been virtually eliminated from this population since the advent of effective prophylaxis.

However, ALL patients who are unable to take TMP-SMZ because of sensitivity to the drug or drug-induced myelosuppression are still at risk for PCP, even if they are treated with inhaled pentamidine. Several cases of PCP have been reported during the use of inhaled pentamidine. These cases tend to be milder and localized to lung segments not reached by an aerosolized drug (e.g., the right upper lobe). Patients who have had lung, mediastinal, or spinal irradiation, who are receiving high-dose corticosteroids, or who are receiving severely myelosuppressive chemotherapy are also at risk for PCP if not given adequate prophylaxis.

Now that the risk of PCP has been lessened with adequate prophylaxis, other causes of interstitial pneumonitis must be considered. Causative microorganisms include 1) atypical bacteria-- *Mycoplasma* and*Legionella* species; 2) viruses—EBV, CMV, varicella, respiratory syncytial virus (RSV), and influenza species; and 3) fungi, especially *Candida* and *Aspergillus* species. Interstitial pneumonitis may also be a direct result of chemotherapy with bleomycin, cyclophosphamide, methotrexate or cytosine arabinoside, or irradiation of the lungs.

PCP presents with fever, cough, tachypnea, and hypoxemia. The degree of hypoxemia is often out of proportion to the adventitial sounds heard on chest examination. Initially, the chest radiograph may be normal in PCP, but a diffuse increase in interstitial markings is usually evident by 24 hours after the onset of symptoms (Fig. 100.8). *Mycoplasma* or viral infection may present in a similar manner, although a history of upper respiratory tract symptoms suggests these diagnoses.*Mycoplasma* and RSV infection in infants may be associated with wheezing. Aspergillus infection can present as a diffuse interstitial pneumonia in the neutropenic patient or with a fungus ball (Fig. 100.9) and hemoptysis in a patient who has had an adequate number of neutrophils to localize the infection. Pleuritic chest pain is sometimes present in fungal pneumonia, probably a result of the presence of pleural-based lesions.

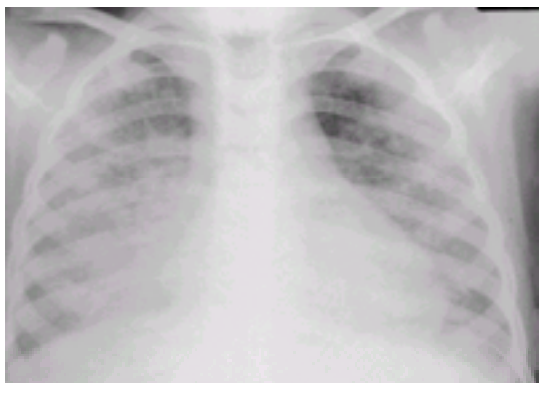

FIGURE 100.8. *Pneumocystis carinii* pneumonitis in a 6-year-old boy with a 24-hour history of fever and cough. Chest radiograph shows bilateral alveolar infiltrates most prominent at the hila. (Courtesy of Dr. Spencer Borden, Department of Radiology, Children's Hospital of Philadelphia.)

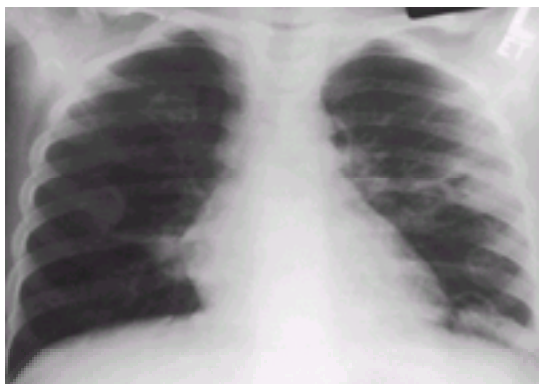

FIGURE 100.9. Fungal pneumonitis in a 6-year-old boy with acute lymphoblastic leukemia; he had received 60 mg/m ^{2} per day of prednisone and broad-spectrum antibiotics and had persistent fever. Physical examination was remarkable for hepatosplenomegaly and normal heart and lungs. Chest radiograph shows bilateral cavitary lesions. (Courtesy of Dr. Spencer Borden, Department of Radiology, Children's Hospital of Philadelphia.)

Interstitial pneumonitis may be a manifestation of bacterial pneumonia in the severely neutropenic patient. It may also follow septicemia by 24 to 48 hours and be a harbinger of adult respiratory distress syndrome.

The evaluation of the oncology patient with interstitial pneumonitis should include a CBC with differential, liver function tests in patients suspected of having disseminated viral infections, blood culture, and sputum culture when feasible. Pulse oximetry should be used to monitor for hypoxemia. CMV and fungi may be grown from urine; urine viral and fungal cultures should be sent if these organisms are suspected. Serologic testing for specific viruses, such as CMV and EBV, may be useful.

Varicella and herpetic pneumonia are rare in the absence of skin or mucosal lesions. When skin lesions are present, scrapings from the base of a vesicle should be submitted for Tzank prep, viral culture, or rapid immunologic detection tests. If no diagnosis is evident from initial noninvasive tests, early bronchoscopy with bronchoalveolar lavage is indicated. Open lung biopsy or thoracoscopic biopsy is indicated when no diagnosis is evident following bronchoscopy.

In the neutropenic patient, initial evaluation and therapy should be directed toward the identification and treatment of presumptive bacterial infection. Treatment guidelines are presented in the previous section on the febrile neutropenic patient. Additional treatment of the neutropenic patient, and initial treatment of the nonneutropenic patient, should be directed toward the most likely organism. If a patient is at risk for PCP and is not receiving systemic prophylaxis, TMP-SMZ (20 mg/kg per day of TMP) should be given. Prednisolone 2 mg/kg per day given intravenously or orally has proven beneficial as an adjunct to anti-*Pneumocystis* therapy in patients older than 13 years of age with human immunodeficiency virus (HIV) infection and Pa_{O_2} less than 70 mm Hg on room air. Intravenous acyclovir is given when herpes (750 mg/m^2 divided every 8 hours) or varicella (1500 mg/m^2 divided every 8 hours) virus is evident or suspected. Ganciclovir (2 to 5 mg/kg every 8 hours) and intravenous gamma-globulin are used for proven CMV infection or when CMV is highly suspected, as in patients who had a bone marrow transplant 1 to 3 months prior. Early treatment with erythromycin is rarely contraindicated.

Supplemental oxygen should be used when transcutaneous oxygen saturation is less than 96%. Packed RBC transfusion is indicated in the setting of hypoxemia and anemia. Ventilatory support should be provided, except when its use is precluded by prior discussions with the family of a terminally ill patient.

Chemotherapy-induced and radiation therapy–induced pneumonitis are diagnoses of exclusion. The emergency physician should concentrate on the identification and treatment of possible infectious causes.

Varicella and Herpes Virus Infection

Up to half the children with Hodgkin's disease develop varicella-zoster during or after therapy. About 1% develop lethal visceral dissemination. In patients with ALL, varicella has replaced PCP as the most common cause of death during remission. Acyclovir 1500 mg/m^2 per day intravenously divided every 8 hours can shorten the course of the illness and lessen the risk of dissemination in the immunocompromised patient.

Any immunocompromised patient who has a vesicular rash should be admitted for intravenous treatment with acyclovir. Oral treatment is inadequate. All chemotherapy should be discontinued. On admission, an appropriate diagnostic test (viral culture or rapid detection test) should be done because herpes simplex virus (HSV) can present as a vesicular skin eruption in the immunocompromised host; for HSV, a lower dosage of acyclovir is used (750 mg/m^2 per day divided every 8 hours). Interstitial pneumonitis may occur in these patients, so a chest radiograph should be obtained when respiratory signs and symptoms are present. Initial liver function tests, to detect evidence of viral hepatitis, and renal function tests as a baseline for monitoring acyclovir toxicity, should be obtained.

Hodgkin's disease and ALL patients are at the highest risk for varicella infection, but any patient with significant immunosuppression and evidence of varicella infection should be treated in the manner previously described. Patients with intraoral or labial HSV infection may also require intravenous acyclovir if they are leukopenic and ill with their infection. Oral acyclovir is sometimes used to suppress HSV reactivation during times of severe leukopenia in patients with a history of HSV infection.

Care should be taken to isolate oncology patients from other ED patients with active varicella or a history of exposure to

the virus. If a child without a history of previous varicella infection or documented titers before treatment is exposed to varicella, varicella-zoster immunoglobulin (VZIG) should be given (125 units/10 kg to a maximum of 625 units) by intramuscular injection. VZIG is most effective if given within 96 hours of exposure. The patient's oncologist should be consulted regarding discontinuation of chemotherapy during the postexposure period.

Postsplenectomy Sepsis

Laparotomy with splenectomy is a part of the diagnostic evaluation of Hodgkin's disease in many pediatric oncology centers. All patients should be given pneumococcal and *Haemophilus influenzae* type b vaccines before splenectomy. However, many patients do not respond adequately to these vaccines. Although penicillin or erythromycin prophylaxis (250 mg orally twice daily) is prescribed for these patients, sepsis from encapsulated organisms not covered by the vaccines or the prophylactic antibiotics can be life-threatening.

In the past, hyperacute infection occurred in as many as 10% of children with Hodgkin's disease. Most of these children had advanced disease, had experienced multiple relapses, or had received no prophylaxis. Hyperacute infection is less common today. Children with hyperacute infections develop high fever and cardiovascular collapse, followed by death in at least half the patients. If a patient who has undergone splenectomy or who has had more than 3600 cGy of splenic irradiation suddenly develops high fever and appears acutely ill, blood and urine cultures should be obtained, and cefotaxime (100 mg/kg per day) should be started immediately. Careful attention should be given to maintenance of adequate perfusion see Chapter 3). If the patient is neutropenic as a result of therapy, antibiotic coverage should be administered as described in the previous section about the febrile neutropenic patient.

Common Childhood Infections

Children with cancer acquire common pediatric viral and bacterial infections. For the child who is receiving maintenance therapy for ALL, or who is not neutropenic from therapy for other malignancies, diseases such as otitis media, urinary tract infection, streptococcal pharyngitis, impetigo, infectious mononucleosis, mumps, influenza, and the ubiquitous upper respiratory tract infections may be managed as they would be in an otherwise healthy child. However, varicella and measles (rubeola) infection are dangerous in oncology patients regardless of the status of their disease. Varicella should be treated as described in the previous section on varicella and herpes virus infection.

Measles outbreaks have become more prevalent in recent years, placing oncology patients at higher risk of exposure to the disease. Measles can result in fatal pneumonia in the immunocompromised host, gamma-globulin (0.1 mL/kg intramuscularly) provides some protection from measles and should be given to an exposed patient. Measles vaccine is contraindicated during therapy, although patients who were adequately vaccinated before therapy usually retain their immunity.

Measles–mumps–rubella vaccine may be used safely in the contacts of immunocompromised patients; its use should be encouraged in families with a child who is being treated for cancer. Families of children with cancer should also be encouraged to get an influenza vaccination during the course of their child's treatment.

Neurologic Complications

Neurologic complications of cancer therapy may be caused by the compression of vital structures of the CNS by growing tumor or the effects of therapy. Radiation therapy to the brain or spinal cord may have direct effects on CNS function. Chemotherapy may act directly to cause seizures or encephalopathy, or indirectly by causing electrolyte imbalance or a predisposition to thrombosis. In addition, other drugs used during the course of cancer treatment may cause alterations in CNS function: antiemetics may cause extrapyramidal symptoms, headache, or encephalopathy; amphotericin B can cause severe electrolyte imbalance, which may result in CNS symptoms.

Seizures

Seizures in a child with cancer should be evaluated and treated in much the same way as outlined in Chapter 70. A careful history should be taken to see whether the child has a ventriculoperitoneal shunt, what medicines the child is taking, and whether the child has had radiation to or surgery on the brain or spinal cord.

Metabolic abnormalities account for 7% of seizures in children with cancer. Cisplatin, ifosfamide, and amphotericin B can all cause renal wasting of electrolytes, especially potassium, phosphorus, magnesium, and calcium. A rapid assessment of potassium, calcium, and magnesium levels is essential in patients who have received these drugs.

Vincristine, intrathecal methotrexate, and cytosine arabinoside may directly cause seizures. L-Asparaginase predisposes cancer patients to CNS thrombosis, which may result in seizures. Therefore, knowing the child's recent chemotherapy history is essential.

A child in whom a shunt malfunction or primary or metastatic tumor in the CNS is a possibility must have imaging studies performed to localize the problem. Infectious meningitis is rarely a problem except in the child who has a CNS foreign body or who has had a recent lumbar puncture.

Altered Mental Status

A few unique circumstances must be considered when evaluating the cancer patient with coma or encephalopathy. Progressive disease must always be considered in the differential diagnosis. The emergency physician must be concerned with a cerebrovascular accident in those with thrombocytopenia, DIC, or a thrombotic tendency secondary to L-asparaginase use. Viral encephalitis with HSV or varicella is of particular concern in the immunocompromised host.

Ifosfamide, BCNU, 5-fluorouracil, thiotepa, high-dose cytosine arabinoside, and high-dose methotrexate may cause acute coma or encephalopathy.

Phenothiazines are often used as antiemetics and may cause extrapyramidal symptoms, somnolence, or coma. Occasionally, a patient who is using a scopolamine patch for control of emesis will leave a patch in place long enough to cause encephalopathy. Removal of the patch reverses the problem over a few hours.

Many terminally ill patients are maintained on home infusions of opioid narcotics or are taking high-dose oral narcotics for cancer pain. Care should be taken not to rapidly reverse the effects of these drugs with naloxone because acute withdrawal symptoms can be precipitated. Supportive care and gradual withdrawal of the opioid is indicated if coma or encephalopathy occur.

Cerebrovascular Accidents

In children with cancer, a cerebrovascular accident (CVA) may result from direct or metastatic spread of tumor, the effects of chemotherapy or radiation, or as a result of a bleeding diathesis associated with thrombocytopenia or DIC (secondary to leukemia or sepsis). Patients with acute leukemia and hyperleukocytosis are at risk of CVA secondary to leukostasis. The treatment of hyperleukocytosis is discussed in the previous ection on leukemia. Acute promyelocytic leukemia and acute monocytic leukemia predispose to stroke by causing DIC. Treatment with fresh-frozen plasma is discussed in the previous section on leukemia.

L-Asparaginase has a direct effect on anticoagulant factor production. Patients who are receiving the drug three times per week during induction therapy for ALL must be considered at high risk for thrombotic stroke. If stroke occurs, antithrombin III, protein S, and protein C levels should be measured and the patient should then be given 10 mL/kg of fresh-frozen plasma twice daily until the patient is able to produce normal amounts of these factors.

Spontaneous hemorrhagic stroke secondary to thrombocytopenia is rare when the platelet count is maintained at greater than 20,000/mm^3 using platelet concentrates. If intracranial hemorrhage does occur in the thrombocytopenic patient, the platelet count should be raised to greater than 80,000/mm^3 with platelet concentrates. Platelet-refractory patients present a difficult problem. Plasmapheresis, intravenous gamma-globulin, and human lymphocyte antigen (HLA)-matched platelets have varying success in maintaining an adequate platelet count.

CT scan with contrast should be taken for all oncology patients suspected of having a CVA. Although CT may not reveal an early stroke, it will identify bleeding or a progressive tumor that may be the cause of neurologic symptoms. A CT scan repeated at 7 to 10 days after the acute event usually identifies the lesion. If the patient is stable enough to complete the examination, MRI will often be able to identify a stroke that is not apparent on CT scan. MRI offers the advantage of magnetic resonance angiography, which may obviate the need for invasive angiography.

Spinal Cord Compression

Spinal cord compression is almost always a result of direct invasion of tumor into the spinal canal. Occasionally, a patient taking high-dose corticosteroids may develop nerve root compression after the collapse of an osteopenic vertebral body. Radiation-induced spinal cord injury is rare but should be considered in the differential diagnosis if the patient received more than 4500 cGy to the cord.

Careful neurologic examination should include an assessment of sacral nerve function (anal tone and anal wink, postvoiding bladder residual). A spinal sensory level should be sought. A spinal cord lesion may often be localized by percussion tenderness or pain over the involved vertebral segment(s). MRI is the best technique for definitive localization of the lesion.

Spinal cord compression is treated with surgery (see Fig. 100.2 and Chapter 125), radiotherapy, or chemotherapy. The choice of treatment is determined by the type of tumor and the duration and severity of symptoms. Dexamethasone should be given to all patients at an initial dose of 1 to 2 mg/kg intravenously. The restoration of bowel and bladder continence can have a profound impact on quality of life. Therefore, even in a terminally ill patient, spinal cord compression should be considered an emergency.

Gastrointestinal and Genitourinary Complications

Mucositis and Esophagitis

Chemotherapy, especially with methotrexate, high-dose cytosine arabinoside, and anthracyclines can cause mucosal injury throughout the GI tract. Breaks in the mucosal barrier, added to the effects of myelosuppression, predispose patients to fungal and viral infections of the oral and GI mucosa. Radiation therapy to the head, neck, and mediastinum also commonly results in oral or esophageal mucositis. Oral and esophageal mucositis may interrupt normal nutrition and require hospitalization for intravenous fluids, hyperalimentation, and pain relief. Treatment strategies for acute stomatitis are outlined in Table 100.5. *Candida* and HSV should be suspected and treated aggressively if found.

1. Cleanse 2–3 times/day with gauze or disposable sponge brush; use ½ tsp of sodium bicarbonate/cup of water.
2. Use 0.1% chlorhexidine gluconate mouthwash after each cleansing.
3. Avoid commercial mouthwashes.
4. Topical agents used 3–4 times/day:
• Dyclonine HCl 0.5% applied on a cotton applicator or as a spray
• Diphenhydramine or hydroxyzine applied on a cotton applicator (using maximum dose appropriate for age)
• 1:1 mixture of Maalox and diphenhydramine or hydroxyzine swish and spit or applied with cotton applicator
5. Individual lesions: Apply Orabase with benzocaine or Kenalog as needed.
6. Admit for intravenous fluids, hyperalimentation, and analgesia if intake inadequate.
7. Culture suspicious lesions for HSV; KOH preparation for yeast.
8. Treat oral candidiasis (mycostatin, fuconazole) and HSV (acyclovir) aggressively.

HSV, herpes simplex virus; *KOH*, potassium hydroxide.

Table 100.5. Treatment of Acute Stomatitis

Esophagitis presents with dysphagia and substernal chest pain. Fever and acute stomatitis may or may not be present. The diagnosis may be confirmed with barium swallow (Fig. 100.10) or endoscopy. Endoscopy offers the advantage of definitive diagnosis of candidal or herpetic lesions and the ability to examine the gastric and duodenal mucosa during the same procedure.

FIGURE 100.10. Esophageal candidiasis in a 16-year-old boy with acute lymphocytic leukemia who was receiving corticosteroids and broad-spectrum antibiotics. Barium swallow shows ulceration and dissection of mucosa of distal esophagus. (Courtesy of Dr. Spencer Borden, Department of Radiology, Children's Hospital of Philadelphia.)

Esophagitis in an otherwise well patient who can still swallow may be treated using oral ketoconazole (3.3 to 6.6 mg/kg per day) or fluconazole (100 to 200 mg/day). Patients who are neutropenic, febrile, or unable to maintain adequate oral intake should be admitted for intravenous fluids, alimentation, and a 7 to 10 day course of amphotericin B.

Abdominal and Rectal Pain

Abdominal pain is a common complaint in children with cancer. Pain may be caused by inflammation in ny part of the abdominal viscera. Acute abdominal pain may be caused by the usual disorders seen in childhood (e.g., gastroenteritis, appendicitis), but some problems deserve special consideration in oncology patients. Table 100.6 lists these problems, the clinical setting in which they are seen, and the appropriate course of action to be taken.

Problem	Clinical Setting	Action
Stomach pain	Chemotherapy-induced gastritis Corticosteroid-induced gastritis	Trial of oral antacid
Small bowel obstruction	Progressive tumor Postoperative adhesions	Abdominal radiographs Nasogastric drainage Surgical consultation
Right lower quadrant pain/typhlitis	Severe myelosuppression, especially acute myelogenous leukemia	Abdominal radiographs (look for pneumatosis) Nothing-by-mouth status and nasogastric drainage Antibiotics (Gram negative, anaerobic, and fungal coverage) Surgical consultation
Anorectal pain/perirectal abscess	Severe myelosuppression	Rectal examination by pediatric surgeon Antibiotics (Gram negative and anaerobic coverage) Sitz baths
Abdominal pain, vomiting hypotension, and increased amylase/lipase (pancreatitis)	L-Asparaginase use Corticosteroid use	Ultrasound examination Nothing-by-mouth status and nasogastric drainage
Constipation/ileus	Vincristine use, hypokalemia Narcotic-induced	Increased fluids Oral stool softeners and laxatives Suppository/enema if no results with oral therapy (consult oncologist before giving enema) Check serum potassium

Table 100.6. Abdominal Pain in Children with Cancer

Gastritis caused by chemotherapy or corticosteroid use is common in oncology patients. Oral antacids usually provide symptomatic relief. Small bowel obstruction is rare in patients with cancer, but it occasionally results from progressive tumor or postoperative adhesions.

Patients who are receiving weekly doses of vincristine are prone to ileus and constipation. Hypokalemia caused by renal electrolyte loss also causes constipation in the patient who has had cisplatin, ifosfamide, or amphotericin B. Pain can be

severe, and the risk of infection is increased in the patient with severe ileus or obstipation. Stool softeners (e.g., docusate sodium), increased fiber in the diet, and increased fluid intake help prevent constipation. However, if severe ileus or obstipation occurs, rapid resolution with oral laxatives or enemas is important. Enemas should be used only in the most recalcitrant cases and only after consultation with the patient's oncologist.

Typhlitis and perirectal abscess are seen in the setting of neutropenia. Typhlitis is a necrotizing colitis of the cecum seen almost exclusively in patients with AML. It presents with right lower quadrant pain, tenderness on palpation, and fever. Plain films of the abdomen may show ileus, thickened bowel wall, and air in the bowel wall. Ultrasound examination will confirm thickening of the cecal wall. The mortality rate ranges from 50 to 100% regardless of treatment. A patient suspected of having typhlitis should be allowed nothing by mouth and be placed on broad-spectrum antibiotic coverage that includes coverage for anaerobic bacteria.

Perirectal abscess presents with anorectal pain, pain on defecation, a frank abscess on external or rectal xamination, or perirectal cellulitis and fever. Therapy is antibiotics, including coverage for anaerobic bacteria, and sitz baths. Stool softeners should be used to ease the pain of defecation and to help prevent future episodes. Although a pediatric surgeon should be consulted, many patients improve with medical therapy.

Pancreatitis must be considered in a patient taking L-asparaginase or corticosteroids who presents with abdominal pain and vomiting. Determination of the amylase and lipase and ultrasound examination will confirm the diagnosis. The treatment is the same as for any patient with acute pancreatitis.

Hemorrhagic Cystitis

Hemorrhagic cystitis is a complication of treatment with ifosfamide and high dosages of cyclophosphamide. The metabolism of these drugs produces an acrolein dye that is excreted in the urine. Contact between the dye and the bladder wall may lead to mucosal inflammation and resultant bleeding. Hemorrhagic cystitis has become much less common with the use of the uroprotectant MESNA (sodium 2-mercaptoethane sulfonate).

A child with hemorrhagic cystitis presents with pain on voiding and either microscopic or gross hematuria. Lower abdominal pain, when present, represents bladder wall irritation or spasm. Occasionally, the patient passes large clots. Urinary tract infection should be ruled out, especially in the febrile or neutropenic patient. The platelet count and clotting times should be checked and corrected if abnormal.

Initial treatment of microscopic or uncomplicated gross hematuria consists of hydration with 1.5 to 2 times maintenance fluids and frequent voiding. A pediatric urologist should be consulted to remove large clots and to assist with further management if bleeding does not abate with medical therapy. Belladonna and opium suppositories or oral oxybutynin chloride (Ditropan 5 mL twice daily in children older than 5 years of age) will help control bladder spasm and pain. MESNA has no role in the treatment of established hemorrhagic cystitis.

Metabolic Complications

Electrolyte Abnormalities

Patients receiving certain chemotherapeutic agents or amphotericin B may have renal wasting of electrolytes, which requires acute treatment. Tumor lysis syndrome is discussed in the previous section on leukemia (Table 100.3).

Amphotericin B commonly causes moderate to severe hypokalemia. Cisplatin causes loss of calcium, phosphorus, and magnesium. Renal wasting is the mechanism of electrolyte loss for both of these drugs. Patients who are being treated with these drugs are usually taking oral electrolyte supplementation. However, decreased oral intake or noncompliance, especially in adolescent patients, can result in symptomatic electrolyte disturbances.

Hypokalemia can cause symptomatic ileus or cardiac arrhythmias. Bolus intravenous doses of potassium should be reserved for the patient with electrocardiogram (ECG) changes. Otherwise, potassium should be replaced by increased oral supplementation or constant intravenous infusion.

Of the electrolyte problems that cisplatin causes, hypocalcemia is most often associated with symptoms. If tetany or cardiac arrhythmias occur, intravenous calcium should be given (calcium gluconate 100 mg/kg per dose every 6 hours) until the ionized calcium is normal. Cisplatin also causes magnesium wasting; magnesium must also be corrected in the patient with symptomatic hypocalcemia. In these patients, hypomagnesemia and hypophosphatemia are rarely symptomatic before the onset of symptomatic hypocalcemia.

Hyperglycemia

L-Asparaginase or corticosteroids may cause hyperglycemia. L-Asparaginase inhibits production of insulin; corticosteroids stimulate glucose production through catabolic pathways. During induction therapy for ALL, L-asparaginase and corticosteroids are often used in combination, and hyperglycemia is common. Conservative dietary measures—small, frequent meals and decreased concentrated sugar—usually control the transient, mild hyperglycemia most often seen with these drugs. However, when the serum glucose is greater than 250 g/dL or glycosuria or ketonuria are found, a diabetic diet should be instituted.

Patients who are unresponsive to dietary management require insulin. Initial insulin doses should be small because some insulin reserve exists. After a small dose of insulin is given, the patient should be monitored for the recurrence of urine glucose/ketones or a rise in the blood glucose above 250 g/dL before subsequent doses are administered. A single, small dose often provides reasonable control for many hours. Tight diabetic control is not the goal in these

patients because the problem is transient, and overtreatment can result in hypoglycemia. Diabetic ketoacidosis is rare; it should be treated in the usual manner if it occurs (see Chapter 97).

Syndrome of Inappropriate Antidiuretic Hormone

In children with cancer, the syndrome of inappropriate antidiuretic hormone (SIADH) occurs for reasons unrelated to therapy (e.g., stress, pulmonary disease, primary CNS disease) or is a side effect of vincristine or cyclophosphamide treatment. In patients with CNS tumors or renal tubular damage, salt-wasting syndromes must be considered. Hypotonic dehydration must be considered in the differential diagnosis in young children with severe diarrhea. The mainstay of treatment is fluid restriction (see Chapter 97).

Cardiovascular Complications

Superior Vena Cava Syndrome

SVC syndrome occurs in patients with anterior mediastinal tumors. This problem is discussed in the previous section on thoracic tumors.

Cardiac Failure and Rhythm Disturbances

Anthracyclines (adriamycin and daunomycin) can cause rhythm disturbances, cardiomyopathy, and cardiac failure. These problems are related to total dose and increase in frequency with total doses of more than 450 to 500 mg/m ^{2}. Radiation alone or in combination with anthracycline may contribute to cardiac damage. Arrhythmias and conduction abnormalities are the most common acute toxicities of anthracyclines, but carditis-pericarditis syndrome and congestive failure are also seen. Myocardial and pericardial fibrosis may follow large doses of radiation. Chronic cardiomyopathy may be mild or severe; patients may be asymptomatic for many years and then present with congestive failure.

In the anthracycline-treated patient who is in shock and who does not respond to initial fluid resuscitation, cardiogenic shock must be considered. The initial treatment in the acutely ill patient is with oxygen, afterload reduction with diuretics or vasodilators (e.g., furosemide or nitroprusside), and inotropic agents (e.g., dobutamine, amrinone).

In patients who have anthracycline-induced heart damage, hypokalemia or hypocalcemia may exacerbate the problem. Patients who have not had anthracyclines may still have rhythm disturbances on the basis of potassium or calcium abnormalities. The severity of the rhythm disturbance determines the rate at which the electrolyte disturbance is corrected.

Post-Bone Marrow Transplantation

Bone marrow transplantation is increasingly used to treat many hematologic, oncologic, or immunologic diseases. In hematologic malignancies (leukemias), allogeneic marrow transplantation may follow initial remission induction or disease relapse. This approach requires the use of a histocompatible related or unrelated donor. The patient's own marrow is ablated using chemotherapy alone or with radiotherapy, and donor marrow is transfused to reconstitute the hematopoietic system. Allogeneic transplant may result in the complication of graft-versus-host disease.

Graft-versus-host disease (GVHD) may develop as newly engrafted immune cells of the donor react against tissue antigens of the recipient which are perceived to be “foreign.” GVHD risk increases for donor grafts which are less tissue compatible but can occur even in HLA-identical allografts. Acute GVHD causes skin, gastrointestinal, and liver disease and generally has onset at the time of engraftment during hospitalization. Therapy is primarily immunosuppressive using corticosteroids and/or cyclosporine. The ER physician should be aware of these complications as patients may present with exacerbations of acute GVHD. Chronic GVHD involves dermal, hepatic, ocular, pulmonary, gastrointestinal, and neuromuscular systems. Children with chronic GVHD have severe immunologic dysfunction and are at risk of acute hyperinfection with encapsulated organisms (see Postsplenectomy Sepsis, above).

Children with solid tumors which are high-risk for conventional treatment failure, or who have recurrent or refractory disease, may benefit from autologous transplantation. The patient's own marrow is harvested and used to reconstitute their hematopoietic system (bone marrow rescue) following myeloablative chemotherapy. Since the patient's own bone marrow is reinfused, there is no risk of graft-versus-host disease.

Infectious complications of marrow transplantation result from the extreme immunosuppression achieved by myeloablation, the cutaneous and mucosal barrier damage, and the immunologic immaturity of the transplanted marrow. Patients are at risk of exogenous and reactivated endogenous viral infections, including cytomegalovirus or herpes pneumonitis, varicella-zoster, and Epstein-Barr posttransplant lymphoproliferative disease. Adenovirus and cryptosporydium may cause severe diarrhea. Hyperacute pneumococcal sepsis may occur in the first posttransplant year.

Chemotherapy

Table 100.7 lists antineoplastic agents, their uses, and side effects. Most of these agents and irradiation cause some myelosuppression or immunosuppression, and most, except corticosteroids, vinca alkaloids, and bleomycin, cause nausea and vomiting.

Table 100.7. Cancer Chemotherapy

Attempts to control nausea and vomiting are made at the time of drug administration. Ondansetron (0.15 mg/kg intravenously or orally given 30 minutes before and 2 and 6 hours after chemotherapy) is a new serotonin antagonist that provides effective prevention of emesis without sedation. The phenothiazines (promethazine or chlorpromazine 0.25 to 0.50 mg/kg per dose every 4 to 6 hours orally, intravenously, or rectally) and metoclopramide (1 to 3 mg/kg per dose intravenously) are useful in the prevention and control of emesis, but they cause sedation and extrapyramidal symptoms. Diphenhydramine or hydroxyzine (1.0 to 1.25 mg/kg per dose orally or intravenously) are used to control extrapyramidal symptoms.

Pain Management

Pain in children with cancer may be acute and may be the first symptom of the disease or of recurrence of disease. When pain is acute, it is most appropriately managed by diagnosing its cause and treating the cause specifically with surgery, chemotherapy, or irradiation. If a delay in diagnosing the cause and initiating therapy occurs, analgesics are necessary to treat the pain.

Chronic pain in a child with cancer is often difficult to manage, but a child with cancer should never be allowed to suffer. Chronic pain may be caused by several mechanical problems: compression of a nerve by a mass or edema, spasm or compression around a vessel, obstruction of a viscous organ, or distension of the marrow space and subperiosteal hemorrhage. These physical components of pain are complicated by the psychologic experience. Pain itself is frightening, and many children with cancer and their families are frightened both by the suffering and by the association of cancer pain with cancer death. The physician can relieve some of the anxiety by explaining that she or he will look for the cause of the pain and will make every attempt to relieve the pain within a short time. Relaxation and hypnotic techniques are successful in managing chronic pain, but emergency facilities are not often prepared to provide impromptu services of this sort. As with acute pain, a specific cause should be sought. Compression of a nerve may be relieved with corticosteroids or radiation and, if the child is not terminally ill, possibly by surgery. The pain of pathologic fractures can be relieved by immobilization but usually requires narcotic analgesia. Obstruction of a viscous organ may require surgery, unless it appears to be an imminently fatal event in a child who is known to be terminally ill. The extent to which specific measures are appropriate should be clarified with the patient's physician and the family.

If no analgesics have been used and the pain is mild, acetaminophen is usually the preferred analgesic. Aspirin is contraindicated in most cancer patients because it interferes with platelet function. If acetaminophen alone is insufficient, acetaminophen plus codeine (3 mg/kg per day) should be tried for moderate pain. Patients should be warned about the constipating effects of codeine, morphine, or morphine analogues. The dosages, routes of administration, and duration of action of analgesics are listed in Table 100.8. These recommendations are standard; however, they are often considerably less than the dosages needed to relieve pain in a terminally ill cancer patient. Tolerance is acquired rapidly, and pain often breaks through previously successful dosages. Studies of pain management in cancer patients invariably show that physicians use too little medication because of fear of causing addiction and because of conservative recommendations in the available literature. Addiction is not a problem in the terminally ill patient. The most important rule in the management of cancer pain is that the dose of narcotic necessary to relieve the pain is that which relieves it successfully.

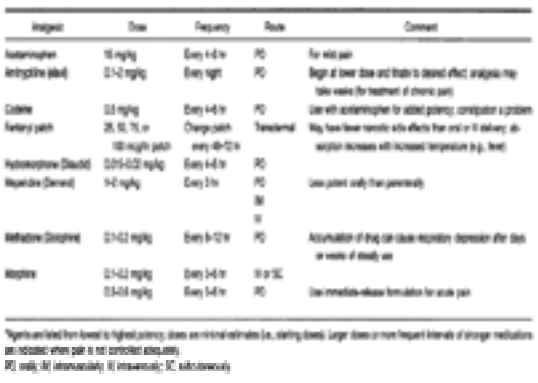

Table 100.8. Analgesics for Use in Acute and Chronic Cancer Pain[a]

If a child is in severe pain, he or she should not be discharged from the ED without control of the pain or without a plan

for means to control the pain. The plan may require admission for titration of narcotics, sedatives, and antiemetics, or consideration of nerve blocks, intrathecal morphine, or even rhizotomy or chordotomy. Alternatively, the plan may involve communication with a hospice facility in the child's home town. Hospice facilities vary in their willingness and capacity to deal with dying children, but in the recent past, many have been able to care for the dying child at home in a medically compassionate and philosophically appropriate way. See Chapter 126 for a discussion of the psychosocial aspects of pain.

Suggested Readings

GENERAL

Bleyer WA: Impact of childhood cancer in the United States and the world. CA 1990;40:355–367.

Foley GV, Hannigan-Whittam E: Care of the child dying with cancer. CA 1990;40:327–354.

Hutton JH: Principles of total care: Physiologic support. In Fernbach DJ, Vietti TJ (eds): Clinical pediatric oncology, 4th ed. St Louis, Mosby-Year Book, 1991:231–248.

Kelly KM, Lange B: Oncologic emergencies. Pediatr Clin N Am 1997;44: 809–830.

Lange B, O'Neill JA Jr, Goldwein JW, Packer RJ, Ross AJ III: Oncologic emergencies. In: Pizzo PA, Poplack DG, eds: Principles and practice of pediatric oncology. Philadelphia, JB Lippincott, 1997:1025–1049.

LEUKEMIA

Bunin NJ, Pui C-H. Differing complications of hyperleukocytosis in children with acute lymphoblastic and acute nonlymphoblastic leukemia. J Clin Oncol 1985;3:1590–1595.

Ebb DH, Weinstein JH: Diagnosis and treatment of childhood acute myelogenous leukemia. Pediatr Clin N Am 1997;44:847–861.

Maurer HS, Steinherz PG, Gaynon PS, et al. The effect of initial management of hyperleukocytosis on early complications and outcome of childhood acute lymphoblastic leukemia. J Clin Oncol 1988;6:1425–1432.

Pui C-H: Acute lymphoblastic leukemia. Pediatr Clin N Am 1997;44: 831–846.

NON-HODGKIN'S LYMPHOMA

Sandlund JT, Downing JR, Crist WM: Non-Hodgkin's lymphoma in childhood. N Engl J Med 1996;334:1238–1248.

HODGKIN'S LYMPHOMA

Hudson MM, Donaldson SS: Hodgkin's disease. Pediatr Clin N Am 1997;44:891–906.

HISTIOCYTIC DISORDERS

Favara BE, Feller AC, with members of the WHO committee on histiocytic/reticulum cell proliferations: Contemporary classification of histiocytic disorders. Med Pediatr Oncol 1997;29:157–166.

Imashuku S: Differential diagnosis of hemophagocytic syndrome: underlying disorders and selection of the most effective treatment. Int J Hematol 1997;66:135–151.

THORACIC TUMORS

Ingram L, Rivera GK, Shapiro DN: Superior vena cava obstruction syndrome associated with childhood malignancy: analysis of 24 cases. Med Pediatr Oncol 1990;18:476–481.

WILMS' TUMOR

Green DM: Wilms' tumor. Eur J Cancer 1997;33:409–418.

NEUROBLASTOMA

Brodeur GM, Pritchard J, Berthold F, et al: Revisions of the international criteria for neuroblastoma diagnosis, staging, and response to treatment. J Clin Oncol 1993;11:1466–1477.

HEPATIC TUMORS

Haas JE, Muczynski KA, Krailo, et al: Histopathology and prognosis in childhood hepatoblastoma and hepatocarcinoma. Cancer 1989;64:1082–1095.

TUMORS OF THE MALE AND FEMALE GENITALIA

Castleberry RP, Kelly DR, Joseph DB, Cain WS. Gonadal and extragonadal germ cell tumors. In Fernbach DJ, Vietti TJ (eds). Clinical pediatric Oncology, 4th ed. St Louis: Mosby-Year Book, 1991:577–594.

RETINOBLASTOMA

Abramson DH: Retinoblastoma 1990: Diagnosis, treatment, and implications. Pediatr Ann 1990;19:387–395.

THYROID CANCER

LaQuaglia MP, Telander RL: Differentiated and medullary thyroid carcinoma in childhood and adolescence. Semin Pediatr Surg 1998;6:42–49.

SOFT TISSUE SARCOMAS

Womer RB: Paediatric update: Soft tissue sarcomas. Eur J Cancer 1997;33:2230–2236.

BONE TUMORS

Grier HE: The Ewing's family of tumors: Ewing's sarcoma and primitive neuroectodermal tumors. Pediatr Clin N Am 1997;44:991–1004.

Meyers PA, Gorlick R: Osteosarcoma. Pediatr Clin N Am 1997;44:973–989.

CENTRAL NERVOUS SYSTEM TUMORS

Kun LE: Brain tumors: challenges and directions. Pediatr Clin N Am 1997;44:907–917.

HEMATOLOGIC COMPLICATIONS

Kobrinsky NL, Tulloch H: Treatment of refractory thrombocytopenic bleeding with 1-desamino-8- D-arginine vasopressin. J Pediatr 1988;6:993–996.

INFECTIOUS COMPLICATIONS

Hughes WT, Armstrong D, Bodey GP, et al: 1997 guidelines for the use of antimicrobial agents in neutropenic patients with unexplained fever. Clin Infect Dis 1997;25:551–573.

Pizzo PA: Management of fever in patients with cancer and treatment-induced neutropenia. N Engl J Med 1993;328:1323–1332.

Pizzo PA, Rubin M, Freifeld A, Walsh TJ: The child with cancer and infection. I. Empiric therapy for fever and neutropenia, and preventive strategies. J Pediatr 1991;119:679–694.

NEUROLOGIC COMPLICATIONS

DiMario FJ Jr, Packer RJ: Acute mental status changes in children with systemic cancer. Pediatrics 1990;85:353–360.

Packer RJ, Rorke LB, Lange B, et al: Cerebrovascular accidents in children with cancer. Pediatrics 1985;76:194–201.

GASTROINTESTINAL AND GENITOURINARY COMPLICATIONS

Nylander WA Jr: The acute abdomen in the immunocompromised host. Surg Clin N Am 1988;68:457–470.

West NJ: prevention and treatment of hemorrhagic cystitis. Pharmacotherapy 1997;17:696–706.

METABOLIC COMPLICATIONS

Kelly KM, Lange B: Oncologic emergencies. Pediatr Clin N Am 1997;44: 809–830.

Silverman P, Distelhorst CW: Metabolic emergencies in clinical oncology. Semin Oncol 1989;16:504–515.

CARDIOVASCULAR COMPLICATIONS

Giantris A, Abdurrahman L, Hinkle A, Asselin B, Lipshultz SE: Anthracycline-induced cardiotoxicity in children and young adults. Crit Rev Oncol Hematol 1998;27:53–68.

Helms Sr, Carlson MD: Cardiovascular emergencies. Semin Oncol 1989;16:463–470.

BONE MARROW TRANSPLANTATION

Sanders JE: Bone marrow transplantation for pediatric malignancies. Pediatr Clin N Am 1997;44:1005–1020.

CHEMOTHERAPY

Krakoff IH: Systemic treatment of cancer. CA Cancer J Clin 1996;46: 134–141.

Roila F, Ballatori E. The efficacy and cost-effectiveness of various antiemetic regimens. Curr Opin Oncol 1998;10:310–315.

PAIN

Berde C, Ablin A, Glazer J, et al: Report of the Subcommittee on Disease-related Pain in Childhood Cancer. Pediatrics 1990;86 (Suppl):818–825.

CHAPTER 101

Rheumatologic Emergencies

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JUVENILE RHEUMATOID ARTHRITIS

Background

Juvenile rheumatoid arthritis (JRA) is now the most common rheumatic disease of children in the developed world, having replaced acute rheumatic fever during the past 20 years. JRA occurs in all races and ethnic groups and, in the United States alone, may affect as many as 100,000 children.

The American College of Rheumatology criterion for diagnosing JRA is persistent unexplained arthritis of 1 or more joints lasting more than 6 weeks in children less than 16 years of age. The JRA diagnostic category thus includes a disparate group of syndromes characterized by chronic synovitis. Because no laboratory abnormalities are specific for JRA, the diagnosis is made clinically after exclusion of other conditions associated with chronic synovitis.

The cause of JRA is unknown. Although it has been reported to occur after trauma or systemic infections, extensive investigations attempting to correlate JRA with specific viral, bacterial, or mycoplasmal agents have not been conclusive. The presence of rheumatoid factor (RF) in the serum of some children with JRA, a lowered level of complement in the synovial fluid, and activated lymphocytes in the synovial tissue, suggest that immunologically mediated injury plays a role in the pathogenesis of this disease. There may also be genetic factors, as suggested by the association of JRA with particular histocompatibility antigens and cytokine alleles.

Pathophysiology

The pathogenesis of JRA may be divided into an initiating phase and a perpetuating phase. It appears that a variety of events, particularly viral infections, may trigger the articular inflammation. For unknown reasons, a process that is self-limited in most children leads to ongoing inflammation in genetically susceptible hosts. This inflammation is characterized by abnormal tissue and circulating levels of proinflammatory cytokines (including interleukin-1 [IL-1], interleukin-6 [IL-6], tumor necrosis factor [TNF], and interferon-g [IFN-g]), leading to activation of lymphocytes and infiltration of synovium.

Pathologically, vasculitis is prominent in early lesions. In established cases of arthritis, light microscopy of the synovium shows fibrin deposits, hyperplasia and hypertrophy of synovial lining cells, and an inflammatory cell response. Increased secretion of synovial fluid results in joint effusions. In uncontrolled and persistent arthritis, synovial villi project into the joint. Fronds of synovium may spread from the edges of the joint and overgrow the articular cartilage (pannus), causing destruction of the cartilage and eventually of the underlying bone.

Clinical Manifestations

A wide variety of demographic and clinical features may accompany the arthritis of JRA, so the condition has been divided into subtypes based on these factors and on the pattern of the disease during the first 6 months after onset. Until better means of classifying and distinguishing these subtypes of JRA become available, what is likely to be several discrete conditions will continue to be grouped on the basis of purely clinical features (Table 101.1).

Subgroup	% of JRA	Sex Ratio	Age at Onset	Joints Affected	Serologic and Genetic Test*	Extra-articular Manifestations	Prognosis
Rheumatoid positive polyarticular	10%	90% female	Late childhood	Any joints (typically hands, wrists	ANA 70% RF 100%	Low-grade fever, anemia, nodules, rheumatoid nodules	>50% severe arthritis
Rheumatoid negative polyarticular	20%	70% female	Younger onset	Any joints	ANA 80% RF negative	Low-grade fever, mild anemia, nodules, growth retardation	20-40% severe arthritis
Type I Pauciarticular	40%	90% female	Early childhood	Few large joints (hips and sacroiliac joints spared)	ANA 90%, RF negative	Few constitutional complaints, chronic iridocyclitis in 50%	Severe arthritis uncommon; 10-20% ocular damage from iridocyclitis if un-treated
Type II Pauciarticular	5%	90% male	Late childhood	Few large joints (hip and sacroiliac involvement common)	ANA negative, RF negative HLA-B27 70%	Few constitutional complaints, acute iridocyclitis in 5-10% during childhood	Clinically similar to spondyloarthritis
Systemic onset	20%	50% female	Any age	Any joints	ANA negative, RF negative	High fever, rash, or lymphadenopathy, polyserositis, hepatosplenomegaly, growth retardation	20% severe arthritis

*Modified with permission from Schaller J. In: Franklin EC, ed. Clinical Immunology. Update. New York: Elsevier North Holland, 1979.
ANA, antinuclear antibody; RF, rheumatoid factor; HLA-B27, histocompatibility antigen B27.

Table 101.1. Subgroups of Juvenile Rheumatoid Arthritis

Pauciarticular arthritis is defined as arthritis involving four or fewer joints. Type I pauciarthritis occurs more often in young girls and is the most common subtype of JRA, accounting for approximately half of all cases. Typically, it involves one or more large joints with swelling, pain, and limitation of movement. Antinuclear antibodies (ANAs) are detectable in the sera of more than 50% of these children, and its presence correlates with a higher risk for developing iridocyclitis.

Type II pauciarticular JRA occurs more often in preadolescent boys. Although they are often classified as having JRA, the predilection for axial involvement is more typical of spondyloarthritis. In fact, some of these children may develop ankylosing spondylitis on long-term follow-up. Typical of spondyloarthritis, the risk for developing chronic iridocyclitis is negligible, but 5 to 10% of these children may develop acute anterior uveitis.

Polyarticular arthritis (both RF positive and RF negative) occurs more commonly in girls than in boys. It is characterized by the insidious onset of symmetric synovitis in both large and small joints, accompanied by low-grade fever, morning stiffness, and malaise (Fig. 101.1). The presence of antibodies to native immunoglobulins in the serum (RF) corresponds to an increased risk of severe, erosive arthritis, as well as to the development of vasculitic complications and subcutaneous nodules. Cervical spine involvement occurs in approximately 30 to 50% of patients with this variety of arthritis, resulting in neck pain, stiffness, and torticollis. Unlike pauciarticular JRA, in which ocular involvement is the cause of the most significant morbidity, polyarticular disease may result in severe musculoskeletal disability. Thus, involvement of the temporomandibular joint may result in restricted ability to open the mouth, involvement of the hips may permanently affect ambulation, and small joint arthritis of the hands may compromise manual dexterity.

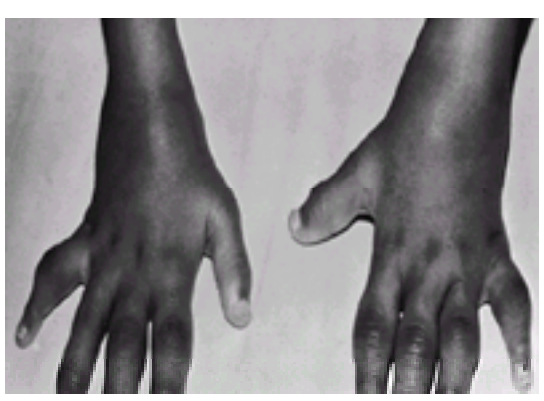

FIGURE 101.1. Symmetric involvement of large and small joints of the hands in a child with polyarticular arthritis.

The least common subtype of JRA is systemic-onset, or Still's disease. This subtype occurs most often in boys less than 5 years of age, although it has been reported even in adults. Clinically, these children often present with a fever of unknown origin; they may have high spiking temperatures (39° to 41°C [102.2° to 105.8°F]) for several weeks or months. Although the child often feels stiff and does not move normally, arthritis may not be a prominent feature at the onset of the disease. Diagnosis therefore generally involves excluding infectious and malignant conditions, especially sepsis, leukemia, and neuroblastoma. A characteristic salmon-pink evanescent maculopapular rash (Fig. 101.2), diffuse lymphadenopathy, and hepatosplenomegaly may also be present in the early stages, offering clues to the diagnosis. Arthralgias and myalgias are common, and pericarditis occurs most typically in this subtype of JRA. With time, systemic features of the disease become less prominent, and polyarticular arthritis becomes the major focus of management.

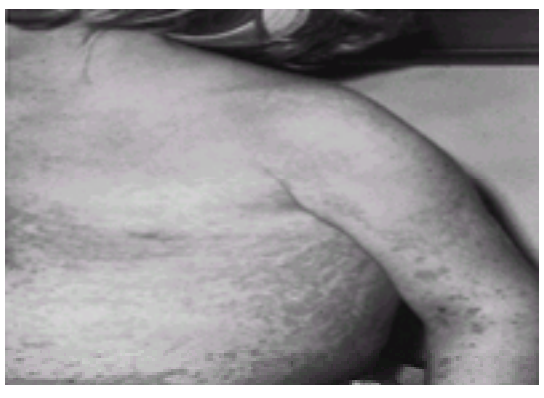

FIGURE 101.2. Macular rash in a child with systemic type of juvenile rheumatoid arthritis.

Laboratory and Radiologic Features

No laboratory test is diagnostic of JRA. JRA is a clinical condition diagnosed on the basis of characteristic findings on history and physical examination, although some laboratory studies may be suggestive of the diagnosis. In polyarticular JRA, one subgroup shows RF in the serum; no other pediatric rheumatologic disease typically has this marker. Mild to moderate anemia is common in all subtypes, particularly the systemic type. The white blood cell (WBC) count is often elevated, again most typically in the systemic type, in which leukemoid reactions may be seen. Platelet counts are often elevated, and urinalysis is usually normal. Levels of acute-phase reactants in the serum are elevated, often in proportion to the number of joints involved, and most prominently in systemic-onset disease. Complement levels may be normal or elevated, whereas immunoglobulins may be increased, leading to a reversal of the albumin:globulin ratio. ANA in the serum, particularly in children with pauciarticular arthritis, is an important marker for increased risk of developing iridocyclitis.

Radiographic features of JRA include soft-tissue swelling and periarticular osteopenia adjacent to affected joints. Later, narrowing of the joint space, bony cysts, erosions, subluxation, and ankylosis may be seen. In rare children in whom physical examination is difficult or inconclusive, ultrasonography may confirm the presence of a joint effusion, and magnetic resonance imaging (MRI) may show both synovial proliferation and increased fluid in the joint space.

Management

General Management

The major goal of therapy of children with JRA is to help the child and his or her family maintain as normal a life as possible. Emotional support, including the information that most children with this disease recover with minimal residual problems, provides reassurance. Simple measures, such as warm tub baths and the use of electric blankets at night, help control morning stiffness. For children with minimal joint involvement, regular daily activities, including participation in physical education classes, are to be encouraged, although high-impact activities should be avoided. In the presence of muscle wasting, weakness, or restricted range of motion in any joint, an active physical therapy program is indicated. Splinting may be used to rest actively inflamed, painful joints and to prevent worsening of deformities.

Nonsteroidal anti-inflammatory drugs (NSAIDs) are now the mainstay of therapy for children with JRA. Aspirin, formerly the preferred agent, has fallen into disfavor because of concerns about Reye syndrome, as well as the need for dosing four times daily. Agents that are approved for use in children include naproxen, ibuprofen, and tolmetin sodium (Table 101.2). These and other drugs in this class share similar toxicity profiles, with gastrointestinal (GI) disturbances being both the most common and the most troublesome.

Drug	Dosing Frequency	Dosage Range	Side Effects
Ibuprofen (Motrin, Advil, Pediaprofen)	TID-QID	Anti-inflammatory doses are 37.5–40 mg/kg/day	Gastric irritation, chemical hepatitis
Indomethacin	TID	Start at 0.5 mg/kg/day; increase to 2.5 mg/kg/day	Gastric irritation, headache, hematuria
Naproxen (Naprosyn)	BID	10–20 mg/kg/day; maximum daily dose, 1000 mg	Gastric irritation, behavioral changes, headache, rash
Tolmetin sodium (Tolectin)	TID-QID	Start at 15 mg/kg/day; increase to 30 mg/kg/day; maximum daily dose, 1800 mg	Gastric irritation, headache, hematuria

Table 101.2. Nonsteroidal Anti-Inflammatory Drugs

For children who respond inadequately to NSAIDs, so-called disease-modifying antirheumatic drugs (DMARDs) are generally added to the regimen. Several agents, including sulfasalazine, hydroxychloroquine, and methotrexate, are available. Practitioners choose among these agents based on a child's age, the severity of the synovitis, and the subtype of arthritis. Methotrexate is the most commonly used agent in both juvenile and adult rheumatoid arthritis, and it appears to be the most effective drug at preventing or reversing joint damage. Sulfasalazine is most often used in children with inflammatory bowel disease but also is effective in various forms of arthritis. It is typically administered at a dosage of 40 to 70 mg/kg per day divided two to three times daily.

Hydroxychloroquine is an antimalarial agent with mild immunomodulatory effects. Dosages should never exceed 7 mg/kg per day to avoid ocular toxicity, including corneal deposition and macular degeneration.

Corticosteroids must be used judiciously in JRA because of the significant toxicity associated with their use. Systemic steroids are typically reserved for children with severe cardiac or pulmonary symptoms, during brief flare-ups of severe arthritis, or while waiting for slower-acting agents to take effect. Topical steroids are also effective for localized manifestations of JRA. Intra-articular steroids may be used in patients with pauciarthritis, or in children with polyarticular

disease in whom selected joints require particularly aggressive management. Ocular steroids are the linchpin of therapy for iridocyclitis.

Management of Complications and Emergencies (Table 101.3)

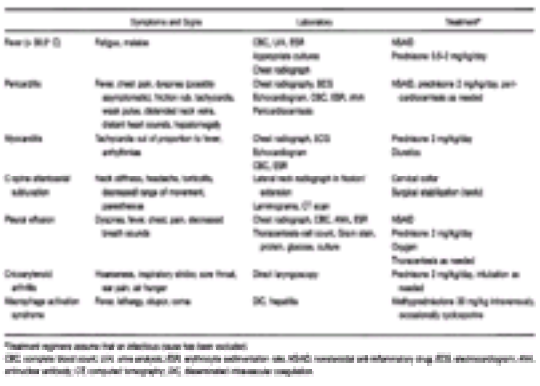

Table 101.3. Complications in Juvenile Rheumatoid Arthritis

Fever

Marked elevation of body temperature is characteristic of systemic JRA, whereas a lower-grade fever often accompanies polyarticular disease. The diagnosis of systemic JRA is one of exclusion, and fever is a common symptom, so diligent efforts should be made to rule out infectious diseases and malignancies. This may require hospitalization for a diagnostic evaluation, particularly in infants and young children. Appropriate cultures, including cultures of cerebrospinal fluid (CSF) if indicated, should be obtained. A bone marrow examination to rule out malignancy is necessary in many patients with systemic JRA.

In a patient being treated for known systemic JRA, the appearance of fever is always of concern. Fever may represent recurrence of JRA, or it may be caused by an intercurrent infection. Fevers in Still's disease typically follow the classic double quotidian pattern, with two peaks above 39°C (102.2°F) daily, as well as periods at or below normal without use of antipyretic medications. The child may be treated for a presumed JRA flare-up if there are no localizing signs of infection; if the complete blood count (CBC) shows the leukocytosis, thrombocytosis, and anemia typical of JRA; and if the urinalysis is normal. If the fever results from a specific infection (e.g., otitis media, urinary tract infection), appropriate antibiotics should be used. If the patient has received corticosteroids for more than 4 weeks within the previous 12 months, appropriate coverage with stress dosages (three times the physiologic dose) is indicated during the period of treatment of the infection.

Fever in systemic JRA, especially within 6 months of onset of disease, may rarely be caused by macrophage activation syndrome (MAS). This life-threatening complication is marked by disseminated intravascular coagulopathy (DIC) with diffuse microthromboses, hepatic inflammation, and central nervous system (CNS) changes progressing to seizures or coma. The cause of MAS is unknown, but it occurs more commonly during intercurrent viral illnesses, as well as in those taking NSAIDs or DMARDs (particularly sulfasalazine) as treatment. Differentiation from sepsis or a flare-up of JRA may be difficult, although a sudden rise in hepatic enzymes or a sudden drop in platelets, red blood cells, or erythrocyte sedimentation rate (ESR) (because of consumption of cellular elements and fibrinogen) are suggestive. Early diagnosis and a high level of suspicion are essential. Treatment with pulse dose methylprednisolone and/or cyclosporine, as well as general support measures for DIC, usually in an intensive care unit (ICU) setting, often lead to full recovery. Delayed diagnosis, on the other hand, is accompanied by a reported mortality rate of 20 to 50%.

Pericarditis, Myocarditis, and Cardiac Tamponade

Cardiac involvement is an important feature of systemic onset JRA but is uncommon in other subtypes of juvenile arthritis. Pericarditis (Fig. 101.3), like other systemic manifestations of Still's disease, most often occurs during the first 2 years of the illness. Common symptoms are fever, chest pain, dyspnea, and inability to lie flat in bed, although at times pericardial effusions may be asymptomatic. On physical examination, a parasternal pericardial friction rub may be heard over the left second and third intercostal spaces, especially with the patient supine. If the child has a moderate to large effusion, the clinician may not hear the friction rub but should look for the following signs of pericardial fluid: edema, tachycardia, weak pulse, distended neck veins, distant heart sounds, palpation of the apical impulse within the border of cardiac dullness, and hepatomegaly. Occasionally, the effusion may be massive, leading to cardiac tamponade as suggested by a pulsus paradoxus greater than 20 mm Hg.

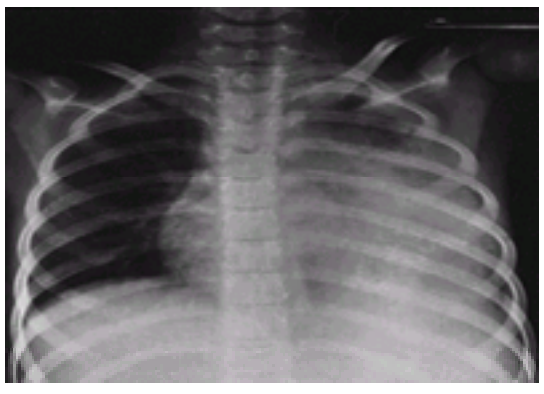

FIGURE 101.3. Pericardial and pleural effusions in a child with systemic type of juvenile rheumatoid arthritis.

Other types of cardiac involvement are unusual. Valvulitis is not typical of JRA and should suggest the possibility of acute rheumatic fever or bacterial endocarditis. Myocarditis is rare but may be seen. Tachycardia out of proportion to the elevation of temperature, arrhythmias, and congestive heart failure (CHF) are the usual clinical indicators of myocarditis.

If pericarditis or myocarditis is suspected, the child should be admitted and observed closely. Diagnostic studies should include electrocardiogram (ECG), chest radiograph, and echocardiogram. In pericarditis, the usual changes noted on ECG are tachycardia, elevated ST segment, and inverted T waves. Radiographs of the chest may show straightening of the left border of the heart and cardiac enlargement. Echocardiogram should be performed to confirm the presence of a pericardial effusion, as well as to quantify ventricular function, particularly if cardiac tamponade or myocarditis is suspected.

Bed rest and therapy with one of the NSAIDs should be adequate for the treatment of mild to moderate pericarditis. Corticosteroids (prednisone 1 to 2 mg/kg per day) are indicated for the treatment of myocarditis, for massive pericarditis causing compromise of cardiac output, or if significant symptoms persist despite therapy with NSAIDs. In the presence of tamponade or progressive deterioration, pericardiocentesis provides temporary relief while anti-inflammatory medications are used to prevent reaccumulation of fluid. If the child is acutely ill, requiring intravenous (IV) fluid support, care should be taken to avoid fluid overload, and diuretics should be added to the regimen.

Pulmonary Emergencies

Pleural effusions are a recognized complication of systemic JRA. Other pleuropulmonary manifestations include pneumonitis, diffuse interstitial disease, lymphoid bronchiolitis, and pulmonary arteritis. Occasionally, pleural fluid collections may be massive, resulting in respiratory distress. The usual clinical features of pleural effusion are chest pain, cough and dyspnea. On physical examination, there is dullness to percussion and diminished breath sounds on auscultation over the area of fluid. Chest radiographs, including lateral decubitus views (involved side down), may be used to document the extent of the effusion. Thoracentesis is indicated for diagnostic purposes, especially to rule out infectious processes, and in severe cases removing pleural fluid may help relieve respiratory compromise. Otherwise, treatment is aimed at the underlying disease process, primarily involving control of inflammation with NSAIDs or corticosteroids. Children with this complication often require admission for the overall severity of systemic features of the disease, rather than for the pleural effusion alone.

Iridocyclitis

Iridocyclitis (inflammation of the iris and ciliary body) occurs in approximately 10 to 20% of all children with JRA. This can be of acute or chronic onset. The chronic type of iridocyclitis occurs primarily in young girls with type I pauciarticular JRA, and it is virtually universal in girls with pauciarthritis and a positive ANA. In contrast, acute iridocyclitis occurs most often in older boys with pauciarticular disease.

The onset of chronic iridocyclitis is insidious and asymptomatic. Late signs are decreased visual acuity, unequal pupils, and band keratopathy. These reflect irreversible damage to the eye, however, and represent a missed opportunity for prevention. Therefore, all children with JRA, particularly those at high risk for iridocyclitis, should have a routine eye examination as soon as the diagnosis is made and at 3- to 6-month intervals thereafter. The emergency physician may be able to recognize evidence of established iridocyclitis such as posterior synechiae or cataracts using a +8 or +10 diopter lens in the ophthalmoscope, but slitlamp examination is required to recognize early inflammation.

Acute iridocyclitis, on the other hand, is characterized by sudden onset of redness, tearing, pain, and photophobia, and urgent management is crucial. Consultation with an ophthalmologist is essential. The usual treatment includes topical corticosteroids and mydriatics.

Flare-Up of a Single Joint in a Patient with JRA

In a patient known to have JRA who is taking anti-inflammatory medication, acute swelling with pain and limitation of range of movement of a single joint raises a major management problem. The differential diagnosis of this situation includes an acute flare-up of JRA versus infectious arthritis, and careful attention to physical examination and historical features are essential to avoid misdiagnosis.

Physical findings characteristic of infection of the joint are extreme pain, tenderness, erythema, and warmth over the joint. The affected joints of JRA may be swollen, warm, and stiff, but they are rarely red. There is usually a pronounced limitation of range of movement of the joint in infectious arthritis; the slightest movement may cause severe pain and muscle spasm. In contrast, some range of motion (5 to 10 degrees) is usually possible even with severely inflamed joints of JRA. If the patient is taking one of the corticosteroid preparations, physical findings of inflammation and/or infection may be masked.

If infection cannot be excluded with confidence, joint fluid must be aspirated, and the fluid sent for cell count, Gram stain, and culture. If there is any doubt about the diagnosis, it is best to obtain a blood culture as well and to initiate treatment for septic arthritis. For the acute swelling and pain in a single joint as a result of JRA, resting for 2 to 3 days and splinting the involved joint may be adequate. Local injection of the joint with a topical steroid preparation such as triamcinolone

hexacetamide (10 to 20 mg) is sometimes indicated after infection has been excluded.

Ruptured Popliteal Cyst

There are six bursae around the knee joint. Of these, the gastrocnemius-semimembranosus bursa is the one that most often communicates with the synovial space. Consequently, in the presence of effusion in the knee joint, fluid may enter the bursa and produce a popliteal cyst (Baker's cyst). Patients with popliteal cysts have a palpable and visible enlargement in the popliteal area, which is best seen in extension.

Rupture of a popliteal cyst with drainage of fluid into the calf muscles may present as an emergency. Affected patients complain of sudden pain in the calf associated with swelling in the leg. On physical examination, they have induration, erythema, warmth, and tenderness of the calf, as well as ankle edema. An effusion in the knee joint and evidence of synovial thickening are often present. Homan's sign may be positive, but other signs of thrombophlebitis, including palpable venous cords, dilation of collateral veins, and arterial spasm, are usually absent.

Differentiation of a ruptured popliteal cyst from thrombophlebitis may be difficult. Imaging techniques such as ultrasonography or MRI may be needed to establish the diagnosis. Intra-articular administration of steroids (triamcinolone hexacetamide, 10 to 20 mg) is the recommended initial treatment. If the response is inadequate or if the syndrome is chronic, surgical excision of the cyst may be necessary.

Cervical Spine Involvement

Cervical spine involvement usually is seen in children with established severe polyarticular JRA. Although this complication is known to occur in 30 to 50% of patients with JRA, atlantoaxial subluxation (AA subluxation) and subluxation of the lower cervical spine are less common in children than in adults, up to half of whom have AA subluxation. Clinical evidence of pressure on the spinal cord is seen in 23 to 65% of adults with radiologic evidence of AA subluxation. Similar figures are not available for children.

Neck stiffness that is worst in the morning is the most common symptom of cervical spine involvement in JRA. Occasionally, torticollis may be the presenting manifestation of cervical arthritis. Severe pain in the neck and referred pain over the occipital and retroorbital areas also may occur. The pain has a dull, aching quality and is often aggravated by neck movement. On physical examination, torticollis and/or loss of lordosis of the cervical spine and limitation of range of movement of the neck are the typical findings.

Paresthesia of the fingers is the most common symptom of spinal cord compression. Weakness of the arms and legs and inability to control the bladder are other complaints that should suggest spinal cord compression. During the initial stages, exaggerated deep tendon reflexes and an extensor plantar reflex are noted. Chronic myelopathy results in muscle atrophy and loss of deep tendon reflexes. Lateral radiographs of the neck in flexion and in extension are required for complete evaluation of the cervical spine. The patient should be asked to actively and slowly flex and extend the neck to tolerance without discomfort; care should be taken not to force these movements. Tomograms and an open-mouth view of the odontoid process may be helpful. On some occasions, computed tomography (CT) or MRI may be indicated.

The distance between the anterior surface of the odontoid and the posterior surface of the anterior arch of atlas when measured in a lateral flexion film is usually 4 mm or less. In the presence of AA subluxation, this may be as wide as 10 to 12 mm (Fig. 101.4). Other radiologic abnormalities characteristic of cervical spine involvement in JRA include loss of curvature, osteoporosis, erosions and sclerosis of joints, disc space narrowing, and altered height:width ratio of the vertebral bodies.

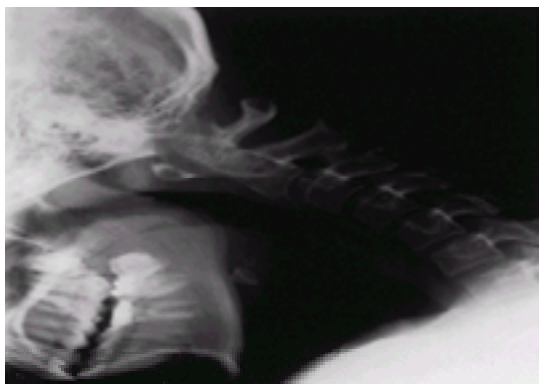

FIGURE 101.4. Atlantoaxial subfixation in a child with juvenile rheumatoid arthritis. (The distance between the anterior arch of the atlas and the odontoid process in the original radiograph was 5 mm.)

Although most children with AA subluxation do not have evidence of spinal cord compression, the physician must be wary of its occurrence with excessive movement, as for endotracheal intubation. Regular use of a light plastic cervical collar is often all that is required to relieve pain and prevent excessive anterior flexion, particularly during automobile rides. In the presence of spinal cord compression with muscle weakness and atrophy, surgical stabilization may be required.

Cricoarytenoid Arthritis

The cricoarytenoid joint is a diarthrodial joint with a synovial membrane. In patients with known polyarticular JRA, cricoarytenoid arthritis may rarely lead to acute airway obstruction. Clinical features of cricoarytenoid arthritis include stridor and hoarseness. The inspiratory stridor may wax and wane and may be present only when the patient is asleep. Some of these patients also may complain of pain in the throat while swallowing and pain in the ears. Many of these symptoms and signs are similar to those of severe acute laryngotracheobronchitis, which at times may be excluded only by direct laryngoscopy. Redness and swelling of the arytenoid eminences may be observed in cricoarytenoid arthritis, rather than the airway inflammation of croup.

Increasing airway obstruction with severe inspiratory retractions demands urgent treatment with respiratory support. High dosages of corticosteroids (methylprednisolone, 2 mg/kg per day intravenously) may control acute inflammation of the joints, avoiding emergency tracheostomy. If significant obstruction occurs, intubation should be attempted to establish an airway until swelling decreases; occasionally, emergency tracheostomy may be necessary. Even if tracheostomy is done, corticosteroid therapy is indicated so that the tracheostomy may be closed as quickly as possible.

Drug Toxicity

Almost all drugs used for the treatment of JRA have the potential for serious toxicity. If a child with JRA is being treated and develops a new symptom, drug toxicity must always be considered as one of the possible causes. Table 101.2 lists the common adverse reactions reported with NSAIDs. GI toxicity is the most common side effect, but significant NSAID gastropathy is actually unusual in children, and gastric or intestinal perforations, a significant problem in older adults, are rare. Nonetheless, children remain at risk for these complications, and only prostinoids such as misoprostil provide true gastroprotection. Antacids and H_2 blockers, although they ameliorate symptoms, do not actually reduce the risk of GI complications.

NSAIDs may cause a variety of other side effects as well. Reversible CNS complaints, particularly headaches, dizziness, and fatigue, occur in about 5% of children. Hepatotoxicity, manifested primarily as elevation of transaminases, and nephrotoxicity, including proteinuria and renal papillary necrosis, are rare but potentially dangerous if overlooked. Friability of the skin and a porphyrialike blistering of sun-exposed areas may be seen with these agents, especially in fair-skinned children taking naproxen.

Unlike salicylates, NSAIDs rarely cause tinnitus or hyperventilation. Reye syndrome, although far less common than with salicylates, has been reported in children taking NSAIDs; therefore, it is prudent to consider suspension of these agents in children with influenza or varicella. Salicylates must be carefully avoided in children who are even exposed to these viruses. Reye syndrome should be considered in the differential diagnosis of any child taking salicylates or other NSAIDs who presents with pernicious vomiting and/or alteration in mental status.

Each of the second-line agents used in the treatment of JRA also has the potential to cause specific forms of toxicity. More than 100,000 Americans now receive low-dose methotrexate for the treatment of arthritis, so questions of potential side effects are most likely to involve this drug. Doses used for JRA are much lower than those employed for treating malignancies—typically 0.3 to 1 mg/kg per week—and the degree of immunosuppression, although controversial, appears to be minimal. Live viral vaccines are nonetheless generally avoided in children receiving methotrexate, but reported cases of opportunistic or unusually severe infections are rare.

Despite its favorable therapeutic profile, methotrexate is an antimetabolite with the potential to cause oral ulcers, nausea, and abdominal pain. These may be minimized by supplementation with folic acid. Children must be monitored regularly for evidence of hepatic toxicity, and persistent elevation of hepatic transaminases identifies those at risk for hepatic fibrosis or cirrhosis. Methotrexate may also cause lymphopenia, especially with prolonged use, or even pancytopenia caused by bone marrow suppression. Concurrent use of other dihydrofolate reductase inhibitors, such as trimethoprim–sulfamethoxazole, potentiates this risk, and should be avoided.

Rarely, use of methotrexate is associated with the development of pulmonary hypersensitivity. This most commonly occurs during the first 6 to 12 months of use and may be marked by dyspnea, cough, fever, and fluffy infiltrates on chest radiograph. Although such symptoms may be conclusively distinguished from viral pneumonitis only by lung biopsy, suspicion of this complication necessitates discontinuation of methotrexate and institution of treatment with systemic corticosteroids. Failure to stop the drug or rechallenge with methotrexate may cause fatal respiratory failure.

Sulfasalazine is a sulfa drug, and its most severe toxicity is typical of this class of medications. Headache and GI upset, especially with preparations that are not enterically coated, are the most common side effects. Rarer, although more concerning, are bone marrow suppression, agranulocytosis, photosensitive eruptions, and hypersensitivity reactions, including Stevens-Johnson syndrome. Sulfasalazine is contraindicated in children with known intolerance of sulfa drugs, as well as in children less than 2 years of age in whom neurotoxicity may occur.

Antimalarial agents such as hydroxychloroquine must be administered judiciously because they can cause irreversible ocular toxicity at high dosages. Even at lower dosages, children may develop rashes, gastric upset, or reversible visual disturbances secondary to altered accommodation. Finally, children with glucose-6-phosphate deficiency who receive hydroxychloroquine may develop hemolytic anemia, especially during intercurrent infections.

The long list of potential side effects of systemic corticosteroids is enumerated elsewhere. In the acute-care setting, immunosuppressive effects of systemic steroids have the greatest impact on clinical management of JRA. It is important to remember that these agents most dramatically increase susceptibility to herpesviruses (especially disseminated varicella) and intracellular pathogens such as *Mycobacteria* and *Listeria*. Although they have little effect on susceptibility to other bacterial infections, their anti-inflammatory effects tend to mask clinical signs of infection, accentuating the need for vigilance on the part of clinicians.

A variety of other agents are rarely used in the United States, although occasionally, patients taking intramuscular or oral gold or d-penicillamine, may still present for evaluation. The major side effects from gold compounds are skin rash, bone marrow suppression with cytopenias, and proteinuria. D-penicillamine may cause skin rash, bone marrow suppression, nephrotoxicity, myasthenia gravis, and Goodpasture's syndrome. In view of their low risk:benefit ratios and prolonged duration of action, these agents should be discontinued if toxicity is suspected. If subsequent investigations identify another explanation for an apparent drug reaction, the drug may be restarted after a hiatus of days or weeks with little effect on control of the underlying arthritis.

SYSTEMIC LUPUS ERYTHEMATOSUS

Background

Systemic lupus erythematosus (SLE) is a multisystem disease that is both pleomorphic in its presentation and variable in its clinical course. The annual incidence of this disease has been reported to be 7.6 per 100,000 in adults, and it is estimated that there are approximately 10,000 to 15,000 children with SLE in the United States. The prevalence of SLE is greater among women between the ages of 15 and 64 years (1 in 700), particularly among African-American women (1 in 245).

The diagnosis of SLE is based on classification criteria established by the American College of Rheumatology (ACR). The 1971 preliminary criteria were revised in 1982 to include the presence of ANA and antibodies to native DNA; in 1997, a subcommittee of the ACR recommended inclusion of antiphospholipid antibodies as well. Table 101.4 lists the revised criteria and definitions. A patient should meet (any) 4 or more of the 11 criteria, simultaneously or in sequence, during any period of clinical follow-up to be diagnosed as having SLE. It is nonetheless important to remember that these criteria are intended for classification, not diagnosis, so patients may have SLE and not fulfill criteria, or they may meet criteria despite having another illness. Thus, pediatricians experienced in the care of children with SLE should participate in the diagnosis and management of all pediatric lupus patients.

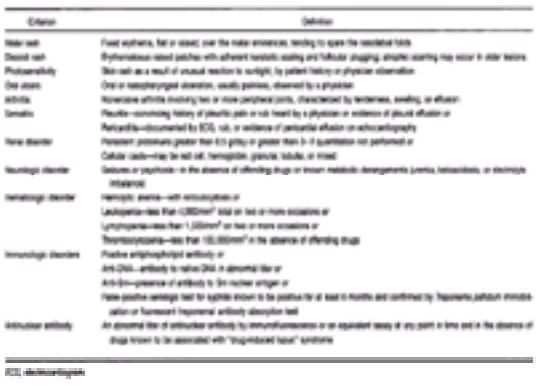

Table 101.4. Criteria for Classification of Systemic Lupus Erythematosus

Pathophysiology

The great variation in manifestations of SLE suggests that several discrete conditions may fall within the overall diagnostic category. A 7-year-old boy with arthritis, positive ANA, photosensitive eruption, and oral ulcerations appears to have little in common with a multiparous woman with seizures, pericarditis, renal failure, and high-titer anti-dsDNA antibodies. In fact, several different animal models for SLE have been developed, and they vary both in their clinical characteristics and in the underlying defect causing disease. The final common pathway unifying all cases of SLE is abnormal production of autoantibodies directed against a variety of antigens, including double-stranded DNA. The resultant circulating and in situ immune complexes deposit in tissues and activate the complement cascade. Release of activation products of complement, such as C5a, as well as other chemotactic, opsonic, and proinflammatory mediators, ultimately leads to tissue damage.

Current concepts of the pathogenesis of SLE envisage an interplay of environmental and genetic factors. Exposure to sun or to certain drugs (e.g., procainamide), and various types of infections (especially Epstein-Barr virus [EBV]), precipitate or exacerbate SLE in a predisposed host. On the other hand, family clusters of SLE, the occurrence of lupuslike syndromes with certain types of complement deficiencies, and the association between SLE and specific human leukocyte antigen (HLA) haplotypes, support a genetic component to the pathogenesis. The net result of these factors is functional abnormalities of both T and B cells, including diminished T-cell proliferative and stimulatory responses, polyclonal B-cell activation, defects in lymphocyte apoptosis, immunoglobulin isotype derangements, and defective cytokine production and response.

Histopathology of affected tissues reflects these mechanisms. In the skin, lesions vary from nonspecific vasculitis in the maculopapular variety of rash, to deeper lesions showing thinning of the epidermis, disruption of the dermal–epidermal junction, edema of the dermis, lymphocytic infiltration, and fibrinoid degeneration of the connective tissue. On immunofluorescent staining, localization of immunoglobulins and complement in the dermal–epidermal junction of the skin is seen in more than 75% of patients, although these findings may be present in other collagen vascular diseases as well.

Joints affected by arthritis show synovitis with fibrinoid degeneration of connective tissue but no pannus formation or cartilage destruction. Histologic features of lupus nephritis, even in the absence of clinical renal involvement, include

cellular proliferation and crescent formation, leukocyte and mononuclear cell infiltrates, and hyaline thrombi, as well as more chronic changes such as interstitial fibrosis and glomerular sclerosis. Analogous lesions in the CNS include microinfarcts and perivascular lymphocytic accumulation, as well as true vasculitis with inflammatory cells infiltrating vessel walls. Microhemorrhages into the subdural space have been documented as well.

Clinical Manifestations

The onset of SLE may be insidious or acute. The initial presentation usually includes constitutional features such as fever, malaise, and weight loss, in addition to features of specific organ involvement such as rash, pericarditis, arthritis, and seizures. Because virtually any part of the body may be affected by SLE, lupus may present with a bewildering variety of signs and symptoms. Although many of these are nonspecific, the examiner's level of suspicion for possible SLE should increase as the number of involved organ systems increases. Furthermore, although SLE is indeed a protean disease, most pediatric cases present with a typical spectrum of findings marked by musculoskeletal, renal, cutaneous, and hematologic involvement.

Arthritis in SLE is usually symmetric, involving both large and small joints. Swollen joints may be painful, but they are usually not erythematous. Patients may lose function because of tendon involvement, but the erosive synovial proliferation seen in JRA is uncommon.

Cutaneous lesions are present in more than 85% of patients with SLE. The typical malar erythematous rash with butterfly distribution occurs in about half of patients at the time of diagnosis. Discoid lesions are less common in children but, when seen, are characteristic of SLE (Fig. 101.5). Vasculitic skin lesions over the extensor surface of the forearm and on the fingertips are reported in about 20% of patients. These lesions are tender and may ulcerate. Nodules are less common. Mucosal lesions (macular and ulcerative) may involve the nose or the mouth, particularly the palate (Fig. 101.6) and are usually painless. Rarer types of mucocutaneous lesions in SLE include livedo reticularis, urticaria, erythema multiforme, and alopecia.

FIGURE 101.5. Adolescent girl with discoid lesions in malar distribution.

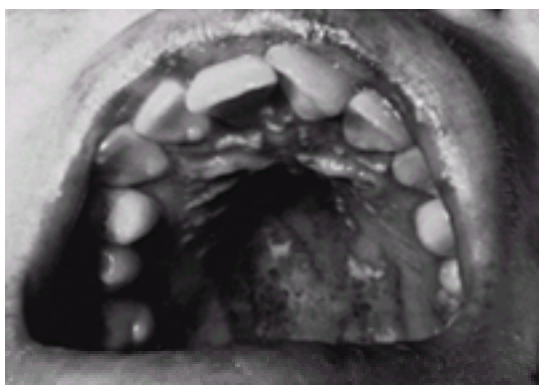

FIGURE 101.6. Mucosal lesions (macules and ulcers) of the palate in an adolescent girl with active lupus.

Evidence of renal disease is present in approximately 50% of children with SLE at the time of presentation. Nearly 90% of affected children will develop some degree of renal involvement during the course of their disease. Lupus nephritis is usually asymptomatic, although occasionally edema or hypertension may be clues to involvement of the kidney. Despite significant improvements in treatment, the extent of renal involvement remains the single most important determinant of prognosis in SLE. Most children will therefore require a renal biopsy before a therapeutic regimen may be established.

Clinical evidence of CNS involvement may occur at onset or later in the disease course. Symptoms and signs referable to the CNS include headache, seizures, polyneuropathy, hemiparesis/hemiplegia, ophthalmoplegia, hallucinations, memory alterations, and psychosis. Chorea is the most common movement disorder and may be a presenting sign; Lyme disease and rheumatic fever must also be considered in such cases. Cranial nerve palsies most commonly involve the optic nerve, trigeminal nerve, and nerves controlling the extraocular muscles. Myasthenia gravis should be excluded if any extraocular muscles are involved.

Pericarditis is the most prevalent form of cardiopulmonary involvement in SLE. Myocarditis occurs less often but is seen more often in SLE than in JRA. Heart murmurs caused by valvular lesions are uncommon, but asymptomatic vegetations

on valve leaflets are seen at autopsy in most patients (Libman-Sachs endocarditis). These do provide a potential nidus for bacterial superinfection, and in fact, patients with SLE are at increased risk of developing subacute bacterial endocarditis.

Pleuropulmonary involvement in SLE occurs in more than 50% of cases. Patients with pleuropulmonary manifestations commonly complain of chest pain, dyspnea, cough productive of small amounts of sputum, and fever. Pleural rub is the most common physical finding; dullness on percussion of the chest and/or diminished breath sounds on auscultation indicate the presence of a pleural effusion. Unilateral or bilateral pleural involvement may occur, with or without pleural effusion. For any SLE patient with pleuropulmonary manifestations, exclusion of intercurrent infection, CHF, aspiration pneumonia, and renal failure are critical for care. Although uncommon, pulmonary hemorrhage also occurs in children with SLE. It is potentially lethal and requires early recognition.

Common GI manifestations include nausea, vomiting, and anorexia. Persistent localized abdominal pain should suggest specific organ involvement, such as pancreatitis or gastric ulcer—both of which may occur from the disease or associated with steroid therapy. Generalized abdominal pain in SLE is not always related to the underlying disease but may stem from other causes such as appendicitis, ruptured ovarian cyst, or pelvic inflammatory disease. Manifestations of any of these conditions may be masked by the corticosteroids most patients receive. Malabsorption syndrome may be an occasional manifestation of SLE. Melena should suggest bleeding from the GI tract and requires immediate evaluation and treatment.

Laboratory Features

Mild to moderate anemia is common in patients with SLE. Hemolytic anemia associated with a positive Coombs' test is most characteristic. An acute decrease in the hemoglobin or hematocrit should alert the physician to the possibility of internal hemorrhage or massive hemolysis. Autoimmune thrombocytopenia, even in the absence of offending drugs, is commonly seen in SLE. Leukopenia and lymphopenia are additional hematologic abnormalities characteristically seen in SLE; few other conditions in children lead to lymphocyte counts under 1000/mm $_3$.

Circulating antibodies to specific clotting factors, deficiencies of one or more clotting factors, and abnormal platelet function may lead to abnormal hemostasis in SLE. A specific circulating anticoagulant, the “lupus anticoagulant,” has been described in up to 10% of patients with SLE. The antibody is so named because in vitro assays of coagulation are prolonged in its presence. In vivo, this antibody predisposes to arterial or venous thrombosis.

Proteinuria, hematuria, and cellular casts are the usual urinary abnormalities. Renal failure is suggested by decreased urine output, elevated levels of blood urea nitrogen (BUN) and creatinine, and reduced creatinine clearance. Nephrotic syndrome is best documented by a 24-hour urine collection for quantitation of protein excretion, although a spot urinary protein:creatinine ratio is a useful screening tool for proteinuria.

The most important single test in children suspected of having SLE is the ANA. Up to 2% of normal children have low to intermediate titers of ANA at any time; in most cases, these antibodies are transient cross-reacting immunoglobulins triggered by a viral infection. In SLE, the titer is often high--1:512 or greater—and is accompanied by antibodies to double-stranded DNA, a more specific marker for the lupus. Nonetheless, it must be remembered that SLE may be diagnosed only in the presence of evidence of multiple organ system involvement, so no laboratory study is pathognomonic.

Total serum hemolytic complement (CH_{50}) is often decreased in patients with active disease; therefore, it may aid in differentiating disease flare-ups from intercurrent illnesses. An elevated level of anti-DNA antibody with hypocomplementemia particularly correlates with active renal disease. Acute-phase reactants are elevated in the serum during active phases of the disease, whereas serum protein analysis may demonstrate elevation of immunoglobulins.

Management

General Management

No specific treatment exists for SLE. Rather, type and intensity of therapy are dictated by the particular organ systems affected. Patients with mild disease (fever and/or arthritis) without nephritis generally receive one of the NSAIDs (e.g., tolmetin sodium 20 to 40 mg/kg per day) (Table 101.2). Severe systemic features usually require treatment with oral or IV corticosteroids, with doses divided three or four times daily in the most florid cases. As disease activity subsides, steroids may be carefully tapered; tapering too rapidly often results in a flare-up of the disease process. Steroids are generally first consolidated into a single morning dose, then the total daily dose is gingerly tapered over weeks to months. Ultimately, when possible, patients are weaned to alternate-day therapy in an attempt to minimize side effects.

Patients with life-threatening disease, particularly those with severe renal or CNS involvement, may require so-called pulse doses of corticosteroids (IV methylprednisolone 30 mg/kg per day), plasmapheresis, or an immunosuppressive agent (especially azathioprine or cyclophosphamide). Symptomatic management may be necessary for the treatment of seizures, psychosis, or acute renal failure. Most patients also receive hydroxychloroquine, which has been shown to prolong disease-free remissions once signs and symptoms of active lupus are controlled. In any event, close follow-up is mandatory to detect clinical and serologic clues to exacerbations as rapidly as possible and to monitor drug toxicity. Finally, as with all chronic diseases, total management should include emotional support for patients and their families.

Management of Complications and Emergencies (Table 101.5)

Table 101.5. Complications of Systemic Lupus Erythematosus

Infections in SLE. Management of emergencies in patients with SLE first and foremost involves distinguishing primary disease manifestations from secondary complications. Infection is the major cause of mortality in children with SLE. Defective phagocytosis, decreased leukocyte chemotaxis, lymphocytotoxic serum factors, hypocomplementemia, and functional asplenia, contribute to the increased susceptibility of these patients to opportunistic infection. Gram-negative bacilli (especially *Salmonella*), *Listeria, Candida, Aspergillus, Cryptococcus, Toxoplasma, Pneumocystis,* and the varicella-zoster virus are some of the organisms associated with severe infections in SLE. Patients with SLE who are taking corticosteroids or cytotoxic drugs are at even higher risk for developing viral, mycotic, and other opportunistic infections. Most of these infections are diagnosed at autopsy, so clinicians must maintain a high level of suspicion in all children with SLE.

All patients with lupus and suspected infection need not be admitted. However, acutely ill children, those with an absolute neutrophil count of less than 1000/mm_3, and those with pneumonia or the possibility of meningitis require hospitalization for IV antibiotics while awaiting culture results. Patients with minor infections who are not acutely ill or neutropenic may be treated with appropriate antibiotics given orally along with frequent follow-up visits. The dosage of corticosteroids should also be increased to provide stress coverage (at least three times the physiologic need) in any acutely ill child who has received more than 20 mg of prednisone for more than 1 month.

Fever. Each febrile episode in a child with SLE represents a potential emergency. It is difficult to determine whether the fever is secondary to infection, to a flare-up of the primary disease, or to a combination of both. A complete physical examination should be performed. A CBC, including total and differential WBC counts and platelet count, urinalysis, Westergren sedimentation rate, quantitative C-reactive protein (CRP), CH_{50} (or C4), C3, ANA, and anti-dsDNA antibody titers, should be obtained. Cultures of blood and urine are mandatory, and clinicians should have a low threshold for culturing CSF and other fluids when indicated. These cultures are particularly critical if no source of fever is apparent after a complete physical examination. In most cases, children with SLE who develop fever without a readily apparent source should be given antibiotics pending culture results; abnormal splenic function places them at increased risk of rapid development of bacteremia and overwhelming sepsis.

Renal Complications. Renal disease is a major cause of morbidity in patients with SLE, so its presence and severity must be established at the time of diagnosis and renal function regularly monitored thereafter. Clinical manifestations of lupus nephritis are often minimal. Gross hematuria or headache resulting from hypertension may be warning signs. In the presence of nephrotic syndrome, the child may be edematous. Laboratory evidence of renal disease includes proteinuria, hematuria, hyposthenuria, casts, and elevated levels of BUN and creatinine. The presence of these findings in a patient with known SLE requires a more thorough investigation, including estimation of the protein in a 24-hour urine collection, creatinine clearance, measurement of C3, ANA, and anti-dsDNA antibodies, and renal biopsy. In a patient with SLE and documented renal disease, hospitalization is necessary only in the presence of rapidly worsening renal status, acute renal failure, hypertensive crisis, or severe complications of therapy.

Treatment of renal disease is aimed at preserving renal function while minimizing medication toxicity. Active disease may often be managed with the usual dosages of corticosteroids (prednisone 1 to 2 mg/kg per day). In the presence of progressive renal failure, the patient should be hospitalized for more aggressive therapy. This generally includes divided doses of IV corticosteroids with or without an immunosuppressive agent such as cyclophosphamide. “Pulse” therapy with methylprednisolone (30 mg/kg in 50 mL of D5W) may be indicated in the presence of rapidly progressive renal disease. Plasmapheresis has been used in the treatment of severe lupus nephritis, especially in patients who fail to respond to conventional therapy with corticosteroids and cytotoxic agents. Although this modality appears to have little effect on long-term outcome, acute disease flare-ups may be rapidly controlled by removing pathogenic autoantibodies, immune complexes, and cytokines. Each of these therapies is associated with significant toxicity, so their use should be limited to centers experienced in the care of acutely ill children with SLE.

Hematologic Complications. Anemia is common in SLE and may have many causes. The most common type is the normocytic, normochromic anemia of chronic disease. Anemia from GI blood loss may be secondary to vasculitis or caused by irritation of the GI tract. These patients often have symptoms of GI distress and occult blood in the stool. They require further investigation, but the urgency depends on the severity of the bleeding.

Hemolytic anemia in SLE may be related to the disease itself (antierythrocyte antibodies) or to medications. Patients with hemolytic anemia often present with pallor, fatigue, jaundice, splenomegaly, and dark-colored urine. Occasionally, these patients develop symptoms of cardiorespiratory distress and CHF after severe hemolysis and a rapid fall in hemoglobin. Laboratory investigation of anemia in SLE should include CBC, reticulocyte count, and examination of the blood smear for red cell size and shape, nucleated red cells, and fragmented red cells. Serum levels of iron, iron-binding capacity, free erythrocyte protoporphyrin, and bilirubin should be obtained. The antibody responsible for autoimmune hemolytic

anemia is of the “warm” variety, most commonly of the IgG type; IgM type antibody is present in only a small percentage of cases. These red cell–bound antibodies may not be demonstrated by the standard Coombs' test, so more sensitive assay systems may have to be used.

Mild to moderate anemia of any cause may be managed using oral iron preparations and by treatment of the primary disease. Children with a hematocrit of less than 20% or compromised cardiac function often require admission to the hospital. Corticosteroids are the most effective agents for the control of autoimmune hemolytic anemia in SLE. Prednisone at 2 mg/kg per day is the preferred initial treatment. Transfusion may be needed for children with a rapidly dropping hemoglobin concentration or CHF.

Leukopenia occurs in about 50% of patients with SLE. It may be caused by a reduction in granulocytes, lymphocytes, or both. Granulocytopenia may be secondary to drugs used in the treatment of SLE or, less commonly, to disease-related destruction of granulocytes. Febrile children with absolute granulocyte counts of less than 1000/mm $_3$ should be admitted for empiric antibiotic coverage, pending results of further studies because they are at higher risk of severe infections.

Thrombocytopenia occurs in approximately 25% of patients with SLE; conversely, more than 5% of children presenting with idiopathic thrombocytopenic purpura (ITP) eventually develop SLE. The usual causes of thrombocytopenia are circulating antibodies to platelets or drug-induced bone marrow suppression. Infection should always be considered as a possible cause. The presence of purpura and ecchymoses requires immediate investigation. Hemorrhage into one of the internal organs, a sudden drop in hemoglobin, and platelet counts of less than 20,000/mm $_3$ are the usual indications for admission to the hospital. Studies should include CBC, examination of the peripheral blood smear, and appropriate cultures. At times, bone marrow examination and testing of serum for antiplatelet antibodies may be helpful in determining the cause of reduced platelet counts.

Patients with SLE are at risk of bleeding from any mucosal surface because of vasculitic ulceration, impaired hemostasis, thrombocytopenia, or a combination of these factors. Patients with life-threatening *epistaxis* may require local packing and platelet replacement in addition to high-dose corticosteroids. Severe pulmonary hemorrhage may necessitate general supportive measures such as transfusions, ventilatory assistance and bronchial lavage, as well as treatment of the underlying pathologic condition with high-dose corticosteroids, immunosuppressive agent, and plasmapheresis.

Although rare, *DIC* may occur in patients with SLE, with or without an associated infection. Therefore, patients with thrombocytopenia and severe bleeding should be investigated with prothrombin time (PT), partial thromboplastin time (PTT), fibrin split products, and examination of the peripheral smear. Lupus appears to predispose patients with SLE to a particularly malignant form of thrombotic thrombocytopenic purpura. Reported mortality rates are extremely high despite general support in ICUs, as well as aggressive treatment with pheresis and immunosuppression.

The presence of a circulating *lupus anticoagulant* does not lead to a bleeding diathesis unless associated with significant thrombocytopenia; on the contrary, these patients are at increased risk of deep venous or arterial thromboses. Prolongation of PTT and chronic false-positive serologic test results for syphilis are the usual clues to the presence of these autoantibodies. Specialized studies such as mixing assays and the Russell viper venom test (RVVT) may confirm the diagnosis. Significant thrombosis or pulmonary embolus in a child with SLE is an indication for immediate anticoagulation with heparin, followed by oral warfarin or subcutaneous low-molecular-weight heparin, pending assays for these circulating anticoagulants.

Neurologic Complications. *Seizures* (see Chapter 70) and altered states of consciousness (see Chapter 13 and Chapter 83) are the most common manifestations of CNS involvement in SLE. Other possible causes of seizures in patients with SLE include hypertension (from the disease itself or as a complication of corticosteroid therapy), infection (acute or indolent meningitis or abscess), and uremia. Coma is not a primary manifestation of SLE but may result from meningitis or CNS hemorrhage secondary to thrombocytopenia. Therefore, patients with SLE who develop seizures or altered states of consciousness require admission for evaluation, which should include a thorough examination with special attention to blood pressure and neurologic findings, as well as the following investigations: CBC with differential and platelet counts, electrolytes, BUN, creatinine, urinalysis, and lumbar puncture (including measurement of opening pressure and CSF cultures). The CSF should be sent for routine studies and for special stains to look for opportunistic organisms such as fungi and acid-fast bacilli.

No study is perfectly sensitive for detecting lupus cerebritis. Measurement of the IgG index [(CSF IgG/Serum IgG) (CSF Albumin/Serum Albumin)] may allow estimation of IgG synthesis within the blood–brain barrier; although this is increased in a variety of chronic infections, it also typically rises in active CNS lupus. In addition, an electroencephalogram (EEG), MRI study, and CT scan with contrast may facilitate elucidation of the cause of CNS signs in children with lupus.

IV diazepam (0.2 to 0.3 mg/kg) is the preferred drug for the initial management of seizures, followed by phenytoin for maintenance of seizure control. Phenytoin is preferred over phenobarbital because the latter may alter mental status acutely. If CNS manifestations are considered to be secondary to active vasculitis, IV corticosteroid therapy should be initiated. In the presence of deteriorating mental function, “pulse” methylprednisolone (30 mg/kg in 50 mL of D5W), IV cyclophosphamide, or plasmapheresis may be beneficial.

Other manifestations of CNS involvement, such as psychosis, also may need inpatient evaluation. *Listeria monocytogenes* may cause an indolent meningitis that is clinically indistinguishable from organic brain syndromes. Similarly, it may be difficult to determine whether psychosis is secondary to corticosteroid therapy, especially because steroids are most likely to induce psychiatric symptoms in patients with underlying psychiatric disease. Clinicians should not hesitate to aggressively pursue a diagnostic evaluation, including lumbar puncture and imaging procedures, so that appropriate therapy may be instituted as expeditiously as possible. When psychosis resulting from SLE is suspected, psychotropic drugs (e.g., haloperidol 0.025 to 0.05 mg/kg per day in divided doses) may be used along with large doses of corticosteroids for 1 to 2 weeks. If no improvement is seen, the steroid dosage may be reduced gradually in an attempt

to rule out steroid-induced psychosis.

Transverse myelitis is a rare complication of SLE thought to result from vascular compromise of the spinal cord. Patients note acute onset of pain and weakness, and they may develop incontinence. Physical examination is remarkable for weakness or flaccid paralysis below the level of the functional transection. In a high percentage of cases, the process is associated with a circulating lupus anticoagulant or antiphospholipid antibodies. Prognosis is related to the duration of symptoms before initiation of therapy, and favorable outcomes are possible only with urgent intervention. Thus, once infection, epidural abscess, and hematoma are excluded with appropriate imaging procedures and lumbar puncture, pulse doses of IV methylprednisolone (30 mg/kg over 1 to 2 hours) plus anticoagulation with IV heparin are begun. Early addition of potent immunosuppressive agents such as IV cyclophosphamide (500 to 750 mg/m_2 intravenously) is often essential.

Pulmonary Complications. Pleural effusion is the most common pulmonary manifestation of SLE. However, pulmonary infections and hemorrhage present more acute management issues. *Pleural effusion* is often bilateral and small, although occasionally, it may be massive. The child is often ill with acute manifestations of systemic disease, such as fever, fatigue, and poor appetite. Symptoms may be minimal (cough, chest pain, and mild tachypnea) or absent. Chest pain aggravated by deep breathing or coughing is suggestive of pleurisy, although pain may be absent. In the presence of a moderate or large effusion, the patient may have dyspnea and tachypnea. On auscultation, a pleural rub may be heard. A radiograph is essential to determine the extent of the effusion.

If the physician knows the child and has no concerns about infection, hospitalization may not be necessary. Increasing the corticosteroid dosage or adding indomethacin (0.5 to 2 mg/kg per day) or tolmetin sodium (20 to 40 mg/kg per day) may be adequate therapy, but arrangements must be made for close follow-up. If the cause of a pleural effusion is in doubt, pleural fluid should be aspirated. Usually, pleural effusions caused by SLE are exudates that show elevated protein levels and cell counts, and decreased levels of lactic dehydrogenase. The presence of fluid is easily demonstrated clinically (diminished breath sounds and dullness to percussion), and the radiograph of the chest establishes the extent of the effusion. Patients with large effusions should be admitted to the hospital for further observation and management. Thoracentesis is often necessary 1) to relieve symptoms, 2) for diagnosis, and 3) to reveal any underlying lesions obscured by the effusion.

Pulmonary hemorrhage is a potentially catastrophic complication of SLE, particularly in the pediatric age group. Early recognition and treatment are critical. A hemorrhage may be related to the disease itself (e.g., pulmonary vasculitis), to the treatment (e.g., drug-induced thrombocytopenia), or to an infection (e.g., aspergillosis). Clinical features of patients with pulmonary hemorrhage include hemoptysis, tachypnea, tachycardia, and dyspnea; rapid deterioration may result in asphyxia within 24 to 48 hours.

Evaluation of patients with unexplained respiratory symptoms should include a chest radiograph. In cases of pulmonary hemorrhage, this shows fluffy infiltrates resembling pulmonary edema. CBCs often reveal a dramatic drop in hemoglobin and a low platelet count. Diagnosis of a pulmonary hemorrhage may be confirmed by pulmonary function testing, including diffusing capacity of carbon monoxide (DL_{CO}). Intra-alveolar blood increases CO absorption and therefore is one of the few conditions that results in an abnormally *high* DL_{CO}. Bronchoalveolar lavage or lung biopsy still may be needed in some patients in whom *Pneumocystis* or *Aspergillus* infection remains a concern.

Management should include transfusions and high dosages of IV corticosteroids. If bleeding is related to thrombocytopenia, platelet transfusion is indicated. Tracheal lavage with epinephrine, oxygen therapy, and intubation with positive end-expiratory pressure (PEEP) ventilation may be needed, depending on the severity and progression of the process.

Occasionally, children with lupus may develop interstitial pneumonitis. Such patients are often ill with high fever, chest pain, cough, and dyspnea. On examination, rales may be heard throughout the chest. Radiographs show a diffuse alveolar infiltrate, unilateral or bilateral, with or without effusion. Cultures of the blood and respiratory secretions, bronchial washings, transtracheal aspirate, or lung biopsy may be necessary to exclude infection. Supportive therapy should include increased concentrations of oxygen, adequate pulmonary toilet, and antipyretic drugs. Measures used to control other manifestations of SLE, including corticosteroids or immunosuppressive agents, may lead to dramatic improvement.

Gastrointestinal Complications. Peritonitis and GI hemorrhage are emergencies associated with SLE. Drug-induced gastric ulcer and pancreatitis also occur. The nature of an intra-abdominal catastrophe is often difficult to determine. Plain radiographs of the abdomen, ultrasonogram, MRI, CT scanning, peritoneal aspiration, and rarely, even exploration may be required to rule out surgical conditions.

Peritonitis may be a feature of the disease itself (serosal inflammation) or may be caused by secondary infection or visceral perforation. Patients with SLE and peritonitis should be admitted at once. Symptoms and signs associated with peritonitis are pain in the abdomen, fever, vomiting, diarrhea, abdominal distension, diffuse tenderness, rigidity of the anterior abdominal wall, and hypoactive or absent bowel sounds on auscultation. However, all of these findings of peritoneal irritation may be masked by corticosteroid therapy.

A radiograph of the abdomen may show dilation of intestinal loops with edema of the wall of the intestines, free air in the peritoneal cavity, or evidence of ileus or obstruction. Aspiration of the peritoneal fluid under strict aseptic conditions is essential if the cause of the peritoneal effusion is in doubt. The fluid should be sent for Gram stain and culture. Cell counts of more than 300/mm_3 should be considered indicative of infection. Peritonitis secondary to GI perforation should be treated aggressively with surgery and IV antibiotics. Peritonitis of the serous type (a feature of SLE) may be treated with one of the NSAIDs; corticosteroids may be added if the response to the anti-inflammatory medication is inadequate or if there is additional evidence of active systemic disease. Prolonged use of both NSAIDs and corticosteroids may lead

to more problems from gastric irritation and/or ulceration.

An acute abdomen in SLE may be the result of bowel ischemia, infarction, or perforation, in addition to the occasional unrelated complication such as intussusception or appendicitis. Symptoms of an acute abdomen include sudden onset of abdominal pain, vomiting, and diarrhea that may be bloody, although corticosteroids may obscure all signs and symptoms. The patient may go into shock rapidly. Localized abdominal tenderness, guarding, and rigidity with absent bowel sounds are often present. Rectal examination must be performed to localize tenderness, palpate any masses, and obtain stool for occult blood testing. The patient should be well hydrated and shock promptly treated in an intensive care setting. A CBC, serum electrolytes, and serum amylase determination should be obtained at once. A plain radiograph of the abdomen may show air–fluid levels or free air under the diaphragm. Abdominal ultrasound or CT may allow greater diagnostic precision. Paracentesis is essential to rule out infection or hemorrhage secondary to perforation. Gram stain and culture of peritoneal fluid, in addition to blood culture, should be obtained immediately. Infection should be treated aggressively, and the ischemic or perforated area of intestine surgically repaired.

Pancreatitis must be considered in children with SLE and abdominal symptoms. SLE is the most common medical cause of pancreatitis in children, and corticosteroids are the medication most often associated with this complication. If the serum amylase is normal and pancreatitis is suspected, one should obtain an amylase clearance. An amylase:creatinine clearance ratio of greater than 5 suggests pancreatitis. If pancreatitis is secondary to SLE, corticosteroid therapy is used. If it is secondary to medications, attempts should be made to withdraw them gradually. During this slow process, the patient may have to be maintained on parenteral hyperalimentation.

GI hemorrhage may be secondary to NSAIDs (stomach), vasculitis of the GI tract (small intestines), or thrombocytopenia. Symptoms include abdominal pain, hematemesis and melena if the bleeding is in the upper GI tract, and abdominal pain with hematochezia or occult blood in the stool if bleeding is from the lower GI tract. The patient may develop massive bleeding, leading to shock. Immediate studies to be obtained in the emergency department include hematocrit, CBC, serum electrolytes, and blood type and crossmatch. Stool obtained by rectal examination should be tested for occult blood even if the stool appears frankly bloody. If bleeding from a gastric ulcer is suspected, endoscopy can confirm the diagnosis. Therapy for a bleeding gastric ulcer includes volume replacement, hourly antacid administration, and H_2 blockers (e.g., cimetidine 20 to 40 mg/kg per day, maximum 1200 mg/day).

If active bleeding caused by vasculitis is suspected, celiac axis angiography or endoscopy with deep intestinal biopsies are required for confirmation. GI vasculitis is rare in pediatric lupus, and most commonly occurs in the setting of chronically active disease. Children typically have an associated peripheral neuropathy, as well as chronic weight loss, anorexia, and inanition.

Cardiac Complications. Pericarditis and myocarditis are two of the important cardiac complications of SLE that may require emergency care. The features of pericarditis are similar to those described in JRA and include chest pain, dyspnea, inability to lie flat in bed, and pericardial friction rub. In the presence of cardiac tamponade, additional signs supervene (weak pulse, distended neck veins, distant heart sounds, and pulsus paradoxus of more than 20 mm Hg). Pericarditis without significant hemodynamic effects may be managed with NSAIDs or corticosteroids. Massive effusion leading to tamponade requires pericardiocentesis in addition to treatment with corticosteroids, which may be injected directly into the pericardium at the time of pericardiocentesis.

Myocarditis is characterized by resting tachycardia out of proportion to fever, cardiomegaly without an effusion, CHF, ST-T wave changes on ECG, and arrhythmias. These patients should be on strict bed rest with monitoring. In addition to treatment of the basic disease, digoxin and diuretic therapy are often indicated.

Raynaud's Phenomenon. Raynaud's phenomenon (RP) is characterized by triphasic color changes of the extremities upon exposure to cold. These color changes proceed from cyanosis to blanching because of microcirculatory compromise and resolve with erythema caused by reactive hyperemia. Severe episodes of RP may cause excruciating pain in the extremities or even digital ulceration and autoamputation. Poor circulation impairs wound healing and clearing of infections, so patients with paronychia or digital cellulitis may require admission for IV antibiotics.

Prophylactic techniques to improve digital circulation (avoidance of cold exposure, biofeedback) are the cornerstones of treatment of RP. Calcium channel blockers (e.g., slow-release nifedipine 30 to 180 mg/day) may decrease the frequency and severity of attacks, whereas oral and topical vasodilator drugs (e.g., prazosin, nitroglycerin) or medical or surgical sympathetic blockade may be necessary during severe episodes. Cases of impending gangrene may also be treated with prostacyclin analogs such as Iloprost. These medications may cause dramatic vasodilation and result in pulmonary edema or cardiac arrhythmias; therefore, they should be used by experienced clinicians only.

Hypertension. Hypertension may be a result of effects of SLE on systemic vasculature, a concomitant of renal involvement, or secondary to steroid therapy. Mild to moderate hypertension is usually controlled by combinations of diuretics, vasodilators, and a- or b-blockers, whereas angiotensin-converting enzyme (ACE) inhibitors are effective for renovascular hypertension. Diazoxide (5 mg/kg) given intravenously as rapidly as possible as a single bolus is the preferred drug for the treatment of hypertensive encephalopathy.

Ocular Complications. Children with SLE may develop blurring or loss of vision. When this is accompanied by headache and vomiting, the differential diagnosis includes meningitis (both septic and aseptic), hypertension, and pseudotumor cerebri. An ophthalmologic consultation should be obtained to exclude other complications such as retinal vasculitis or retinal vascular occlusion. The patient should have an exhaustive neurologic evaluation, and examination of the spinal fluid should be performed with great caution after an emergency CT scan has been obtained. Gradual periodic release of CSF pressure is the treatment of choice for pseudotumor cerebri. High-dose corticosteroid therapy should be added if the intracranial hypertension is believed to result from SLE, whereas it should be tapered if the pseudotumor is secondary to steroid toxicity.

JUVENILE DERMATOMYOSITIS

Background

Juvenile dermatomyositis (JDMS) is a rare rheumatic disorder characterized by inflammation of the skin and striated muscle. The disease has a wide spectrum, from a mild form involving mainly the skin to a severe vasculitic type with a rapidly fulminating course. JDMS may be conceptualized as passing through four overlapping phases that typically last for 2 to 5 years but may persist indefinitely: 1) a prodromal phase of nonspecific aches and pains, 2) a phase of progressive muscle weakness, 3) a phase of persistent active disease, and 4) an indolent phase with development of contractures and calcinosis. The goal of therapy is to compress this natural history into a shorter time period that ends before irreversible sequelae occur.

As with other idiopathic rheumatic diseases of childhood, diagnosis of JDMS depends on fulfillment of clinical criteria. Bohan and Peter's criteria for dermatomyositis (DM)/polymyositis (PM) in adults are typically used for diagnosing this condition, and they are given in Table 101.6. In fact, the condition in children differs significantly from that in adults; it includes a more prominent degree of vascular inflammation and scantier evidence of circulating autoantibodies, and it rarely accompanies malignancies. Furthermore, in children, the appearance of inflamed muscle and soft tissues on MRI is essentially pathognomonic, eliminating the need for muscle biopsy or electromyogram (EMG) to confirm the diagnosis (Fig. 101.7). Nonetheless, in the absence of alternative criteria, Bohan and Peter's scheme remains the gold standard for classification of children with inflammatory myopathies.

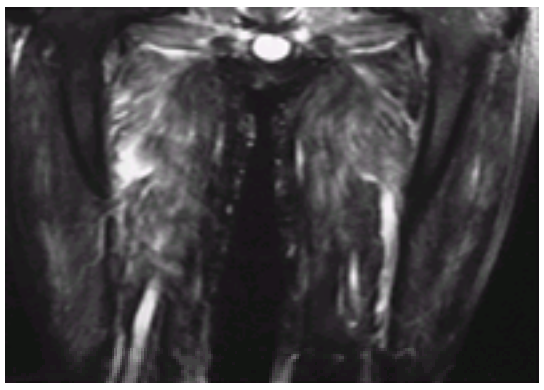

FIGURE 101.7. Coronal fast multiplanar inversion recovery image of the thighs shows areas of increased signal intensity, especially in the adductor muscle groups, in a patient with dermatomyositis.

1. Symmetric weakness of the proximal limb muscles and of anterior neck flexors	
2. Evidence of necrosis of type I and II fibers on muscle biopsy	
3. Elevation of serum levels of skeletal muscle enzymes—creatine phosphokinase and aldolase	
4. Short, small, polyphasic motor unit potentials with fibrillation; insertional irritability; and high-frequency repetitive discharges on electromyography	
5. Skin rash—characteristic heliotrope rash, scaly erythematous rash over extensor aspects of the joints, and periungual erythema	
Definite:	4 criteria (PM) 3 or 4 criteria + rash (DM)
Probable:	3 criteria (PM) 2 criteria + rash (DM)
Possible:	2 criteria (PM) 1 criterion + rash (DM)

From Bohan A, Peter JB: Polymyositis and dermatomyositis. N Engl J Med 1975;292:344.

Table 101.6. Criteria for the Diagnosis of Dermatomyositis (DM)/Polymyositis (PM)

Pathophysiology

Microscopically, the skin in JDMS shows dermal atrophy, obliteration of appendages, and lymphocytic infiltration. Characteristic lesions in the muscle include a mixture of degenerating and regenerating muscle fibers, variations in muscle fiber size, perivascular lymphocytic infiltration, and perifascicular atrophy of muscle fibers. Angiopathy involving small arteries, venules and capillaries of the skin, muscle, fat, and GI tract is characteristic of childhood DM. Viral infections, particularly coxsackie B, vasculitis caused by immune complex deposition, and cell-mediated cytotoxicity against muscle fibers, have all been implicated in the pathogenesis of JDMS. Associations of juvenile DM with the presence of HLA-B8/DR3 antigens have also been reported. Theories of pathogenesis thus center on an as yet unexplained perpetuation of muscle inflammation in susceptible hosts following what is generally a self-limited illness in most children.

Clinical Manifestations

The onset of JDMS is often insidious, with aches and pains in the limbs, low-grade fever, general weakness, and edema of the hands, feet, and eyelids. A diffuse and nonspecific rash may be seen. This prodromal stage evolves into the acute phase, when the characteristic features of JDMS become evident. Classical skin manifestations include a violaceous heliotrope rash in the periorbital region and occasionally on the forehead; dusky red or atrophic lesions over the extensor aspects of the knees, elbows, and knuckles (Gottron's papules); and periungual erythema (Fig. 101.8). Skin findings may

precede or follow the onset of muscle weakness.

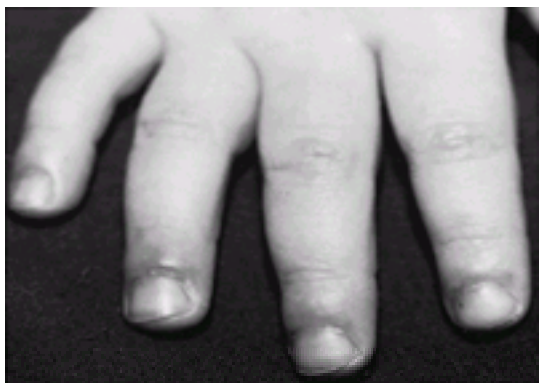

FIGURE 101.8. Atrophic, hypopigmented lesions overlying extensor surfaces of interphalangeal joints, with periungual erythema typical of juvenile dermatomyositis.

The muscular involvement is characterized by pain, tenderness, and weakness of proximal muscles in a symmetric fashion with prominent involvement of the anterior neck flexors and sparing of the facial muscles. The disease may progress to involve the muscles of the palate and pharynx, resulting in regurgitation, nasal voice, and aspiration. Involvement of the respiratory muscles may lead to a poor cough, pneumonia, and respiratory failure. Risk of GI hemorrhage and perforation are increased at this stage and are associated with abdominal pain, hematemesis, and melena.

The clinical course of JDMS is variable. Traditionally, patterns such as “limited” or “monocyclic,” with a single period of disease activity, were differentiated from a “chronic” or “polycyclic” pattern of exacerbations and remissions. It now appears that these distinctions are an artifact of inadequate therapy, however, and with newer approaches to treatment, disease manifestations may be controlled within a few months in most patients.

Children who continue to have florid or smoldering muscle inflammation for more than 6 to 12 months are at risk of developing late complications of JDMS. These complications include pronounced muscle wasting, contractures, lipodystrophy, and pigmentary changes of the skin. The rash over the extremities often becomes dry, scaly, and atrophic. Subcutaneous calcifications have historically occurred in up to 30 to 40% of children during this phase, although aggressive treatment of inflammation from the onset of JDMS dramatically lowers this figure. Calcifications are most typically discrete nodules around large joints, but a diffuse encasement of the soft tissues by a shell of calcium, called calcinosis universalis, may occur. Occasionally, children pass through the early stages insidiously and come to the attention of the physician with contractures and calcinosis.

Laboratory Features

Weakness is a consistent manifestation of JDMS, but it is a late, variable, and subjective clinical sign. Objective evidence of muscle inflammation should also be sought by measuring serum levels of muscle enzymes that are released into the circulation when myocytes are injured. A wide variety of enzymes may be elevated in JDMS, including creatine kinase (CK), aldolase, lactate dehydrogenase (LDH), and transaminases (ALT and AST). Interpretation of these markers must be done with caution, however, because none is specific for muscle. Elevated levels may be derived from damage to a variety of other tissues, including hepatocytes, brain cells, and the GI tract. Furthermore, for unknown reasons, many children do not reliably demonstrate elevated muscle enzymes despite significant myositis. This is particularly true during later stages of the disease, when LDH and aldolase may rise during a disease flare-up, but CK levels often remain normal.

Because JDMS is a microangiopathic process, disease activity may also be demonstrated through the presence of elevated levels of factor VIII–related antigen (von Willebrand factor antigen), which is released by damaged endothelial cells. On the other hand, evidence of systemic inflammation may be absent, and acute phase markers (including ESR and CRP) and CBCs may be normal. In particularly difficult cases, MRI of the thighs may be the most sensitive method of documenting muscle inflammation (Fig. 101.7). Evidence of cardiac involvement should also be sought, particularly with an ECG and an echocardiogram. Serologic markers of myocardial involvement are unreliable because both the CK MB fraction and troponin level are often elevated in JDMS due to myoblast proliferation in skeletal muscles.

Management

General Management

The sine qua non of treating JDMS is aggressively controlling muscle inflammation. The more rapidly markers of myocyte damage, such as CK and aldolase, can be normalized, the less the chance that acute and chronic complications will occur. During the initial evaluation of JDMS, it is essential to monitor the function of the palatopharyngeal and respiratory muscles; palatal weakness increases the risk of aspiration. Eating only in the upright position, frequent suctioning, or placement of a nasogastric tube may be necessary to avoid aspiration. Support of weak muscles (e.g., wearing a soft neck collar while riding in automobiles) may also be necessary to avoid complications until children regain their strength.

Recognition of the importance of expeditious disease control is leading to modifications in the medical management of JDMS. Thus, virtually all children with clinical or biochemical evidence of muscle inflammation begin treatment with prednisone at a dosage of 1 to 2 mg/kg per day in two or three divided doses as they traditionally have. If muscle

enzymes, weakness, or GI symptoms do not rapidly improve, however, or if significant steroid toxicity develops, other agents are introduced within 4 to 8 weeks. IV methylprednisolone (30 mg/kg, maximum 1.5 g) infused over 1 to 2 hours ("pulse therapy") is the agent of choice, even when high oral dosages of corticosteroids are ineffective. This drug may prove effective because it bypasses the GI tract, where absorption of orally administered medications may be impaired because of vasculitis.

In more recalcitrant cases, it may be necessary to add cytotoxic drugs such as cyclophosporine (2 to 7 mg/kg per day), azathioprine (1 to 3 mg/kg per day), or methotrexate (0.5 to 1.0 mg/kg weekly). Plasmapheresis may be beneficial in particularly severe cases. Under all circumstances, the goal is to rapidly control disease activity while minimizing toxicity from medications. Fortunately, active disease generally does not recur if a complete remission can be induced and maintained for 1 to 2 years.

Management of Complications and Emergencies (Table 101.7)

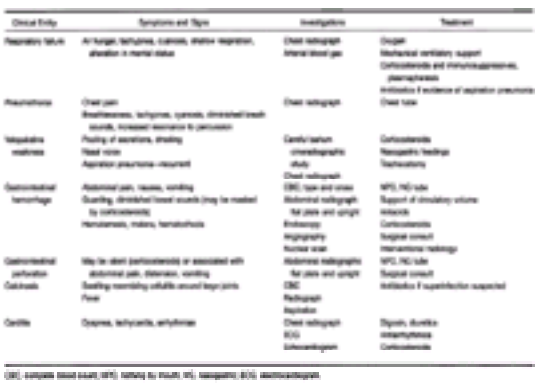

Table 101.7. Complications of Juvenile Dermatomyositis

The most serious emergencies in JDMS relate to the respiratory and GI tracts. In addition, complications occur as a result of therapy with corticosteroids (e.g., infection, GI hemorrhage).

Respiratory Complications. Respiratory emergencies seen in childhood DM have diverse causes. Entities to be considered include 1) aspiration pneumonia secondary to weakness of velopalatine muscles; 2) atelectasis and pneumonia secondary to difficulty in clearing secretions as respiratory muscles become involved; 3) respiratory failure secondary to profound involvement of respiratory musculature, including the diaphragm; 4) progressive interstitial lung disease; and 5) opportunistic infection (tuberculosis, fungi, viruses, or *Pneumocystis*) in the immunocompromised host.

The history should provide clues for differentiating between these possibilities. On physical examination, the child with a respiratory complication is often acutely ill with an elevated temperature. Because fever also may occur with active DM, fever caused by infection must be differentiated from fever caused by underlying disease. The patient with respiratory complications is often dyspneic and tachypneic and has a weak cough with impaired production of sputum. Pooling of secretions in the mouth and a nasal voice should suggest the presence of palatal weakness. Each breath is shallow, with poor air entry on auscultation. The child cannot complete a sentence in one breath and often pauses between words. On auscultation, crackles may be heard. Cyanosis and alteration of consciousness imply impending respiratory failure.

Children with JDMS who develop respiratory problems are usually hospitalized for observation and diagnosis. Those at risk of developing respiratory failure should be cared for in an ICU. Preliminary investigations should include a CBC, urinalysis, serum electrolytes, measurement of muscle enzymes (including CK, aldolase, and LDH), and chest radiograph. Depending on the seriousness of the symptoms and the cooperativeness of the child, blood gas analysis and pulmonary function studies may be obtained. These latter studies should be compared with baseline pulmonary function tests (PFTs) on an individual basis because more than two-thirds of children with DM show restrictive disease and a diffusion abnormality.

If the cause of the respiratory deterioration remains in doubt, more sensitive tests of disease activity, including factor VIII–related antigen and MRI, may be necessary to determine whether more aggressive control of the underlying myositis is necessary. Corticosteroids are the preferred treatment for weakness of respiratory muscles and interstitial lung disease. If the weakness seems to be worsening, maximum efficacy may be obtained with pulse dose methylprednisolone 30 mg/kg (maximum of 1.5 g) in a single IV bolus dose of 50 mL of D5W over 1 to 2 hours. During this pulse therapy, blood pressure and cardiac rhythm should be monitored and the infusion stopped if sudden hypertension or hypotension or a rhythm disturbance occurs. Plasmapheresis is reserved for children who deteriorate even after pulse steroid therapy. Frequent suctioning, nasogastric feeding, and occasionally, tracheostomy may be necessary to avoid aspiration pneumonia.

Aspiration pneumonia can be recognized on a clinical and radiographic basis. A chest radiograph with a severe interstitial or reticulonodular pattern may indicate progression of underlying lung disease or opportunistic infection. Lung biopsy may be helpful in such situations. If pulmonary problems are suspected to result from infection, treatment with IV antibiotics should be initiated after appropriate cultures are obtained. In addition, sufficient corticosteroids (three times physiologic need) are given to cover the child for iatrogenic adrenal insufficiency if he or she has recently received high dosages of steroids.

Pneumothorax is another complication known to occur during the course of childhood DM. The usual symptoms are sudden onset of chest pain and tachypnea. Physical examination shows deviation of the trachea to the opposite side of

the chest, and increased resonance and diminished breath sounds on the affected side. A radiograph of the chest shows air in the pleural cavity. A chest tube should be placed and connected to underwater seal.

Gastrointestinal Complications. Vasculitic changes characterized by intimal hyperplasia, and arteriolar occlusion by fibrin thrombi, are characteristic of severe or poorly controlled JDMS. Arteries and veins of the skin, muscles, and GI tract may be involved. Resultant ulcerations and perforations may occur anywhere from the esophagus to the large intestine, and they may disrupt the integrity of the integument. Symptoms and signs of these complications depend on the site of the lesion. For example, bleeding from the esophagus is uncommon, but perforation may cause mediastinitis. On the other hand, bleeding from ulceration of the stomach or duodenum typically leads to abdominal pain with vomiting and melena. If the bleeding is severe, hematemesis with a sudden drop in the hemoglobin will be the presenting manifestation. Laboratory studies to be obtained include a CBC, electrolytes, and BUN. Endoscopy may prove useful in locating the site of bleeding. Treatment of upper intestinal hemorrhage includes support of circulatory volume and hematocrit, antacids, and H_2 blockers (e.g., cimetidine 20 to 40 mg/kg per day, maximum 1200 mg).

Evidence of bleeding from the lower portion of the GI tract includes abdominal pain, vomiting, a distended abdomen, and melena or bright red blood in the stool. The hematocrit can fall precipitously, and radiographs of the abdomen may show free air in the peritoneum. If bleeding is active, a technetium scan to locate the area of hemorrhage is the initial step. This may be followed by an angiogram to localize the actual vessel that is bleeding. The details of the management of hemorrhage from the GI tract are discussed under SLE and in Chapter 93.

In a patient with JDMS, intestinal perforation may go unnoticed and present with pneumatosis intestinalis. This finding may also precede clinical perforation and pneumoperitoneum. Thus, any patient with JDMS and persistent abdominal pain should be examined radiographically for the presence of pneumatosis intestinalis. In the presence of acute perforation, usual physical findings are abdominal tenderness, guarding of the abdominal wall, and distant or absent bowel sounds. It should be stressed that corticosteroids may mask these physical findings. Supine and erect abdominal radiographs are indicated to demonstrate intramural gas or subdiaphragmatic air. Patients with this diagnosis require admission to the hospital and emergent surgical evaluation.

Calcinosis. During the period of formation of subcutaneous calcification, children with JDMS may develop high fever, chills, and one or more areas of swelling under the skin. The inflammation caused by the subcutaneous calcium deposit may be indistinguishable from that of cellulitis or abscess formation, with warmth, erythema, and tenderness. Eventually, the lesion may spontaneously extrude calcium, at which time the fever often subsides. Although this is the natural history of subcutaneous calcifications, it is often hard to exclude an infectious cause for the swelling. If doubt exists, needle aspiration of the site may be performed and the fluid examined for calcium crystals and organisms. In the face of uncertainty, it is best to treat for infection with antibiotics until culture reports are available. Incision and drainage or surgical debridement should be avoided, however, because the inflamed skin rarely heals satisfactorily.

Cardiac Emergencies. One of the less common complications of childhood JDMS is myocarditis, although ECG abnormalities may be seen in up to 50% of children. Tachycardia out of proportion to fever may be the earliest evidence of this complication. Involvement of the conduction system by edema and fibrosis leads to electrical abnormalities and arrhythmias. All patients with myocarditis should be admitted for an evaluation that includes an ECG, chest radiograph, and echocardiogram. Supportive management includes judicious and careful use of diuretics and cardiotonic drugs while aggressively treating the primary disease.

SCLERODERMA

Background

Systemic sclerosis (SS) is an uncommon disorder occurring in 2 to 12 adults per million per year. It is even less common in children. Morphea and linear scleroderma are more likely to be seen than SS in this age group. Various conditions included under the term scleroderma are given in Table 101.8.

I. Systemic Sclerosis
  A. With diffuse skin thickening
  B. With limited skin thickening (including CREST syndrome)
  C. With overlapping manifestations of other connective tissue disease, including systemic lupus erythematosus, dermatomyositis, rheumatoid arthritis
II. Chemically Induced Conditions
  A. Systemic syndromes seen with vinyl chloride, bleomycin, trichloroethylene
  B. Localized syndromes seen with eosinophilia-myalgia, toxic oil, pentazocine, graft-versus-host disease
III. Localized Scleroderma
  A. Morphea
  B. Linear (includes "en coup de sabre")
  C. Eosinophilic fasciitis
IV. Pseudosclerodermа
  A. Edematous as associated with diabetes mellitus, myxedema
  B. Indurative as in porphyria cutanea tarda, phenylketonuria, acromegaly
  C. Atrophic as in progeria, acrodermatitis chronica atrophicans

Modified with permission from Medsger TA Jr, Steen V. Systemic sclerosis and related syndromes. In: Schumacher HR, ed. Primer on the Rheumatic Diseases. 10th ed. Atlanta, Arthritis Foundation;1993.
CREST, calcinosis, Raynaud's esophageal dysfunction, sclerodactyly, and telangiectasia.

Table 101.8. Classification of Scleroderma and Related Conditions

Pathophysiology

The most characteristic microscopic feature of affected areas of the skin is increased thickness and density of collagen in the dermis. In addition, flattening of rete pegs, mononuclear cell infiltrate around small blood vessels, obliteration of skin

appendages, and hyalinization and fibrosis of arterioles are seen.

The cause of this disease is unknown. Increased collagen production by fibroblasts, perhaps in response to disordered immune regulation and cytokine release, appears to be a final common pathway for a clinical entity with numerous genetic and environmental triggers. Similarities between this disorder and graft-versus-host disease after bone marrow transplantation, and to chronic Lyme disease (acrodermatitis chronica atrophicans) have suggested many areas of research. However, clear understanding of the underlying abnormalities in scleroderma remains elusive.

Clinical Manifestations

Localized scleroderma is more common in children than in adults. The lesions may be one of three types. Morphea is a focal ivory-white patch with a violaceous or erythematous rim; it is often a single lesion on the trunk, although generalized morphea also occurs in children. The linear form of scleroderma causes scarring, fibrosis, and atrophy (not corresponding to dermatomes) of involved skin with firm binding of the subcutaneous tissues to deeper structures ("hidebound" appearance). It may extend to involve an entire extremity (Fig. 101.9) and to affect underlying muscle and bone, leading to flexion contractures, leg-length discrepancies, and atrophy of an extremity. A variant affecting the forehead is called scleroderma en coup de sabre.

FIGURE 101.9. Linear scleroderma involving left lower extremity in a 12-year-old girl.

SS often presents with cutaneous changes such as RP (90% of patients), edema, induration, increased pigmentation, and tightening of the skin. Some of these children also may develop arthritis resembling JRA, muscle weakness resembling JDMS, and nodules along tendon sheaths. If these features are seen, the clinician should consider the possibility of undifferentiated or mixed connective tissue disease, in which features of SLE, SS, JDMS, and JRA overlap.

Serious illness and death can occur in SS. Severe, uncontrolled hypertension and rapidly progressive renal failure (scleroderma renal crisis) have been a major source of mortality, although introduction of ACE inhibitors has dramatically improved short-term survival. Primary myocardial disease with conduction disturbances, pericarditis, and intractable CHF, as well as pulmonary hypertension secondary to fibrosis, remain significant sources of morbidity and mortality. Additional complications of SS include 1) digital gangrene and nonhealing ulcers most often involving the fingers, elbows, and malleoli secondary to vascular occlusion; 2) disordered motility of the distal esophagus with dysphagia and reflux esophagitis (60% of affected children); 3) malabsorption syndrome; 4) thrombocytopenia with subsequent cerebral hemorrhage; 5)interstitial lung disease; and 6) cranial nerve involvement with trigeminal sensory neuropathy, facial weakness, and tinnitus.

Management

General Management

Specific therapy for SS is nonexistent at present. Virtually every medication, from antihistamines to potent immunosuppressives, has been used in patients with this disease, although none has been shown to offer clear benefit. During the inflammatory, prefibrotic stages of interstitial lung disease and pulmonary vascular involvement, corticosteroids (prednisone 2 mg/kg per day or the equivalent) are indicated. Cyclophosphamide appears to forestall pulmonary fibrosis if added early to the treatment regimen. If the esophageal sphincter is involved, patients should be advised to sleep with the head comfortably elevated, and an antacid may be prescribed. Minor episodes of Raynaud's syndrome are managed with prophylactic measures such as the avoidance of cold exposure and the use of warm clothing. Biofeedback training and calcium channel blockers such as nifedipine may be helpful in decreasing the frequency of attacks. Aggressive physical therapy is indicated to prevent contractures and maintain normal function. Despite these measures, linear scleroderma with involvement of deep structures may lead to contractures of the extremities requiring surgery, whereas the mortality rate of systemic scleroderma appears to approach 50% at 7 years.

Management of Complications and Emergencies (Table 101.9)

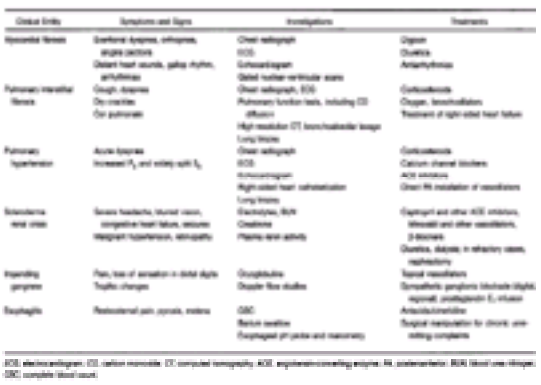

Table 101.9. Complications of Systemic Sclerosis

Cardiac Complications. Signs and symptoms of myocardial fibrosis are essentially those of a cardiomyopathy with dyspnea, orthopnea, and fatigue. Angina pectoris and myocardial infarction also occur. Fibrosis of the conduction system may result in arrhythmias, presenting as palpitations, syncope, or sudden death. Pericarditis is usually silent and valvular involvement in scleroderma is rare. Even in the absence of symptoms or physical findings, cardiac involvement eventually develops in most patients with systemic sclerosis, and sensitive imaging or functional studies reveal some cardiac involvement early in the course of disease in most cases. Management of cardiac dysfunction is symptomatic, including inotropic support and afterload reduction. Extensive diuresis should be avoided because of potential adverse effects on renal cortical perfusion. No specific drugs are available to arrest the progress of cardiac involvement.

Pulmonary Complications. Pulmonary involvement in SS may have three manifestations: pleurisy, interstitial lung disease, or pulmonary artery fibrosis. Diffuse interstitial lung disease is often asymptomatic. A dry cough may be the earliest symptom. Early in the course of the disease, even before symptoms appear, PFTs in these patients show a restrictive pattern and diffusion abnormalities. Later, radiographs of the chest show increased reticulation, a so-called honeycombed appearance, mainly basilar and bilateral. Other diagnostic modalities, including high-resolution CT scanning, bronchoalveolar lavage, and lung biopsy may identify earlier, prefibrotic states of disease more responsive to anti-inflammatory therapy. With progression of the disease, cough and dyspnea become prominent. On examination, crackles over both sides of the chest, particularly over the infrascapular area, may be the only finding. With the onset of right-sided heart failure, these patients may have increasing dyspnea, although edema of the lower extremities may not be appreciated because of hidebound skin. Patients with right-sided heart failure require admission and symptomatic management.

Patients with irreversible pulmonary fibrosis and chronic respiratory failure also may need admission. If such patients contract intercurrent respiratory infections, they should be treated promptly. Supplemental oxygen, bronchodilators, and corticosteroids may be helpful. If residual inflammation is demonstrable after further investigations such as those previously noted, these patients should receive corticosteroids (prednisone 2 mg/kg per day) for 6 to 8 weeks, although the value of this therapy is doubtful in established fibrosis. In addition, treatment of right-sided overload is indicated.

Pulmonary hypertension is the most common cause of dyspnea in patients with SS. On auscultation, the pulmonic component of the second heart sound is accentuated, and there is a wide or fixed splitting of the second heart sound. The ECG shows right ventricular hypertrophy. Echocardiography and catheterization of the right side of the heart are often indicated to document the etiology of respiratory deterioration. Corticosteroids and cyclophosphamide (50 mg/day orally or 500 to 750 mg/m_2 by monthly IV infusion) are the treatment of choice in patients without established interstitial fibrosis, in addition to supportive measures. Calcium channel blockers, ACE-inhibitors, and prostaglandin analogs may provide temporary symptomatic improvement in individual cases.

Renal Complications. Sclerodermatous involvement of the vessels of the kidney is the most common cause of renal failure in adults with SS. Proteinuria, hypertension, rapid progression of skin thickening early in the illness, anemia, pericardial effusion, and CHF are all markers of the patient at risk for renal scleroderma. The development of a microangiopathic hemolytic anemia suggests imminent renal failure. These complications appear to be less common in children than in adults.

Renal failure may develop gradually or acutely in a patient with known renal disease, and use of corticosteroids may precipitate its appearance. The combination of rapidly progressing azotemia with malignant hypertension (scleroderma renal crisis) requires hospitalization and urgent management. Characteristically, a patient displays a sudden rise in blood pressure to levels as high as 150 to 200 mm Hg diastolic, often without symptoms or heralded only by headache. Evaluation reveals hypertensive retinopathy (flame hemorrhages, cotton wool exudates, and papilledema), elevated plasma renin activity, and rapid deterioration of renal function. Immediate investigation should include urinalysis, measurement of urine output and urinary electrolytes, serum electrolytes, BUN, creatinine, and plasma renin level.

A major advance in the pharmacologic management of scleroderma renal crisis has been the use of ACE inhibitors such as captopril. Patients who fail to respond to this drug may still respond to potent vasodilators such as minoxidil, along with b-blockers and diuretics; regimens involving multiple drugs may also be necessary (see Chapter 35). Renal dialysis and, rarely, bilateral nephrectomy may be indicated in hypertension unresponsive to pharmacologic therapy. Maintenance of blood volume is an essential part of management of these patients.

Peripheral Vascular Complications. RP often can be incapacitating, particularly in cold weather. Symptoms include severe pain in the extremities and loss of sensation in the tips of the digits. Treatment with calcium channel antagonists such as slow-release nifedipine (30 to 180 mg/day) may decrease the frequency or severity of attacks. In urgent cases with impending gangrene, systemic or topical vasodilators (e.g., nitroglycerin paste or intra-arterial reserpine) or

sympathetic ganglion block may be tried, although these forms of therapy have not been validated in well-constructed studies. They also may be associated with complications, so these procedures should be performed only with intensive monitoring. If gangrene has set in, it is best left alone if no infection is present. Spontaneous separation of the tips of the digits will occur and carries less risk and morbidity than surgical amputation.

Gastrointestinal Complications. Abnormal esophageal motility with reflux may result in esophagitis. The major symptom of this condition is retrosternal pain that is made worse by certain foods and recumbent positioning. The pain may be severe and incapacitating. Although children with the complaint of retrosternal pain do not require admission to the hospital, they need an evaluation of their lower esophageal sphincter with esophageal manometry. Those with mild pain and objective manifestations of reflux (lower esophageal sphincter pressure of less than 10 mm Hg, evidence of esophagitis on endoscopy) are usually treated with simple measures such as antacids 1 hour after meals and 1 hour before bedtime, and elevation of the head during sleep. If symptoms are severe, H_2 blockers such as cimetidine (20 to 40 mg/kg per day, maximum daily dosage 1200 mg) may be prescribed.

VASCULITIS

Background

Vasculitis is rare in children, and one result of this has been insubstantial and inconsistent data on the classification and prognosis of childhood vasculitides. Classification schema useful in adults have limited applicability to children because certain illnesses are unknown in the young (e.g., temporal arteritis, Churg-Strauss disease), whereas other vasculitides occur only in children (e.g., Kawasaki disease). Classification is hampered further by a paucity of pathogenetic data on the vasculitides; distinction between the entities is based instead on a combination of clinical and histologic criteria. Finally, the recent discovery that antineutrophil cytoplasm antibodies (ANCA) are central to several forms of vascular inflammation has caused many old categorization schema to become outdated. One classification that is suitable for use in children is shown in Table 101.10. The most common of the life-threatening vasculitides (polyarteritis nodosa [PAN] and Kawasaki disease) are discussed here.

Leukocytoclastic vasculitis	Polarteritis nodosa
Henoch-Schönlein purpura	Systemic
Hypersensitivity vasculitis	Cutaneous
Allergic granulomatous angiitis	Kawasaki disease
Churg-Strauss syndrome	Giant cell arteritis
ANCA-positive vasculitis	Temporal arteritis
Wegener's granulomatosis	Takayasu's arteritis
Microscopic periarteritis	Isolated central nervous system angiitis
Pauci-immune glomer-	Vasculitis associated with rheumatic
ulonephritis	disease

ANCA, antineutrophil cytoplasm antibody.

Table 101.10. Necrotizing Vasculitis

Polyarteritis Nodosa

Background

The annual incidence of PAN in adults is approximately 0.3 per 100,000; no comparable data are available for children. Before the introduction of corticosteroids for the treatment of PAN, mortality rates as high as 100% were reported; today, fewer than 1 in 5 cases is believed to have a fatal outcome.

Pathophysiology

PAN is characterized by focal, panmural, necrotizing inflammation of small- and medium-sized muscular arteries. Sites of bifurcation are particularly prone to involvement, presumably because of hemodynamic turbulence at these points. Biopsies reveal a cellular infiltrate initially predominated by polymorphonuclear leukocytes and fibrinoid necrosis. As lesions mature, mononuclear cells, thrombosis, and recanalization mark the healing process.

The cause of PAN is unknown, although it is considered an archetype of immune complex–mediated vascular damage. Most children with PAN have serologic evidence of an antecedent streptococcal infection; up to one-third of adults have PAN associated with hepatitis B or C, including chronic antigenemia with viral proteins demonstrable in the circulating and fixed immune complexes. The incidence of hepatitis-associated PAN in children is significantly lower.

Clinical Manifestations

Childhood PAN occurs in both cutaneous and generalized forms, and distinguishing between them may be difficult. Both types display systemic manifestations, including fever, malaise, and myalgias. However, generalized PAN is significantly more likely to also involve the renal, GI, and central nervous systems. Common to both are rashes, although these are more likely to be nodular or lacy (so-called livedo reticularis) in the cutaneous form, and urticarial, petechial, or ischemic in the systemic form. Renal involvement (including proteinuria, abnormal urinary sediment, and hypertension), abdominal pain (often a manifestation of gut vasculitis), arthritis, mononeuritis multiplex, and CNS involvement (seizures, hemiparesis) typify generalized PAN. Less commonly, children may have cardiac disease (pericarditis, cardiomegaly,

ECG changes, myocardial infarction) or pulmonary involvement (diffuse infiltrates, pulmonary hemorrhage, or hemothorax). A rare subtype of PAN, Cogan's syndrome, is characterized by interstitial keratitis and sensorineural hearing loss.

Laboratory Features

Laboratory findings in polyarteritis are nonspecific. Most children have WBC counts greater than 15,000/mm $_3$, hemoglobin less than 10 g/dL, broadly elevated acute-phase reactants, and hypergammaglobulinemia. Complement levels are usually normal or increased, and ANA and rheumatoid factor levels are elevated only slightly if at all. Some children have evidence of ANCA, although other autoantibodies are usually absent.

Diagnosis of PAN generally requires tissue confirmation. Acute necrotizing inflammation of small and medium arteries is demonstrable in renal, cutaneous, muscular, or GI tissues. At times, biopsy may not be practical, and angiographic visualization of aneurysms may provide an alternative to tissue samples. Prognosis seems to improve if treatment is initiated early, so absence of tissue confirmation should not delay therapy. Vasculitis in general, and PAN in particular, is a pleomorphic condition with no diagnostic laboratory findings. Clinicians must have a high level of suspicion and be willing to subject their patients to invasive diagnostic procedures to avoid significant morbidity from delayed diagnosis.

Management

General Management

The initial management of PAN should include corticosteroids (generally divided doses of prednisone 2 mg/kg per day to a maximum of 80 mg/day). Rash and constitutional symptoms improve first, followed by control of end-organ involvement. Pulse doses of methylprednisolone (30 mg/kg in 50 mL of D5W by IV infusion over 1 to 2 hours, maximum dose 1500 mg) may offer an alternative for the treatment of acute exacerbations, provided that blood pressure and cardiac rhythm are closely monitored.

Cutaneous PAN may require lower dosages of steroids to suppress disease activity, or alternative agents such as dapsone or colchicine may adequately control the rash. On the other hand, children with systemic PAN may not tolerate a reduction in their steroid dosage or may not respond adequately to steroids. In such cases, addition of cytotoxic agents (e.g., oral or IV cyclophosphamide) may improve the outcome. Other modalities, including NSAIDs for fever and arthritis, anticonvulsants, antihypertensives, and physical therapy, should be used when appropriate.

Management of Complications and Emergencies (Table 101.11)

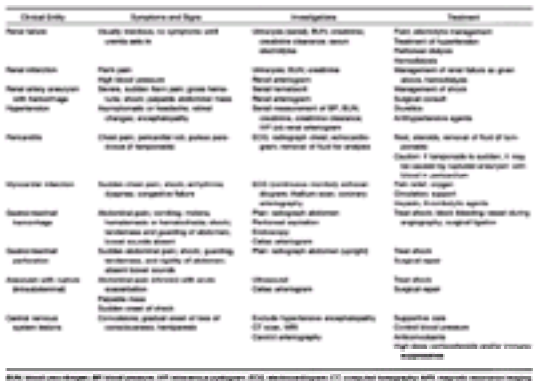

Table 101.11. Complications of Polyarteritis Nodosa

The most serious emergencies in childhood polyarteritis are 1) renal insufficiency; 2) severe hypertension; 3) cardiac complications such as CHF, myocardial infarction, and arrhythmias; 4) GI vasculitis resulting in bowel infarction, intestinal perforation, or cholecystitis; and 5) CNS manifestations, such as seizures and cranial nerve palsies.

Renal Emergencies. Although medical management of PAN has resulted in a significantly improved prognosis, azotemia and hypertension at the time of diagnosis continue to identify children with extremely aggressive disease. Arteritis of medium-sized vessels of the kidney may lead to renal infarction and ischemia or to glomerulonephritis manifested by hematuria, hypertension, and uremia. Sudden flank pain associated with gross hematuria, falling blood pressure, and an expanding abdominal mass suggest the possibility of aneurysmal dilation and rupture, with renal artery hemorrhage.

Serial urinalyses, measurements of BUN and creatinine levels, and determination of creatinine clearance are essential components of the investigation of all patients with PAN. Management of renal failure includes correction of fluid and electrolyte abnormalities, as well as high dosages of corticosteroids to control the underlying disease process (e.g., prednisone 2 mg/kg per day). Rupture of a renal artery aneurysm initially is managed with treatment of shock and replacement of volume, followed by surgical repair of the aneurysm once the patient is stabilized.

Hypertension. A mild to moderate elevation of blood pressure is noted in more than 90% of children with generalized PAN. Diuretics, hydralazine, and b-blockers are preferred drugs for the management of hypertension. Severe hypertension associated with encephalopathy or CHF requires inpatient management.

Cardiac Emergencies. Pericarditis may be asymptomatic. Alternatively, chest pain (particularly in the recumbent

position), shortness of breath, pericardial friction rub, and pulsus paradoxus may be present. The ECG may reveal depression of the ST segment and T-wave inversion, and chest radiograph may demonstrate globular enlargement of the cardiac silhouette. However, echocardiographic demonstration of pericardial fluid is the most sensitive means of confirming the presence of pericarditis.

Chest pain with tachycardia, arrhythmia, and dyspnea may herald the occurrence of myocardial infarction in a patient with PAN. Pericardial tamponade caused by a ruptured coronary aneurysm may present similarly. Occasionally, a patient with coronary disease may present with CHF. Characteristic ECG changes (deep Q waves) and areas of ischemia on myocardial nuclear scanning may be seen. Echocardiogram is indicated to study the function of the myocardium and the status of the valves. Coronary arteriography is essential to establish the size, location, and extent of aneurysms and occlusions.

Patients with CHF, myocardial ischemia, and arrhythmias will require continuous monitoring and urgent management in an intensive care setting. Patients with pericarditis without effusion may be treated with bed rest, careful monitoring, and corticosteroids. Pericardiocentesis is indicated in the presence of tamponade. Patients with myocardial infarction need careful monitoring, pain relief (morphine), treatment of shock, antiarrhythmic agents, and treatment of CHF. A radiograph of the chest, ECG, and echocardiogram should be obtained as soon as possible; thallium scan and coronary angiography may be indicated in certain patients.

Supportive medical management includes careful monitoring of cardiorespiratory status, judicious use of IV fluids, diuretics, and cardiotonics when needed. Treatment of the primary disease with steroids and cytotoxic agents should be continued as described. If hypertension does not respond to diuretic therapy, other antihypertensive agents may have to be added. Serum electrolytes should be monitored because most patients will be taking high dosages of steroids, diuretics, and antihypertensive agents, and electrolyte imbalances increase the risk of toxicity.

Gastrointestinal Complications. Abdominal pain is the most common manifestation of GI involvement in PAN. It may be diffuse and nonspecific or localized and severe. Hematemesis and melena suggest ulceration and hemorrhage. Patients with persistent abdominal pain, hematemesis, and melena require immediate admission.

Visceral perforation should be suspected in cases of active systemic disease and unrelenting abdominal pain. Tenderness on palpation of the abdomen, guarding of the abdominal wall, and absent bowel sounds are the usual physical findings, although they may be masked by steroid therapy. Arteritis of specific organs may lead to cholecystitis, pancreatitis, appendicitis, and hepatitis. These complications are generally manifested by vomiting and localized abdominal pain and tenderness.

Mesenteric thrombosis with infarction of the bowel may present with sudden abdominal pain, vomiting, hematemesis or hematochezia, and shock. Exquisite tenderness of the abdomen and absent bowel sounds are the major findings. Hemorrhage from a ruptured aneurysm (mesenteric, hepatic, or renal) with hemoperitoneum is heralded by sudden onset of severe pain, vomiting, tachycardia, and shock. The abdomen is tender and tense, and bowel sounds are diminished or absent.

Initial management of each of these GI catastrophes includes volume replacement, gastric decompression, and stress doses of corticosteroids. All such patients will require measurement of intake and output and serial determination of hematocrit, BUN, and electrolytes. Abdominal radiographs (supine and upright), abdominal ultrasound, technetium scan, angiography of the celiac axis vessels, and peritoneal aspiration may be indicated in some cases. In selected instances, direct examination of the GI tract by endoscopy may yield valuable information concerning the nature, location, and extent of lesions. Surgical consultation should be obtained immediately, and in the presence of bleeding aneurysms or infarcted bowel, surgical exploration is mandatory.

Central Nervous System Complications. Clinical signs of CNS disease are less common than those of peripheral nervous system involvement. Seizures and hemiparesis are the most common manifestations of CNS involvement in PAN and require immediate hospitalization. A complete neurologic evaluation, including measurement of blood pressure, and funduscopic examination for evidence of hypertension or intracranial bleeding, should be performed. CT and/or carotid angiography may help localize the lesion.

Management of hypertensive encephalopathy and increased intracranial pressure are described elsewhere (see Chapter 35 and Chapter 83). Surgical correction of a ruptured aneurysm should be undertaken if the bleeding vessel can be localized and is accessible.

KAWASAKI DISEASE

Background

Mucocutaneous lymph node syndrome was first described by a Japanese pediatrician in 1967, and it has become known as Kawasaki disease in his honor. In fact, the condition certainly predates this description: a preserved heart from the 19th century shows pathologic changes characteristic of Kawasaki disease, and the entity of “infantile polyarteritis” probably represents the same syndrome. Kawasaki disease is an idiopathic vasculitis of small- and medium-sized vessels, which has become a leading cause of acquired heart disease in children in the United States. Characteristically, children with Kawasaki disease have fever, conjunctivitis, rash, mucosal inflammation, lymphadenopathy, and extremity changes. However, the major morbidity of Kawasaki disease occurs in the heart. Coronary artery aneurysms or ectasia develop in approximately 15 to 25% of untreated children and may lead to myocardial infarction, sudden death, or chronic coronary artery insufficiency. IV gamma-globulin decreases the incidence of coronary artery aneurysms by threefold to fivefold if given within 10 days of disease onset. Therefore, management of children with suspected Kawasaki disease requires accurate and expeditious diagnosis and close monitoring of the cardiovascular system.

Pathophysiology

In Kawasaki disease as in other vasculitides, blood vessel damage appears to result from an aberrant immune response leading to endothelial cell injury and vessel wall damage. Humoral factors, such as antiendothelial cell antibodies or circulating immune complexes, may be critical. On the other hand, a direct cell-mediated attack on endothelial cells that are infected with an as yet unidentified infectious agent may underlie the vascular injury. For unknown reasons, Kawasaki disease has a predilection for the coronary arteries.

The pathologic changes of coronary arteries in Kawasaki disease have been classified by Fujiwara and Hamashima into four stages, depending on the duration of illness at the time of examination (Table 101.12). Initially, endothelial swelling is accompanied by a neutrophilic infiltrate. Lymphocytes and plasma cells replace polymorphonuclear cells by the subacute stage (beginning 2 weeks after onset), accompanied by destruction of the internal elastic lamina; coronary artery aneurysms characteristic of Kawasaki disease first become apparent at this time. Finally, during the convalescent state of Kawasaki disease, healing of the vascular lesions occurs with fibroelastic proliferation and scar formation, along with expansion of aneurysms caused by hemodynamic forces.

Stage I—Disease duration <10 days
Acute perivasculitis of coronary arteries
Microvascular angiitis of coronary arteries and aorta
Pancarditis with pericardial, myocardial, endocardial inflammation
Inflammation of the atrioventricular conduction system
Stage II—Disease duration 12–28 days
Acute panvasculitis of coronary arteries
Coronary artery aneurysms present
Coronary obstruction and thrombosis
Myocardial and endocardial inflammation less intense
Stage III—Disease duration 28–45 days
Subacute inflammation in coronary arteries
Coronary artery aneurysms present
Myocardial, endocardial inflammation much decreased
Stage IV—Disease duration >50 days
Scar formation, calcification in coronary arteries
Stenosis and recanalization of coronary vessel lumen
Myocardial fibrosis without acute inflammation

From Melish ME. Kawasaki syndrome. Pediatr Rev 1980;2:111.

Table 101.12. Pathology of Kawasaki Disease (Adapted from Fujiwara)

The cause of Kawasaki disease is unknown, although many lines of evidence point toward an infectious origin. The fact that the disease often occurs in epidemics, that boys are more susceptible than girls, and that studies in Japan suggest that household contacts of children with Kawasaki disease are at increased risk for developing the disease, all point to a transmissible agent. Nonetheless, although many putative causes have been proposed during the past three decades, suggestions that certain viruses (EBV, parvovirus, human immunodeficiency virus [HIV]-2) or bacterial toxins (streptococcal erythrogenic toxin, staphylococcal toxic shock toxin) account for most cases have not been substantiated. Many researchers now believe that Kawasaki disease represents a final common pathway of immune-mediated vascular inflammation after a variety of inciting infections.

Clinical Manifestations

Kawasaki disease is a clinical syndrome diagnosed on the basis of fever and four of five signs of mucocutaneous inflammation (Table 101.13). These guidelines were established by Tomisaku Kawasaki in 1967, and they remain the sine qua non for diagnosing Kawasaki disease. Nonetheless, as with all clinical criteria, these should be regarded as imperfect guidelines with less than 100% sensitivity and specificity. Children who do not meet criteria may indeed have Kawasaki disease, and some children with other conditions may nonetheless manifest 5 or 6 criteria of Kawasaki disease.

I. Fever ≥5 days unresponsive to antibiotics If the fever disappears because of intravenous gamma-globulin (IVGG) therapy before the fifth day of illness, a fever of <5 days' duration fulfills fever criterion for case definition.
II. At least four of the five following physical findings with no other more reasonable explanation for the observed clinical findings: 1. Bilateral conjunctival injection 2. Changes in the oropharyngeal mucous membranes (erythematous and/or fissured lips, strawberry tongue, injected pharynx) 3. Changes of peripheral extremities, including erythema and/or edema of the hands or feet (acute phase) or periungual desquamation (convalescent phase) (Fig. 101.12) 4. Polymorphous rash, primarily truncal; nonvesicular 5. Cervical lymphadenopathy ≥1.5 cm diameter

Modified from Centers for Disease Control and Prevention. Kawasaki disease—New York. MMWR 1990;39 (RR-13):17–18.

Table 101.13. Diagnostic Criteria for Kawasaki Disease

Fever is probably the most consistent manifestation of Kawasaki disease. It reflects the elevated levels of proinflammatory cytokines (e.g., TNF, IL-1), which are also thought to mediate the underlying vascular inflammation. A diagnosis of Kawasaki disease should be considered in all children with prolonged, unexplained fever, irritability, and laboratory signs of inflammation, especially in the presence of mucocutaneous inflammation. Conversely, the diagnosis must be suspect in the absence of fever.

The remaining cardinal manifestations of Kawasaki disease vary considerably in frequency. Up to half of children with Kawasaki disease do not have cervical lymphadenopathy, especially children under 2 years of age. When present, lymphadenopathy tends to involve the anterior cervical nodes overlying the sternocleidomastoid muscle. Diffuse lymphadenopathy, as well as other signs of reticuloendothelial involvement such as splenomegaly, should prompt a search for an alternative diagnosis.

Bilateral, nonexudative conjunctivitis is present in more than 90% of patients. A predominantly bulbar injection typically begins within days of the onset of fever, and eyes eventually develop a brilliant erythema, which spares the limbus. Children are also commonly photophobic, and five of six patients have evidence of anterior uveitis during the first week of illness. Consequently, in ambiguous cases, slitlamp examination may be helpful in confirming a diagnosis of Kawasaki disease.

Cracked, red lips and a strawberry tongue are characteristic of the mucositis typically seen during the first week of Kawasaki disease (Fig. 101.10). Discrete oral lesions, such as vesicles or ulcers, and tonsillar exudate, suggest a viral or bacterial infection rather than Kawasaki disease. The cutaneous manifestations of Kawasaki disease are polymorphous. The rash typically begins as perineal erythema and desquamation, followed by macular, morbilliform, or targetoid lesions of the trunk and extremities. Vesicular or bullous lesions are rare. Changes in the extremities are generally the last clinical manifestation of Kawasaki disease to develop. Children demonstrate an indurated edema of the dorsum of their hands and feet, and a diffuse erythema of their palms and soles (Fig. 101.11). In addition, one-third of children develop arthritis. This is typically a small joint polyarthritis during the first week of illness, followed by a large joint pauciarthritis. During the convalescent phase of Kawasaki disease, sheetlike desquamation that begins in the periungual region of the hands and feet, and linear nail creases (Beau's lines) are characteristic (Fig. 101.12).

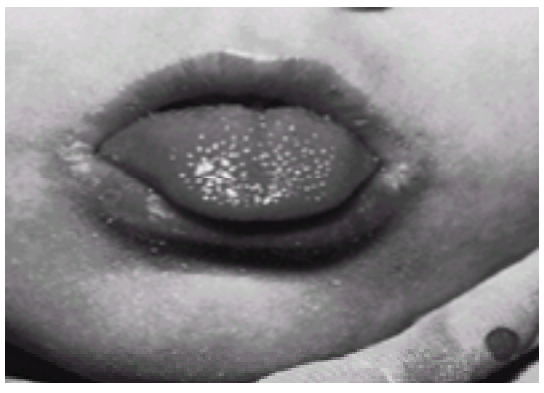

FIGURE 101.10. Cracked, erythematous lips and “strawberry” tongue in Kawasaki disease.

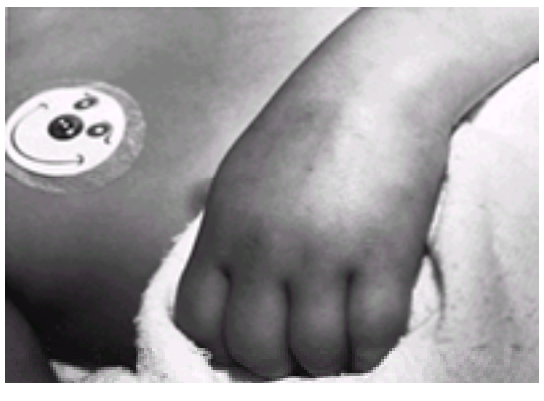

FIGURE 101.11. Brawny edema of dorsum of hand and small joint polyarthritis in Kawasaki disease.

FIGURE 101.12. Peeling of the skin over the thumbs in Kawasaki disease.

Kawasaki disease is most commonly confused with exanthematous infections of childhood (Table 101.14). Measles, echovirus, and adenovirus may share many of the signs of mucocutaneous inflammation, but they typically have less

evidence of systemic inflammation and generally lack the extremity changes seen in Kawasaki disease. Toxin-mediated illnesses, especially b-hemolytic streptococcal infection and toxic shock syndrome, generally lack the ocular and articular involvement typical of Kawasaki disease. Finally, drug reactions such as Stevens-Johnson syndrome or serum sickness may mimic Kawasaki disease but with subtle differences in the ocular and mucosal manifestations.

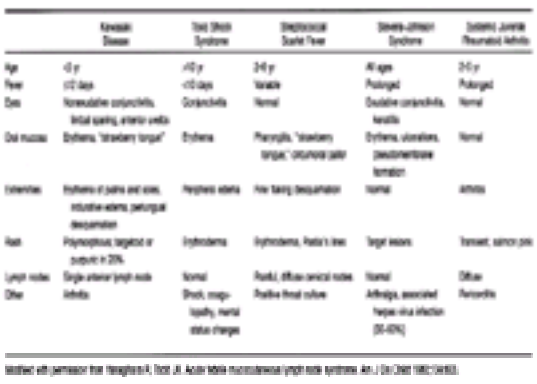

Table 101.14. Differential Diagnosis of Kawasaki Disease

The conventional diagnostic criteria are particularly useful in preventing over diagnosis, but they may result in failure to recognize incomplete forms of the illness. Clinical manifestations of Kawasaki disease tend to be most incomplete and atypical in the youngest patients, the subgroup at highest risk for development of coronary artery abnormalities. Thus, Kawasaki disease should be considered in any infant with prolonged, unexplained fever. On the other hand, alternative explanations for the child's symptoms must be carefully excluded before treating empirically with intravenous immunoglobulin (IVIG). Consideration should be given to referring such children to a regional Kawasaki disease center for further evaluation.

Laboratory Features

No laboratory studies are included among the diagnostic criteria for Kawasaki disease, but certain findings may support the diagnosis. Most characteristic is the systemic inflammation, with widespread elevation of acute-phase reactants (including CRP and ESR), leukocytosis, and a shift to left in the WBC count. By the second week of illness platelet counts rise as well, reaching 1,000,000/mm_3 in the most severe cases.

Children with Kawasaki disease often present with a normocytic, normochromic anemia; hemoglobin concentrations more than 2 standard deviations below the mean for age are noted in approximately half of patients within the first 2 weeks of illness. Urinalysis commonly reveals WBCs on microscopic examination; the cells are mononuclear and therefore are not detected by dipstick tests for leukocyte esterase. They also originate in the urethra, so they will be missed on urinalyses obtained by bladder tap or catheterization. Measurement of liver enzymes often reveals elevated transaminase levels or mild hyperbilirubinemia caused by intrahepatic congestion. In addition, a minority of children may develop obstructive jaundice from hydrops of the gallbladder. If sampled, other body fluids demonstrate inflammation as well: CSF typically displays a mononuclear pleocytosis (less than 100 cells/mm_3) with normal glucose and protein concentrations, whereas arthrocentesis of involved joints demonstrates 50 to 300,000 WBCs/mm_3, primarily neutrophils.

Management

General Management

IVIG has truly revolutionized the care of children with Kawasaki disease; treatment within 10 days of onset significantly shortens disease duration and minimizes the incidence of complications. Overall, prompt diagnosis and appropriate therapy prevent aneurysm formation in approximately 95% of children and result in rapid symptomatic improvement in about 90%. Use of anti-inflammatory medications, such as aspirin or NSAIDs, improves patient comfort and complements the disease-modifying effects of IVIG.

Studies in Japan were the first to suggest relative protection from coronary artery aneurysms when IVIG is administered early in the course of Kawasaki disease. Since then, further trials in the United States and Japan have confirmed this finding and documented the safety of high-dose infusions of immunoglobulin. Most recently, a single large infusion of IVIG (2 g/kg) administered over 8 to 12 hours has become the standard of care for Kawasaki disease. This is somewhat more effective, and equally as safe, as traditional 4-day infusions, and significantly shortens the duration of hospitalization as well.

Therapy with IVIG also has other benefits. Treatment results in a reduced prevalence of giant aneurysms, the most serious form of coronary abnormality caused by the disease, and accelerates normalization of abnormalities of left ventricular systolic function and contractility. Finally, high-dose IVIG reduces fever and laboratory indices of inflammation, suggesting a rapid, generalized anti-inflammatory effect in addition to specific cardioprotective effects. Several different preparations of IVIG are currently available, but data are insufficient to determine whether all are equally effective.

Despite its advantages, IVIG is an expensive and potentially toxic intervention. The greatest long-term concern is of transmission of bloodborne pathogens. Elaborate sterilization procedures, including lyophilization, pasteurization, and addition of solvent detergents, are generally effective in rendering the product free of infectious agents. Nonetheless,

technical errors apparently led to more than 100 cases of hepatitis C in recipients of a single brand of IVIG in 1994, although none were children with Kawasaki disease. Overall, however, significant toxicity is rare, and benefits clearly outweigh risks in children with confirmed Kawasaki disease.

Aspirin

Aspirin was the first medication to be used for treatment of Kawasaki disease, both for its anti-inflammatory and its antithrombotic effects. High-dose (more than 80 mg/kg per day) and lower-dose regimens (30 mg/kg per day) are still used in conjunction with IVIG during the acute phase of the illness, despite the fact that aspirin has no known effect on development of coronary artery aneurysms. Once fever resolves, patients are generally switched to antiplatelet doses of aspirin (3 to 5 mg/kg per day). Unless coronary artery abnormalities are detected by echocardiogram, aspirin is discontinued once laboratory studies return to normal, usually within 2 months of the onset of Kawasaki disease.

The risks of aspirin appear to be similar to those reported in other settings: transaminitis, chemical hepatitis, transient hearing loss, and rarely, Reye syndrome. These risks may even be increased in Kawasaki disease: 1) Aspirin-binding studies have suggested that the hypoalbuminemia of children with Kawasaki disease predisposes them to toxic free salicylate levels despite measured (bound) values within the therapeutic range. 2) At least one case of Reye syndrome has been reported after 6 days of aspirin therapy for Kawasaki disease. 3) Alternative antipyretic and anti-inflammatory agents, such as ibuprofen, may be used for prolonged or debilitating fever, and aspirin should be rapidly discontinued on exposure to or signs of varicella or influenza.

Cardiovascular Complications and Emergencies

Cardiac abnormalities dominate the pathology of Kawasaki disease. Clinical examination is often remarkable for tachycardia and gallop rhythms that are more prominent than expected from the degree of fever and anemia. The ECG in acute Kawasaki disease may show mild abnormalities consistent with myocarditis, most commonly a prolonged P-R interval and nonspecific ST- and T-wave changes. Echocardiographic evaluation of myocardial function early in the course of the disease often reveals reduced left ventricular function and contractility. Rarely, myocardial inflammation may progress to frank CHF. The severity of myocarditis does not correlate with the risk of coronary artery aneurysms or with the pericardial effusion that may develop during the second week of illness. The effusion only rarely progresses to tamponade and resolves spontaneously in most instances. Valvulitis presenting as either aortic or mitral regurgitation is seen in a percentage of children during the early phases of Kawasaki disease. Late-onset mitral regurgitation, from papillary muscle dysfunction or myocardial infarction, may also complicate the clinical course.

Most characteristic of Kawasaki disease is inflammation of the coronary arteries. This progresses to ectasia or aneurysm formation in 15 to 25% of untreated children. Male gender, age less than 1 year, prolonged fever, dramatic elevation of CRP and absolute band count, and pronounced depression of albumin level identify children at greatest risk for developing coronary artery aneurysms. Dilation of coronary arteries may be detected by echocardiography as early as 6 days after the appearance of fever and usually peaks 3 or 4 weeks after onset of illness. Cardiac catheterization need not be performed in patients with normal echocardiograms and ECGs throughout the disease course because the likelihood of finding unsuspected lesions is negligible.

Coronary aneurysms in early Kawasaki disease usually occur in the proximal segments of the major coronary vessels; abnormalities that occur distally are almost always associated with proximal coronary dilation. Aneurysms may also occur in arteries outside the coronary system, most commonly the subclavian, brachial, axillary, iliac, or femoral vessels, and occasionally in the abdominal aorta and renal arteries. Therefore, abdominal aortography and subclavian arteriography are often performed in patients undergoing coronary arteriograms for Kawasaki disease. However, visceral vessels are almost never involved.

Management of Complications and Emergencies

Myocardial infarction caused by thrombotic occlusion of an aneurysmal and/or stenotic coronary artery is the principal cause of death in Kawasaki disease (Fig. 101.13 and Fig. 101.14). Mortality resulting from Kawasaki disease has decreased from almost 2% to less than 0.1% as a result of improved treatment. Nonetheless, most deaths continue to occur during the first 6 months after disease onset, when myocardial and coronary artery inflammation are greatest. A Japanese registry of 195 children with myocardial infarction revealed that almost 40% infarcted within 3 months of disease onset, and 74% had their infarctions during the first year after onset of Kawasaki disease. About two-thirds of myocardial infarctions were associated with symptoms (shock, crying, chest or abdominal pain, vomiting, dyspnea, or arrhythmia), but only three patients had a history of antecedent angina.

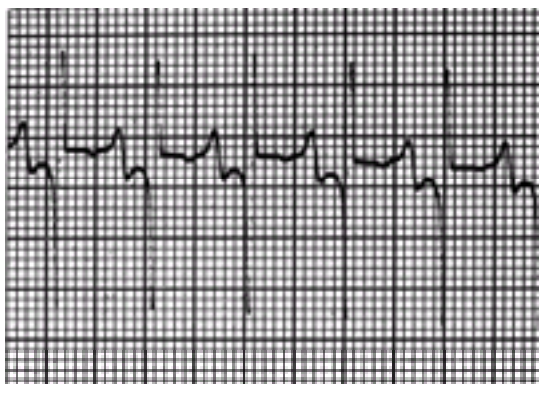

FIGURE 101.13. Lead III from an electrocardiogram on a 15-month-old boy showing deep Q waves in Kawasaki disease.

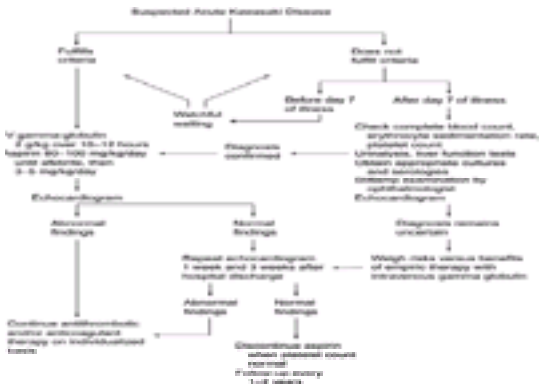

FIGURE 101.14. Algorithm for evaluation of patients with suspected Kawasaki disease. (Modified with permission from Sundel RP, Newburger JW. In: Cooke JP, Frohlich ED, eds. Current Management of Hypertensive and Vascular Diseases. St Louis: Mosby,1992:258.)

Treatment with thrombolytic agents in adults with myocardial infarction has been demonstrated to decrease mortality and improve function. In children with Kawasaki disease and coronary artery thrombosis, thrombolytic agents—mainly urokinase and streptokinase, either IV or intracoronary—have been used with variable success. Thrombolytic therapy for coronary artery thrombosis is most effective if begun within 3 to 4 hours of symptom onset. Immediately after clot lysis, systemic heparin is begun in combination with aspirin. Maintenance of reperfusion then requires long-term oral antithrombotic therapy (e.g., warfarin with dipyridamole), although the ideal regimen has not been established.

CHF may rarely complicate the acute phase of Kawasaki disease. When this is caused by myocarditis, routine treatment with IVIG generally results in rapid clinical improvement. Although IVIG therapy involves infusing large volumes of isotonic solution-- 2 g/kg of 5% IVIG delivers 40 mL/kg over 8 to 12 hours—improvements in myocardial contractility compensate for the volume load, and treatment rarely leads to circulatory deterioration. By the second week of illness, and especially in children with coronary artery dilation, ischemia or infarction must be excluded as causes of new myocardial dysfunction. Characteristic ECG and echocardiographic changes allow this distinction to be made rapidly in most patients.

Vascular Obstruction

Children with severe Kawasaki disease, especially infants or those in whom treatment is delayed, may develop other complications related to arterial occlusion. Peripheral obstruction leads to ischemia and gangrene; this complication generally accompanies other manifestations of critical disease such as giant coronary artery aneurysms and aneurysms in peripheral arteries. Various therapies have been attempted to restore circulation, although control of vascular inflammation with sufficient IVIG and/or corticosteroids is an essential prerequisite to arterial reperfusion. Thereafter, treatments may include thrombolytic therapy if arterial thrombosis is present or vasodilators if tissue viability is primarily threatened by vasospasm. Peripheral arterial obstruction may be corrected by thrombolysis with urokinase, streptokinase, or tissue-type plasminogen activator, after which perfusion is maintained with heparin followed by a long-term oral anticoagulant regimen.

Other Complications

Arthritis occurs in approximately one-third of children with Kawasaki disease and may add supportive evidence for the diagnosis in ambiguous cases. The arthritis tends to involve the small joints of the extremities during the acute phase of illness, and the large joints during the second and third weeks. The arthritis of Kawasaki disease is always nondeforming and self-limited, generally resolving within 30 days. Anti-inflammatory medications such as ibuprofen are usually effective in relieving symptoms until spontaneous resolution occurs.

Kawasaki disease may recur in 1 to 2% of children within 12 months of diagnosis, and an additional 5 to 10% may respond poorly to IVIG treatment during the initial bout of illness. In fact, patients who fail to respond completely to IVIG pose the greatest therapeutic dilemma. Prolonged fever itself correlates with increased risk of developing coronary artery abnormalities, and fever lasting for more than 14 days identifies a group of children at risk for developing giant coronary artery aneurysms (internal diameter larger than 8 mm), the group at highest risk for infarction and sudden death.

In cases of persistent, recurrent, or recrudescent Kawasaki disease, most clinicians repeat treatment with IVIG 2 g/kg over 8 to 12 hours. The risk of additional IVIG seems to be minimal, and several studies show a dose response to IVIG in Kawasaki disease. However, it is extremely important to confirm the diagnosis; failure to respond to IVIG might indicate that the child has a bacterial or viral infection or systemic JRA.

Approximately two-thirds of children with Kawasaki disease who fail to respond to an initial dose of IVIG improve with a second course. A small number seem to be resistant to IVIG, and approaches to these children vary. One regimen that appears to be safe and effective is the use of pulse doses of methylprednisolone, as used in PAN and other vasculitides.

LYME DISEASE

Background

Current knowledge of Lyme disease is the result of discoveries that have spanned most of the 20th century. Afzelius in Sweden first described the chronic migrating erythematous skin rash of Lyme disease in 1909, in association with an Ixodid tick bite. Twenty-one years later, Bannworth reported a tickborne syndrome of lymphocytic meningitis, neuritis, radicular pain, and an expanding erythematous rash (erythema migrans). In 1975, an outbreak of arthritis in Lyme, Connecticut, allowed identification of the tick vector associated with late disease. Finally, in 1981 the spirochete that is transmitted by ticks and causes the illness, *Borrelia burgdorferi,* was characterized.

Lyme disease is now the most common tickborne illness in the United States. More than 16,000 cases were reported in 1996, and outbreaks have been documented in 48 of the United States. Endemic foci are located in the New England, Middle Atlantic, Upper Midwest, and Western regions, as well as in Canada, Europe, China, Japan, and Russia. Most cases of Lyme disease occur between April and October, and up to 40% of cases occur in children.

Clinical Manifestations

Symptoms of *B. burgdorferi* infection may be classified into three stages, similar to the progression of syphilis. Stage 1 or early infection consists of a localized erythema migrans rash (Fig. 101.15). This begins as a small, red, indurated papule at the site of the tick bite, and then expands centrifugally for days or weeks. Lesions ultimately may reach an average diameter of 15 cm and may be accompanied by mild flulike symptoms, including fever, regional lymphadenopathy, and malaise.

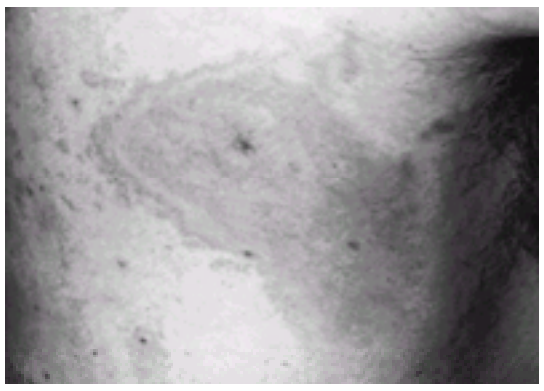

FIGURE 101.15. Primary erythema chronicum migrans lesion. (Courtesy of James Leyden, MD.)

Stage 2 of Lyme disease results from hematogenous dissemination of the spirochete. Integumental, musculoskeletal, and central nervous systems are most commonly affected. Approximately half of patients develop secondary annular skin lesions similar to erythema migrans but smaller and without central punctae. Debilitating fatigue may accompany myalgias and migratory arthralgias, followed in somewhat more than 50% of patients by a large-joint oligoarthritis. Severe headache and meningismus, cranial neuritis (especially Bell's palsy), and peripheral radiculoneuropathy may supervene. Finally, approximately 5% of patients demonstrate cardiac involvement, including conduction abnormalities or, more rarely, pancarditis. These symptoms generally resolve within weeks or months, but they may recur or persist.

Stage 3 disease is characterized by persistent infection and symptoms of prolonged latency. A sclerodermalike skin rash, acrodermatitis chronica atrophicans, is seen most commonly in Europe. A potentially erosive chronic oligoarthritis may be seen months to years after the tick bite. Subtle neurologic findings, including peripheral neuropathies and organic brain syndromes, may become apparent long after other manifestations of spirochete infestation have resolved.

Laboratory Features

B. burgdorferi is extremely difficult to grow in culture, and spirochetes generally cannot be identified in infected tissues. Diagnosis of Lyme disease therefore is made on the basis of characteristic clinical features accompanied by confirmatory serologic markers. Two caveats accompany serologic testing for Lyme disease. First, these tests are relatively difficult to perform, and standardization has been difficult to achieve. Many laboratories are plagued by both false-positive and false-negative results, so only experienced reference labs, preferably state or regional centers, should be used. Second, serologic tests for LD depend on the patient's antibody response. Titers may not be measurable until the second month after a tick bite in up to 85% of cases, and they may be abrogated by early antibiotic therapy. Early Lyme disease is thus a clinical condition based on a typical erythema migrans rash, and serology should not be relied on to confirm the diagnosis.

Current recommendations for testing use a two-tiered approach to optimize efficiency and accuracy. Patients are screened using an enzyme immunoassay (EIA) or immunofluorescent assay (IFA). If these tests are negative, generally no further testing is indicated. Positive results by these methods, however, require confirmation because a variety of viral illnesses and autoimmune conditions may cause false-positive results.

The second level of serologic evaluation involves Western or immune blotting. Any positive or equivocal antibody screening study should be confirmed by demonstrating the presence of at least five IgG bands directed against discrete *B. burgdorferi* proteins. This test is not foolproof, and false-positive studies may be seen in the setting of EBV and HIV infections and in SLE. In addition, up to 10% of residents of endemic regions have positive Lyme titers without evidence of true infection. Consequently, only those with compatible clinical findings, as well as positive serologies, require treatment.

Other laboratory data are not specific for Lyme disease. Hemoglobin, WBC counts, and platelet counts are generally normal. The ESR is elevated in approximately 50% of cases. ANA and RF are negative. Serum IgM is elevated in one-third of cases and correlates with the severity and chronicity of illness; IgM cryoglobulins are similarly associated. Immune complexes may be demonstrated in serum and synovial fluid.

In Lyme arthritis, synovial fluid analysis typically reveals elevated leukocyte counts ranging from 2,000 to 100,000 cells/mm_3. Polymorphonuclear leukocytes usually predominate, but Lyme disease is one of the few conditions in which a significant number of eosinophils may be identified in the synovial fluid. Total protein is elevated. Synovial biopsy reveals nonspecific synovial hypertrophy and mononuclear cell infiltration. In cases of neuroborreliosis, CSF analysis may reveal a mononuclear pleocytosis ranging from 25 to 500 cells/mm_3. At times, however, CSF may be entirely normal despite the presence of neurologic symptoms; even polymerase chain reaction (PCR) testing might fail to reveal evidence of borrelial DNA. In such cases, MRI of the brain might be useful, although other causes of the symptoms must also be considered. Suspected cardiac involvement may be confirmed by ECG, which reveals varying degrees of atrioventricular block and nonspecific ST-T wave changes.

Management

General Management

The cornerstone of treatment of Lyme disease is antibiotics. Oral antibiotic therapy is beneficial in early stages of the infection, whereas IV medications have a lower failure rate in chronic Lyme disease. Late manifestations of Lyme disease may represent a host autoimmune response rather than direct effects of the spirochete, but antibiotics may nonetheless be beneficial.

Current treatment guidelines are shown in Table 101.15 Clinical trials have resulted in the recommendation that patients with erythema migrans be treated with 14 to 21 days of doxycycline (100 mg twice daily). Children less than 8 years old should receive amoxicillin (25 to 50 mg/kg per day, maximum 2 g). Erythromycin (30 mg/kg per day) is reserved for allergic patients. Therapy of early constitutional symptoms is supportive and includes bed rest, analgesics, and antipyretics. As with other spirochetal infections, a Jarisch Herxheimer reaction may be seen after initial antibiotic therapy.

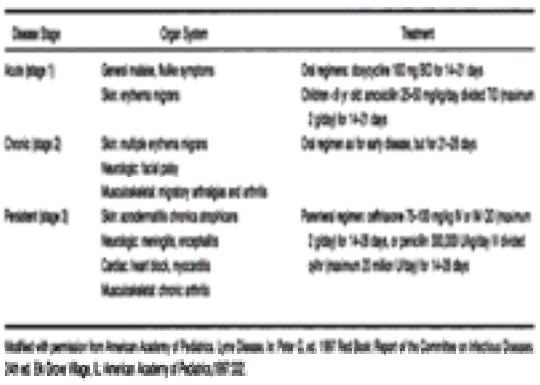

Table 101.15. Treatment of Lyme Disease

Management of Complications and Emergencies (Table 101.15)

Neurologic Complications. The presentation of neuroborreliosis is varied and may be categorized according to duration of infection. Early neurologic involvement, occurring during the first month after exposure, is seen in approximately 15% of untreated patients. Symptoms of aseptic meningitis and encephalitis, including severe headache, stiff neck, nausea and vomiting, photophobia, lethargy, and poor memory, are most notable. Kernig's and Brudzinski's signs are generally negative.

Facial nerve palsy is the most common cranial neuropathy of early neuroborreliosis. This is most often unilateral but may be bilateral. An isolated facial nerve palsy often presents difficult diagnostic decisions. With a history of tick exposure or erythema migrans rash, the diagnosis may be clear. More often, however, families will not recall such specifics, in which case facial nerve involvement caused by *B. burgdorferi* must be distinguished from that of viral, autoimmune, and idiopathic Bell's palsy.

Lyme-induced facial nerve symptoms tend to resolve spontaneously, but oral antibiotics are recommended to prevent dissemination and late sequelae. Because similar CSF findings may be seen regardless of the cause, lumbar puncture is generally not recommended in cases of isolated facial nerve palsy thought to result from Lyme disease. When a lumbar puncture is performed, CSF typically shows a lymphocytic pleocytosis with or without elevated protein. A CSF index, obtained by comparing the ratio of albumin to Lyme antibodies centrally and peripherally, is currently the most sensitive marker of neuroborreliosis. Evidence of intrathecal production of antiborrelial antibodies may be found in 80 to 90% of patients with Lyme meningoencephalitis and is treated with parenteral ceftriaxone or penicillin for 14 to 21 days.

Months or years after infection, some patients may develop late or chronic peripheral neuropathies. These manifest as paresthesias, symmetric or asymmetric; radicular pain and muscle weakness are generally less intense than in early

neuroborreliosis. Treatment with IV antibiotics, generally for 2 to 4 weeks, usually results in gradual improvement.

Most vexing of the late neurologic sequelae of Lyme disease is chronic Lyme encephalopathy. Symptoms are largely nonspecific, including debilitating fatigue, cognitive slowing, memory impairment, sleep disturbances, and depression. Distinction from idiopathic chronic fatigue syndrome or psychiatric conditions is extremely difficult. In cases attributable to Lyme disease on the basis of positive serologic markers of borrelial infection in addition to antibody, PCR, or MRI evidence of CNS involvement, symptoms often improve gradually with IV antibiotic therapy.

Cardiac Complications. Lyme carditis may present weeks to months after the initial infection in 5 to 10% of untreated children. Atrioventricular (AV) block is the most common manifestation; pericarditis, intraventricular conduction disturbances, and heart failure also may be seen. Patients may be asymptomatic with low-grade AV block, or they may complain of dyspnea, chest pain, palpitations, dizziness, or syncope with higher-grade disturbances. Physical signs include those of CHF with gallop rhythm, bibasilar crackles, and hepatojugular reflux, or those of pericarditis with friction rub and pulsus paradoxus (greater than 10 mm Hg). The heart rate may be elevated in the face of myocarditis or CHF; bradycardia as low as 30 beats/minute can be seen in patients with conduction abnormalities. ECG reveals varying degrees of AV block and ST-T wave changes. Chest radiographs may reveal cardiomegaly. Echocardiography is useful in documenting small pericardial effusions and ventricular dysfunction.

Treatment decisions are made on the basis of the severity of the carditis. Symptomatic patients or those with high-grade conduction disturbances should be admitted to the hospital for close monitoring with telemetry. This course should also be considered for those with significant prolongation of the P-R interval. Placement of a temporary pacemaker may be necessary in patients with complete heart block. There is no evidence that antibiotic therapy speeds recovery from Lyme carditis, but these agents are nonetheless used to eradicate infection and prevent additional complications. Oral antibiotics are used in early or mild cases, whereas IV regimens are indicated for more severe or chronic disease. Anti-inflammatory therapy with either aspirin (80 to 100 mg/kg per day) or occasionally prednisone (1 to 2 mg/kg per day) is recommended in cases of cardiomegaly, high-grade block, or significantly prolonged P-R interval (more than 300 ms) caused by Lyme disease.

Arthritis. Lyme arthritis occurs in more than 50% of untreated children, making it the most common late manifestation of infection. The classic presentation is a pattern of intermittent episodes of joint swelling beginning weeks to months after exposure. Less typical presentations of Lyme arthritis may also be seen. Patients may develop an apparent septic arthritis, with the acute onset of fever and joint pain, swelling, and erythema. Alternatively, synovitis might be migratory, simulating serum sickness or a postinfectious process, or chronic and persistent as in JRA. There is a propensity for large joint involvement, and often there is relatively little discomfort for the degree of swelling. The knee is affected in approximately 90% of cases, but more than two joints, or joints other than the knee, hip, ankle, or wrist, are involved in fewer than 10% of cases.

Lyme arthritis is clinically indistinguishable from JRA, septic arthritis, and postinfectious reactive synovitis, and it should be diagnosed only in the presence of convincing serologic evidence of borrelial infection. Current guidelines recommend initial treatment with oral antibiotics. Joint symptoms may only gradually improve over several weeks, but if joint pain and swelling persist for more than 2 months despite treatment, patients should be changed to IV therapy. Adjunct treatment with NSAIDs and physical therapy are important means of minimizing disability and accelerating recovery from Lyme arthritis.

Suggested Readings

JUVENILE RHEUMATOID ARTHRITIS

Athreya BH, Cassidy JT. Current status of the medical treatment of children with juvenile rheumatoid arthritis. Rheum Dis Clin North Am 1991;17:871.

Athreya BH, Doughty RA, Bookspan M, et al. Pulmonary manifestations of juvenile rheumatoid arthritis. A report of 8 cases and review. Clin Chest Med 1980;1:361.

Bland JH. Rheumatoid arthritis of the cervical spine. J Rheumatol 1974; 1:319.

Bray VJ, Singleton JD. Disseminated intravascular coagulation in Still's disease. Semin Arthritis Rheum 1994;24:222.

Cassidy JT, Petty RE. Juvenile rheumatoid arthritis. In: Textbook of Pediatric Rheumatology. 3rd ed. Philadelphia: WB Saunders,1995.

Cron RQ, Sherry DD, Wallace CW. Methotrexate-induced hypersensitivity pneumonitis in a child with juvenile rheumatoid arthritis. J Pediatr 1998;132:901.

Giannini EH, Calkwell GD. Drug treatment in children with juvenile rheumatoid arthritis. Pediatr Clin North Am 1995;42:1099.

Jacobs JC, Hui RM. Cricoarytenoid arthritis and airway obstruction in juvenile rheumatoid arthritis. Pediatrics 1977;59:292.

Keenan GF, Giannini EH, Athreya BH. Clinically significant gastropathy associated with nonsteroidal antiinflammatory drug use in children with juvenile rheumatoid arthritis. J Rheumatol 1995;22:1149.

Leitman PS, Bywaters EGL. Pericarditis in juvenile rheumatoid arthritis. Pediatrics 1963;32:855.

Lindsley CB. Uses of nonsteroidal anti-inflammatory drugs in pediatrics. Am J Dis Child 1993;147:229.

Martinez-Cordero E, Orosco Barocio G, et al. Evaluation of disease activity by laboratory tests in juvenile rheumatoid arthritis. J Invest Allerg Clin Immunol 1995;5:216.

Miller JJ, French JW. Myocarditis in juvenile rheumatoid arthritis. Am J Dis Child 1977;131:205.

Mouy R, Stephan JL, Pillet P, et al. Efficacy of cyclosporine A in the treatment of macrophage activation syndrome in juvenile arthritis: report of

five cases. J Pediatr 1996;129:750.

Murray K, Thompson SD, Glass DN. Pathogenesis of juvenile chronic arthritis: genetic and environmental factors. Arch Dis Child 1997;77:530.

Padeh S, Passwell JH. Intraarticular corticosteroid injection in the management of children with chronic arthritis. Arthritis Rheum 1998;41:1210.

Schaller JG. Juvenile rheumatoid arthritis. Pediatr Rev 1998;18:337.

Schmidt MC, Workman, JB, Barth WF. Dissection or rupture of a popliteal cyst. Arch Intern Med 1974;134:694.

Van Rossum MA, Fiselier TJ, Franssen MJ, et al. Sulfasalazine in the treatment of juvenile chronic arthritis: a randomized, double-blind, placebo-controlled, multicenter study. Arthritis Rheum 1998;41:808.

Wolf MD, Lichter PR, Ragsdale CG. Prognostic factors in the uveitis of juvenile rheumatoid arthritis. Ophthalmology 1987;94:1242.

Yancey CL, Doughty RA, Cohlan BA, et al. Pericarditis and cardiac tamponade in juvenile rheumatoid arthritis. Pediatrics 1981;68:369.

SYSTEMIC LUPUS ERYTHEMATOSUS

Aisen AM, Gabrielsen TO, McCune WJ. MR imaging of systemic lupus erythematosus involving the brain. Am J Radiol 1985;144:1027.

Austin HA III, Klippel JH, Balow JE, et al. Therapy of lupus nephritis: controlled trial of prednisone and cytotoxic drugs. N Engl J Med 1986;314:614.

Caramaschi P, Riccetti MM, Pasini AF, et al. Systemic lupus erythematosus and thrombotic thrombocytopenic purpura. Report of three cases and review of the literature. Lupus 1998;7:37.

Connolly B, Manson D, Eberhard A, et al. CT appearance of pulmonary vasculitis in children. Am J Roentgen 1996;167:901.

Esdaile JM, Abrahamowicz M, Joseph L, et al. Laboratory tests as predictors of disease exacerbations in systemic lupus erythematosus. Why some tests fail. Arthritis Rheum 1996;39:370.

Friedman DM, Lazarus HM, Fierman AH. Acute myocardial infarction in pediatric systemic lupus erythematosus. J Pediatr 1990;117:263.

Green L, Vinker S, Amital H, et al. Pseudotumor cerebri in systemic lupus erythematosus. Semin Arthritis Rheum 1995;25:103.

Hoch S, Schur PH. Methylprednisolone pulse therapy for lupus nephritis: a follow-up study. Clin Exp Rheumatol 1984;2:313.

Khamashta MA, Cuadrado MJ, Mujic F, et al. The management of thrombosis in the antiphospholipid-antibody syndrome. N Engl J Med 1995;332:993.

Lacks S, White P. Morbidity associated with childhood systemic lupus erythematosus. J Rheumatol 1990;17:941.

Lehman TJA, Sherry DD, Wagner-Weiner L, et al. Intermittent intravenous cyclophosphamide therapy for lupus nephritis. J Pediatr 1989; 114:1055.

McCune WJ, Golbus J, Zeldes W, et al. Clinical and immunologic effects of monthly administration of intravenous cyclophosphamide in severe systemic lupus erythematosus. N Engl J Med 1988;318:1423.

Medina F, Fraga A, Lavalle C. Salmonella septic arthritis in systemic lupus erythematosus. The importance of chronic carrier state. J Rheumatol 1989;16:203.

Mok CC, Lau CS, Chan EY, et al. Acute transverse myelopathy in systemic lupus erythematosus: clinical presentation, treatment, and outcome. J Rheumatol 1998;25:467.

Parikh S, Swaiman KF, Kin Y. Neurologic characteristics of childhood lupus erythematosus. Pediatr Neurology 1995;13:198.

Platt JL, Burke BA, Fish AJ, et al. SLE in the first two decades of life. Am J Kidney Dis 1982;11:212.

Rajani KB, Ashbacher LV, Kinney TR. Pulmonary hemorrhage and systemic lupus erythematosus. J Pediatr 1978;93:810.

Ravelli A, Martini A. Antiphospholipid antibody syndrome in pediatric patients. Rheum Dis Clin North Am 1997;23:657.

Roldan CA, Shivley BK, Crawford MH. An echocardiographic study of valvular heart disease associated with systemic lupus erythematosus. N Engl J Med 1996;335:1424.

Saab S, Corr MP, Weisman MH. Corticosteroids and systemic lupus erythematosus pancreatitis: a case series. J Rheumatol 1998;25:801.

Silverman ED, Lang B. An overview of the treatment of childhood SLE (Editorial). Scand J Rheumatol 1997;26:241.

Spahn JD, Kamada AK. Special considerations in the use of glucocorticoids in children. Pediatr Rev 1995;16:266.

Stahl N. Fever in systemic lupus erythematosus. Am J Med 1979;67:935.

Tan EM, Cohen AS, Fries JF, et al. 1982 Revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1982;25:1271.

Uziel Y, Laxer RM, Silverman ED. Persistent pulmonary hemorrhage as the sole initial clinical manifestation of pediatric systemic lupus erythematosus. Clin Esp Rheum 1997;15:697.

Ward MM, Pyun E, Studenski S. Causes of death in systemic lupus erythematosus. Long-term followup of an inception cohort. Arthritis Rheum 1995;38:1492.

Yell JA, Mbuagbaw J, Burge SM. Cutaneous manifestations of systemic lupus erythematosus. Br J Dermatol 1996;135:355.

DERMATOMYOSITIS

Banker BQ, Victor M. Dermatomyositis (systemic angiopathy) of childhood. Medicine 1966;45:261.

Bohan A, Peter JB. Polymyositis and dermatomyositis. N Engl J Med 1974;292:344,403.

Bowyer SL, Blane CE, Sullivan DB, et al. Childhood dermatomyositis: factors predicting functional outcome and development of dystrophic calcification. J Pediatr 1983;103:882.

Crowe WE, Bove KE, Levinson JE, et al. Clinical and pathogenetic implications of histopathology in childhood polydermatomyositis. Arthritis Rheum 1982;25:126.

Falcicni F, Trapini S, Ermini M, et al. Systemic vasculitis in juvenile dermatomyositis: a fatal case (Letter). Clin Exp Rheum 1995;13:531.

Gonzalez-Lopez L, Gamez-Nava JI, Sanchez L, et al. Cardiac manifestations in dermato-polymyositis. Clin Exp Rheum 1996;14:373.

Hernandez RJ, Keim DR, Sullivan DB, et al. Magnetic resonance imaging appearance of the muscles in childhood dermatomyositis. J Pediatr 1990;117:546.

Laxer RM, Stein L, Petty RE. Intravenous pulse methylprednisolone treatment of juvenile dermatomyositis. Arthritis Rheum 1987;30:328.

Magill HL, Hixson SD, Whitington G, et al. Duodenal perforation in childhood dermatomyositis. Pediatr Radiol 1984;14:28.

Pachman LM. Juvenile dermatomyositis. Pathophysiology and disease expression. Pediatr Clin North Am 1995;42:1071.

Pachman LM, Hayford JR, Hochberg MC, et al. New-onset juvenile dermatomyositis: comparisons with a healthy cohort and children with juvenile rheumatoid arthritis. Arthritis Rheum 1997;40:1526.

Olsen NJ, Park JH. MRS and NIRS for muscle disease evaluation. Bull Rheum Dis 1995;44:4.

Quecedo E, Febrer I, Serrano G, et al. Partial lipodystrophy associated with juvenile dermatomyositis: a report of two cases. Pediatr Dermatol 1996;13:477.

Rider LG, Miller FW. Classification and treatment of the juvenile idiopathic inflammatory myopathies. Rheum Dis Clin North Am 1997;23:619.

Singsen BH, Tedford JC, Platzker ACG, et al. Spontaneous pneumothorax: a complication of juvenile dermatomyositis. J Pediatr 1978;92:771.

Zeller V, Cohen P, Prieur AM, et al. Cyclosporin A therapy in refractory juvenile dermatomyositis. Experience and long-term follow-up of 6 cases. J Rheum 1996;23:1424.

SCLERODERMA

Alstaedt HO, Berzewski B, Taschke C, et al. Treatment of patients with peripheral arterial occlusive disease Fontaine stage IV with intravenous iloprost and PGE1: a randomized open controlled study. Prostaglandins Leukot Essent Fatty Acids 1993;49:573.

Armstrong GP, Whalley GA, Doughty RN, et al. Left ventricular function in scleroderma. Br J Rheum 1996;35:983.

Botstein CR, LeRoy EC. Primary heart disease in systemic sclerosis (scleroderma): advances in clinical and pathologic features, pathogenesis and new therapeutic approaches. Am Heart J 1981;102:913.

Cassidy JT, Sullivan DB, Dabich L, et al. Scleroderma in children. Arthritis Rheum 1977;20:351.

Fine LG (chairman). Systemic sclerosis: current pathogenetic concepts and future prospects for targeted therapy. Lancet 1996;347:1453.

Fujita Y, Yamamori H, Hiyoshi K, et al. Systemic sclerosis in children: a national retrospective surgery in Japan. Acta Paedatr Jpn 1997; 39:263.

Garty B-Z, Athreya BH, Wilmott R, et al. Pulmonary functions in children with progressive systemic sclerosis. Pediatrics 1991;88:1161.

Haber PL. Clinical manifestations of scleroderma. Pediatr Rev 1995;16:49.

Harrison NK, Glanville AR, Strickland B, et al. Pulmonary involvement in systemic sclerosis: the detection of early changes by thin section CT scan, bronchoalveolar lavage and 99mTc-DTPA clearance. Respir Med 1989;83:403.

Harrison NK, Myers AR, Corrin B, et al. Structural features of interstitial lung disease in systemic sclerosis. Am Rev Respir Dis 1991;144:706.

Medsger TA Jr, Steen V. Systemic sclerosis and related syndromes. In: Schumacher HR, ed. Primer on the Rheumatic Diseases. 10th ed. Atlanta, Arthritis Foundation, 1993.

Roberts NK, Cabeen WR, Moss J, et al. The prevalence of conduction defects and cardiac arrhythmias in progressive systemic sclerosis. Ann Intern Med 1981;94:38.

Rodeheffer RJ, Rommer JA, Wigley F, et al. Controlled double-blind trial of nifedipine in the treatment of Raynaud's phenomenon. N Engl J Med 1983;308:880.

Seely JM, Jones LT, Wallace C, et al. Systemic sclerosis: using high resolution CT to detect lung disease in children. Am J Roentgen 1998;170:691.

Singsen BH. Scleroderma in childhood. Pediatr Clin North Am 1986;33:1119.

Sjogren RW. Review: gastrointestinal motility disorders in scleroderma. Arthritis Rheum 1995;34:3.

Steen VD, Constantino JP, Shapiro AP, et al. Outcome of renal crisis in systemic sclerosis: relation to the availability of converting enzyme inhibitors (ACE). Ann Intern Med 1990;113:352.

Steen VD, Lanz JK Jr, Conte C, et al. Therapy for severe interstitial lung disease in systemic sclerosis. Arthritis Rheum 1994;37:1290.

Steen VD, Medsger JA, Osial JA, et al. Factors predicting development of renal involvement in progressive systemic sclerosis. Am J Med 1984;76:779.

Teasdall RD, Frayha RA, Shulman LE. Cranial nerve involvement in systemic sclerosis (scleroderma): a report of 10 cases. Medicine 1980;59:149.

Vancheeswaran R, Black CM, David J, et al. Childhood-onset scleroderma: is it different from adult-onset disease? Arthritis Rheum 1996;39:1041.

POLYARTERITIS NODOSA

Athreya BH. Vasculitis in children. Pediatr Clin North Am 1995;42:1239.

Blau EB, Morris RF, Yunis EJ. Polyarteritis nodosa in older children. Pediatrics 1977;60:227.

Camilleri M, Pusey CD, Chadwick VS, et al. Gastrointestinal manifestations of systemic vasculitis. Q J Med 1983;52:141.

Chalopin JM, Rifle G, Turc JM, et al. Immunological findings during successful treatment of HBs-Ag-associated polyarteritis nodosa by plasmapheresis. Br Med J 1980;280:368.

Cupps TR, Fauci AS. The vasculitides. In: Smith LH Jr, ed. Major Problems in Internal Medicine. Philadelphia: WB Saunders,1981;XXI.

Dahlberg PJ, Lockhart JM, Overholt EL. Diagnostic studies for systemic necrotizing vasculitis. Arch Intern Med 1989;149: 161.

Dillon MJ. Rare vasculitic syndromes. Ann Med 1997;29:175.

Dillon MJ, Ansell BM. Vasculitis in children and adolescents. Rheum Dis Clin North Am 1995;21:1115.

Fauci AS, Doppman JL, Wolff SM. Cyclophosphamide-induced remissions in advanced polyarteritis nodosa. Am J Med 1978;64:890.

Fink CW. Vasculitis. Pediatr Clin North Am 1986;33:1203.

Gillespie DN, Burke EC, Holley KE. Polyarteritis nodosa in infancy: a diagnostic enigma. Mayo Clin Proc 1973;48:773.

Magilavy DB, Petty RE, Cassidy JT, et al. A syndrome of childhood polyarteritis. J Pediatr 1977;91:25.

Moore PM, Cupps TR. Neurological complications of vasculitis. Ann Neurol 1983;14:155.

Reimold EW, Weinberg AG, Fink CW, et al. Polyarteritis in children. Am J Dis Child 1976;130:534.

Sack M, Cassidy JT, Bole GG. Prognostic factors in polyarteritis. J Rheumatol 1975;2:411.

Smoller BR, McNutt NS, Contreras F. The natural history of vasculitis. Arch Dermatol 1990;126:84.

KAWASAKI DISEASE

Barron KS. Kawasaki disease in children. Curr Opin Rheum 1998;10:29.

Beiser AS, Takahashi M, Baker AL, et al. A predictive instrument for coronary artery aneurysms in Kawasaki disease. US Multicenter Kawasaki disease study group. Am J Cardiol 1998;81:1116.

Burns JC, Mason WH, Glode MP, et al. Clinical and epidemiologic characteristics of patients referred for evaluation of possible Kawasaki disease. J Pediatr 1991;118:680.

Durongpisitkul K, Gurunaj VJ, Park JM, et al. The prevention of coronary artery aneurysm in Kawasaki disease: a meta-analysis on the efficacy of aspirin and immunoglobulin treatment. Pediatr 1995;96:1057.

Friter BS, Lucky AW. The perineal eruption of Kawasaki syndrome. Arch Dermatol 1988;124:1805.

Fujiwara T, Fujiwara H, Hamashima Y. Frequency and size of coronary arterial aneurysms at necropsy in Kawasaki disease. Am J Cardiol 1987;59:808.

Fukushige J, Takahashi HN, Ueda Y, et al. Incidence and clinical features of incomplete Kawasaki disease. Acta Paediatr 1994;83:1057.

Fukushige J, Takahashi N, Ueda K, et al. Long-term outcome of coronary abnormalities in patients after Kawasaki disease. Pediatr Cardiology 1996;17:71.

Horrigome H, Sekijima T, Miyamoto T. Successful thrombolysis with intracoronary administration of tissue plasminogen activator in an infant with Kawasaki disease. Heart 1997;78:517.

Kato H, Sugimua T, Akagi T, et al. Long-term consequences of Kawasaki disease. A 10- to 21-year follow up study of 594 patients. Circulation 1996;94:1379.

Kawasaki T, Kosaki F, Okawa S, et al. A new infantile acute febrile mucocutaneous lymph node syndrome (MLNS) prevailing in Japan. Pediatrics 1974;54:271.

Landing BH, Larson EJ. Are infantile polyarteritis nodosa with coronary involvement and fatal mucocutaneous lymph node syndrome the same? Comparison of 20 patients from North America with patients from Hawaii and Japan. Pediatrics 1977;59:651.

Marasini M, Pongiglione G, Gazzolo D, et al. Late intravenous gamma globulin treatment in infants and children with Kawasaki disease and coronary artery abnormalities. Am J Cardiol 1991;68:796.

Nakamura Y, Yanagawa H, Kato H, et al. Mortality rates for patients with a history of Kawasaki disease in Japan. Kawasaki disease follow-up group. J Pediatr 1996;128:75.

Naoe S, Takahashi K, Masuda H, et al. Kawasaki disease with particular emphasis on arterial lesions. Acta Pathol Jpn 1991;41:785.

Newburger JW, Sanders SP, Burns JC, et al. Left ventricular contractility and function in Kawasaki syndrome. Circulation 1989;79:1237.

Newburger JW, Takahashi M, Beiser AS, et al. A single intravenous infusion of gamma globulin as compared with four infusions in the treatment of acute Kawasaki syndrome. N Engl J Med 1991;324:1633.

Newburger JW, Takahaski M, Burns JC, et al. The treatment of Kawasaki syndrome with intravenous gamma globulin. N Engl J Med 1986;315:341.

Rosenfeld EA, Corydon KE, Shulman ST. Kawasaki disease in infants less than one year of age. J Pediatr 1995;126:524.

Rowley AH. Controversies in Kawasaki syndrome. Adv Pediatr Infect Dis 1997;13:127.

Schneider L, Geha R. Outbreak of hepatitis C associated with intravenous immunoglobulin administration—United States, October 1993–June 1994. MMWR 1994;43:505.

Smith LBH, Newburger JW, Burns JC. Kawasaki syndrome and the eye. Pediatr Infect Dis J 1989;8:116.

Sundel RP, Burns JC, Baker A, et al. Gamma globulin re-treatment in Kawasaki disease. J Pediatr 1993;123:657.

Von Planta M, Fasnacht M, Holm C, et al. Atypical Kawasaki disease with peripheral gangrene and myocardial infarction: therapeutic implications. Eur J Pediatr 1995;154:830.

Westphalen MA, McGrath MA, Kelly W, et al. Kawasaki disease with severe peripheral ischemia: treatment with prostaglandin E1 infusion. J Pediatr 1988;112:431.

Wright DA, Newburger JW, Baker A, et al. Treatment of immune globulin-resistant Kawasaki disease with pulsed doses of corticosteroids. J Pediatr 1996;128:146.

Yanagihara R, Todd JK. Acute febrile mucocutaneous lymph node syndrome. Am J Dis Child 1980;134:603.

LYME DISEASE

Albisetti M, Schaer G, Good M, Boltshauser E, Nadal D. Diagnostic value of cerebrospinal fluid examination in children with peripheral facial palsy and suspected Lyme borreliosis. Neurology 1997;49:817.

American Academy of Pediatrics, Lyme Disease. In: Peter G, ed. 1997 Red Book: Report of the Committee on Infectious Diseases. 24th ed. Elk Grove Village, IL: American Academy of Pediatrics, 1997.

Athreya BH, Rose CD. Lyme disease. Curr Probl Pediatr 1996;26:189.

CDC. Case definition for public health surveillance. MMWR 1990;39 (RR-13):19.

Bingham PM, Galetta SL, Athreya B, et al. Neurologic manifestations in children with Lyme disease. Pediatr 1995;96:1053.

Dattwyler RJ, Halperin JJ, Volkman DJ, et al. Treatment of late Lyme borreliosis-randomised comparison of ceftriaxone and penicillin. Lancet 1988;i:1191.

Eichenfield AH, Goldsmith DP, et al. Childhood Lyme arthritis: experience in an endemic area. J Pediatr 1986;109:753.

Gerber MA, Shapiro ED. Diagnosis of Lyme disease in children. J Pediatr 1992;121:157.

Kaplan RF. Lyme encephalopathy: a neuropsychological perspective. Semin Neurol 1997;17:31.

Nagi KS, Joshi R, Thakur RK. Cardiac manifestations of Lyme disease: a review. Can J Cardiol 1996;12:503.

Shapiro ED. Lyme disease. Pediatr Rev 1998;19:147.

Steere AC, Bartenhagen NH, Craft JE, et al. The early clinical manifestations of Lyme disease. Ann Intern Med 1983;99:76.

Steere AC, Malawista SE, Syndam DR, et al. Lyme arthritis: an epidemic of oligoarticular arthritis in children and adults in three Connecticut communities. Arthritis Rheum 1977;20:7.

Williams CL, Strobino B, Lee A, et al. Lyme disease in childhood: clinical and epidemiologic features of ninety cases. Pediatr Infect Dis J 1990;9:10.

CHAPTER 102

Problems of the Very Early Neonate

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No aspect of the medical system's transition to shortened inpatient hospital stays has received more attention than that of early discharge to home of the postpartum mother with her newly delivered offspring. Discharge often occurs before the parents have received adequate instruction in care and before they have established a level of comfort and rapport with their baby. As a result, even experienced parents may feel inadequate in determining whether their newborn is ill or has a significant defect.

If a primary care contact has not yet been established, immediate access to medical evaluation is provided only by a visit to the hospital emergency department (ED). Thus, ED physicians are seeing increasing numbers of very early neonates who, in prior years, would still have been housed in the newborn nursery. This chapter serves as a guide to evaluating

and managing the parents' concerns during the first week to 10 days with their new baby.

Many of the problems will be insignificant and can be dealt with by judicious reassurance, but it must be kept in mind that seriously ill newborns may present with only subtle, nondescript symptoms. The parent's expressed complaint may be only a clue to a serious problem far removed from their area of concern, so a relevant perinatal history should be queried and, in most instances, a thorough physical examination performed.

The chapter is structured to allow the physician quick access to the complaint that precipitated the emergency visit. For conciseness and for easier access to information, the description of the presenting complaints is divided into subsections, based on physical, behavioral, and etiologic concerns. In most subsections, there is a delineation of those findings that are normal variants, those that might need further evaluation, and those that call for immediate attention. References are made to other chapters in this book where the more complicated problems are discussed in greater detail.

HISTORY

The emergency physician must be able to elicit relevant information needed to make decisions regarding diagnosis and management within a relatively short period. Time constraints, level of acuity within the ED, and the absence of long-term relationships can make communication in this setting particularly challenging. In the newborn period, not only is it essential to evaluate the health and physiologic stability of the baby, but often it is equally important to assess the psychosocial, emotional, and physical well-being of the mother; her ability and confidence in caring for her baby; the adequacy of support systems for the mother and family; and the access to follow-up care.

Because current concerns may have their origins in issues related to pregnancy or parturition, knowledge of the immediate history, a review of the events of labor and delivery, history of prior and current pregnancies, and a review of the family history are all essential components of the newborn history. Neonatal mortality is defined as the death of an infant between birth and 1 month of age. The leading causes of neonatal mortality—congenital anomalies, disorders related to short gestation and low birth weight, respiratory distress syndrome, and maternal complications of pregnancy—all have an association with pregnancy or parturition. Disclosure or identification of certain maternal, pregnancy, or infant-related factors (Table 102.1) place the newborn at increased risk and alert the practitioner of the potential for morbidity and mortality.

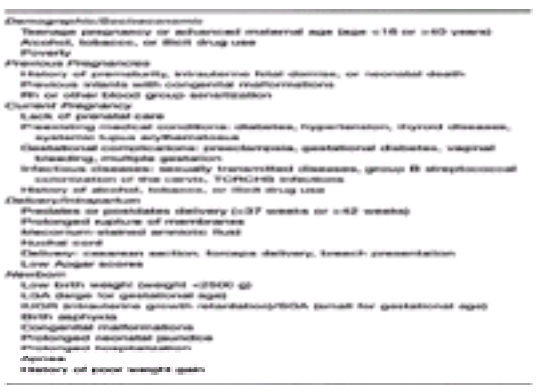


Table 102.1. High-Risk Factors for Neonatal Morbidity and Mortality

Certain points should be included in the history of all newborn infants, regardless of the presenting complaint. Always query regarding the infant's gestational age because size of an infant at birth does not necessarily reflect gestational age. An infant who is born “small for gestational age” (SGA) or with “intrauterine growth retardation” (IUGR) is at higher risk for morbidity and mortality than an infant of the same gestational age who is “appropriate for gestational age” (AGA). An infant whose birth weight is below the 10th percentile for gestational age is SGA. SGA infants have a more than fourfold increase in perinatal mortality than AGA infants of the same gestational age. Conversely, an infant whose birth weight is above the ninetieth percentile for gestational age is “large for gestational age” (LGA). The most common causes for LGA infants are familial, infants of diabetic mothers, and overgrowth syndromes such as the Beckwith-Wiedemann syndrome. Hypoglycemia, hyperbilirubinemia, hypocalcemia, and polycythemia are all associated acute complications that may be seen in the macrosomic infant. The clinician should inquire regarding the infant's general level of alertness, diet, and elimination patterns. Inadequate alimentation or infrequent urination may be clues to more serious problems. Parental concern about excessive crying or inability to soothe their infant requires attention because this excessive fussiness may have an underlying organic origin (see Chapter 17). Nonspecific complaints such as increased crying, sleepiness, and decreased appetite are sometimes the only symptoms of serious illness in the newborn.

Additional history should be guided by and focused specifically on the presenting complaint. Relevant suggestions are included in each of the “presenting complaint” subsections.

VITAL SIGNS

Growth

Weight gain serves as an important indicator of general well-being during the newborn period. Failure of a newborn to gain weight appropriately may be a sign of underfeeding or significant underlying illnesses such as heart disease,

metabolic problems, or malabsorption. Similarly, gaining weight according to age-specific norms can be one of the best indicators that the infant is well, despite nondescript symptoms such as fussiness.

The average newborn infant weighs 7.7 lb (3.5 kg), is about 20 inches (50 cm) long, and has a head circumference of 14 inches (35 cm). The newborn will lose about 5 to 10% of his or her birth weight during the first several days of life and then regain this weight by 10 to 14 days of age. Thereafter, the newborn should gain about 25 to 35 g/day (roughly 1% of the birth weight per day). The average newborn takes 2 to 3 ounces of formula (about 10 minutes on each breast) every 2 to 3 hours and has 10 to 12 wet diapers and 1 to 2 bowel movements (as often as once after each feeding for breast-fed infants) per day. Because breast milk is easier to digest and passes out of the stomach quicker than formula (on average 1½ hours versus up to 4 hours for formula), the breast-fed infant will want to feed more frequently, with an increased number of nighttime feedings as well, than the formula-fed infant.

Temperature

The young infant's immature autonomic thermoregulatory responses, larger body surface area to mass ratio, immature sweating response, and limited ability to move away from or modify adverse environments all limit his or her thermoregulatory ability. Temperature instability, either *hypothermia* or *hyperthermia,* may be the only sign of significant infectious illness. It is unusual to see high temperatures in the newborn. Even a septic newborn may develop only a slight elevation in temperature or temperature instability. Therefore, any temperature greater than 38°C (100.4°F) should be regarded as a fever in the newborn and receive appropriate evaluation (see Chapter 28 and Chapter 84).

Heart Rate (see Chapter 82)

Normal resting heart rate is between 120 to 160 beats/minute. It varies with respiration (increasing with inspiration) and activity (increasing significantly with crying and appreciably slower during sleep). Cardiac output in the infant is primarily increased by increasing the heart rate rather than stroke volume. Sinus *tachycardia* (heart rate greater than 180 beats/minute) is a common response to many types of stress, such as pain, hypovolemia, fever, or cardiac disease (see Chapter 74). *Sinus tachycardia* must be differentiated from *paroxysmal supraventricular tachycardia (SVT)* (see Chapter 82) when the electrocardiogram (ECG) demonstrates a narrow QRS complex tachycardia with a P wave preceding each QRS complex. SVT is usually associated with a more rapid heart rate (usually greater than 220 beats/minute) than is sinus tachycardia. Infants may tolerate SVT for a variable period, but eventually they develop irritability, tachypnea, poor feeding, and poor perfusion. The development of *bradycardia* (heart rate less than 80 beats/minute) usually signals the presence of significant cardiorespiratory compromise and is an ominous sign that requires immediate attention (see Chapter 2 and Chapter 3).

Respiratory Rate

Normal resting respiratory rate is usually between 40 to 60 breaths/minute. During sleep, most newborn infants will exhibit some degree of *periodic breathing,* in which normal respiration is interrupted with short pauses. This breathing pattern is especially common in premature infants. Periodic breathing must be differentiated from *pathologic apnea* (see Chapter 10). In the simplest of terms, apnea is an absence of respiration that can have a central, obstructive, or mixed cause. Short periods of central apnea (less than 15 seconds) can be normal at all ages. However, pathologic apnea is a prolonged respiratory pause (more than 20 seconds) or a shorter pause associated with cyanosis, pallor, bradycardia, or hypotonia. Underlying disorders that must be considered in the evaluation of the apneic infant, even the premature infant, include septicemia, severe anemia, intracranial hemorrhage, seizures, gastroesophageal reflux, metabolic disturbances such as hypoglycemia, or infant or maternal ingestion of narcotics or other central nervous system (CNS) depressants.

Varying degrees of *expiratory grunting, chest retractions, nasal flaring, crackles,* or *rales* are all signs of respiratory distress in the newborn. In addition to a primary pulmonary cause, respiratory distress in the newborn can also be a presenting sign of *congestive heart failure.* Among term infants, especially those born by cesarean section, a common cause of respiratory distress presenting within the first 24 hours, usually beginning between 2 to 6 hours after birth, is *transient tachypnea of the newborn* (TTN). TTN is believed to be caused by a delay in the absorption of the normal fetal lung fluid. The differential diagnosis of TTN includes meconium aspiration pneumonitis, respiratory distress syndrome (unlikely in the term newborn), pneumonia, and bronchiolitis. Symptoms of TTN typically resolve within 72 hours.

Blood Pressure

Normal systolic blood pressure in the term newborn after the first few days of life ranges between 60 to 90 mm Hg. Blood pressure is lower during the first few days of life and in premature infants is related to weight and gestational age. Congenital renal abnormalities, renal tumors, and complications of umbilical artery catheters are some of the more common causes of *hypertension* (see Chapter 35) in the neonatal period. *Coarctation of the aorta* may be diagnosed by the combination of increased upper extremity blood pressure with low blood pressure or diminished pulses in the lower extremities.

COLOR CHANGES

Normal Variants

The skin of a normal Caucasian early neonate is a pink, flushed color. This in itself may be a cause for alarm to some parents but can be dismissed with reassurance if the remainder of the history and physical examination is not remarkable. A hemoglobin or hematocrit determination offers assurance that the baby is not abnormally polycythemic. Racial and ethnic factors may result in variation of the baby's skin color, but this usually can be determined by comparing the baby with the parent's pigmentation.

Alterations of the flushed appearance to blue, deep yellow, orange, or pale may precipitate an ED visit. Evaluation and

management of these changes are discussed here.

Cyanosis/Acrocyanosis (see Chapter 16)

The presenting complaint may be that “the baby is blue.” Relevant questions to be asked include the following: When was the blueness first noted? Is it persistent or does it come and go? Does it involve all of the body or only the distal extremities and lips? Does it increase or lessen with crying or feeding? Is there emesis or diarrhea?

Physical examination should be complete. The distribution of the blueness should be carefully noted; particularly, check the color of the tongue. Does the intensity of the blueness decrease or increase with crying or with effort? Are the vital signs normal for a neonate? Is the baby responsive to stimulation? Is mottling present? Is there a cardiac murmur? Are respirations labored? Are the lungs clear? Is the liver enlarged? Are the femoral pulsations palpable?

The degree of cyanosis should be documented by pulse oximetry. The more likely causes are discussed in the following sections.

Acrocyanosis

An otherwise healthy baby has cyanosis confined to the hands, feet, and lips. The tongue is pink; pulse oximetry is normal. It may be associated with cool ambient temperature. Disposition: Parent(s) should be reassured that the acrocyanosis is self-limited.

Cyanotic Congenital Heart Disease (see Chapter 16, Chapter 33, and Chapter 82)

Cyanosis is diffusely distributed and increases with crying. Pulse oximetry shows diminished saturation at rest, worsening with crying; cyanosis responds only minimally to oxygen therapy. A cardiac murmur is usually, but not necessarily, present. ECG and echocardiography, if accessible, should be performed. Neonates with cyanotic heart disease only rarely go into cardiac failure. Disposition: Very early neonates with a strong suspicion of cyanotic cardiac defect should be admitted if cardiac consultation is not readily available.

Congestive Cardiac Failure (see Chapter 33 and Chapter 82)

Cyanosis is diffusely distributed, and pulse oximetry shows desaturation but improves somewhat with oxygenation. The infant is tachypneic, possibly with retraction. Rales may be apparent in lung fields. Cardiac murmur may be present or femoral pulsations may be absent. The liver is enlarged. Disposition: Emergency treatment and admission are indicated.

Respiratory Disease (see Chapter 16, Chapter 68, and Chapter 95)

Respiratory disease is associated with tachypnea and possibly retractions. Pulse oximetry shows desaturation but usually improves significantly with rest, crying, and oxygenation. The clinician should think of respiratory infection or congenital intrathoracic defect. A chest radiograph should be done. Disposition: After appropriate emergency treatment, the patient should be admitted.

Hypovolemia, Acidosis, and Shock (see Chapter 3 and Chapter 69)

Hypovolemia, acidosis, and shock are characterized by cyanosis accompanied by mottling in an extremely lethargic, hypotonic baby with marked tachycardia and possibly hypotension. Pulse oximetry is desaturated. Disposition: After appropriate emergency workup and treatment, the patient should be admitted.

Methemoglobinemia (see Chapter 16 and Chapter 87)

Methemoglobinemia is characterized by cyanosis diffusely distributed in the absence of cardiac or pulmonary disease. There may be a recent history of gastroenteritis. Pulse oximetry gives factitiously normal readings. A drop of blood on filter paper remains reddish brown, even after oxygenating it by waving in room air. Disposition: Emergency and laboratory evaluation to determine the intensity of involvement are indicated; treatment with methylene blue and admission should be considered.

Blue Sclerae

Blue color confined to the sclerae may be associated with osteogenesis imperfecta. The neonate's sclerae commonly may be normally blue, although the intensity is less. If in doubt, the clinician should take a careful family history and examine for fractures of the extremities, ribs, and pelvis.

Jaundice

The parent may be concerned because the newborn appears yellow or orange. Because this *may be a normal variant* in Pacific rim or Native American racial or ethnic groups, the parents' coloring should be compared with the baby's.

Bilirubin is formed by the catabolism of hemoglobin and may accumulate when there is either excessive hemolysis, failure of conjugation with glucuronic acid in the liver, or inadequate excretion through the liver canaliculi or the bile ducts. Bilirubin that has not been conjugated in the liver tests as indirect and is related to excessive hemolysis of red cells; conjugated bilirubin is reported as direct and is elevated when excretion is obstructed.

If jaundice is identified, relevant questions to be asked should include the following: When was the jaundice first noticed?

What is the color of stools and urine? What type of feeding is being used (breast or formula)? Are feedings adequate? What are the mother's and baby's blood types, if known? Has the infant vomited? Is there a family history of jaundice? Is the mother diabetic?

The most precise way of determining whether the color change is truly jaundice is by examining the sclerae—yellow color of the sclerae is jaundice. A complete physical examination should be performed, with emphasis on vital signs, intensity and distribution of icterus, presence of cephalohematoma, or hepatosplenomegaly.

The intensity of jaundice is determined by the level of bilirubin in the blood and by its distribution. As a rule, jaundice is not discernible in infants at levels less than 5 mg/dL. Jaundice is usually first discerned in the face and becomes more obvious caudally as the total serum bilirubin (TSB) level increases. In each patient, a TSB level and a complete blood count (CBC) with blood smear should be performed. If subsequent TSB levels are indicated in the baby, the direct bilirubin level should be determined at least once.

Physiologic Jaundice (see Chapter 41)

Physiologic jaundice is icterus that is not pathologic. Discernible jaundice in the first 24 hours of life of a healthy term newborn is probably pathologic, so a cause must be sought and therapy inaugurated. Beyond the first day, however, if the CBC and smear are not abnormal and the physical examination is not remarkable, a moderate degree of jaundice can be assumed to be physiologic. Infants of diabetic mothers and babies with congenital hypothyroidism or those with resorption from large cephalohematomata may have higher levels of icterus than one would expect physiologically.

Management and disposition can be governed by the recommendations in Table 102.2 and will vary with the baby's age and the bilirubin level. Babies with TSB nearing the levels at which therapy is suggested should be followed with at least one repeat determination within 24 hours to get a sense of the rate of rise.

Age (hr)	Consider Phototherapy[b]	Phototherapy	Exchange Transfusion if Intensive Phototherapy Fails[c]	Exchange Transfusion and Intensive Phototherapy
<24	—	—	—	—
25–48	≥12 (170)	≥15 (260)	≥20 (340)	≥25 (430)
49–72	≥15 (260)	≥18 (310)	≥25 (430)	≥30 (510)
>72	≥17 (290)	≥20 (340)	≥25 (430)	≥30 (510)

Adapted from the report of the Committee for Quality Improvement of the American Academy of Pediatrics. Pediatrics 1994;94:558–565.
TSB, total serum bilirubin.
[b]TSB level, mg/dL (μmol/L).
[c]Phototherapy at these TSB levels is a clinical option, meaning that the intervention is available and may be used on the basis of individual clinical judgment.
[d]Intensive phototherapy should produce a decline of TSB of 1–2 mg/dL within 4–6 hr, and the TSB level should continue to fall and remain below the threshold level for exchange transfusion. If this does not occur, it is considered a failure of phototherapy.

Table 102.2. Management of Hyperbilirubinemia in the Healthy Term Infant According to TSB and Baby's Age[a]

Breast Milk Jaundice (see Chapter 41)

In breast-fed babies, jaundice may be initiated by inadequate caloric intake and the dehydration that may occur before there is sufficient milk. It usually begins in the early neonatal period and may continue for weeks. If TSB is not approaching levels at which phototherapy needs to be considered, breast-feeding should be continued and even encouraged more frequently. Higher TSB levels should be followed with at least one additional determination.

Blood Type Incompatibility (see Chapter 41 and Chapter 87)

If the TSB level is at or near levels requiring therapy (Table 102.2) and particularly if the jaundiced baby has a lower hemoglobin than expected, direct Coombs' test should be done. Mother and baby should be blood typed for ABO and Rh factors. If there is an incompatibility and if the bilirubin is rising rapidly despite phototherapy, blood should be prepared for possible exchange transfusion. Rh incompatibility is uncommon in those mothers who have had ongoing obstetric care. ABO incompatibility is usually more insidious in onset and less severe than Rh.

Sepsis (see Chapter 69 and Chapter 84)

Sepsis need not be a consideration in the alert, well-appearing, jaundiced newborn. If the jaundiced baby is lethargic or extremely irritable, hypotonic, or tachycardiac and has been feeding poorly, sepsis and/or urinary tract infection should be considered. The bilirubin may be indirect or direct.

Congenital Red Cell Defects (see Chapter 41 and Chapter 87)

Careful examination of the blood smear may provide a clue to red cell membrane defects. These babies may have indirect bilirubinemia and anemia and may have a positive family history for spherocytosis or elliptocytosis. More severe forms of glucose-6-phosphate dehydrogenase deficiency also may present with early jaundice and anemia, especially if the mother has received possibly precipitating drugs during late pregnancy. Elevated reticulocytes with a negative Coombs' test would make one suspicious.

Other Congenital Problems (see Chapter 41 and Chapter 42)

The group of congenital infections that falls under the acronym of TORCHS (TOxoplasmosis, Rubella, Cytomegalovirus, Herpes virus, Syphilis) may present with jaundice. These should be thought of in the baby with elevated direct bilirubin who presents with microcephaly, jitteriness, seizures, organomegaly, or petechiae. Elevated direct bilirubin also may be an early manifestation of galactosemia and cystic fibrosis. Babies with the severe form of the enzymatic defect of Crigler-Najjar syndrome may present with extremely high levels of unconjugated bilirubin, which may rise despite phototherapy.

Pallor

The neonate may be brought to the physician because he or she appears pale to the parents. A careful history and physical examination should be done to consider the possibilities of septic or cardiogenic shock, severe chemical or electrolyte imbalance, or significant anemia.

Questions should be asked regarding the mother's perinatal history, the baby's suck and feeding, the occurrence of vomiting or diarrhea, fever, and the baby's responsiveness. Physical examination should include thorough evaluation of the vital signs. Is the baby extremely lethargic? The clinician should look at the inner palpebral conjunctivae, and check the skin for petechiae or rashes. Is there a large cephalohematoma? Is there hepatosplenomegaly?

A CBC should be done and consideration given to a stat glucose and electrolyte panel and total serum bilirubin level. Normal levels at this age for hemoglobin are in the range of 13 to 20 g/dL with a mean of 16; for hematocrit, 42 to 65% with a mean of 50%. Diagnostic considerations include the topics discussed in the following sections.

Fair Complexion

If the vital signs and the physical examination are normal and the CBC does not indicate anemia, the presenting concern probably relates to the baby's normal fair-skinned coloring. Offer appropriate explanation and discharge home.

Anemia (see Chapter 59 and Chapter 87)

Anemia in the neonate is not usually recognized as pallor until the hemoglobin falls below 10 g/dL. It is most commonly, but not necessarily related to excessive hemolysis of the red blood cells and often is accompanied by jaundice. The more common etiologies might include the following:

- *Hemolytic disease of the newborn:* Complete maternal and neonate blood typing should be done, as well as a Coombs' test. Depending on the degree of anemia and the baby's functional status, crossmatching for possible packed cell or exchange transfusion should be inaugurated.
- *Congenital erythrocyte membrane defects:* These defects can usually be suggested by careful evaluation of the blood smear.
- *Malignancy:* Congenital leukemia is a rare entity that should be discernible on the blood smear.
- *Congenital and aplastic anemias:* Diamond-Blackfan syndrome, transient erythroblastopenia of childhood, sickle cell disease, and Fanconi's syndrome do not usually appear in the early neonatal period.
- *Blood loss:* Blood loss can occur through the gastrointestinal tract (Meckel's; clotting disorder) and should be suspected if a stool is positive for blood. Imaging evaluation is indicated. Significant blood loss can also occur into a large cephalohematoma or an intracranial hemorrhage in an infant.
- *Disseminated intravascular coagulopathy (DIC):* DIC may occur in association with bacterial sepsis, viremia, and shock, accompanied by petechiae and purpura. Platelet count is significantly diminished in these ill children. Evaluation is directed at finding a cause and therapy at replacing deficient elements (see Chapter 3, Chapter 65, and Chapter 87).

Ill Children (see Chapter 3)

Pallor may be the most striking presenting complaint in the neonate who is ill. Children in septic or cardiogenic shock may appear pale because of poor perfusion, without necessarily being anemic. Hypoglycemic infants and those with severe electrolyte disturbances may also present with pallor. Rapid diagnosis and emergency administration of glucose and volume restoration are essential.

Mottling

Mottling in the neonate is the patchy appearance of the body surface, resulting from prominent dilation of the superficial veins showing through the thin skin and causing a mosaiclike, patchy appearance. Mottling may be a normal variant when it appears in an otherwise normal baby, undressed in a cool ambient temperature. It is more likely to appear in a preterm baby with thin skin.

However, mottling can be an ominous diagnostic sign in a neonate. It may be indicative of hypovolemia and poor perfusion in a baby in shock or a septic baby. A careful history and complete physical examination with cautious evaluation of the vital signs need to be done. If there is doubt about the baby's status, an electrolyte panel and stat glucose should be drawn. Treatment of the underlying cause and restoration of hemodynamic stability should be inaugurated in the ED.

SKIN FINDINGS

The quality of the newborn's skin varies with gestational age. The premature infant's thin, almost translucent skin is in sharp contrast to the dry, cracked, peeling skin of the postterm infant. The term infant's skin is usually pale pink in color

with some diffuse superficial peeling noted at several days of age. Normal peeling of the superficial layers of the skin should be differentiated from the full-thickness skin loss that is associated with *staphylococcal scalded skin syndrome*. *Excoriations* when noted in an irritable or jittery infant, especially when they are located primarily on the nose or knees, may be a sign of withdrawal in an infant exposed to narcotics prenatally.

Seborrheic dermatitis (see Chapter 99) is a localized scaling or crusting eruption that most commonly involves the scalp ("cradle cap"), forehead, and area behind the ears. Occasionally, the diaper area may be involved. The eruption may have a greasy appearance and the lesions are generally nonpruritic.

Hair

Soft, downy, fine body hair, *lanugo,* located primarily on the back and shoulders, is a normal finding. However, a tuft of coarse, dark hair located in the midline lumbosacral region may be associated with spina bifida occulta.

Papular Rashes (see Chapter 62 and Chapter 63)

A variety of papular rashes may be observed in the healthy newborn. Characteristic body distribution patterns and age at appearance of these typical rashes may help differentiate them from more worrisome conditions.

Milia are small 1 to 2 mm ivory or yellow papules located primarily on the forehead, nose, and cheeks of the newborn. Milia are keratin retention cysts that require no treatment because they will spontaneously rupture and disappear during the first 3 to 4 weeks of life.

Erythema toxicum is a more generalized eruption of small papules or pustules on an erythematous base that may occur anywhere on the body. Usually presenting during the first 3 to 4 days of life, these lesions may be noted as late as 2 weeks of age. Herpes simplex infections, although usually vesicular, and impetigo, usually pustular or crusted, may be entertained in the differential diagnosis of this eruption. If the diagnosis is in question, a smear of the papular contents will show a predominance of eosinophils with a relative absence of neutrophils and no organisms.

Erythematous papules or pustules (rarely comedones) confined primarily to the cheeks, chin, and forehead are characteristic of *neonatal acne.* Lesions are secondary to the influence of circulating maternal hormones and usually appear at 3 to 4 weeks of age and disappear within a few weeks.

Vesicular Rashes (see Chapter 67)

The most important condition to consider when evaluating a vesicular eruption is a herpes simplex virus (HSV) infection because it has the most significant associated morbidity and mortality. Most neonatal *HSV infections* are caused by HSV type 2. Although transplacental transmission of HSV can occur, most neonatal disease is acquired from infection of the maternal genitourinary tract. Primary maternal infection at the time of delivery is associated with a 40 to 50% risk of disease transmission to the newborn, whereas recurrent disease has a much lower risk of transmission (less than 5%). Clinical disease in the newborn may range from localized infection of the CNS, skin, eyes, or oropharynx to disseminated viremia with multisystem involvement. The infant generally appears well at birth but then becomes ill about day 4 to 7, at which time a vesicular eruption may be noted. Unfortunately, not all HSV-infected infants have the typical vesicular eruption, making early diagnosis difficult. Small (1- to 2-mm) vesicles on an erythematous base, which may become pustular in 24 to 48 hours, are the most common lesions. At times, only 1 or 2 vesicles may be present; lesions are noted most often on the scalp and face, the presenting part of the infant in a normal delivery. Early diagnosis and prompt initiation of antiviral therapy can improve morbidity and mortality.

Neonatal varicella may develop when maternal varicella infection occurs during the last 2 to 3 weeks of pregnancy or the first few days postpartum. The severity of neonatal disease depends on the timing of the maternal infection. If maternal disease onset is 5 or more days before delivery, the infection in the newborn is usually mild because of the transplacental passage of maternal varicella IgG antibody. In contrast, if maternal disease onset is within 4 days before delivery or within 48 hours after delivery, the neonate is at risk of developing severe infection with up to a 30% mortality usually caused by pulmonary or visceral involvement. Diagnosis can usually be made by the history and the characteristic rash, vesicles on an erythematous base (dew drops on a rose petal). The primary differential diagnosis includes HSV-1 or HSV-2 infection. Acyclovir may be considered for moderate or severe cases of neonatal varicella. Varicella-zoster immunoglobulin (VZIG) is recommended for uninfected infants born to mothers whose onset of chickenpox was within 5 days before to 2 days after delivery.

Incontinentia pigmenti is an X-linked dominant disorder with both skin and systemic lesions (affecting the eyes, CNS, and bone). It presents at birth or shortly thereafter with an inflammatory vesicular or bullous rash that develops in crops over the trunk and extremities. The cutaneous lesions have four phases (inflammatory vesicles or bullae, verrucous lesions, whorled hyperpigmentation, and hypopigmented patches) that may overlap and occur in an irregular sequence. Suspected cases should be referred to a dermatologist for evaluation because of the potential for systemic involvement.

Miliaria, or neonatal prickly heat, is caused by sweat retention and is characterized by easily ruptured, tiny (1- to 2-mm) vesicles located primarily on the face, chest, and back.

Impetigo appears as superficial vesicular, pustular, or bullous lesions on an erythematous base. In the newborn, lesions tend to occur primarily in the diaper area, folds of the neck, or axillae.

Vascular Lesions

The *salmon patch (nevus simplex)* is the most common vascular lesion of infancy. It is a pale pink macular lesion that is

found most commonly on the nape of the neck (often called a stork bite), forehead, nasolabial region, or upper eyelids. With the exception of the lesions on the nape of the neck, most will fade within the first year of life.

Nevus flammeus (port-wine stain) consists of mature dilated dermal capillaries and presents at birth as pink to purple macular lesions that can vary tremendously in size, sometimes involving a significant portion of the body (Klippel-Trenaunay-Weber syndrome should be considered when the port-wine stain involves a lower limb). Unilateral facial port-wine stains in a trigeminal nerve distribution may be associated with the Sturge-Weber syndrome (seizures, intracranial calcifications, and hemiparesis). These lesions generally will not fade with time.

Although *capillary hemangiomas (strawberry hemangioma)* may be present at birth, most develop during the first few weeks of life. Lesions may occur anywhere on the body and typically begin as small, well-demarcated telangiectatic macules that subsequently develop into raised bright red or purple tumors with distinct borders. Most lesions will go through a period of rapid growth over the first 6 months of life, followed by a static period and then spontaneous involution, usually by 5 years of age.

Cavernous hemangiomas are deep-seated capillary hemangiomas that usually present at birth as a diffuse swelling with little change in the color of the overlying skin or a bluish hue. Most involute spontaneously with time.

Hemangiomas that may require intervention during the neonatal period are those that by location or size may compromise vital structures such as the eyes, nares or auditory canals; lesions that by their size or location (e.g., perianal or labial lesions) are susceptible to trauma, ulceration and secondary infection; and large, rapidly enlarging hemangiomas associated with thrombocytopenia and a consumption coagulopathy (Kasabach-Merritt syndrome).

Pigmentary Changes

Mongolian spots are poorly circumscribed blue–black, gray, or brown large macular lesions generally located over the lumbosacral region, buttocks, and lower limbs in over 80 to 90% of African-American, Native American, Hispanic, or East Asian infants. The incidence is less than 10% in Caucasian infants. Lesions will usually fade during the first few years of life.

Café-au-lait macules are round or oval, brown macular lesions varying in size from less than 1 cm to greater than 20 cm. Although normal individuals may have these lesions, they may be a sign of neurocutaneous disease, most commonly neurofibromatosis.

Ash-leaf macules are irregular hypopigmented macules often with an oval or “ash-leaf” appearance found in 70 to 90% of individuals with tuberous sclerosis. This is an autosomal-dominant condition characterized by CNS lesions, seizures (infantile spasms), retinal lesions, cardiac rhabdomyomas, and renal lesions (hamartomas or cystic kidneys).

HEAD AND NECK PROBLEMS

Head Size and Shape

Shape, size, and symmetry are all factors that must be considered in the evaluation of the neonate's head. *Microcephaly* refers to a head circumference that is less than 2 standard deviations below the mean, or less than the 3rd percentile for age and sex. Microcephaly is usually a sign of a severe underlying abnormality of brain growth or development and is often associated with mental retardation. It may be secondary to a variety of causes, including Down syndrome, congenital TORCHS infections, and fetal alcohol syndrome. *Macrocephaly* is a head circumference that is greater than 2 standard deviations above the mean, or greater than the 97th percentile for age and sex. An excessively large head may be familial or suggestive of hydrocephalus, storage disease, or intracranial hemorrhage.

Molding of the skull bones during the vaginal delivery process is a common cause of temporary asymmetry and scalp edema, *caput succedaneum.* Caput succedaneum is an ill-defined generalized swelling of the soft tissues of the scalp that extends across suture lines. Generally, both caput succedaneum and skull molding spontaneously resolve by 7 to 10 days of age. Trauma during the birth process may produce a *cephalohematoma,* a subperiosteal hemorrhage, distinguished from a caput by the fact that the swelling never crosses suture lines. However, the diagnosis can be difficult in the immediate newborn period if overlying scalp edema is present. Most commonly, a cephalohematoma is unilateral, but it can be bilateral. Cephalohematomas resolve slowly over 4 to 6 weeks with possible calcification and the formation of a hard bump on the scalp that is often a source of great concern to parents. Occasional complications resulting from the breakdown and resorption of large hematomas are anemia or jaundice.

Overriding cranial sutures, caused by the pressures exerted on the skull during its descent through the pelvis, may be noted for the first several days of life. Overriding sutures that are palpable beyond this time may be a sign of underlying brain pathology and deserve further evaluation. Ridging or prominence of cranial sutures may be a sign of *craniosynostosis,* a premature fusion of cranial sutures. Overriding sutures are ballottable, but if the sutures are rigid and have a heaped-up solid closure, radiographs or even a computed tomography (CT) scan should be done to rule out craniosynostosis. Soft areas, *craniotabes,* are occasionally found on palpation of the parietal bones during the first several days of life, especially in premature infants. Soft areas noted in the occipital region may be suggestive of osteogenesis imperfecta or other syndromes and should be investigated.

At birth, the newborn has two *fontanels*. The anterior fontanel, situated at the junction of the coronal and sagittal sutures, usually measures about 2 × 2 cm (can be up to 5 to 6 cm in its largest diameter), and normally closes between 9 to 18 months. The posterior fontanel, situated at the junction of the lambdoidal and sagittal sutures, generally measures between 0.5 to 1 cm (may be closed at birth in some cases), and usually closes to palpation by 3 to 4 months of age. Enlarged fontanels may be associated with a variety of conditions, including prematurity, hypothyroidism, or hydrocephalus. Increased intracranial pressure produces a full or bulging fontanel, whereas dehydration produces a

depressed fontanel. A fontanel that appears full while the infant is supine or crying should be reassessed while the infant is held upright and sleeping or feeding before it is determined to be full or bulging.

Face

Facial asymmetry is usually secondary to in utero position. Commonly, when the face and neck are pressed against the shoulder in utero, a characteristic flattening of the face and angle of the jaw is noted on that side because of displacement of the mandible. This facial asymmetry will resolve spontaneously in a few weeks.

Neck (see Chapter 45 and Chapter 46)

Congenital muscular torticollis is a positional abnormality of the neck resulting in abnormal tilting and rotation of the head. It is believed to be secondary to intrauterine positioning or trauma to the soft tissues of the neck during delivery, with resulting ischemia of the sternocleidomastoid muscle secondary to venous occlusion. This leads to edema and degeneration of the muscle fibers with eventual fibrosis of the muscle body. Although congenital muscular torticollis may be noted at birth, it usually manifests at 2 to 4 weeks of age. The incidence is increased in breech presentations and difficult deliveries. Unilateral contracture and fibrosis of the sternocleidomastoid muscle results in a characteristic head tilt toward the affected side and the chin pointing toward the opposite side. On examination, a firm, nontender mass may be felt within the body of the sternocleidomastoid muscle. Treatment consists of passive stretching exercises of the neck and repositioning toys and mobiles in the crib to stimulate the infant to look toward the side opposite the preferred gaze. Occipitocervical spine anomalies, such as the Klippel-Feil syndrome (congenital fusion of two or more cervical vertebrae; clinical triad of short neck, limited neck motion and low occipital hairline), are rare causes of torticollis that present in the newborn period.

Congenital neck lesions may present during infancy or sometimes much later in childhood. The most common lesions include, *thyroglossal duct cysts* (midline in the neck and inferior to the hyoid bone), *branchial cleft cysts* (along the lateral neck), and *cystic hygromas* (usually located behind the sternocleidomastoid muscle in the supraclavicular fossa; two-thirds of cystic hygromas are present at birth).

Redundant skin on the back of the neck or webbing in a female infant are suggestive of *Turner's* syndrome and may be associated with lymphedema of the dorsum of the hands and feet in the newborn.

EYE PROBLEMS

The newborn is very nearsighted at birth, with a visual acuity of about 20/400. The eyelids are closed most of the time, and any attempt to force them open usually meets with marked resistance and causes blepharospasm. Holding the infant upright and gently swaying him or her from side to side or up and down often induces the eyes to open spontaneously. Common neonatal ophthalmologic concerns include leukokoria, neonatal conjunctivitis, excessive tearing, scleral and subconjunctival hemorrhages, and uncoordinated eye movements.

Leukokoria

The pupillary light reflex is a simple test that takes only moments and should be performed on all newborns. In the normal newborn, a “red reflex” is seen when the ophthalmoscope is held 10 to 12 inches in front of the eyes. A white pupillary light reflex, or leukokoria, is never normal in the newborn. Leukokoria may be a sign of several conditions of variable severity and prognosis such as colobomas, cataracts, retinal detachment, retinopathy of prematurity, or retinoblastoma (the most common signs are leukokoria [60%] and strabismus [20%]). Therefore, all infants with an abnormal pupillary light reflex should be referred to an ophthalmologist for a prompt evaluation.

Conjunctivitis (see Chapter 24 and Chapter 120)

The major causes of *neonatal conjunctivitis, or ophthalmia neonatorum,* are chemical, chlamydial, bacterial, and viral. The time of onset of symptoms after birth can help identify the causative agent. Mild inflammation of the conjunctivae that begins 12 to 24 hours after birth is typically caused by the prophylactic eye drops instilled at birth. This *chemical conjunctivitis* usually resolves by 48 hours of age. *Neisseria gonorrhoeae* conjunctivitis generally appears 2 to 5 days after birth, whereas conjunctivitis caused by *Chlamydia trachomatis* presents between 5 to 14 days after birth because of its longer incubation period. Gonococcal infection may be delayed beyond 5 days of age because of partial suppression by the prophylactic drops instilled at birth. Gonococcal infection usually manifests as marked inflammation of the eyelids, chemosis, and copious purulent discharge. Presentation of chlamydial infection, which is primarily localized to the palpebral conjunctiva, can vary from mild inflammation to severe swelling of the eyelids with copious discharge. Of neonates with chlamydial conjunctivitis, 10 to 20% have chlamydial pneumonia, which can either occur simultaneously with the eye infection or up to 4 to 6 weeks later. Gonococcal conjunctivitis is considered a medical emergency because the infection can spread to the cornea, producing corneal ulceration and perforation. HSV is a less common cause of neonatal conjunctivitis. The presence of characteristic skin lesions can help in the diagnosis. Gram stain and cultures are essential in the evaluation of neonatal conjunctivitis. Treatment is discussed in detail in Chapter 24 and Chapter 120.

Excessive Tearing (see Chapter 24 and Chapter 120)

Congenital obstruction of the nasolacrimal duct, dacryostenosis, is the most common cause of excessive tearing in the newborn. Dacryostenosis should be differentiated from *congenital or infantile glaucoma,* a serious but fortunately rare cause of excessive tearing. Most cases of infantile glaucoma presenting during the first 3 months of life are bilateral, whereas dacryostenosis is usually unilateral.

Increased wetness of the affected eye relative to the normal eye, excessive tearing, mucoid eye discharge, and crusting

along the eyelid margins are the usual presenting symptoms of dacryostenosis. Gentle pressure along the medial canthal region over the lacrimal sac may produce a reflux of tears or purulent material onto the surface of the eye, confirming the diagnosis. In addition to excessive tearing, infants with glaucoma also present with rhinorrhea, photophobia, and corneal haziness. The cornea may be inspected after instillation of fluorescein dye to rule out a *corneal abrasion* as the reason for the excessive tearing. Uncomplicated cases of nasolacrimal duct obstruction should be managed with gentle cleansing of the eyes with warm water followed by local massage of the nasolacrimal duct several times per day. Topical antibiotic ointments should be prescribed if there is associated conjunctivitis or purulent discharge. Suspected cases of infantile glaucoma require immediate ophthalmologic evaluation.

Scleral and Subconjunctival Hemorrhage

Scleral and subconjunctival hemorrhage are often noted in the newborn secondary to birth trauma. These lesions are common and spontaneous resolution within 1 to 2 weeks is the rule. If the funduscopic examination is performed, similar hemorrhages may be noted on the retina in about 25% of newborns. The presence of retinal hemorrhages should also raise the possibility of intentional trauma. Specifically, the *shaken baby syndrome* has been associated with flame-shaped retinal hemorrhages and subdural hematomas.

Transient Neonatal Strabismus (see Chapter 25)

Intermittent esotropia or exotropia may be noted in normal infants during the first 2 to 3 months of life. These deviations are believed to be secondary to neuromuscular immaturity and generally resolve spontaneously by 4 months of age. If such eye deviations are constant instead of intermittent, the infant should be referred for an ophthalmologic examination. In many infants, a broad, flat nasal bridge and prominent epicanthal folds may obscure a portion of the sclera near the nose and create the appearance of esotropia. This *pseudostrabismus,* or apparent deviation of the eyes, is an illusion that can be dispelled by the finding of symmetric bilateral pupillary light reflexes.

MOUTH PROBLEMS (SEE CHAPTER 49)

Normal Findings

Common normal findings in the oropharynx include natal teeth and benign gingival cysts. The incidence of *natal teeth* (teeth present at birth) is about 1 in every 3000 live births. The mandibular central incisors are the most commonly affected teeth. Because most natal teeth are primary teeth that have erupted early, they should be extracted only if they are loose and pose a danger of aspiration, cause discomfort to the mother or child during nursing, or are confirmed to be supernumerary by radiographic examination.

Benign gingival cysts (see Chapter 124) are found in 75% of newborns. *Epstein's pearls* are usually single, small, white, keratin-filled cysts found along the midline of the palate. *Bohn's nodules* are mucous gland cysts that appear as multiple, firm, grayish white lesions along the gums and occasionally on the palate. *Dental lamina cysts* are formed by remnants of dental lamina epithelium and appear as small, cystic lesions along the crests of the mandibular and maxillary mucosa. They are usually larger and more lucent than either Epstein's pearls or Bohn's nodules. These cysts generally disappear by 4 weeks of age.

Thrush

Thrush is caused by *Candida albicans.* Diagnosis may be based on clinical examination. Creamy white plaques located on the buccal mucosa or tongue that are difficult to remove and may cause bleeding when scraped are characteristic of candidiasis. Treatment consists of local application of nystatin suspension four times a day. Topical application of nystatin ointment to the mother's nipples may be indicated in recurrent or refractory cases if the infant is breast-feeding.

CHEST AND BACK FINDINGS

This section discusses those external lesions on the thorax and back of the neonate for which a parent may bring the baby to emergency care. Intrathoraciclesions and diseases which present with secondary symptomatology are discussed in detail in other contexts elsewhere in this book (see Chapter 68, Chapter 95, and Chapter 119).

Normally, the term newborn's thorax is symmetric and barrel-shaped. It is graced with two nipples anteriorly, each about 10 mm in diameter and slightly elevated and stippled. Respiratory excursion should be symmetric and accompanied by simultaneous movement of the abdomen. In the midline of the back, the tips of the vertebrae can be palpated but not visualized. The rib cage is not flared or depressed. There are a number of variations of this normal anatomy, some normal and some not, which may be striking enough for a parent to bring the child to the ED.

Respiratory Excursion (see Chapter 68, Chapter 84, and Chapter 95)

Intrathoracic disease or anomaly may be suggested by variations in the normal symmetric excursion of the thorax. If thoracic excursion is asymmetric or if there is significant tachypnea or retraction accompanied by grunting and either excessive or paradoxic excursion of the abdomen, a chest radiograph is warranted and pulse oximetry should be checked.

Fractured or Absent Clavicle(s)

In the course of vaginal delivery, the clavicle may be fractured, resulting in asymmetry at the shoulder girdle area. Palpation of the clavicle may reveal a “drop-off” in the continuity of the bone and possible crepitation when gentle pressure is applied. A confirmatory radiograph should be taken and appropriate reassurance offered for the healing

process. Much more rarely, clavicles may be absent bilaterally with resultant low positioning of the shoulders. This positioning may be indicative of the dominant genetic defect known as cleidocranial dysostosis and requires orthopedic and genetic evaluation.

Pectus Excavatum

Relative depression of the lower sternum and rib cage is usually a normal variant unless accompanied by signs of respiratory distress.

Xiphoid Process

Parents may feel a firm small mass in the midline at the distal end of the sternum. This is the xiphoid process, angled outwardly, and is a normal variant.

Breast (see Chapter 12 and Chapter 84)

Supernumerary Nipples

A round, possibly slightly elevated or slightly depressed lesion, about 10 mm in diameter, lighter in shade than the nipple and located about 2 to 3 cm below, is a supernumerary nipple. This is a normal variant and will remain permanently. Uncommonly, these may be associated with renal lesions, but that possibility need not be investigated in a child who is otherwise well.

Enlarged

- *Breast buds:* Prominent, nontender breast tissue in the neonate of either sex is a normal variant, probably related to maternal estrogen. This subsides with time, and no therapy is indicated. Often, colostrumlike material can be extruded, but efforts to do this should be gentle and conducted under hygienic conditions.
- *Breast cellulitis/abscess:* Breast tissue that is hypertrophied, reddened, and tender is probably infected. The child may or may not be febrile. Warm compresses and intravenous antibiotic treatment, including coverage for probable staphylococcal origin, should be inaugurated after appropriate cultures are taken. The baby should be admitted for continuing therapy.

Absent

An absent nipple may be associated with an ipsilateral absent pectoralis muscle. Chest radiograph should be taken and the baby referred for genetic and orthopedic evaluation.

Spinal Column Defects

Spina Bifida

A grossly apparent spina bifida lesion, complete with lower extremity flaccidity and meningeal extrusion in the midline of the back, obviously should not have been missed in the hospital nursery. The baby delivered at home however, may be referred into the ED for initial management. Sterile, moist dressing should be applied and the baby admitted for neurosurgical, orthopedic, and urologic management. If the home delivery occurred under less than sterile conditions, cultures should be taken and inauguration of appropriate antibiotic therapy considered.

Sacral Dimple

A midline dimple of the lower back, with or without a tuft of hair, or a lipomatous intracutaneous or subcutaneous lesion in that area may be clues to the presence of spina bifida occulta, a less obvious form of spina bifida. This may or may not be associated with lower extremity deformity. The dimple may also be the external manifestation of a sinus tract connecting to the intradural space without vertebral anomaly, which would leave the infants susceptible to meningeal infection. On the other hand, the dimple may be a normal skin indentation. If in doubt, scheduling of magnetic resonance imaging and neurosurgical referral should be considered.

ABDOMINAL AND PERINEAL FINDINGS

The neonatal abdomen is full but is neither distended nor scaphoid. The liver is normally palpable 2 to 3 cm below the right costal margin; the spleen is not usually palpable; the lower edges of the kidneys may be felt with deep palpation. There should be no palpable extraneous masses. Constipation, meconium passage, and vomiting are discussed in Chapter 14, Chapter 78, Chapter 96, and Chapter 118. This section discusses those external findings in the abdomen and perineum that might cause a parent to bring the new baby to the ED.

Umbilicus

The umbilical cord is tied or clamped at the time of delivery and usually sloughs off by the tenth day. Careful umbilical care consists of gentle hygienic measures and cleansing with isopropyl alcohol several times a day. Still, the umbilical area may be a source of concern for the parents, who may appear in the ED with their neonate.

Discharge

Discharge from the umbilical area may occur and is benign if it is clear or yellow-tinged and thin. Reassurance and instruction in hygienic measures are all that is necessary. A thick, purulent discharge, accompanied by intense redness and apparent tenderness, however, suggests infection. The discharge should be cultured and treated vigorously with antibiotics because the umbilical vessels are potential entry points for systemic invasion.

Granuloma

Granulation tissue, lumped into a small ball about 1 cm across and attached in the umbilical area, can be cauterized with a silver nitrate stick. The parents should be forewarned that the area will turn transiently black. Often, this treatment has to be repeated in a week or so.

Umbilical Hernia

Umbilical hernia is a result of incomplete merging of the recti muscles at the ring through which the cord had been protruding. It is often accompanied by a larger rectus diastasis extending superiorly, sometimes to as high as the xiphoid process. The size can vary from as little as a few millimeters to as much as 4 or 5 cm. It is covered by skin. With crying or straining, portions of the intestine and omentum can be palpated, but not visualized, within the hernia. No treatment is necessary in the neonatal period because these almost always close as the baby becomes ambulant and strengthens the rectus muscles. Abdominal bandages are unnecessary. Rarely, at a later age, an umbilical hernia may strangulate and require surgery.

Omphalocele

An omphalocele is essentially a large hernia into the base of the cord, but it is covered only by peritoneum, not skin. It contains a significant amount of intestine and, rarely, a lobe of the liver. The child should be admitted for early surgery.

Genital Area

The penis should be at least 1 cm in length with a urethral opening at the tip. The testes are usually palpable within the scrotal sac. The labia majora overlie and cover the labia minora.

Vaginal Discharge and Bleeding (see Chapter 76 and Chapter 77)

White mucoid discharge, which may be thick, in the vaginal opening is a normal finding. Vaginal bleeding after the first day or two and during the first week is also a normal occurrence. It is the result of postpartum estrogen withdrawal.

Inguinal Mass

A mass palpable in the scrotum may be an inguinal hernia or a hydrocele or a combination of the two. Hernias are usually easily reducible in the neonate. Hydroceles are fluid-filled and transilluminate readily. A mass within the labia majora is most likely ovary or intestine that has passed through an inguinal hernia. These are somewhat more likely to incarcerate than male hernias.

Hypospadias

When the urethral opening is not at the tip of the penis but on the glans, the baby has first-degree hypospadias. When the opening is on the shaft, it is second degree and on the perineum, third degree. Infants with second- and third-degree hypospadias should be referred for urologic evaluation and imaging of the genitourinary tract.

Ambiguous Genitalia

The possibility of ambiguous genitalia should be considered in a male if the apparent penis is small and there is third-degree hypospadias with a cleft in the scrotum; in the female, genitalia are ambiguous if there appears to be an unusually long clitoris with partial or complete fusion of the labia majora and if there is a firm mass in the labia. In the female, but not in the male, such pseudohermaphroditism may be associated with *congenital adrenal hyperplasia,* with or without “salt-losing” symptomatology (see Chapter 97). To establish gender identity and to evaluate for the possibility of congenital adrenal hyperplasia, electrolyte, imaging, and chromosomal studies need to be done early in the child's neonatal period under the supervision of a urologist and geneticist and possibly an endocrinologist.

Imperforate Anus

An imperforate anus may not be obvious on external examination. The finding of an anus that appears to be located considerably more anteriorly than expected might suggest a fistula from the lower rectum to the skin, detouring around the anus. Meconium may actually pass through this fistula, simulating normal rectal passage. The area should be examined carefully, looking for the normal perianal–anal puckering, which will not be present if the anteriorly placed opening is a fistula. The rectal examination should be carefully performed. The clinician should consider imaging studies and surgical referral.

ORTHOPEDIC CONCERNS (SEE CHAPTER 123)

Most neonatal orthopedic problems are deformities secondary to intrauterine positioning. Some problems (e.g., metatarsus adductus) require only parental reassurance and expectant management, whereas others (e.g., congenital

clubfoot and hip dysplasia) require early orthopedic attention.

Developmental Hip Dysplasia

Developmental dysplasia of the hip (DDH) applies to a range of hip pathology, ranging from instability to frank dislocation, that may either be present at birth or develop during infancy. The Ortolani and Barlow maneuvers may be used in the neonatal period to evaluate for hip instability. Both tests are performed with the infant in a supine position and the hips and knees flexed to 90 degrees. Each leg is examined separately, not simultaneously. In the Ortolani maneuver, gentle abduction and lifting of the femoral head anteriorly produces a palpable “thunk” or “clunk” as the examiner relocates a dislocated hip. Nonpathologic processes such as ligamentous snapping can produce hip clicks that differ from the pathologic “clunk” associated with DDH. In the Barlow maneuver, the examiner attempts to dislocate the hip by gentle adduction and posterior axial pressure on the thigh. Confirmatory radiographs should be taken and orthopedic consultation obtained in a timely manner.

In-Toeing

The differential diagnosis of in-toeing is guided by the age at presentation. In the newborn period, the most common causes of in-toeing are metatarsus adductus and clubfoot. *Metatarsus adductus* is a functional deformity, resulting from intrauterine positioning, in which the forefoot is in adduction with respect to the hindfoot. Most cases resolve spontaneously by 3 to 4 months of age. If the forefoot cannot be brought into the neutral position either by stroking the lateral border of the foot or by gently straightening it, referral to an orthopedic surgeon for cast correction is indicated.

Congenital clubfoot is a pathologic deformity consisting of three components: forefoot varus; heel varus; and ankle equinus. Clubfoot may be either an isolated deformity or seen in association with other neuromuscular anomalies such as arthrogryposis, cerebral palsy, myelomeningocele, or amniotic band syndrome. Orthopedic treatment should begin in the first week of life.

Brachial Plexus Injuries (see Chapter 36)

Lateral traction on the head and neck during delivery can result in injury to the brachial plexus. Clinical signs relate to the site of the traumatic injury. *Erb's palsy,* the most common birth injury of the brachial plexus, results from injury to the upper plexus affecting the C5 and C6 roots, the upper trunk, or its divisions. The affected arm is held with the shoulder adducted and internally rotated, the elbow in extension and pronation, and the wrist in flexion (“waiter's tip posture”). On examination, the Moro reflex (allowing the infant's head to drop back suddenly results in abduction and upward movement of the arms followed by adduction and flexion) is asymmetric; there is weakness of shoulder abduction, flexion, and supination; the biceps reflex is decreased; and there is slight weakness of wrist and finger extensors. In addition, when the C4 root is involved, ipsilateral hemidiaphragmatic paralysis may be appreciated by fluoroscopic examination.

Klumpke's paralysis results from injury to the lower plexus affecting the C8 and T1 roots, the lower trunk or its divisions. The injury primarily affects the muscles of the hand. The infant presents with clawing of the affected hand (hyperextension at the metacarpophalangeal [MCP] joints and flexion of the interphalangeal joints), the elbow is held in flexion and the wrist is usually held in extension, unless there is injury to the middle trunk. On examination, the palmar grasp is decreased, and the triceps reflex is decreased.

Treatment consists of immobilization and appropriate positioning to prevent contractures. Orthopedic referral is indicated.

NEUROLOGIC CONCERNS

Neonatal Seizures (see Chapter 70 and Chapter 83)

Neonatal seizures may be difficult to recognize clinically because it is rare for newborns to have symmetric, generalized tonic-clonic convulsions. It is much more common to see seizure episodes that present as focal abnormalities or as subtle findings. Subtle seizures can be difficult to distinguish from the normal spectrum of newborn behaviors, jitteriness, or benign myoclonic movements. Benign myoclonic movements are isolated jerky movements of an extremity that occur primarily during sleep. Jitteriness may be differentiated from seizures by its disappearance when the affected extremity is touched or held (Table 102.3).

Clinical Characteristic	Seizure	Jitteriness
Stimulus sensitive	No	Yes
Movement ceases with restraint	No	Yes
Accompanied by autonomic changes	Yes	No
Speed of movements	Slower	Faster
Abnormal eye movements	Common	No

Table 102.3. Clinical Differentiation of Neonatal Seizures from Jitteriness

Four clinical seizure types are recognized in the newborn (Table 102.4): subtle, tonic, clonic, and myoclonic. Generalized tonic-clonic seizures tend not to occur in the first month of life because the newborn's immature nervous system is unable to produce and sustain this type of activity. Not all neonatal seizure types are associated with electroencephalogram (EEG) seizure activity (Table 102.4). It has been hypothesized that these seizures may be originating from areas of the CNS that cannot be detected by surface electrodes (e.g., brainstem or spinal cord).

Seizure Type	Electroencephalogram Seizure Correlation
Subtle	
Sucking or chewing motion	Uncommon*
Lip smacking	
Bicycling of legs	
Apnea	
Eyelid fluttering	
Eye deviations	
Laughter	
Tonic posturing	
Tonic	
Focal	Common
Generalized	Uncommon
Clonic	
Focal	Common
Multifocal	Common
Myoclonic	
Focal	Uncommon
Multifocal	Uncommon
Generalized	Common

*Except for tonic eye deviation, which often has an electroencephalographic correlation.

Table 102.4. Classification of Neonatal Seizures

Subtle seizures, perhaps the most common type of neonatal seizures, are stereotypical repetitive movements such as eye blinking, eye deviations, chewing motions, lip smacking, and bicycling or pedaling movements. Most subtle seizures, especially in term infants, are not consistently associated with EEG seizure activity.

Focal *tonic seizures* present as sustained posturing of a limb, whereas generalized tonic seizures are characterized by either tonic extension of all extremities or, occasionally, flexion of the upper extremities with extension of the lower extremities, mimicking decerebrate or decorticate posturing, respectively. These seizures are often seen in association with severe hypoxic brain injury or with intraventricular hemorrhage, which is most common in premature infants but may also be seen in term infants.

A *clonic seizure* involves rhythmic jerking of one or more parts of the body. Clonic seizures can be focal (affecting only one extremity or both the upper and lower extremity on one side of the body) or multifocal (clonic activity in one extremity that randomly migrates to another part of the body—e.g., left arm jerking followed by right leg jerking). Although focal clonic seizures can result from focal brain lesions, they can also be caused by a generalized metabolic disturbance such as hypoglycemia.

Benign myoclonic jerks are often noted in sleeping infants, especially premature infants, during the first 6 months of life. Unlike this benign sleep-related phenomena, *myoclonic seizures* occur during waking and are single or repetitive rapid jerks of either the entire body or a particular extremity. They are distinguished from clonic seizures by their more rapid speed and a predilection for flexor muscle groups. These seizures usually indicate severe underlying brain pathology or injury such as hypoxic brain injury. Infants with these seizures may later develop infantile spasms. Focal myoclonic seizures typically involve the upper extremity. Multifocal myoclonic seizures are characterized by asynchronous twitching of several areas of the body while generalized myoclonic seizures present as bilateral flexion jerks of the upper extremities and sometimes also the lower extremities.

Although there are a variety of causes for neonatal seizures (Table 102.5), only a few causes (perinatal asphyxia, intracranial hemorrhage, metabolic disturbances, and infection or malformations of the brain), account for most cases. Benign or idiopathic neonatal seizures occur, but this diagnosis should be made only after other causes are thoroughly investigated.

First 24 Hours
Hypoxic-ischemic encephalopathy
Infection (meningitis, TORCHS infection, sepsis)
Direct drug effects (inadvertent anesthetic injection)
Metabolic (hypoglycemia, hypocalcemia)
Intracranial hemorrhage (preterm-intraventricular hemorrhage; term-subdural/subarachnoid hemorrhage)
Pyridoxine dependency
>24 Hours
Infection (meningitis, sepsis, herpes simplex virus)
Intracranial hemorrhage
Metabolic (inborn errors of metabolism, hypocalcemia [dietary])
Intracranial malformations
Drug withdrawal

Table 102.5. Common Causes of Neonatal Seizures by Gestational Age

Perinatal asphyxia is the most common cause of neonatal seizures. During the first several days to weeks of life, signs of acute *hypoxic-ischemic encephalopathy (HIE)* are lethargy, hypotonia, and decreased spontaneous movements. Mild asphyxia is often marked by a transient state of hyperalertness and irritability. Seizures, if they occur, generally begin within the first 24 hours of life and may be difficult to control. The diagnosis of HIE should be strongly considered in a

hypotonic infant with increased deep tendon reflexes.

Intracranial hemorrhage is the second most common cause of neonatal seizures. Intraventricular hemorrhages are seen primarily in premature infants, whereas subarachnoid or subdural hemorrhages are most often seen in large term infants and are caused by birth trauma.

A variety of metabolic disturbances are associated with neonatal seizures. SGA infants and infants of diabetic mothers are at risk for hypoglycemia during the first 24 hours of life. These infants may have a variety of findings, ranging from jitteriness to seizures. Infants of diabetic mothers are also at risk for hypocalcemic seizures during the first 24 hours. Premature infants and infants with perinatal asphyxia are also at risk for early-onset (within the first 2 days of life) hypocalcemia. Late-onset hypocalcemic seizures (after day 2 to 3 of life) may be caused by an imbalance in dietary intake (e.g., cow's milk–based formula), hypomagnesemia, or hypoparathyroidism. Inborn errors of metabolism, pyridoxine dependence, and mitochondrial disorders are less common metabolic causes of seizures in the newborn. Inborn errors of metabolism should be suspected when seizures are associated with vomiting, failure to thrive, hepatomegaly, and altered tone or consciousness.

Several bacterial and viral CNS infections can cause seizures in the newborn. Common bacterial causes are group B *Streptococcus* and *Escherichia coli* infections. HSV encephalitis is an important viral source that must be considered. A newborn with a history of a seizure associated with fever requires a comprehensive evaluation for the cause and should not be diagnosed with simple febrile seizures.

The evaluation of newborn seizures should include a detailed history of prenatal, perinatal, and postnatal events. The examination should be directed toward identifying treatable causes such as infectious or metabolic disturbances. Minimal laboratory evaluation for all newborns with seizures includes a CBC; both bedside and laboratory measurement of serum glucose; serum electrolytes, including calcium, phosphorus and magnesium; blood culture; and cerebrospinal fluid analysis and culture. Cranial ultrasound is especially useful in identifying suspected intraventricular hemorrhages. Measurements of serum ammonia, serum amino acids, and urine organic acids should be obtained if a metabolic defect is suspected. Urine testing with the Clinitest reaction (detects excess excretion of galactose and glucose) can screen for galactosemia.

Appropriate *medical management* includes correction of any metabolic abnormalities and institution of empiric antibiotic therapy. If the results of serum glucose and calcium measurements will not be available in a timely manner, empiric therapy may be instituted if the infant is actively seizing. Hypoglycemia (serum glucose less than 40 mg/dL) is treated with a 2 to 3 mL/kg bolus over 20 minutes of a 10% dextrose solution. Treat symptomatic hypocalcemia (serum calcium less than 8 mg/dL) with 1 to 2 mL/kg of elemental calcium as a 10% calcium gluconate solution by slow intravenous drip. If the infant is actively seizing, a trial of pyridoxine (50 to 100 mg intravenously) may be considered. If the seizures are still not controlled, empiric anticonvulsant therapy may be instituted before consultation with a neurologist. Subtle seizures should not be treated with anticonvulsants before consultation with a neurologist. The anticonvulsant of choice in neonates is phenobarbital and is generally given as an initial intravenous loading dose of 20 mg/kg of body weight. Phenytoin is the secondary drug of choice and may be added if the phenobarbital fails to stop the seizures. Prognosis of neonatal seizures is related to the cause.

Hypotonic Infant (see Chapter 83)

A healthy term newborn normally moves his or her extremities spontaneously and has a dominance of flexor tone. Compared with the term newborn's tone, the premature infant's tone is relatively hypotonic, so corrected gestational age must be taken into consideration during evaluation.

Decreased spontaneous movements, poor head and trunk control, and a preponderance of extensor tone are all characteristics of the hypotonic infant. The healthy term newborn when supported by the trunk in an outstretched prone position, also known as ventral suspension, will flex all extremities against gravity, keep the back straight, and support the head in a neutral position with relation to the rest of the body. In the hypotonic infant, the forces of gravity will allow the back and head to droop downward and the extremities to hang in extension. When held by the axillae in vertical suspension, the hypotonic infant will “slip through” the hands of the examiner instead of contracting the upper extremities to maintain position. Because weakness is often associated with hypotonia, the newborn may present with resultant weak cry, poor suck, or respiratory effort. Weakness should also be suspected if the infant does not briskly withdraw a limb that is subjected to a painful stimuli.

The causes of hypotonia depend on the level of the nervous system that is affected (Table 102.6). Motor dysfunction at any level from the CNS to the muscle itself may result in hypotonia. Central hypotonia involves pathology of the cerebral cortex down to the level of the lower motor neuron or can be caused by systemic disease affecting motor function. Neuromuscular disease can be caused by dysfunction at any of four anatomic sites: anterior horn cell (lower motor neuron), peripheral nerve, neuromuscular junction, and muscle.

Central Nervous System Disease
Perinatal asphyxia (hypoxic-ischemic encephalopathy)
Intracranial hemorrhage
Infection
Hyperbilirubinemia
Neonatal drug withdrawal
Metabolic diseases
Organic and aminoacidemias
Hypercalcemia
Chromosomal abnormality
Down syndrome
Prader-Willi syndrome
Neuromuscular Disease
Anterior horn cell diseases (lower motor neuron)
Type I spinal muscular atrophy (Werdnig-Hoffman disease)
Neonatal poliomyelitis
Type II glycogen storage disease (Pompe's disease)
Peripheral nerve diseases
Leukodystrophies
Guillain-Barré syndrome
Neuromuscular junction diseases
Infantile botulism
Neonatal myasthenia gravis (congenital or acquired transient myasthenia)
Muscle diseases
Congenital myopathies
Mitochondrial myopathies
Glycogen storage disease
Hypothyroidism

Table 102.6. Differential Diagnosis of Neonatal Hypotonia

Central Hypotonia

If muscle weakness is not a significant accompanying feature, a central source for the hypotonia should be considered. Features characteristically associated with central hypotonia include a decreased level of alertness, seizures, and a weak cry; muscle bulk is normal, and deep tendon reflexes are either normal or increased. Perinatal asphyxia and intracranial hemorrhage are the two most common CNS causes of hypotonia that presents in the neonatal period. Depressive symptoms are usually present during the first 1 to 2 days of life in these conditions. An infant who is normal at birth and then develops lethargy, hypotonia, or seizures at several days of age after ingestion of milk protein and carbohydrate may have an inborn error of metabolism. Metabolic diseases should always be taken into consideration in the evaluation of the hypotonic infant, especially when the clinical presentation does not readily fit into any distinct diagnosis or if any unusual odors (see Chapter 47) of the infant or urine are noted (e.g., mustiness—phenylketonuria PKU], sweaty feet—isovaleric acidemia, maple syrup—maple syrup urine disease).

Disorders of the Lower Motor Neuron

Disorders of the lower motor neuron are characterized by hypotonia, muscle weakness, and hypoactive to absent deep tendon reflexes in an otherwise alert infant. Werdnig-Hoffman disease is the most common of the lower motor neuron diseases. In the classic, early-onset form of the disease, infants may present at birth or during the first several weeks of life with generalized weakness; absent deep tendon reflexes; muscle atrophy; fasciculations; and cranial nerve abnormalities, including disordered sucking and swallowing. Infants have a characteristic posture with flaccid tone, abducted limbs, and little spontaneous movement. Respiratory distress may be present. The disease is unfortunately rapidly progressive, and death usually occurs in most patients by 2 years of age.

Disorders of the Neuromuscular Junction

Disorders of the neuromuscular junction are important to recognize because supportive and therapeutic interventions are available. Infants with neuromuscular disorders have hypotonia and weakness like infants with lower motor neuron disease, but infants with neuromuscular disease have normal muscle bulk and normal deep tendon reflexes. Infantile botulism (see Chapter 84) is a toxic abnormality of the neuromuscular junction that is seen in infants under 12 months of age. It is caused by the ingestion of *Clostridium botulinum* spores (from soil, honey, or corn syrup), which germinate in the gastrointestinal tract and produce botulinum toxin. Early symptoms include constipation and poor feeding as a result of poor sucking and swallowing. A descending paralysis develops over the next several days with loss of head control, weak cry, flat facial expression, and eventually, generalized hypotonia. Treatment consists primarily of supportive care; ventilatory support may be required for respiratory failure.

Disorders of Muscle

A variety of primary muscle disorders may present during the neonatal period with the nonspecific features of hypotonia, weakness, decreased muscle bulk, and normal to decreased deep tendon reflexes. Metabolic myopathies are caused by abnormal energy metabolism in the muscle and include disorders of the mitochondria and carnitine metabolism. A mitochondrial myopathy should be considered in a hypotonic infant with lactic acidosis.

Laboratory evaluation of hypotonia is guided by the level of the nervous system believed to be affected. If CNS disease is suspected, brain imaging, an EEG, and endocrine and metabolic determinations may be appropriate. When neuromuscular disease is suspected, muscle enzyme determinations, nerve conduction velocities, electromyography, and nerve or muscle biopsies can be done on a scheduled basis.

Most importantly, the possibility that the hypotonia is really lethargy related to shock or sepsis should be considered.

CONGENITAL INFECTIONS

There are a group of transplacentally transmitted congenital infections often referred to jointly by the acronym TORCHS, which vary in their postpartum presentation according to the time during the pregnancy when they were acquired and with the intensity of the inoculum. These include toxoplasmosis, rubella, cytomegalovirus (CMV), herpes virus infections (herpes simplex and varicella), and syphilis.

Fetal infection with these agents acquired in midpregnancy may result in a neonate born with a complex of findings that may include low birth weight or SGA; jaundice, purpura, and thrombocytopenia; hypotonia; microcephaly; cataracts; microphthalmia; chorioretinitis; hepatosplenomegaly; intracerebral calcifications; congenital heart disease; hypoplastic limbs; hearing loss; cicatricial scarring of the skin; and seizures. Obviously, not all, not even most, of these findings will be present in any one affected infant, but their presence should at least arouse suspicion for the TORCHS syndrome.

Clinically, it is often difficult to distinguish one of these infections from the other. Rubella is more likely to be associated with cataracts and cardiac lesions; toxoplasmosis with chorioretinitis and cerebral calcifications; herpes simplex and varicella with vesicular or cicatricial skin lesions; CMV with hearing loss; and syphilis with bone lesions, snuffles, and palm and hand bullae. However, symptoms overlap. The essentials in management include a careful maternal history and appropriate serologic testing and culturing of both mother and infant.

However, the clinician should keep in mind the possibility that aspects of this symptom complex, particularly purpura and jaundice in a hypotonic baby, may be associated with an active acute septic infection.

MISCELLANEOUS CONCERNS

Neonatal Drug Withdrawal

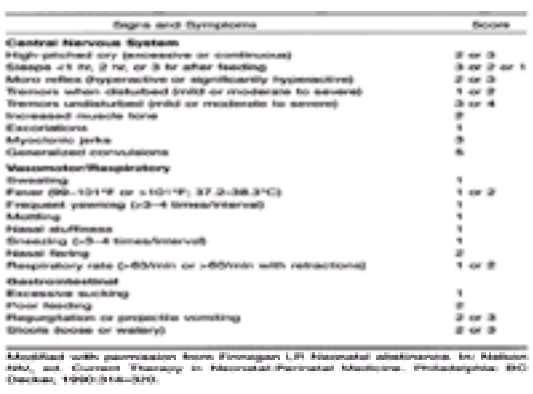
Maternal substance abuse during pregnancy places the newborn at risk for a variety of medical, developmental, behavioral, and psychosocial problems. The particular effects on the newborn infant depend on the type of drug or drugs, the timing of the exposure during gestation, and the frequency of the exposure. Many infants are exposed to cigarettes and alcohol in addition to illicit drugs.

- *Narcotics:* Prenatal exposure to heroin and methadone results in physiologic addiction in the newborn. The symptoms of withdrawal are nonspecific and may not be detected in the newborn nursery, especially when the maternal history of drug abuse is unknown or when the infant is discharged home within 24 hours of delivery. Narcotic antagonists such as naloxone may precipitate withdrawal and should not be used at the time of delivery. Heroin has a short serum half-life, so clinical signs of withdrawal are generally apparent on the first day of life, whereas clinical signs of methadone withdrawal seldom occur before 24 to 48 hours of age because of its long half-life. Symptoms of narcotic withdrawal include irritability, jitteriness, tremors, seizures, disorganized suck and poor feeding, vomiting, diarrhea, sweating, and sneezing. Additional signs of withdrawal that may be noted in the older infant include failure to gain weight and abraded marks along the nose, shins, or occiput caused by the infant's tremulousness and inability to comfort himself or herself.
- *Cocaine:* In utero cocaine exposure has been associated with an increased incidence of abruptio placentae, low birth weight, and preterm delivery. Classically, the cocaine-exposed infant is not so much jittery as he or she is disorganized in sleeping and feeding. These infants are lethargic and poorly responsive but are easily overstimulated and become irritable when alert, making feeding a challenge for many of them. Gavage feeding may be necessary in some instances. Any history of feeding intolerance, vomiting, or abdominal distension needs to be investigated because necrotizing enterocolitis has been noted in term, cocaine-exposed infants. Stool occult blood may be positive secondary to bowel necrosis caused by the vasoconstrictive actions of cocaine. Small CNS bleeds have been described in the basal ganglia and frontal lobes.
- *Amphetamines*: Methamphetamine-exposed infants are often described as being too quiet and may need to be awakened regularly for feedings. Prolonged sleep, depression, and voracious appetite when awakened are characteristic of amphetamine withdrawal.

Evaluation

In general, toxicologic screening can be performed on infants on medical grounds without parental consent. Metabolites of cocaine can be detected for 1 to 2 days after use in the adult and 5 to 7 days in the newborn; amphetamine is present for 1 to 2 days in the adult; and marijuana can be detected in the urine for up to 7 days after use. Most urine toxicology screens use immunoassays and are inexpensive and sensitive but not specific (e.g., antihistamines can cross-react with amphetamines). Newer methods using hair or meconium samples to check for the presence of illicit drugs, especially cocaine, provide a broader window on drug use during pregnancy and are able to document the presence of drugs in the time before delivery.

The Neonatal Abstinence Score (NAS) developed by Finnegan is an objective way of assessing the severity of narcotic withdrawal symptoms (Table 102.7). Infants should be scored at 4-hour intervals for the first several days of life. Three consecutive scores greater than 8 or two scores greater than 12 are an indication for pharmacologic therapy. The NAS can also be used to guide pharmacologic therapy.



Signs and Symptoms	Score
Central Nervous System	
High-pitched cry (excessive or continuous)	2 or 3
Sleeps <1 hr, 2 hr, or 3 hr after feeding	3 or 2 or 1
Moro reflex (hyperactive or significantly hyperactive)	2 or 3
Tremors when disturbed (mild or moderate to severe)	1 or 2
Tremors undisturbed (mild or moderate to severe)	3 or 4
Increased muscle tone	2
Excoriations	1
Myoclonic jerks	3
Generalized convulsions	5
Vasomotor/Respiratory	
Sweating	1
Fever (98–101°F or >101°F; 37.2–38.3°C)	1 or 2
Frequent yawning (>3–4 times/interval)	1
Mottling	1
Nasal stuffiness	1
Sneezing (>3–4 times/interval)	1
Nasal flaring	2
Respiratory rate (>60/min or >60/min with retractions)	1 or 2
Gastrointestinal	
Excessive sucking	1
Poor feeding	2
Regurgitation or projectile vomiting	2 or 3
Loose (loose or watery)	2 or 3

Modified with permission from Finnegan LP: Neonatal abstinence. In: Nelson NM, ed. Current Therapy in Neonatal-Perinatal Medicine. Philadelphia: BC Decker, 1985:314–319.

Table 102.7. Neonatal Abstinence Score

Management

Swaddling and minimizing sensory stimulation are two of the simplest and most effective techniques in managing neonatal drug withdrawal. A blanket or sheet can be draped over the infant's bed to minimize light exposure. Often, these infants do well when placed in a “snugly or front pack” over the mother's chest and abdomen. The mother's regular, monotonous cardiorespiratory sounds can be soothing. Demand feeding with hypercaloric formula (24 to 27 calories per

ounce) may be necessary to maintain weight.

Medications, such as phenobarbital, may be used for narcotic withdrawal if the symptoms do not respond to the above techniques and the infant has significant irritability interfering with sleep and/or feeding. Pharmacologic therapy is generally not required when cocaine or amphetamines are the only drugs of abuse. Phenobarbital is especially good for controlling irritability and insomnia. It requires an initial loading dose of 20 mg/kg. A blood level should be obtained in 24 hours and the maintenance dose adjusted according to the infant's symptoms. The usual maintenance dose is 2 to 6 mg/kg per 24 hours divided every 12 hours. Once a maintenance dose has been established, the dosage is decreased approximately 10% per day based on the NAS.

Prognosis

Drug-exposed infants are at increased risk for abuse and neglect because of a combination of environmental (chaotic family environments, limited financial resources, and limited parenting skills because of the impact of the substance abuse) and infant risk factors (poor attachment, high-pitched cry, irritability, difficult to console, and poor feeding).

Suggested Readings

American Academy of Pediatrics. Management of hyperbilirubinemia in the healthy term infant. Pediatrics 1994;94:558–565.

Bays J. The care of alcohol- and drug-affected infants. Pediatr Ann 1992;21:485–495.

Braveman P, Egerter S, Pearl M, et al. Early discharge of newborns and mothers: a critical review of the literature. Pediatrics 1995;96:716–726.

Brown ER, Zuckerman B. The infant of the drug-abusing mother. Pediatr Ann 1991;20:555–563.

Cheng JCY, Au AWY. Infantile torticollis: a review of 624 cases. J Pediatr Orthop 1994;14:802–808.

Driscoll DJ. Evaluation of the cyanotic newborn. Pediatr Clin North Am 1990;37:1–8.

Elhassani SB, Mease AD. Visible neonatal masses: separating the serious from the benign. Contemp Pediatr 1990;Mar:42–60.

Finnegan LP. Neonatal abstinence. In: Nelson NM, ed. Current Therapy in Neonatal-Perinatal Medicine. Philadelphia: BC Decker, 1990:314–320.

Fleisher GR. Management of children with occult bacteremia who are treated in the emergency department. Rev Infect Dis 1991;13:156–159.

Fuerst RS. Use of pulse oximetry. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1996:823–828.

Hoffmann JIE. Cyanotic heart disease. In: Rudolph AM, ed. Rudolph's Pediatrics. 20th ed. Stamford, CT: Appleton & Lange, 1996:1485–1516.

Molnar GE: Brachial plexus injury in the newborn. Pediatr Rev 1984; 6:110–115.

Newman TB, Maisels MJ. Evaluation and treatment of jaundice in the term newborn: a kinder, gentler approach. Pediatrics 1992;89:809–818.

Shaw KN, Gorelick MH, McGowan KL, et al. Prevalence of urinary tract infection and occult bacteremia in febrile young children in the emergency department. Arch Pediatr Adolesc Med 1996;150:37.

Spitzer AR. Examination of the newborn. In: Athreya BH, Silverman BK, eds. Pediatric Physical Diagnosis. Norwalk, CT: Appleton, 1985:289–298.

Stafstrom CE. Neonatal seizures. Pediatr Rev 1995;16:248–255.

Thilo EH, Townsend SF. Early newborn discharge: have we gone too far? Contemp Pediatr 1996;13:29–46.

Volpe JJ. Neonatal seizures: current concepts and revised classification. Pediatrics 1989;84:422–428.

CHAPTER 103

An Approach to the Injured Child

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Physicians delivering care in an emergency department (ED) must be prepared to treat injured children. Although pediatric trauma victims have needs distinguishing them from adults, it is only in the past decade that investigators have begun to systematically look at the care of the injured child. It is the intent of this chapter to help prepare physicians in the ED for the triage, assessment, and initial care of children with the spectrum of injury from minimal to life-threatening.

Among children aged 1 to 19 years, injuries cause more deaths than all diseases combined. In 1995, more than 20,000 children up to age 19 years died from trauma in the United States. The pyramid of injuries includes more than 500,000 hospitalizations in children and 20 to 30 times as many ED visits. Injuries remain the major factor in 40% of the deaths among children 1 to 4 years of age, and 70% from 5 to 19 years. Of all hospitalizations in pediatrics, 20% result from trauma. An estimated 30,000 children who are injured experience permanent disabilities annually. The economic cost of trauma in childhood is $5 to $8 billion annually in the United States. Although death rates for injuries have declined 25% among children in the last quarter century, those for diseases have decreased by 56%.

The most common cause of traumatic death during childhood is motor vehicle crashes, which account for close to half. Unlike adults, a large proportion of the children injured by automobiles are pedestrians. Overall, homicide and suicide rank second and third as causes of mortality from trauma in the first 19 years of life. Among minority male teenagers, homicide from gunshot wounds is the leading cause of death. More than 5000 of the reported deaths from trauma in children during 1995 were from gunshot wounds.

The approach discussed here provides a framework to decrease morbidity and mortality and is aimed at secondary prevention—after the “impact” has occurred. Secondary prevention is performed in the field by emergency medical services (EMS) providers or after arrival at the hospital. Injury severity scales have been developed to assist in the care of the trauma patient. The two most commonly quoted are the Revised Trauma Score and the Pediatric Trauma Score. These scales have served as useful tools to assess the appropriateness of care provided to injured children. Because both scales are similarly predictive of severity, many recommend use of the Revised Trauma Score because it is suitable for both children and adults. However, the approach to the injured child requires more than the use of a scale for retrospective evaluation.

SPECTRUM OF TRAUMA AND INITIAL TRIAGE

Trauma causes injuries that run the gamut from minimal to fatal, from the splinter lodged in the sole of the foot to the multiply injured victim of an automobile crash. To assist in the initial sorting or diagnostic process, several categorizations of injury are useful: 1) extent—multiple or local; 2) nature—blunt or penetrating; and 3) severity—mild, moderate, or severe (Table 103.1).

Local	Multiple
Blunt	Penetrating
Mild	Severe

Table 103.1. Categorization of Trauma

Trauma can be multiple or local. From a surgical point of view, multiple trauma defines significant injury to two or more body areas: the head and abdomen, the chest and extremities, or other combinations. Although this definition serves a useful purpose in comparing outcomes retrospectively among various centers, it does not suit the needs of emergency physicians who must evaluate and treat children before all diagnostic information has become available. For the purpose of triage in the ED, *multiple trauma* is defined as apparent injury to two or more body areas. Thus, the child who fell from a bicycle, sustaining a forehead laceration and a forearm fracture would be classified in the ED as having multiple trauma (head and extremity), even though the completed evaluation may not uncover any additional or serious injuries.

Localized trauma involves only one anatomic region of the body: head, neck, chest, abdomen/pelvis, or extremities. Again, the designation of local trauma is assigned initially on the basis of even superficial injuries in a given anatomic area, despite the fact that subsequent evaluation might not detect any deeper involvement.

The initial diagnostic task confronting the emergency physician is to decide whether trauma is local or multiple. In some cases, the distinction between local and multiple trauma is obvious. The 13-year-old boy who twisted his ankle coming down with a rebound has a local musculoskeletal injury. The 2-year-old boy who is cyanotic, pulseless, and apneic after a plunge from a ninth story window has sustained critical multiple trauma.

At times, however, the distinction between local and multiple trauma may not be straightforward. A 7-year-old girl who flies over the handlebars of her bicycle onto the sidewalk and is brought to the ED covered with blood, who is thrashing and screaming at every touch, may be judged initially to have serious multiple trauma. After complete and rapid assessment, she may instead have only a superficial forehead laceration and an acute anxiety reaction. The next child with the same history and constellation of symptoms may have a concussion and a ruptured spleen. A third child in a similar scenario may have a rib fracture leading to a pneumothorax; a fourth child may have a buckle fracture of the radius and a depressed skull fracture.

The distinction between local and multiple trauma may be difficult to ascertain initially because 1) some serious injuries are occult in their early phase; 2) some children are difficult to examine because they are nonverbal, uncooperative, frightened, or in pain; and 3) some parents will have played a role in the trauma (i.e., child abuse) and will try to conceal the extent of the injury. Differentiation between local and multiple trauma is thus a dynamic process, and the emergency physician's first impression may change as new evidence accumulates. Generally, it is best to consider trauma multiple until proved otherwise; when the skin is contused over several body parts, the patient is categorized as having multiple trauma until it is known that all the injuries are superficial.

Once it has been decided that multiple trauma has not occurred, the emergency physician should then focus on the specific anatomic region or regions involved. For each region, it is important to ascertain whether the injury is the result of blunt or penetrating trauma and to determine the severity of the wound.

Trauma may be caused by either blunt or penetrating forces. Although most civilian injuries, particularly in childhood, result from blunt trauma, as much as 15% of serious trauma in some centers is related to gunshots, stabbings, and other penetrating wounds. The distinction between these two mechanisms of injury is an important one because it will determine the evaluation based on the expected internal injuries. In this chapter, the management of blunt and penetrating trauma is reviewed for each specific anatomic region.

Finally, the seriousness of injury may vary from mild to severe. Table 103.2 reviews a classification of severity based on history and physical examination, as well as a general schema for disposition of the child with trauma, based on the number of laboratory tests required to diagnose or manage the trauma. Whereas only a few patients with penetrating injuries are considered to have mild wounds, most of the more common blunt injuries seen in the ED are minor. The first categorization by severity depends initially on the history of the incident and the physical examination. An important factor is that a history of a significant or critical force applied during impact increases the index of severity. The schema varies somewhat for each anatomic region; thus, Table 103.2 provides only a general overview. Assessment of severity is essential in the ED because it will determine whether the child is discharged after an examination, receives further observation, undergoes a diagnostic evaluation, or requires immediate intervention. The more laboratory studies required to assess and care for trauma victims, the more likely a child will need admission.

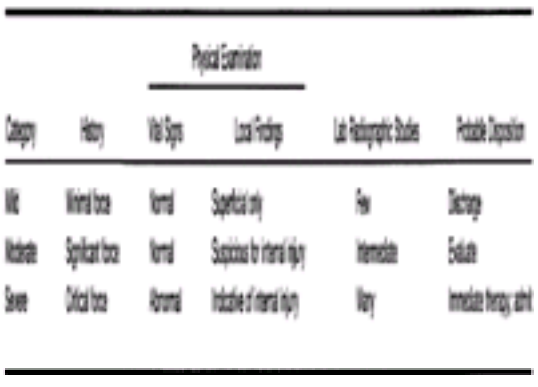

Table 103.2. Classification and Disposition of Trauma by Severity

GENERAL PRINCIPLES OF MANAGEMENT

The child who has sustained more than a trivial injury must be considered at risk of dying; thus, an immediate decision must be made regarding the severity of the trauma (Table 103.2). The clinical approach includes primary assessment, resuscitation (initial treatment), secondary assessment, and definitive care. This approach provides a set of principles for efficiently diagnosing and treating life-threatening conditions without neglecting less severe but important injuries. The primary assessment includes the vital signs and a quick review of the essential functions of all organs; the emphasis is on uncovering treatable injuries and preventing complications (e.g., paralysis from an unstable cervical spine fracture). Concomitantly, resuscitation (initial treatment) attempts to normalize vital functions and prevent further deterioration such as hypoxia or blood loss. Primary assessment and resuscitation occupy the first 5 to 10 minutes of the encounter in most cases. As soon as possible, reassessment of the entire patient (secondary assessment) should take place to fine-tune the details of management. The secondary assessment includes the use of radiographs and laboratory tests. To be effective, physical examinations must be repeated serially and then compared. Although resuscitation of unstable patients is critical and requires a strong team approach, the close surveillance of the apparently stable patient at risk of single or multiorgan trauma may be even more demanding. Definitive care includes stabilization of specific local injuries, preparation of the patient for the operating room, and surgery, as indicated. At the completion of care for trauma patients, a tertiary survey is performed. This is a last thorough check for occult injuries. It is done upon discharge, when a patient goes home after assessment, or in the light of day for admissions.

MULTIPLE TRAUMA

Classification

Multiple trauma (see Chapter 104), as defined for the emergency physician, may vary from mild to severe (Table 103.3). The child with a history of an injury caused by minimal force and a physical examination that shows only superficial lesions in two or more areas of the body would be assigned to the mild category—for example, a 7-year-old child who fell while running and is found to have an abrasion on the forehead and right elbow and tenderness of the right flank. The discovery on examination of signs suggestive of deeper injury places the child in at least the moderate category. Detection of a serious injury or abnormal vital signs (unrelated to anxiety alone) make for classification as severe. Unfortunately, classification of injury by mechanism alone is not a uniformly useful predictor in blunt trauma in children or adults.

		Physical Examination	
Category	History	Vital Signs	Local Findings
Mild	Minimal force	Normal	Abrasions/contusions
Moderate	Significant force	Normal	Refer to Tables 103.7 to 103.9 by anatomic region
Severe	Critical force	Abnormal	Refer to Tables 103.7 to 103.9 by anatomic region

Table 103.3. Severity of Multiple Trauma

Management

Mild Multiple Trauma

The major goal in the management of a child with apparent mild multiple trauma is to confirm the initial impression of lack of severity. If there is any question of more severe injury, a large-bore peripheral intravenous line should be inserted and blood studies obtained, including a complete blood count (CBC) and type and crossmatch. However, in cases that obviously seem to involve minimal trauma, the physician can proceed directly to the examination. Initially, the vital signs should be obtained. Subsequent examination includes a complete assessment with special attention to the level of consciousness; tenderness or limitation of motion of the cervical spine; auscultation of the heart and lungs; palpation of the abdomen, back, and pelvis; and extremities tenderness. The complete physical examination should include vital signs with capillary refill; Glasgow Coma Scale score; inspection and palpation of the head for injuries, pupillary reactions, extraocular muscle function, nasal tenderness, and dental/oral trauma; cervical spine motion (if the child is alert, not in a cervical collar, and without complaint and tenderness); neck vein distension; auscultation of the breath and heart sounds; inspection and palpation of the chest; evaluation of bowel sounds; inspection and palpation of the abdomen; palpation and inspection of the back, flank, and pelvis; rectal and genital examination; evaluation of extremities for deformity or tenderness; palpation of peripheral pulses; neurologic evaluation; and careful survey of the skin and soft tissues.

Laboratory evaluation of a child with a history of minimal multiple trauma and a normal examination may require no studies. If there is any concern, a CBC and urinalysis should be obtained. No other laboratory or radiographic studies are routine.

Moderate Multiple Trauma

The child with multiple trauma categorized as moderate requires immediate intervention, as well as a thorough diagnostic evaluation. A child in this category has an obvious history of involvement of several areas of the body, but initially may have only evidence of musculoskeletal or several superficial local injuries. A 3-year-old child who has been hit by an

automobile and has a significantly deformed femur and a few ecchymoses on the upper extremities, or an older child who fell off a second-story roof but appears well, may fit this group. As a first step, a minimum of one large-bore peripheral intravenous catheter should be placed. If the child is in respiratory distress, supplemental oxygen should be administered. Any suggestion of cervical spine injury mandates immobilization of the neck with a semirigid collar or sandbags. If the vital signs and primary survey are normal for age, the physician then should proceed with a thorough examination, as outlined in mild multiple trauma.

Most patients with moderate multiple trauma require ancillary studies in addition to a urinalysis. These might include a CBC and radiographs of the chest and cervical spine or abdomen. A type and screen for red blood cells is indicated. In fully awake patients, a completely normal examination may be relied upon to exclude the need for all screening studies. Many patients in this category require admission to the hospital. However, an older child with a history of a moderately severe impact, who has an unremarkable examination and normal studies, may be discharged from the ED after observation for several hours.

Severe Multiple Trauma

The management of the child with severe multiple trauma demands immediate action. The initial approach assumes either obvious life-threatening injury or a reasonable likelihood that such an injury exists. An alteration of vital signs (hypotension, tachycardia), diaphoresis, or depressed consciousness automatically categorizes the injury as severe. Although helpful as an initial guide, mechanism alone (e.g., a fall from a two-story building) is not a highly accurate predictor of risk. To adequately manage the child with severe multiple trauma, the physician must understand the need to institute treatment before completing a full examination and to continually intersperse detailed reassessments into an intensive treatment protocol. Table 103.4 provides an outline for organizing the initial approach to severe multiple trauma in the ED. It uses a four-pronged strategy: assessment, treatment, monitoring, and diagnostic testing. The protocol is laid out over time in an idealized fashion; obviously, limitations in the number of personnel or unusually difficult technical procedures may slow the progression.

Table 103.4. Management of Severe Multiple Trauma

LOCALIZED HEAD TRAUMA

Classification

Head trauma (see Chapter 38 and Chapter 105) can be divided into penetrating and nonpenetrating. Cases that involve penetration of the cranial vault entail severe injuries and often require operative intervention. Nonpenetrating head trauma can be classified as mild, moderate, or severe (Table 103.5).

		Physical Examination	
Category	History	Vital Signs	Local Findings
Mild	Minimal force No/momentary LOC	Normal	Glasgow =15 Abrasions/contusions
Moderate	Significant force LOC 1–5 min	Normal	Glasgow ≥13 Drowsiness
Severe	Critical force LOC >5 min	Abnormal	Glasgow ≤12 Focal neurologic abnormalities

LOC, loss of consciousness.

Table 103.5. Severity of Blunt Head Trauma

Management

Penetrating Trauma

Wounds limited to the scalp and not entering the cranial vault are appropriate for primary repair in the ED. Minor wounds

from sharp objects, when the likelihood of penetration is high, should have radiologic evaluation and local exploration before primary closure. All other penetrating injuries require initial stabilization and neurosurgical consultation, as discussed under severe blunt head trauma. Protruding objects should be left in place until definitive management.

Blunt Trauma—Mild

Most children seen in the ED have sustained mild head trauma and have at most momentary loss of consciousness (less than 1 minute), arrive awake, and primarily need a thorough physical examination. The head should be palpated for evidence of local injury, assessing for evidence of a depressed fracture. Bruises around the eyes or behind the ear or a hemotympanum suggest a basilar fracture. The pupils will be equal and reactive and the extraocular muscle function intact, unless the severity of injury has been misjudged. Ideally, the fundi should be visualized; however, this is not essential in most cases. Almost never will an alert child have any significant funduscopic pathology from trauma, nor will visualization of the fundi always be possible in uncooperative infants and young children. In general, papilledema is a late sign of increased intracranial pressure. Although the finding of focal neurologic abnormalities is unlikely, a careful neurologic examination is mandatory. Skull radiographs and computed tomography (CT) are generally unnecessary, but either may be indicated in selected situations, such as palpation of a potentially depressed fracture (see Chapter 38).

Patients with minor head trauma may be discharged from the ED with specific instructions to watch for changes indicative of increased intracranial pressure or hemorrhage. Albeit unlikely, these symptoms include depression of mental status, progressive vomiting (greater than 4 hours from the trauma), visual disturbances, ataxia, or seizures. Postconcussion seizures are unusual but may occur within a few days of mild head injury. In general, they are not prognostic for recurrent seizures.

Blunt Trauma—Moderate

The child with moderate head trauma has sustained a concussion or, perhaps, a cerebral contusion. Moderate head injury includes any clear-cut prolonged loss of consciousness (1 to 5 minutes) or a history suggesting a severe injury, even without specific physical findings to confirm it. Once again, a thorough examination is required to search for signs of intracranial hemorrhage. The most important feature of the examination is a serial evaluation such as the Glasgow Coma Scale. The initial score serves as the baseline for the detection of subsequent deterioration. Radiographs are reserved for the same indications as for mild trauma; CT scanning is often but not necessarily performed in awake patients upon arrival, but it becomes mandatory upon deterioration in mental status or in the presence of focal abnormalities. Because of the small chance of subsequent intracranial hemorrhage or worsening cerebral edema, prolonged observation in the ED or admission is warranted in many cases with moderate injury.

Blunt Trauma—Severe

The child with severe head trauma is at risk for sudden intracranial catastrophe, acute respiratory insufficiency, or a secondary insult (e.g., brain swelling) to the central nervous system. After initial steps to assess the adequacy of respiration and circulation, these functions should be supported as necessary. The cervical spine should be stabilized with a semirigid collar or sandbags. At times, gentle opening of the airway with maintenance of the head in the neutral position will allow adequate ventilation. If intubation is required immediately, extension of the neck should be avoided, and an assistant should stabilize the cervical spine during the procedure. In less urgent situations, intubation may be deferred until a cross-table lateral radiograph of the cervical spine is obtained. In all cases, supplemental oxygen should be administered and two intravenous cannulas inserted. Most children with serious head injury will hyperventilate spontaneously if their airway is patent and decrease their cerebral blood flow, which will help maintain normal intracranial pressure. Intubated, apneic children should be hyperventilated manually to achieve a $PaCO_2$ of 30 to 35 mm Hg. Ideally, continuous noninvasive end-tidal CO_2 monitoring with an arterial blood gas to maximize accuracy should be the goal in the ED. An arterial catheter can be placed as needed. If the patient has an isolated head injury, parenteral fluid administration should be no greater than two-thirds of the daily maintenance rate unless there is evidence of hypovolemia. Corticosteroids are not recommended by most authorities. Osmotic agents are not used prophylactically, but mannitol (0.5 to 1.0 g/kg of a 20% solution) occasionally is necessary to decrease intracranial pressure when acute herniation is suspected or proved.

Standard skull radiographs are time consuming and provide little useful information in the patient with serious head injury. More efficient management calls for an immediate CT to evaluate the intracranial space. Rarely, neurosurgical intervention must precede imaging of the cranial contents. See Chapter 105 for more specific management.

LOCALIZED NECK TRAUMA

Classification

Within the confined anatomic space of the neck pass the larynx and trachea, the carotid arteries, the jugular veins, the spinal cord, and the esophagus. Thus, both penetrating and nonpenetrating insults can cause devastating injuries. All penetrating trauma, with the exception of tangential wounds superficial to the platysma muscle, should be considered serious and be referred promptly for surgical evaluation and possible exploration. Weapons or objects protruding from the neck should be left in place. Children with neck trauma (see Chapter 106) should be carefully examined in the ED for thoracic injuries, such as pneumothorax.

Isolated blunt trauma to the neck does not occur often in children. However, the potential for major disruptions of the airway or large vessels demands a thorough evaluation. Particularly, the examiner should palpate for crepitus, unequal carotid pulses, expanding hematomas, and cervical spine tenderness. Based on the history and physical findings, an estimate of the severity of the injury can be made (Table 103.6). A thorough neurologic examination, with particular

emphasis on spinal cord disruption, is essential.

		Physical Examination	
Category	History	Vital Signs	Local Findings
Mild	Minimal force	Normal	Abrasions/contusions
Moderate	Significant force	Normal	Refusal to move head
			Cervical spine tenderness
Severe	Critical force	Abnormal	Crepitus
			Expanding hematoma
			Unequal carotid pulse
			Paralysis or sensory loss

Table 103.6. Severity of Blunt Neck Trauma

Management

Penetrating Trauma

Wounds clearly superficial to the platysma muscle are appropriate for repair in the ED. Children with penetrating injuries deep to the platysma require stabilization and subsequent surgical evaluation. Initial measures are directed at establishing a patent airway, providing adequate ventilation, tamponading hemorrhage, and restoring the circulation. Protruding objects should be left in place by the physician in the ED.

Blunt Trauma—Mild

If the history is one of a minimal force and no physical findings are indicative of trauma to the deeper structures, the child is symptomatically treated and discharged from the ED. Exceptions might be patients with underlying illnesses such as hemophilia, who are at risk for delayed complications. Follow-up after discharge should be defined clearly to ensure that intervention occurs before compromise to internal structures.

Blunt Trauma—Moderate

The child with an apparent moderate injury to the neck by definition has no evidence of respiratory or vascular compromise. However, either the history of the amount of force involved or the local findings may raise the possibility of cervical spine or other injuries. Such patients require immobilization of the cervical spine with a semirigid collar or sandbags and a meticulous neurologic examination. As a first step, a cross-table lateral radiograph of the cervical spine should be obtained with the child immobilized, usually in the ED. If this first radiograph shows all seven cervical vertebrae to be intact and properly aligned, a complete radiologic evaluation of the cervical spine can be performed, which may include anteroposterior, oblique, and open-mouth views. The discovery of a bony or ligamentous injury requires consultation with appropriate specialists. Spinal cord injury without radiographic abnormality may be present and should be pursued, when symptoms or signs are suggestive. The neurologically intact child with a normal cervical spine evaluation and no other neck trauma who remains well on repeat physical examination may be discharged after observation.

Moderate trauma to the anterior neck requires careful evaluation for possible disruption of the major vessels, trachea, and esophagus. Cervical spine radiographs may outline the airway adequately. However, it is important to pay attention to the alignment of the larynx and trachea and to check for air in the soft tissues from a tear in the airway or esophagus. The carotid triangle must be palpated carefully. If there is a hematoma or abnormality of the pulse, referral for possible arteriogram should be made.

Blunt Trauma—Severe

Classification of blunt neck trauma as severe indicates concern for overt injury to the airway, the major vessels, or the spinal cord. The initial goals of management are establishment of a patent airway, stabilization of the cervical spine, and intravenous access. The first choice for establishment of the airway is orotracheal intubation, with maintenance of the head in the neutral position. The inability to intubate a critical airway requires an immediate surgical approach to the trachea. Blood should be sent for a type and crossmatch; if vascular injury is suspected, multiple units of blood should be available. A surgical consultant should decide whether to proceed with an exploration in the operating room or to rely on further diagnostic studies such as bronchoscopy, arteriography, or esophagoscopy.

LOCALIZED THORACIC TRAUMA

Classification

Penetrating chest injuries (see Chapter 107) are extremely relevant to physicians in the ED because they may be rapidly life-threatening if untreated, yet usually respond to fairly straightforward therapeutic maneuvers. Any object that enters the thoracic cavity will result in significant injury. Patients with large open wounds or instability of vital signs should be considered to have sustained life-threatening trauma.

Blunt chest trauma is seen more often than penetrating injury in civilian practice in general and in children in particular. Although there are no studies on the percentage of chest injuries classified as mild in childhood, statistics are available for older patients. Newman et al. found that 53% of chest injuries sustained by adults in motor vehicle accidents were merely bruises or abrasions. As with trauma to other anatomic regions, blunt thoracic trauma can be divided into mild, moderate, and severe categories (Table 103.7).

		Physical Examination	
Category	History	Vital Signs	Local Findings
Mild	Minimal force	Normal	Abrasions/contusions
Moderate	Significant force	Tachypnea	Splinting
		Normal pulse and blood	Bony tenderness
			Decreased breath sounds
Severe	Critical force	Abnormal pulse or blood pressure	Flail chest
			Distant heart tones
			Absent breath sounds

Table 103.7. Severity of Blunt Chest Trauma

Management

Penetrating Trauma

Patients with mild injury, in which the wound clearly entered only the superficial tissues and not the thoracic cavity, may need only ED management. However, for any knife or gunshot injuries, it is advisable to have an experienced physician explore the wound and to obtain radiographs to determine the extent of injury.

Patients with deeper wounds require chest radiography, CBC, and type and crossmatch. An arteriogram may be necessary in the stable patient with a suspected aortic injury. Tube thoracostomy should be performed to drain a hemothorax or pneumothorax. Hemorrhage can be managed starting with crystalloid, followed by blood replacement. For the child sustaining a cardiopulmonary arrest in the ED after a penetrating thoracic injury, immediate resuscitative thoracotomy may be lifesaving.

Blunt Trauma—Mild

The child with a history of a minimal blow to the chest, normal vital signs, and no local signs of trauma other than abrasions or contusions has sustained a mild injury. The absence of bony tenderness, tachypnea, or decreased breath sounds obviates the need for radiologic evaluation of the thoracic cage or its contents.

Blunt Trauma—Moderate

If the history indicates significant force, bony tenderness is elicited, or there is a question of decreased breath sounds, a chest radiograph, electrocardiogram (ECG), and CBC are often helpful. Particularly in children, a pneumothorax may follow blunt injury with or without a rib fracture. Widening of the mediastinum on chest radiograph suggests disruption of the aorta. The detection of a solitary rib fracture is not important per se because no specific treatment is necessary. However, it raises the suspicion of visceral or vascular disruption. In particular, fracture of the first rib is correlated in adults with injuries to the great vessels. Although the data are scant in pediatrics, an injury to the first rib may require arteriography. The chest radiograph may provide a clue to the diagnosis of pericardial hemorrhage (by showing a slightly enlarged cardiac silhouette), but it is more often normal in this condition. Bleeding into the pericardial space leading to tamponade will invariably manifest on the physical examination at some point; findings include tachycardia, followed by hypotension, distended neck veins, and muffled heart tones. A pulmonary contusion or aspiration may produce a consolidation on chest radiograph. The ECG is obtained as an aid in the diagnosis of myocardial contusion; elevated ST segments are characteristic of this entity.

In the setting of moderate blunt chest injury, it is advisable to achieve venous access and order a type and crossmatch. Sophisticated diagnostic studies, such as CT scan and arteriography, are reserved for children with abnormal findings on the preliminary evaluation. Admission for observation is often warranted; however, the child who shows improvement on examination over the observation interval and does not have an abnormal chest radiograph, ECG, or CBC may often be discharged.

Blunt Trauma—Severe

The child with abnormal vital signs or local findings indicative of internal injuries has sustained an immediate life-threatening injury. Initial therapy includes airway management, the institution of two large-bore intravenous lines, and the administration of supplemental oxygen. Depending on the condition of the child, chest tube insertion may be necessary for treatment of hemopneumothorax before radiographic studies are obtained. In selected circumstances, resuscitative thoracotomy in the ED may be beneficial, although in blunt trauma to the chest the outcome is almost uniformly poor if there has been cardiopulmonary arrest. Admission to the hospital is mandatory, and a full diagnostic

evaluation should be performed to ascertain the need for surgery.

LOCALIZED ABDOMINAL TRAUMA

Classification

Penetrating abdominal injuries (see Chapter 108 and Chapter 109) often cause moderate to severe trauma. However, the physician may cautiously define a small category of mild injuries, depending on the weapon involved.

All gunshot wounds must be considered at least moderate because almost all penetrate the peritoneum. Of those that penetrate the peritoneum, most cause visceral injury. If the vital signs are abnormal after a gunshot, the trauma should be considered severe.

Stab wounds, on the other hand, may be superficial to the peritoneum. The patient with stable vital signs and an apparent superficial stab wound may be judged to have a mild injury if local exploration confirms the clinical impression. Stab wounds that violate the peritoneum should be considered moderately serious and the patient should be referred for surgical consultation. By definition, stab wounds that lead to unstable vital signs have produced severe trauma.

Overall, blunt abdominal trauma is much more common than penetrating injury in children. Most children evaluated in the ED for blunt trauma to the abdomen will have relative minor injuries.

Management

Penetrating Trauma

All gunshot wounds are of at least moderate severity. Thus, these children require two large-bore intravenous lines (preferably inserted above the diaphragm), a nasogastric tube; radiographs of the abdomen and chest; and laboratory studies, including CBC, urinalysis, amylase, AST, ALT, and type and crossmatch. In the child with unstable vital signs, appropriate resuscitation should be initiated and laparotomy urgently considered. Otherwise, an initial CT scan may be preferable to delineate the extent and location of internal injury. All patients will require hospitalization.

Stab wounds produce variable degrees of internal injury, and the approach to management differs among institutions. Patients with abnormal vital signs require stabilization in the ED, including appropriate resuscitative measures, intravenous access, a nasogastric tube, radiographs, and laboratory studies. Transfer to the operating room may be necessary on an urgent basis. Patients whose vital signs are stable are evaluated further by local exploration, lavage, or laparotomy.

Blunt Trauma—Mild

Mild blunt abdominal injuries often occur when there is contusion of the abdominal wall from local trauma (e.g., a fist, a fall). After a careful history and physical examination, including rectal palpation and testing of the stool for blood, usually only a urinalysis for red blood cells is required.

Blunt Trauma—Moderate

Moderate blunt abdominal trauma is often seen in patients with multiple injuries or those in whom there has been an isolated but forceful blow to the abdomen. These patients should have a CBC, amylase, aspartate transaminase (AST), alanine transaminase (ALT), radiographs, and type and crossmatch. At least one intravenous catheter should be inserted, preferably above the diaphragm. A CT scan is often warranted. A diagnostic ultrasound may be useful to reduce the need for immediate CT in some settings. In many cases, hospitalization or prolonged observation in the ED is indicated. Other imaging studies, such as intravenous pyelography, are obtained in selected situations (see Chapter 108 and Chapter 109).

Blunt Trauma—Severe

Severe blunt abdominal trauma usually warrants prompt surgery after an initial stabilization of the vital signs. The issue of whether to perform peritoneal lavage before exploratory laparotomy or whether to do a CT scan is discussed at length in Chapter 108. In general, patients who have stable vital signs after initial fluid resuscitation may be evaluated with CT scanning to define intra-abdominal bleeding. The scans help define the need for surgery, especially if a hepatic or splenic injury producing limited hemorrhage is identified. Peritoneal lavage is usually reserved for children with a decreased level of consciousness, requiring CT scanning of the brain, who experience hemodynamic instability or deterioration in the ED.

In the presence of significant hematuria or strong suggestion of renal injury, a CT scan should be performed (see Chapter 109). In symptomatic patients with severe blunt trauma, the absence of red blood cells in the urine is not by itself sufficient evidence of an intact genitourinary tract. Avulsion of the renal pedicle may occur without hematuria.

EXTREMITY TRAUMA

Classification

Most injuries to the extremities of children (see Chapter 115) seen in the ED are mild. A few injuries are of moderate extent, and occasionally, extremity trauma may be life- or limb-threatening. Both penetrating (Table 103.8) and nonpenetrating trauma (Table 103.9) run the gamut in terms of severity. With penetrating wounds, the major immediate

concern is hemorrhage, although impairment of neurovascular or musculoskeletal integrity with concomitant loss of long-term function is also a consideration. On the other hand, nonpenetrating trauma may cause vascular insufficiency without external bleeding.

		Physical Examination	
Category	History	Vital Signs	Local Findings
Mild	Minimal force	Normal	Laceration
Moderate	Significant force (e.g., stab)	Normal	Laceration of tendon or nerve Significant venous hemorrhage
Severe	Critical force (e.g., gunshot)	Abnormal	Partial/complete amputation of arm or leg Arterial hemorrhage Open fracture

Table 103.8. Severity of Penetrating Extremity Trauma

		Physical Examination	
Category	History	Vital Signs	Local Findings
Mild	Minimal force	Normal	Contusions/point tenderness
Moderate	Significant force Crush injury	Normal	Obvious dislocation of major joint Displaced fracture
Severe	Critical force	Abnormal	Decreased or absent pulses

Table 103.9. Severity of Blunt Extremity Trauma

Management

Penetrating Trauma

The child with mild penetrating trauma requires appropriate wound care and tetanus prophylaxis (see Appendix D) in the ED. A radiograph is indicated if a radiopaque foreign body (including glass) is suspected in the wound. Moderate injuries require careful physical examination and often local exploration to define the extent of the trauma and the degree of functional impairment. At times, prompt surgical consultation or follow-up with a specialist is indicated. Some injuries require repair in the operating room; however, others—even extensive lacerations or extensor tendon disruptions—may be handled, time permitting, in the ED by an experienced physician. Surgical referral is mandatory for children in the severe category, and it is important to proceed as rapidly as possible when vascular damage is suspected.

Blunt Trauma—Mild

Children with mild nonpenetrating injuries often require a radiograph to detect underlying fractures. Particularly when there is tenderness at the end of long bones, careful consideration should be given to radiologic evaluation for growth plate (epiphyseal) injuries, keeping in mind that a normal radiograph does not exclude a nondisplaced Salter-Harris type I fracture.

Blunt Trauma—Moderate

Obvious dislocations should be repositioned as expeditiously as possible, usually after radiographs confirm the diagnosis. Depending on the joint involved, discharge is acceptable after reduction and radiologic reevaluation (e.g., shoulder, patella, metacarpal, phalangeal, or interphalangeal joints). Crush-type injuries may initially manifest with pain and swelling. Of particular concern is the possibility that crush injury may lead to a compartment syndrome over the ensuing 6 to 24 hours.

Blunt Trauma—Severe

Extremity injury associated with hemodynamic disturbances or disruption of the vascular supply are severe. These include de-gloving and crush (i.e., wringer-type) injuries, as well as some long-bone fractures with high energy transfer. Patients with severe extremity injuries should receive a rapid but thorough overall assessment, followed by prompt surgical consultation.

SUMMARY

The approach to the injured child requires great care and clinical acumen to establish the extent of the trauma and

institute appropriate treatment. Loss of life from occult internal hemorrhage, or neurologic sequelae from a missed unstable cervical spine injury, are devastating. Yet, physicians in the ED must also know which children need only a careful physical examination and when laboratory testing or admission is unwarranted. This chapter has described a brief schema for providing appropriate care to children with trauma in such a way that specific issues about management can be approached reasonably by the emergency physician. The subsequent chapters in this section provide a wealth of additional detail for each anatomic area of the body.

Suggested Readings

Barlow B, Memirok M, Garrdler RP. Ten years' experience with pediatric gunshot wounds. J Pediatr Surg 1982;17:927.

Breaux CW, Smith G, Georgeson KE. The first two tears: experience at a pediatric trauma center. J Trauma 1990;30:37.

Chadwick DL, Chin S, Salerno C, et al. Death from falls in children: how far is fatal? J Trauma 1991;31:1353.

Connors JM, Ruddy RM, Martin J, et al. Delayed diagnosis in pediatric blunt trauma. Pediatr Emerg Care 1997; 13:298.

Division of Injury Control, Centers for Disease Control. Childhood injuries in the United States. Am J Dis Child 1990;144:627.

Friedland LR, Kulick RM. Emergency department analgesic use in pediatric trauma victims with fracture. Ann Emerg Med 1994;23(2):203–207.

Furnival RA, Woodward GA, Schunk JE. Delayed diagnosis of injury in pediatric trauma. Pediatrics 1996;98:56–62.

Greenspan L, McLellan BA, Greig H. Abbreviated injury scale and injury severity score: a scoring chart. J Trauma 1985;25:60.

Guy J, Haley K, Zuspan SJ. Use of intraosseous infusion in the pediatric trauma patient. J Pediatr Surg 1993;28:158–161.

Holmes MJ, Reyes HM. A critical review of urban pediatric trauma. J Trauma 1984;24:253.

Jubilerer RA, Nikheleshwer NA, Beyer FC, et al. Pediatric trauma triage: review of 1307 cases. J Trauma 1990;30:1544.

Keller MS, Sartorelli KH, Vane DW. Associated head injury should not prevent nonoperative management of spleen or liver injury in children. J Trauma 1996;41:471–475.

Lieu TA, Fleisher GR, Mahboubi S, et al. Hematuria and clinical findings as indications for intravenous pyelography in pediatric blunt renal trauma. Pediatrics 1988;82:216.

Lindsay JN, Rodarte A, Peterson BP, et al. Reduced staffing of a pediatric trauma center at night. Ann Emerg Med 1988;17:434.

Meller JL, Little AG, Shermeta DQ. Thoracic trauma in children. Pediatrics 1984;74:813.

Musemeche CA, Barthel M, Cosentino C, et al. Pediatric falls from heights. J Trauma 1991;31:1347.

Newman RS, Jones IS. A prospective study of 413 consecutive car occupants with chest injuries. J Trauma 1981;24:129.

O'Gorman M, Trabulsy P, Pilcher DB. Zero-time pre-hospital IV. J Trauma 1989;29:84.

Ordog G, Prakash A, Wassenberger J, et al. Pediatric gunshot wounds. J Trauma 1987;27:1272.

Pohlgeers T, Ruddy RM. Pediatric trauma. In: Reisdorff EJ, ed. Emerg Med Clin North Am 1995;13:267–289.

Ramenofsky ML, Moulton SL. The pediatric trauma center. Semin Pediatr Surg 1995;9:128–134.

Saladino R, Lund D, Fleisher G. The spectrum of liver and spleen injuries in children: failure of pediatric trauma score and clinical signs to predict isolated injuries. Ann Emerg Med 1991;20:636.

Snyder JL, Jain VN, Saltzman DA, et al. Blunt trauma in adults and children: a comparative analysis. J Trauma 1990;30:1239.

Tunell WB, Knost J, Nance FC. Penetrating abdominal injuries in children and adolescents. J Trauma 1975;15:720.

Valentine J, Blocker S, Chang JHT. Gunshot wounds in children. J Trauma 1984;24:952.

Wears RL, Winton CN. Load and go versus stay and play: analysis of pre-hospital IV fluid therapy by computer simulation. Ann Emerg Med 1990;19:163.

Wesson DE, Williams JI, Salmi R, et al. Evaluating a pediatric trauma program: effectiveness versus preventable death rate. J Trauma 1988;28:1226.

Widone MD, ed. Injury Prevention and Control for Children and Youth. Elk Grove, IL: American Academy of Pediatrics, 1997.

CHAPTER 104

Major Trauma

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Accidents continue to account for close to half of all deaths in children from the ages of 1 to 14 years, exceeding all other causes of childhood mortality and accounting for more than 20,000 deaths per year in children less than 19 years of age in the United States. More than half of these deaths are related to motor vehicle injury. Nearly 22 million children are injured each year in the United States, surpassing all major diseases in children and young adults. Two of three childhood accidents occur in males. The peak accidental age range is between 4 and 12 years, with the highest frequency at age 8 years. Accidental injury also accounts for approximately 30% of infant deaths. The societal impact of years of life lost from childhood mortal accidental injury is staggering.

The mortality rate for children hospitalized after an accident is reported to be low, but that is because 80% of all trauma deaths occur either at the accident scene or in the hospital emergency department (ED). The most common preventable cause of death in injured children is failure to secure the airway. As much as 18% of hospital trauma deaths are avoidable if a correct diagnosis is made and a treatment regimen is instituted. The most common single injury associated with death in injured children is head trauma, which alone or in association with other injuries is responsible for 80% of trauma deaths. Combined thoracoabdominal injuries produce an 82% mortality or condition of extremis upon arrival at the hospital ED. More than 50% of major injuries have associated injuries of the head, chest, and musculoskeletal system. Such multisystem injuries require the use of multiple medical disciplines, with varied diagnostic and treatment modalities, to achieve optimal care. These problems clearly place a large burden on the emergency physician to effect an improvement in outcome from childhood trauma.

A critical factor that influences outcome for the pediatric trauma patient is the recognition that a child's physiologic needs are not the same as those of a small adult. Although the child will usually mount an appropriate physiologic and endocrinologic response to stress, the greater surface area relative to body size results in greater susceptibility to body heat loss and insensible fluid loss. Water, minerals, trace elements, fat, and vitamins are all needed in greater maintenance portions in the child. Critically, the growing child has a significantly higher energy-calorie requirement than that of an adult. Equally important to proper tissue repair are greater total protein and essential amino acid nitrogen requirements, which are age dependent. Whether reversal of the malnourished state will influence morbidity and mortality in childhood trauma is speculative, but if data examining this question for adults are applicable to the child, attention to the patient's nutritional status is certainly important. Despite these concerns, children have amazing recuperative powers and are prime candidates for subsequent medical rehabilitation.

CAUSES OF MAJOR TRAUMA

Motor vehicle crashes account for as many as half of all childhood trauma deaths. Motor vehicle occupant death rates begin to climb steeply at age 13 years and peak at 18 years. Falls from heights and falls against fixed objects account for 25 to 30% of deaths, drownings 10 to 15%, and burns 5 to 10%.

Societal violence as a cause of death in children is increasing at an alarming rate. An estimated 1077 child maltreatment fatalities occurred in the 50 states and the District of Columbia in 1996. Homicide rates in children have two peaks: from age 0 to 3 years and from 14 to 18 years. Death by homicide afflicts African-American citizens most severely. Presently, 1 of every 28 African-American males born today will die as a result of homicide.

In infancy, the most common causes of accidental death are aspiration, suffocation, and motor vehicle crashes. In a 10-year period, more than 50,000 accidental deaths will be reported in children under age 5 years. Annually, there will be more than 70,000 injuries as a result of automobile accidents in the same younger population. Accident mortality statistics show that the youngest occupant in an automobile will be the most vulnerable to injury in the event of a crash.

Children from 5 to 9 years of age are most likely to be pedestrian injury victims, and in this age group, such injuries are

the most common cause of motor vehicle fatalities. Overall, pediatric pedestrian injury accounts for 46% of motor vehicle fatalities. Boys in densely populated urban areas represent the largest group at risk. Injury from bicycle crashes is particularly common in children 6 to 16 years. There are 700 bicycle crash deaths in this age group alone, principally from head injury.

Because the United States is a nation in which vehicles are driven on the right side of the road, injuries to the left side of the pedestrian are the most common. The resulting frequency of injured organs includes, in order, the spleen, genitourinary tract, gastrointestinal (GI) tract, liver, pancreas, pelvis, and major vessels.

The predominant mechanism of major injury in children is blunt trauma, with only 10 to 20% penetrating wounds. This is strikingly different from adult trauma patients, for whom as many as 50% will have a penetrating injury. More than half of the deaths from such blunt abdominal trauma have an associated major liver injury.

ORGANIZATION OF THE TRAUMA SERVICE

The regionalization of trauma care in the United States is still in its evolutionary stage. Hospitals have been designated as Level I, II, or III institutions based both on their capability and on their desire to care for the multiply injured patient. Under the guidance of the American College of Surgeons' Committee on Trauma, the eventual benefit of such a designation plan is to triage Category I–, Category II–, or Category III–injured patients to appropriate, qualified facilities. Because no one hospital can be expected to have in its immediate geographic area the childhood trauma volume necessary to be an independent and freestanding trauma center, regionalization becomes a necessity, using rapid transportation to transport children over distances.

Each institution must develop its own in-house organizational tree for a trauma service. Such a service needs a well-established chain of command with an appropriately designated leader, a responsibility that may change hands as additional personnel arrive at a resuscitation in the ED. The role of this leadership position in a hospital trauma service is to accept responsibility for patient care and organize the multisystem specialists needed to care for the patient with multisystem injury. Such organization begins at the scene of an accident, includes transport, and involves patient triage after initial evaluation and care once the patient arrives in the hospital ED. Subsequently, a decision to transfer the patient to another hospital, admit him or her to an intensive care unit, or take him or her straight to the operating room will be made by the team leader after consultation with the other varied specialists involved. If at this time it is clear that the predominant injury is to a single body system, it is appropriate for the team leader to transfer patient care responsibility to the designated head of a given subspecialty. Figure 104.1 demonstrates a flow diagram of a response to the traumatized child, an example of organizational schema put into action when or before a victim of serious trauma arrives in the ED.

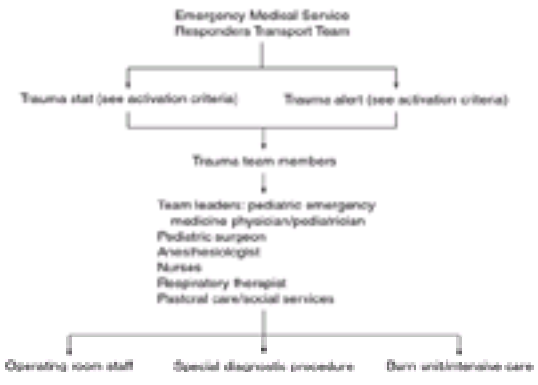

FIGURE 104.1. Sample flow diagram of a response to a traumatized child that is placed into action when or before a victim of serious trauma arrives in the emergency department.

ASSESSMENT AND MANAGEMENT

Initial Evaluation

Guidelines for the street-side evaluation and treatment of accident victims are well documented by the American College of Surgeons. Obviously, a list of priorities needs to be established for the logical approach to the trauma victim—priorities that are no different from those for the severely ill ED or hospitalized patient. It is also imperative that during injury assessment a simultaneous patient management protocol be initiated in a logical sequence. After prehospital evaluation and care of the injured child, the victim is transported to the hospital. A rapid and reproducible schema of immediate, simultaneous, and subsequent evaluation and treatment principles should be applied to every child with major multiple trauma admitted to the ED (Table 104.1). This initial assessment includes a primary survey, resuscitation, a secondary survey, and eventual triage. Two key principles must be followed in the initial assessment of the trauma patient. First, if any physiologic threat to the patient is identified, that threat must be treated immediately. The order of priority is airway, breathing, and circulation (ABCs). For example, relief of a tension pneumothorax takes precedence over intravenous (IV) access. Second, if at any point in the patient's secondary survey or subsequent care there is an unexpected physiologic deterioration, rapidly repeat the primary survey in order of priority (A, B, C). A “trauma stat,” alerting appropriate personnel, should be called upon notification of an impending arrival or simultaneously with an unexpected arrival of a child with multiple injuries in the ED, and all participants of the trauma care team should participate in this initial evaluation and treatment of the patient (Fig. 104.1). The indication for declaring a trauma stat is any one of the criteria listed in Table 104.2. The patient should be admitted to a generously sized area of the ED and placed on a multipurpose stretcher to be undressed and examined. It is important that the same examiners serially assess the patient.

I. Primary survey	C. Intubations—urinary tract, gastrointestinal tract
A. Airway maintenance, cervical spine control	III. Secondary survey
B. Breathing	A. Head
C. Circulation	B. Neck
D. Disability	C. Chest
E. Exposure	D. Abdomen
II. Resuscitation	E. Extremities
A. Oxygenation, airway management, and ventilation	F. Neurologic
B. Shock management	IV. Triage

Table 104.1. Initial Assessment and Management Guidelines for the Injured Child

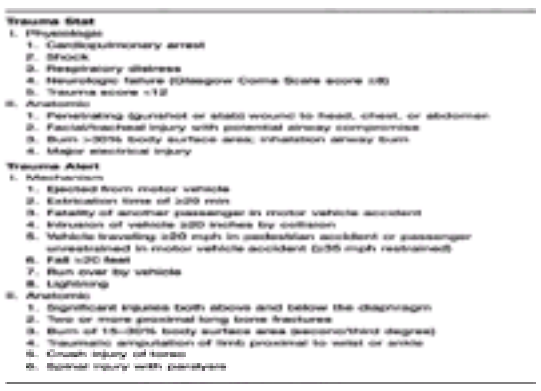

Table 104.2. Declaration of Trauma Stat and Alert

Primary Survey

The first priority in assessment and management is to secure an adequate airway while concomitantly stabilizing the neck to protect the cervical spinal cord from a yet-to-be-diagnosed cervical spine injury. The chin-lift or jaw-thrust maneuver and cleaning the oropharynx of accumulated foreign debris and secretions are the initial steps in establishing an airway. A cervical spine injury should be assumed to be present in all patients with major trauma, especially those injured above the clavicle. Adequate cervical spine radiographs and a normal clinical examination are required to exclude this problem. Therefore, until such films are taken, the head and neck should be neither hyperextended nor hyperflexed during maneuvers to secure the airway. Before any effort at intubation, artificial ventilation may need to be established using a bag-valve-mask device (see Chapter 1). An experienced physician should be able to exclude cervical spine injury clinically only in cases of minor trauma, when a patient is old enough to communicate, or in the absence of a distracting injury.

Breathing is deemed acceptable only in the face of a patent airway and adequate air exchange with normal oxygen saturation and carbon dioxide excretion. Early monitoring with pulse oxymetry is important. Compromise of ventilatory function in an injured child can occur with a depressed sensorium, airway occlusion, restriction of lung expansion, and direct pulmonary injury (see Chapter 95). Compromise of diaphragmatic excursion is a special hazard because of the increased importance of the diaphragm in ventilation in children. Gastric distension, a common event in an injured child, can significantly limit diaphragmatic excursion. In these situations, cricoid pressure should be performed while preparations are made for an artificial airway. This maneuver prevents possible aspiration of gastric content caused by passive regurgitation and/or increased intragastric pressure. Therefore, the early use of a nasogastric or an orogastric tube to decompress the stomach may need to be considered. If the child is obtunded or comatose, ventilation may require use of a bag-valve device that is connected to a mask or endotracheal tube delivering sufficient oxygen-enriched air (approximately 12 to 15 mL/kg) to produce an appropriate rise in the chest and an adequate oxygen saturation. Prompt attention to a hemopneumothorax, especially with mediastinal shift secondary to tension, is essential to the assessment of breathing (see the following section, Secondary Survey).

Circulation is initially assessed by examining the pulse, skin color, and capillary refilling time; from this information the peripheral perfusion and oxygenation may be estimated. A palpable peripheral pulse will generally correlate with a pressure greater than 80 mm Hg; a palpable central pulse indicates a pressure greater than 50 to 60 mm Hg, and a normovolemic patient capillary refilling time, as assessed by color return after blanching, will be within 2 seconds. External hemorrhage should be controlled by direct pressure or pneumatic splints, but the application of extremity tourniquets or hemostats to bleeding vessels is less useful. A mild Trendelenburg position may be of benefit in mild low-perfusion states to restore the central circulation.

To assess patient disability, a rapid neurologic examination is completed to establish the level of consciousness, as well as pupillary size and reaction. Table 104.3 lists the AVPU (alert, verbal stimuli response, painful stimuli response, unresponsive) method of assessing level of consciousness, in addition to pupillary assessment as previously mentioned. The Glasgow Coma Scale (Table 104.4) provides a quantitative measure of the level of consciousness.

A—Alert	P—Responds to painful stimuli
V—Responds to vocal stimuli	U—Unresponsive

Table 104.3. AVPU Method for Assessing Level of Consciousness

	Glasgow Coma Scale	Modified Infant Coma Score	
Eye Opening	Spontaneous	Spontaneous	4
	To voice	To voice	3
	To pain	To pain	2
	None	None	1
Verbal Response	Oriented	Coos, babbles	5
	Confused	Irritable cry, consolable	4
	Inappropriate	Cries to pain	3
	Garbled	Moans to pain	2
	None	None	1
Motor Response	Obeys commands	Normal movements	6
	Localizes pain	Withdraws to touch	5
	Withdraws to pain	Withdraws to pain	4
	Flexion	Flexion	3
	Extension	Extension	2
	Flaccid	Flaccid	1

Table 104.4. Pediatric Coma

To facilitate both assessment and treatment, the patient should be undressed and exposed, while careful attention is paid to the maintenance of body heat. Radiant warmers, air shields, and IV fluid warmers are useful tools in maintaining adequate temperature control in the pediatric patient.

Resuscitation

All patients with major trauma should receive supplemental oxygen therapy. Vascular access is another early necessity in resuscitation, and percutaneous or cutdown cannulation of upper or lower extremity veins should be of first priority, establishing two IV lines to facilitate resuscitation. Although large-bore cannulas are ideal, the size of the available veins should guide the choice of which cannulas to use. In a hypotensive child, the visible veins may be small. Successful placement of a 22- or 20-gauge cannula is preferable to a failed attempt to place a larger cannula. In a small child, one can give large volumes of fluids and blood through a small cannula. Improvement in vascular volume may then permit placement of a larger cannula at an alternative site. Early resuscitation also may be begun by intraosseous infusion into the tibial marrow space by a transtibial needle. We prefer percutaneous cannulation of an antecubital vein plus a saphenous vein at the ankle. In a hypotensive child in whom peripheral access is quickly found to be unsuccessful, the femoral vein provides a safe site for insertion of a central line, often accomplished rapidly using a guidewire technique. Rapid cutdown access is best done on an antecubital vein or the saphenous vein at either the ankle or in the groin just below the saphenofemoral junction.

At cannulation, blood should be sent for a type and crossmatch and for baseline hematologic and chemical parameters (Table 104.5). Central vein cannulation above the diaphragm is not a preferred primary access route, and in children, such access should be done only by experienced personnel.

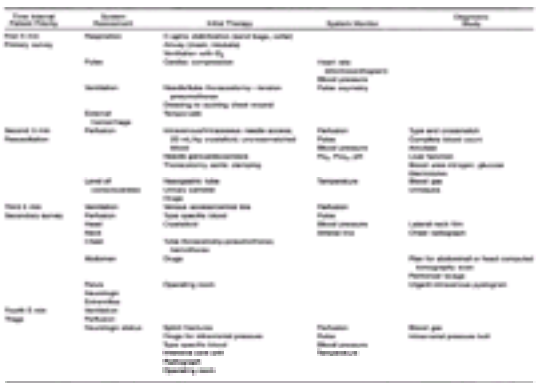

Table 104.5. Rapid Approach to the Pediatric Trauma Patient

The presence of shock must be assessed by appreciating whether or not inadequate organ perfusion exists. Shock after trauma is usually hypovolemic but may be cardiogenic or neurogenic. As a rule, isolated head trauma is not a cause of

shock. Any injured patient who is cool and tachycardic is in shock until proved otherwise.

Hypovolemic shock is most common after major trauma and is usually secondary to a significant acute loss of the 8 to 9% of body weight made up by the child's blood volume. To quantify the extent of the problem and decide on treatment priorities, hemorrhagic shock can be classified according to severity (Table 104.6). Reliance on the hematocrit alone may prove unreliable because a near-normal hematocrit level does not exclude the possibility of significant blood loss. Class I hemorrhage occurs with up to 15% acute blood volume loss (approximately a 250-mL blood loss in a 20-kg child), and physiologic changes will be minimal. Primary fluid replacement will stabilize the circulation. Class II hemorrhage, 15 to 30% blood loss (approximately 250- to 500-mL blood loss in a 20-kg child), is associated with a mild tachycardia and tachypnea along with a fall in pulse pressure as catecholamine release produces elevation of peripheral vascular resistance. Such patients may have impaired capillary refilling, and they may be either frightened or belligerent. These patients initially may be stabilized with crystalloid infusion, although blood transfusion eventually may be necessary. Class III hemorrhage is physiologically more significant, the 30 to 40% blood loss corresponding to 500 to 650 mL of blood in a 20-kg child. These patients have obvious signs of shock with altered mental status, tachycardia, tachypnea, and a measurable diminution in systolic pressure. Crystalloid resuscitation should be begun promptly, and most patients also will require blood products. Class IV hemorrhagic shock is immediately life-threatening. Patients are mentally depressed, cold, and pale; they have profound tachycardia and tachypnea, the pulse pressure is narrow, and there is no urine output. After rapid transfusion, such patients usually require prompt operative intervention.

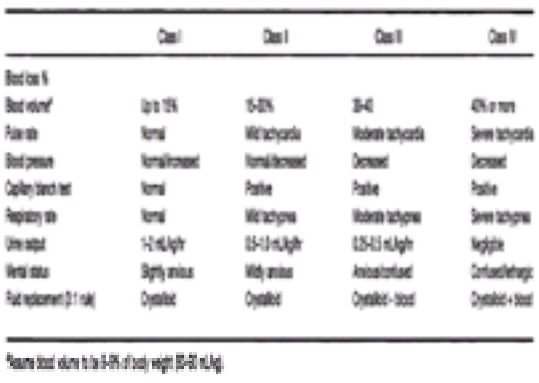

Table 104.6. Therapeutic Classification of Hemorrhagic Shock in the Pediatric Patient

Cardiogenic shock after major childhood injury is rare, but it could be a result of cardiac tamponade or a direct cardiac contusion. Dilated neck veins in a patient with a decelerating injury, sternal contusion, or a penetrating thoracic injury should arouse such suspicion (see Chapter 82 and Chapter 107). Neurogenic shock classically presents with hypotension without tachycardia or vasoconstriction; however, isolated head injuries do not produce shock, and causes of hypovolemia should always be sought in such patients. Septic shock rarely occurs immediately after injury, even in the face of abdominal content contamination.

Crystalloid isotonic solution, preferably Ringer's lactate or normal saline solution, is the initial resuscitative fluid of choice. The initial infusion is given as rapidly as possible in a dose of 20 mL/kg with careful monitoring of patient physiologic response to the fluid. Table 104.6 emphasizes the anticipated fluid needs, depending on the degree of shock, and the formulation is based on the premise that the patient will require 300 mL of crystalloid for each 100 mL of blood loss—the 3:1 rule. The restoration of perfusion may be clinically assessed, and at times invasive monitoring with elective placement of a central venous line or a pulmonary artery catheter facilitates this assessment. A most useful practical guide is the monitoring of urinary output; 1 mL/kg per hour is optimum, although for children less than 1 year of age, preferred output should approach 2 mL/kg per hour.

Blood transfusion preferably is done with fully crossmatched warmed blood passed through a 160-µm macropore filter. In the face of a transient or absent benefit from a rapid crystalloid infusion, fully crossmatched, type-specific, or type O negative blood should be given as a whole-blood transfusion. In summary, fluid and blood are given rapidly enough to maintain stable vital signs and adequate urine output. Vasopressors, steroids, and sodium bicarbonate do not play a role in the initial treatment of hypovolemic shock.

After perfusion has been restored, if not already accomplished, placement of urinary and gastric catheters should be done. Urinary catheterization should not be attempted before a retrograde urethrogram has proved urethral integrity if blood has been noted at the urethral meatus or in the scrotum, or if there is abnormal prostate placement on rectal examination. The urinary specimen should be immediately analyzed. In the presence of blunt head trauma with blood coming from the ears, nose, or mouth, care must be taken in inserting a nasogastric tube to avoid passage into the brain through a cribriform plate fracture. Such patients are better intubated through the mouth or through a soft nasopharyngeal airway.

Secondary Survey

A systematic assessment of organ systems must be done after securing the airway, breathing, and circulation. A pertinent history includes allergies, medications, past illnesses, time of the last meal, and the events preceding the injury.

A head examination includes reevaluation of pupillary size and reactivity, a conjunctival and fundic examination for hemorrhage or penetrating injury, and a quick assessment of visual acuity. A thorough palpation of skull and mandible will detect fracture-dislocations, but if the airway is secure, maxillofacial bony trauma is of low priority in the total

treatment plan.

Injury to the cervical spine is uncommon in children (see Chapter 106), but risk of injury must still be considered. This is especially true for any child with injury above the clavicles. It is also true for young children who fall one or more floors, are hit by a motor vehicle at 30 mph or more, and who are unrestrained or poorly restrained occupants of a motor vehicle involved in a crash. In older children, sports injuries are the second most common cause of cervical spine injury. In a child with a low risk for cervical spine injury (e.g., a fall when running), the neck can be cleared with a normal lateral cervical spine film, showing all seven cervical spine vertebrae. Also, a normal clinical examination in an awake, cooperative child (active but controlled flexion and extension and rotatory motion with no symptoms or signs of spasm, guarding pain, or tenderness) is the most important part of the assessment in ruling out neck injury. Ligamentous disruption and dislocation injuries of the cervical spine without radiographic evidence of bony injury are not uncommon in children because of the weakness of the soft tissue of the neck and the incomplete development of the bony spine. In approximately 40% of children with spinal cord injury, injury occurs without radiologic abnormality (SCIWORA). In children with a high-risk mechanism of trauma, it is best to obtain an anteroposterior, an odontoid, and lateral views. If the patient has an altered sensorium, a cervical collar should be left in place even if the three survey films are negative. When the patient recovers sufficiently to permit a fuller evaluation of the neck, the collar can be removed.

Three situations require a special approach. First, if a seriously injured child has had an endotracheal intubation placed, one can get a computed tomographic (CT) scan of C1–C2 when getting a head CT scan. Second, if the patient is brought to the hospital with a helmet on (e.g., football or motorcycle) and if there is no respiratory distress or other problem requiring immediate intubation, an initial cervical spine series can be done before helmet removal. If it is necessary to remove the helmet before the neck is cleared, a two-person technique ensuring neck immobilization should be used. Third, in the case of penetrating injuries to the neck, wounds deep to the platysma usually warrant operative exploration. In the case of missile injuries, the entry site should be denoted with an opaque marker, and anteroposterior and lateral spine films should be obtained.

Visual inspection of the chest will identify a sucking chest wound, best treated by immediate application of a sterile occlusive dressing; a major flail component, treated by splinting or endotracheal intubation; or a penetrating wound (see Chapter 107). Palpation of all thoracic bony parts must be done quickly, and auscultation may reveal a pneumothorax, hemothorax, or cardiac tamponade. The former two should be treated by tube thoracostomy. The latter may best be detected by muffled heart sounds, distended neck veins, and a narrow pulse pressure, and should be relieved by prompt pericardiocentesis (see Procedures, Section VII). A diagnosis of tension pneumothorax is supported by observing a contralateral tracheal shift and distended neck veins in addition to diminished breath sounds. Needle thoracentesis should provide relief, but tube thoracostomy should follow, with placement of the tube to water seal drainage plus suction. If an impaled object is protruding from the chest or any part of a patient, it is best debrided from surrounding clothing and left in place until definitive operation. If the history suggests a severe deceleration injury and the chest radiograph demonstrates a widened mediastinum with or without a fractured first rib, a thoracic aortic injury is suggested, and aortography is promptly indicated. If the chest radiograph reveals air lucencies suggesting intestine, a ruptured diaphragm is a possibility.

The purpose of the secondary abdominal examination is to establish whether an injury exists (see Chapter 104 and Chapter 108); it is not to give an exact diagnosis. Injury should be suspected in the presence of abdominal wall contusion, distension, abdominal or shoulder pain, signs of parietal peritoneal irritation, and/or shock. Hence, in most patients a baseline imaging study such as an abdominal CT scan should be done as soon as possible. Diagnostic peritoneal lavage (DPL) should be considered in the ED to expedite the decision about whether a patient needs an immediate laparotomy. In children, this is primarily of benefit in a patient who is unstable despite appropriate resuscitation; who has penetrating trauma that does not clearly involve the abdomen (e.g., gunshot wound of the lower chest); or who requires urgent, nonabdominal operative intervention (e.g., decompression of an epidural hematoma). In fact, most children with documented intra-abdominal or retroperitoneal injury who remain stable or rapidly become so do not require surgery. A positive DPL with greater than 100,000 red blood cells (RBCs) per cubic millimeter may correlate with a laceration of the spleen or liver but is not necessarily an indication for surgery in a stable patient. Of such patients, 80% will stop bleeding without operative intervention. However, diagnosis of injury is important in planning for care. Table 104.7 summarizes the indications and contraindications for this procedure. After emptying the urinary bladder, a midline approach above or below the umbilicus is used (see Procedures, Section VII). If the initial aspirate is grossly bloody or if the aspirate after instilling 10 mL/kg of Ringer's lactate reveals greater than 100,000 RBCs/mm ^{3}; greater than 500 white blood cells (WBCs) per cubic millimeter; a spun effluent hematocrit greater than 2%; or the presence of bile, bacteria, or fecal material, the paracentesis is positive. Lavage is most commonly falsely positive in the face of a pelvic fracture, or it may be falsely negative despite injuries to pancreas, duodenum, genitourinary tract, aorta, vena cava, and diaphragm.

Indications
Potential false-negative examination—head injury, medication overlay, spinal cord trauma
Potential false-positive examination—rib, pelvic, or spine fractures
Unexplained hypovolemia
Anesthetic need for unrelated injury
Selected penetrating injury
Absolute Contraindications
Multiple previous operations by history
Already determined need for laparotomy
Relative Contraindications
Pregnancy

Table 104.7. Peritoneal Lavage in the Evaluation of Abdominal Trauma in Childhood

A rectal examination is essential, assessing sphincter tone, rectal integrity, prostatic position, pelvic fracture, and presence of blood in the stool.

A thorough extremity examination should assess deformity, contusions, abrasions, penetration, and perfusion, including pulse palpation. The presence of a distal extremity pulse does not exclude a concomitant proximal arterial injury. Soft-tissue injuries should be thoroughly inspected for both wound foreign bodies and the presence of devitalized tissue. Long bones should be palpated with rotational or three-point pressure for tenderness, crepitation, or abnormal movement, and pressure must be applied to the pubis and anterior iliac spines to assess for the presence of a pelvic fracture. Severe extremity angulations should be straightened, joints should be immobilized, and traction splints should be applied. Compound fracture sites should be covered with sterile dressings. Generous irrigation and debridement of open wounds is beneficial in early wound care to minimize contamination before considering primary or delayed wound closure (see Chapter 123).

Hypothermia is a special risk in injured children who have relatively more surface area than an adult. Hypothermia can develop in the prehospital setting but can further occur in the ED, where proper assessment and treatment may require full exposure of the patient. The dangers of hypothermia are impaired circulatory dynamics, impaired coagulation, increased peripheral vascular resistance, and increased metabolic demand. The use of overhead radiant warmers, warm blankets, and warmed IV solutions are important measures in combating the deleterious effects of hypothermia. These measures should be used as soon after arrival in the ED as possible.

The burned child needs to have an initial appraisal of burn severity including the depth, location, and type of burn (Table 104.8); an appraisal of the extent of the burned area, using the percentage of surface area for children up to or older than 10 years as defined in Table 104.9; and a determination of whether the injury includes pulmonary, soft tissue, or bony damage as well. Burns may be classified in the following categories: 1) critical, in which there is a respiratory tract injury; a partial thickness burn exceeding 20% of body surface area (BSA); a full thickness burn exceeding 10% of the BSA; the hands, face, feet, or genitalia are included; there is a complicating fracture or soft tissue injury; electrical burns are part of the mechanism; or deep acid burns are present; 2) moderate, in which partial thickness is less than 20% and full thickness is less than 2% of BSA and does not include the hands, feet, or genitalia; and 3) minor, in which partial thickness is less than 15% and full thickness is less than 2% of BSA. The burn victim should be undressed, and sterile covers should be placed over the burn wounds. IV fluid resuscitation is necessary immediately if the burn exceeds 20% of BSA (see Chapter 1 and Chapter 114). Referral to a burn center should be considered in critical burns or cases with the possibility of devastating cosmetic and functional deficits.

Anatomic Area	Percentage Adult Surface (Age >10 yr)	Percentage Infant Surface
Head	9	16
Right upper extremity	9	9
Left upper extremity	9	9
Right lower extremity	18	13
Left lower extremity	18	13
Anterior trunk	18	18
Posterior trunk	18	18
Neck	1	4
Total	100	100

Table 104.8. Burn Wound Assessment: Body Surface Estimation (Adult versus Child)

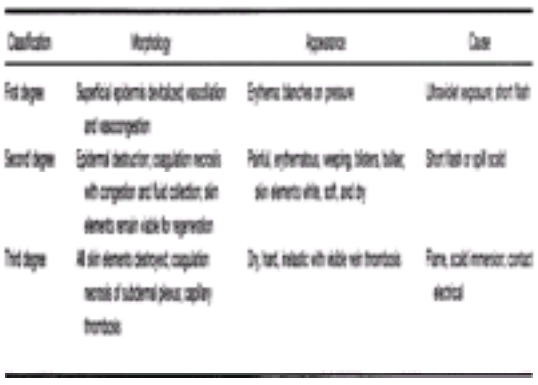

Table 104.9. Burn Injury

Neurologic assessment includes a reevaluation of the level of consciousness, a repeat pupillary examination, and a thorough sensorimotor examination. Not only is serial reassessment critical, but also quantification of the findings using a Glasgow Coma Scale is of benefit to detect early changes (see Chapter 105; Table 104.4). Any evidence of paralysis or paresis suggests a major neurologic injury. Until spinal cord injury is determined or ruled out in any patient with signs of central nervous system injury, maintain the patient in a semirigid cervical collar and immobilize him or her on a long spinal board.

Supplemental studies (including regular and contrast imaging studies; biochemical analyses of liver, pancreatic, and

renal function; and electrocardiographic analysis of cardiac function) may now be done. Tetanus prophylaxis should be considered (see Chapter 103), and antibiotics should be administered if specifically indicated.

Imaging the Pediatric Trauma Patient

In any child with major trauma caused by a blunt mechanism, a basic radiographic survey series should be considered. Traditionally, these included cervical spine, chest, and pelvic radiographs. Recent studies have demonstrated that patients with a Glasgow Coma Scale score of 15, no distracting injury, and no pain in the pelvic region have a low incidence for pelvic fractures. In these patients, the routine use of pelvic radiographs is not necessary but also rarely helpful in the acute initial management of the traumatized child. In a stable, cooperative patient, the clinician should review the lateral cervical spine film first and then proceed with the other films. In a less stable or neurologically compromised patient, it is best to obtain all films at once while continuing to protect the cervical spine. In patients with trauma to the torso, a pelvic film is important as a clinical indicator; 80% of children with multiple fractures of the pelvis have concomitant abdominal or genitourinary injuries. Additional survey films of the thoracolumbar spine and extremities depend on clinical findings and the mechanism of trauma.

The primary and secondary survey may then suggest the need for more definitive imaging studies. For example, any patient who is normotensive but has an abnormal Glasgow Coma Scale score, especially of 13 or less, should undergo a CT scan of the head. Other indications include a history of posttraumatic seizures, prolonged lethargy or loss of consciousness, or an underlying medical risk factor such as hemophilia. A CT scan of the abdomen is indicated in a hemodynamically stable victim of blunt trauma who has clinical signs of intra-abdominal injury, transaminase elevation of greater than 100 U/L, hematuria greater than 20 RBCs per high-powered field, or a worrisome mechanism of trauma in the presence of neurologic compromise. Most CT scans of the abdomen for trauma should be performed with single contrast IV. Double contrast (GI + IV) may delay the process and increase the risk for aspiration. If a Foley catheter is in place, it should be clamped during the abdominal CT scan to provide information about the bladder. Because of the potential for injury to the liver or spleen in children with blunt abdominal trauma, an abdominal CT scan is more comprehensive in evaluating a patient with significant hematuria.

The likelihood of positive findings in abdominal CT scans is significantly increased if three or more of the following indicators are present: 1) gross hematuria, 2) lap belt injury, 3) assault or abuse as a mechanism of trauma, 4) abdominal tenderness, and 5) trauma score less than or equal to 12. However, certain indicators alone, such as positive abdominal findings, worrisome mechanism of trauma (e.g., ejected from a motor vehicle), and neurologic compromise (e.g., Glasgow Coma Scale score of less than 10), warrant obtaining a double-contrast abdominal CT scan to document possible intra-abdominal or retroperitoneal injury. In general, the accuracy of CT scans in diagnosing intraperitoneal or retroperitoneal injuries is 95% or better. The use of newer modalities of scanning, such as ultrafast CT scans, will increase the diagnostic accuracy of CT scans because of less motion artifact and better contrast enhancement. Ultrafast scans will also enhance patient safety: less sedation required, lower radiation dosage, and improved accessibility of the patient. When emergency CT scans of the abdomen are not available, ultrasonography of the abdomen is a useful alternative. In the hands of an experienced examiner, it can be performed in the ED and will document most injuries to the liver, spleen, and/or kidney, as well as demonstrate intraperitoneal fluid.

The limitations of CT scans of the abdomen in trauma patients are in diagnosing injuries to hollow viscus organs, such as perforation of the bowel and bladder. One other striking limitation in the use of abdominal CT scans is in motor vehicle crash occupants who have clinical evidence of lap belt injuries to the abdomen and/or spine; transverse ecchymoses of the abdominal wall, abdominal pain, and/or tenderness; and symptoms or signs of lumbar spine injury with or without spinal cord injury. CT scans in such patients will miss certain injuries: free intraperitoneal air caused by bowel perforation in 75% of cases, and lumbar spine injuries in 77% of cases. In hemodynamically stable patients in this group, the workup should include complete thoracolumbar spine films, especially the lateral view, an abdominal film designed to show the presence of free air (e.g., a left lateral decubitus film), a cystogram, and a DPL. If injury to the bowel or bladder is confirmed, a laparotomy is indicated. If these studies are negative and if there is evidence suggesting possible retroperitoneal injury, a double-contrast CT scan of the abdomen is indicated.

Finally, selective urologic contrast studies are indicated in two situations. First, in a patient with gross blood at the meatus, especially if clinical and radiographic studies suggest a pelvic fracture, one should get a retrograde urethrogram. Gross blood at the meatus often correlates with clinical and/or radiologic evidence of a pelvic fracture. If the urethra is damaged, a surgical or urologic consultation is essential. If the urethra is not damaged, one can then get a cystogram after carefully advancing the catheter into the bladder. Second, if a patient with blunt or penetrating abdominal trauma is too unstable for a CT scan, perform a one-shot intravenous pyelogram in the ED. After IV administration of a bolus of 2 to 4 mL/kg of 50% diatrizoate sodium (Hypaque-Winthrop), the clinician should obtain a survey film of the abdomen 5 minutes after injection. This study will usually confirm the function or malfunction of both kidneys and occasionally the upper ureters. This information can be of great assistance to the surgeon at the time of laparotomy.

Triage

Definitive care may occur in the prehospital setting (e.g., endotracheal intubation), in the ED (e.g., chest tube placement), or in the intensive care unit or operating room. Triage is a process of patient assessment, prioritization of treatment, and selection of appropriate treatment location. In the early stages of patient assessment precise diagnosis of anatomic injury is often impossible. To identify patients with a potential for major morbidity or at risk of dying, a variety of physiologic scoring systems have been developed. In pediatric trauma, the most useful are the Glasgow Coma Scale score, the Trauma Score (TS), which uses the Glasgow Coma Scale (Table 104.4), and the Pediatric Trauma Score (PTS) (Table 104.10). For the purposes of prehospital triage, admission to a designated trauma center is indicated in any patient with a Glasgow Coma Scale score of 12 or less, a TS of 12 or less, or a PTS of 8 or less. Field studies of the TS showed that at night it is difficult to assess capillary refill and respiratory effort. Therefore, the most common tool used in prehospital triage is the Revised Trauma Score (RTS), which deletes these two variables. Admission to a trauma center is then indicated for any one of the following criteria: 1) Glasgow Coma Scale score of 12 or less, 2) systolic blood of 89 mm Hg or less, or 3) respiratory rate above 29 or less than 10. In the ED setting, a complete TS or PTS is usually obtained, but

for trauma outcome studies, the RTS is the most commonly used tool.

Component	Category		
	2+	1+	−1
Size	>20 kg (40 lb)	10–20 kg	<10 kg
Airway	Normal	Maintainable	Unmaintainable
Systolic blood pressure	>90 mm	50–90 mm Hg	<50 mm Hg
Central nervous system	Awake	Obtunded/LOC	Coma/decerebrate
Skeletal	None	Closed fracture	Open/multiple fractures
Cutaneous	None	Minor	Major/penetrating
			Sum _____ (PTS)

[a]Pediatric trauma score for prehospital and in-hospital use. (Reprinted with permission from Tepas JJ III, Ramenofsky ML, Mollitt DL, et al. The pediatric trauma score as a predictor of injury severity: an objective assessment. J Trauma 1988; 28:425–429.
LOC, loss of consciousness.

Table 104.10. Pediatric Trauma Score[a]

The PTS is designed to give added emphasis to the importance of patient sizes and airway control in injured children. Indeed, studies confirm the validity of the PTS as a predictor of outcome: 9% mortality for PTS above 8 and 100% mortality for PTS of 0 or less. From 8 to 0, there is a linear relationship between decreased PTS and an increasing potential for mortality. Nevertheless, studies comparing TS, RTS, and PTS do not show any statistical advantage of PTS over the other two for the purposes of triage. Therefore, which ever physiologic scoring system is selected, it should be used consistently and it should be used sequentially. For example, a repeat TS 1 hour after baseline TS shows how the patient is responding to treatment. Alternatively, it may reveal any delayed deterioration of the patient's condition and may suggest the need for more urgent intervention.

SUMMARY

Table 104.5 and Table 104.11 describe a rapid approach to the patient. Over a 20-minute interval, the patient may be sequentially and simultaneously assessed, treated, monitored, and subjected to further diagnostic study while the format of primary survey, resuscitation, secondary survey, and triage is followed. Each physician dealing with the injured child should have such a format within his or her armamentarium. In addition, one should be able to recognize the need for and be able to perform an emergency resuscitative thoracotomy in a patient with chest trauma who is deteriorating despite maximum fluid resuscitation (see Chapter 107). With these treatment formats well in hand, the clinician can optimize the subsequent care of the patient after admission or can provide for successful transfer of the patient to a tertiary care trauma center.

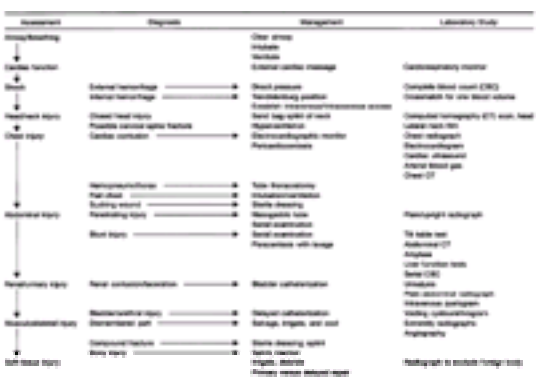

Table 104.11. Emergency Department Assessment and Management Plan for the Injured Child

Suggested Readings

GENERAL

Agran PF, Dunkle DE, Winn DG. Motor vehicle accident trauma and restraint usage patterns in children less than 4 years of age. Pediatrics 1985;76:382–386.

Agran PF, Dunkle DE, Winn DG. Motor vehicle childhood injuries caused by noncrash falls and ejections. JAMA 1985;253:2530–2533.

American College of Surgeons. Advance Trauma Life Support Program. Chicago: American College of Surgeons, 1997.

American College of Surgeons Committee on Trauma. Multiple trauma; early management in the emergency department. Bulletin of the American College of Surgeons, 1980;Feb:20–21.

Baker SP, O'Neill B, et al. The Injury Fact Book. 2nd ed. New York: Oxford University Press, 1992.

Child Maltreatment 1996: Reports from the States to the National Child Abuse and Neglect Data System. Washington, DC: US Government Printing Office, 1998.

Eichelberger MR. Pediatric trauma. In: Trunkey DD, Lewis FR, eds. Current Therapy of Trauma. 3rd ed. St. Louis: BC Decker Mosby-Year Book, 1991:21–39.

Jaffe D, Wesson D. Emergency management of blunt trauma in children. N Engl J Med 1991;324:1477–1482.

Kaback KR, Sanders AB, Meislin HW. MAST suit update. JAMA 1984;252:2598–2603.

Karwacki JJ, Baker SP. Children in motor vehicles; never too young to die. JAMA 1979;242:2848–2851.

Kaufman CR, Rivara FP, Maier RV. Pediatric trauma: need for surgical management. J Trauma 1989;29:1120–1126.

Kronick JB, Kisson N, Frewen TC. Guidelines for stabilization of the critically ill child before transfer to a tertiary care facility. Can Med Assoc J 1988;139:213–220.

Lloyd-Thomas AR. ABC of major trauma. Paediatric trauma: primary survey and resuscitation—I & II. Br Med J 1990;301:334–336, 380–382.

Mayer T, Matlak ME, Johnson DG, et al. The modified injury severity scale in pediatric multiple trauma patients. J Pediatr Surg 1980;15:719–726.

Moulton SL, Lunch FP, Peterson B. Resuscitation of the pediatric victim. In: Mancini ME, Klein J, eds. Decision Making in Trauma Management: A Multi-Disciplinary Approach. Philadelphia: BC Decker, 1991:214–219.

Pokorny WJ, Haller JA. Pediatric trauma. In: Moore EE, Mattox KL, Feliciano DV, eds. Trauma. 2nd ed. Norwalk, CT: Appleton & Lange, 1991;689–702.

Robertson LS. Present status of knowledge in childhood injury prevention. In: Preventing Childhood Injuries. Report of the Twelfth Ross Roundtable on Critical Approaches to Common Pediatric Problems. Columbus: Ross Laboratories, 1982.

Tanz RR, Christoffel KK. Pedestrian injury, the next motor vehicle injury challenge. Am J Dis Child 1985;139:1187–1190.

Templeton JR Jr, O'Neill JA Jr. Pediatric trauma. In: Cayten CG, ed. Emergency Medical Clinics of North America, Multiple Trauma. Philadelphia: WB Saunders, 1984:2(4):899–912.

DIAGNOSTIC PERITONEAL LAVAGE

Hawkins ML, Scofield WM, Carraway RP, et al. Diagnostic peritoneal lavage in blunt trauma. South Med J 1988;81:293–296.

Rothenberg S, Moore EE, Marx JA, et al. Selective management of blunt abdominal trauma in children—the triage role of peritoneal lavage. J Trauma 1987;27:1101–1106.

RADIOGRAPHIC IMAGING OF THE TRAUMA PATIENT

Beaver BL, Colombani PM, Fal A, et al. The efficacy of computed tomography in evaluating abdominal injuries in children with major head trauma. J Pediatr Surg 1987;22:1117–1122.

Bond SJ, Gotschall CS, Eichelberger MR. Predictors of abdominal injury in children with pelvic fracture. J Trauma 1991;31:1169–1173.

Brody AS, Seidel FG, Kuhn JP. CT evaluation of blunt abdominal trauma in children: comparison of ultrafast and conventional CT. Am J Roentgenol 1989;153:803–806.

Haftel AJ, Lev R, Mahour GH, et al. Abdominal CT scanning in pediatric blunt trauma. Ann Emerg Med 1988;17:684–689.

Jones TK, Walsh JW, Maull KI. Diagnostic imaging in blunt trauma of the abdomen. Surg Gynecol Obstet 1983;157:389–398.

Kane NM, Cronan JJ, Dorfman GS, et al. Pediatric abdominal trauma: evaluation by computed tomography. Pediatrics 1988;82:11–15.

Mahboubi S. Abdominal trauma in children: role of computed tomography. Pediatr Emerg Care 1985;1:37–39.

Sivit CV, Taylor GA, Bulas DI, et al. Blunt trauma in children: significance of peritoneal fluid. Radiology 1991;178:185–188.

Sivit CJ, Taylor GA, Newman KD, et al. Safety-belt injuries in children with lap-belt ecchymosis: CT findings in 61 patients. Am J Roentgenol 1991;157:111–114.

Taylor GA, Eichelberger MR. Abdominal CT in children with neurologic impairment following blunt trauma. Ann Surg 1989;210:229–233.

Taylor GA, Eichelberger MR, O'Donnell R, et al. Indications for computed tomography in children with blunt trauma. Ann Surg 1991;213:212–218.

Taylor GA, Fallat ME, Potter BM, et al. The role of computed tomography in blunt abdominal trauma in children. J Trauma 1988;28:1660–1664.

TRAUMA SCORING AND TRIAGE

American College of Surgeons Committee on Trauma. Field categorization of trauma patients and hospital trauma index. Bulletin of the American College of Surgeons 1980;Feb:28–33.

Aprahamian C, Cattey RP, Walker AP, et al. Pediatric trauma score. Predictor of hospital resource use? Arch Surg 1990;125:1128–1131.

Champion HR, Sacco WJ, Copes WS. Trauma scoring. In: Moore EE, Mattox KL, Feliciano DV, eds. Trauma. 2nd ed. Norwalk, CT: Appleton & Lange, 1991:47–65.

Eichelberger MR, Gotschall CS, Sacco WJ, et al. A comparison of the trauma score, the revised trauma score, and the pediatric trauma score. Ann Emerg Med 1989;18:1053–1058.

Kaufman CR, Maier RV, Kaufman EJ, et al. Validity of applying adult TRISS analysis to injured children. J Trauma 1991;31:691–698.

Kaufman CR, Maier RV, Rivara FP, et al. Evaluation of the pediatric trauma score. JAMA 1990;263:69–72.

Nayduch DA, Moylan J, Rutledge R, et al. Comparison of the ability of adult and pediatric trauma scores to predict pediatric outcome of following major trauma. J Trauma 1991;31:452–458.

Tepas JJ, Ramenofsky ML, Mollitt DL, et al. The pediatric trauma score as a predictor of injury severity: an objective assessment. J Trauma 1988;28:425–429.

CHAPTER 105

Neurotrauma

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HEAD TRAUMA

Head trauma accounts for approximately 250,000 hospital admissions and nearly 5 million visits to emergency departments (EDs) for pediatric patients each year. Brain injury is the leading cause of death and disability among pediatric trauma patients. Because morbidity and mortality after head trauma can be lessened by prompt stabilization, emergency physicians need to be thoroughly familiar with the manifestations of significant head injury and the necessary diagnostic and therapeutic maneuvers.

The approach to the child with a head injury is outlined in Chapter 38. This chapter focuses on the clinical anatomy, pathophysiology, clinical manifestations, diagnosis, and management of specific traumatic lesions to the head.

Clinical Anatomy

The anatomy of the head can perhaps best be considered initially in layers, traveling from the scalp inwards toward the brain parenchyma.

Injuries to the scalp, including hematomas and lacerations, are common, although not usually serious. The scalp is well vascularized. Therefore, even minor scalp lacerations may result in vigorous bleeding. Rarely, there is enough bleeding to cause some hemodynamic compromise, especially in infants. The rich vascularity of the scalp also makes it prone to the development of impressive hematomas.

Beneath the skin and subcutaneous fat of the scalp is a strong layer of tissue known as the galea aponeurotica. Deep lacerations of the scalp can be associated with a laceration to the galea as well. Hematomas beneath the galea are common after blunt impact to the cranium, especially in cases associated with skull fracture. As the clotted hematomas begin to liquefy several days after the injury, large, boggy subgaleal hematomas will become evident.

The bony “skullcap” or calvarium is composed of the frontal, parietal, occipital, and temporal bones, each of which is joined to one another by cranial sutures. Portions of the temporal and occipital bones, along with the sphenoid, palatine, and maxillary bones, comprise the skull base. Any portion of the cranium may fracture, although fractures are most likely where the bone is thinnest, as in the temporal and parietal regions, and in the skull base. Fractures of the skull base may in some cases involve the mastoid air cells, the sphenoid sinus, or the cribriform plate, and they may also be associated with a tear in the underlying meninges. In such cases, there is a direct communication between the cerebrospinal fluid (CSF) system and the nasopharynx or middle ear, posing a risk for intracranial infection.

Just beneath the skull is the dura mater, which tightly adheres to the skull at the suture lines. Between the dura mater and the skull is a potential space known as the epidural space. The meningeal arteries are embedded between two layers or “leaves” of the dura. As the body matures through childhood, some vessels—most notably the middle meningeal artery—begin to groove into the overlying bone. Therefore, traumatic impacts to the skull are particularly likely to injure these vessels. In addition, layers of the dura mater split away from each other to form the venous channels known as dural sinuses. These sinuses, which drain all of the venous blood from the brain, may be lacerated by trauma to the skull as well. The result of a laceration to the dural sinus or to the meningeal vessels is an epidural hematoma.

The next layer of meningeal tissue is the arachnoid mater. The arachnoid is a thin layer of tissue that is closely associated with the cerebral cortex, but which does not course into the brain sulci. The arachnoid mater separates the CSF-containing cisterns and subarachnoid space below from the subdural space above. The subdural space is traversed by the cerebral veins as they course from the brain to the dural sinuses. These so-called bridging veins may be sheared by acceleration/deceleration forces that violently move the brain relative to the position of the skull. The collection of blood that results is a subdural hematoma.

The subarachnoid space, which separates the arachnoid mater from the pia mater below, contains the CSF that bathes the brain and spinal cord. The pia mater is a layer of tissue that is essentially inseparable from the underlying brain, coursing with it over all gyri and sulci. The pia mater is highly vascularized with small vessels that may be injured when

shear forces or direct blows are applied to the brain. Localized bleeding from these vessels may result in subpial, subarachnoid, or subdural hemorrhage.

Just beneath the pia mater is brain parenchyma. The brain is not adherent to the skull at any point; rather, it is able to move freely within the skull, cushioned to some extent by the CSF in which it bathes. Direct blows to the head, associated with some deformation to the skull, may lead to bruising or hemorrhage in the cortex at the point of impact. In other cases, in which a blunt impact causes the brain to move against a relatively stationary skull, a contrecoup injury to the cortex, on the side opposite the site of impact, may occur.

In other cases, shear forces (as in severe acceleration/deceleration injury) can lead to diffuse injury to the axons comprising the subcortical white matter.

The brain is separated by bony prominences and by projections of the dura into three compartments: the anterior, middle, and posterior fossae. Clinically, the most important separation is that made by the tentorium cerebelli, a projection of dura mater that separates the cerebellum below from the cerebral cortex above. A notch in the tentorium allows passage of the midbrain. Cranial nerve III, the oculomotor nerve, courses along the edge of this tentorial notch. The parahippocampal gyrus and uncus of the temporal lobe lie just above the tentorial notch. When there is an increase in intracranial volume (as from a mass lesion, or from cerebral edema), the temporal lobe is pushed down through the tentorial notch, compressing cranial nerve III and the midbrain and brainstem in the process. The tentorial herniation syndrome results.

Another projection of the dura, the falx cerebri, separates the two cerebral hemispheres. Mass lesions in either hemisphere can rarely cause herniation beneath the falx to the opposite side. Herniation can also rarely occur when mass effect in the frontal lobe pushes the frontal brain posteriorly across the lesser wing of the sphenoid bone, which separates the anterior from the middle cranial fossa.

Deep within the subcortical white matter of the brain lies the ventricular system, comprised of two lateral ventricles, the third ventricle, and the fourth ventricle. The ventricular system is in communication with the subarachnoid space via connections from the fourth ventricle to the subarachnoid space at the levels of the pons and medulla. A mass lesion or cerebral edema may cause compression of the third or fourth ventricles, or of the outflow tracts, thereby blocking CSF egress and causing acute hydrocephalus.

Finally, it is important to consider the foramen magnum, the opening that allows passage of the neural tissue at the level of the junction between the medulla and the spinal cord. Mass lesions in the posterior fossa can lead to herniation of the cerebellar tonsils through the foramen magnum, with resultant compression of the medulla and potentially devastating consequences.

Pathophysiology

Primary versus Secondary Brain Injury

Discussions of traumatic brain injury typically divide the injury into two main components: primary and secondary brain injury. Primary brain injury refers to neural damage that is attributed directly to the traumatic insult itself. Shearing of neuronal axons, contusion or laceration of cerebral tissue, or direct penetration of the brain by a missile, for instance, all constitute primary brain injury.

Secondary brain injury refers to subsequent injury, after a trauma has occurred, to brain cells not injured by the initial traumatic event. Secondary brain injury may result from numerous causes, including hypoxia, hypoperfusion, excitotoxic damage, free radical damage, or metabolic derangements. In some cases, the effect of secondary brain injury is far more devastating than was the primary brain injury itself. Because many of the causes of secondary brain injury are at least theoretically preventable, most of the efforts in neurotrauma care are directed at monitoring for, and attempting to prevent, these complications.

Cerebral Ischemia

Probably the most important cause of secondary brain injury is brain ischemia, resulting from inadequate cerebral blood flow (CBF). Adequate CBF depends first on the presence of patent cerebral vessels to deliver blood to the brain. Rarely, severe head injury can be associated with shear, dissection, compression, or thrombosis of the major cerebral vessels, leading to tissue infarction. Vasospasm of the cerebral vasculature can also contribute to secondary brain injury. Vasospasm is not uncommon in cases of severe head injury, especially in those cases associated with subarachnoid hemorrhage.

Adequate CBF depends not only on patent vessels but also on adequate cerebral perfusion pressure (CPP). The cerebral perfusion pressure reflects a balance between the mean arterial pressure (MAP) of blood flowing to the brain, and the intracranial pressure (ICP), which acts as a counterforce, limiting blood flow to the brain. The relationship between these forces can be described mathematically: CPP = MAP – ICP.

In healthy children, the ICP is less than 20 mm Hg, and MAP is 70 to 80 mm Hg or greater (depending on the patient's age), yielding a CPP of 50 to 60 mm Hg or greater. The CPP fluctuates, but the healthy body maintains constant CBF in the face of minor fluctuations in CPP through autoregulation. Autoregulation is a process of reflex vasoconstriction or vasodilation in response to changes in CPP, thereby modulating resistance to blood flow in the cerebral vasculature to maintain a constant CBF. However, if the CPP drops too low (i.e., less than 40 or 50 mm Hg), the body will not be able to maintain adequate CBF despite maximal vasodilation. At this point, cerebral ischemia ensues.

Increased Intracranial Pressure

Severe drops in CPP can result from systemic hypotension (as in the multiply traumatized patient with exsanguinating injuries) or from significant increases in ICP. Increases in ICP are common in patients with serious head injuries, and they account for much of secondary brain injury.

Increased ICP may result from any process that increases the volume of the intracranial contents. Because the cranium has a fixed size and is relatively noncompliant, it can only accommodate a certain volume of intracranial contents at low pressure. An idealized pressure-volume curve (as seen in Fig. 105.1) represents the relationship between intracranial volume and ICP. In the normal state, small increments in intracranial volume can be made without significant change in the ICP (point 1 on the curve in Fig. 105.1). At this point, the intracranial contents are not particularly “tightly” packed into the cranium, and there is room for additional volume. After a certain critical point is reached, however (as indicated by point 2 on the curve), additional volume begins to lead to increases in ICP. At some point soon thereafter, the compliance of the intracranial space is exhausted (point 3 on Fig. 105.1), and the pressure-volume curve becomes steep, with even tiny increments in intracranial volume leading to massive increases in ICP. For patients on this steep part of the curve, the addition or removal of even 1 mL of intracranial volume may cause significant changes in the clinical status.

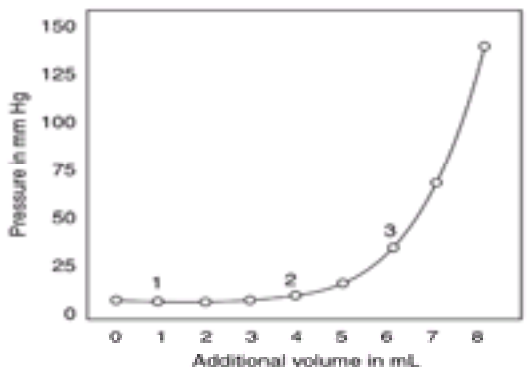

FIGURE 105.1. Effect of additional intracranial volume on intracranial pressure.

Causes of Increased ICP

Increased ICP may result from any abnormal increase in intracranial volume. This increase in volume is often a result of intracranial hemorrhage. When intracranial blood vessels are sheared or lacerated, the blood that is extravasated may accumulate to such an extent that it becomes a sizable intracranial mass. The increase in ICP that results may compromise CPP and lead to global ischemia. In addition to this global effect on ICP, the hematoma may compress the local underlying brain tissue, leading to local ischemia or metabolic derangements. As the mass expands in size, it may lead to significant shift of the brain structures within the cranium, with associated stretching or kinking of the intracranial blood vessels, and resultant ischemia to areas of brain served by these vessels. If the hematoma continues to expand, it may ultimately become so large that it leads to cerebral herniation, with resultant compression of the brainstem, neurologic deterioration, and ultimately death from cessation of brainstem functions.

A similar mass effect may occasionally be seen with large cerebral contusions, even in the absence of frank hemorrhage. Increases in ICP may also be a result of diffuse brain swelling, in the absence of focal mass lesions. Progressive brain swelling, if left unchecked, may ultimately lead to cerebral herniation.

Cerebral Herniation Syndromes

Cerebral herniation refers to the abnormal passage of brain tissue into an anatomic space in which it does not normally reside. Cerebral herniation occurs when the brain tissue is displaced by a large intracranial hematoma and/or massive brain swelling. Several distinct herniation syndromes exist, each correlated with distinct anatomic sites of herniation.

The best known herniation syndrome is tentorial herniation, which refers to the herniation of the parahippocampal gyrus and often the uncus of the temporal lobe through the tentorial notch, from the middle into the posterior fossa. Tentorial herniation most typically results from a focal mass lesion in or overlying the ipsilateral cerebral hemisphere, although it may also result from massive diffuse brain swelling. As the mass lesion or swelling expands, it pushes the brain tissue down, until a portion of the temporal lobe begins to slide through the tentorial notch. As the temporal cortex passes through, it becomes pressed against the brainstem structures and against cranial nerve III, which runs along the edge of the tentorial notch. In addition, the feeding vessels (branches of the basilar artery system) in this region may be stretched and distorted.

Most discussions of tentorial herniation describe a stereotyped sequence of clinical events that follows from the anatomic progression described previously. In many patients, however, the constellation of clinical findings varies considerably from this “classic” presentation. Classically, the patient first complains of headache, which may reflect stretching of the dura or the basal blood vessels. Next, a depression in the level of consciousness occurs, as the reticular activating system is compressed. The ipsilateral third nerve is compressed next, with resulting pupillary dilation (“blown pupil”) and eventually loss of third nerve motor function (ptosis, and loss of medial gaze). As the process continues and the cerebral peduncle is compressed, hemiparesis or decerebrate posturing develops (usually contralateral to the herniating cortex, but sometimes ipsilateral). Brainstem control of vital signs is also affected, with the development of bradycardia, hypertension, and irregular respirations (Cushing's triad). As herniation and brainstem compression continue to progress,

the patient typically loses function of both pupils and develops decerebrate posturing or flaccid paresis bilaterally. Ultimately, respiratory arrest ensues. Even if the cause of the herniation is relieved before cardiorespiratory arrest occurs, prolonged compression of brainstem structures may be associated with hyperemia of the brainstem and fatal brainstem hemorrhages after the compression is relieved.

Some other brain herniation syndromes should also be recognized. One site of herniation is the foramen magnum, through which the cerebellar tonsils may herniate. This form of herniation is usually a result of the progression of a posterior fossa mass lesion. The herniation process produces compression of the cervicomedullary junction. As the brainstem and aqueduct of Silvius are compressed, ventricular outflow obstruction may occur, with the acute onset of hydrocephalus, which will severely worsen the increased ICP, exacerbating the herniation process. Patients with herniation at the foramen magnum may present with symptoms of neck pain, vomiting, depressed mental status, bradycardia, or hypertension. In other cases, the patient may be relatively asymptomatic until sudden cardiorespiratory arrest occurs.

Another herniation syndrome is subfalcine herniation, which occurs when one cerebral hemisphere herniates beneath the falx cerebri to the opposite side. This form of herniation typically results from the progression of a unilateral supratentorial mass lesion. It is associated with symptoms of unilateral or bilateral leg weakness, and disturbances of bladder control, which result from compression and ischemia in the territory of the anterior cerebral artery.

Finally, the clinician should consider the retroalar herniation syndrome, which results from herniation of frontal lobe tissue posteriorly across the lesser wing of the sphenoid bone, usually as a result of frontal lobe mass lesions, or of swelling of the frontal lobes. The herniation may lead to distortion or compression of one or both intracranial carotid arteries, with resultant ischemia and infarction in the territories of the anterior and middle cerebral arteries.

Metabolic Derangements

Other physiologic mechanisms of secondary brain injury are important as well. Hypoxia, resulting from thoracic injuries, airway obstruction, or inadequate respiratory effort can be an important cause of brain injury. Hyperthermia increases cerebral metabolism and magnifies the severity of ischemia to an already compromised brain. There is increasing evidence that hyperglycemia also contributes to cerebral injury in the compromised brain.

The role of other substances in contributing to secondary brain injury has been carefully evaluated by investigators in the past decade. It appears that excess concentrations of the neurotransmitter glutamate, released from injured neurons into the synaptic cleft, can lead to injury through excess excitation of otherwise healthy postsynaptic neurons. Oxidizing agents and oxygen free radicals, released from injured neurons or elaborated as part of the brain's inflammatory response to injury, also appear to play a role in causing secondary brain injury.

Management and General Principles

Initial Resuscitation

Management of the patient with a head injury focuses on the prevention of secondary brain injury. As with all emergency care, management begins with the ABCs (airway, breathing, and circulation) of resuscitation. A patient with a head injury who has altered sensorium may require assistance with positioning of the airway or suctioning of oral and pharyngeal secretions. The clinician must recognize the potential for cervical spine injury in all patients with head injuries, and cervical spine precautions must be taken during airway management. Immobilization of the cervical spine with a semirigid cervical collar or with in-line manual stabilization must be maintained until the clinician can be certain that no cervical spine injury has occurred.

Breathing may be impaired if the patient's neural control of respiratory function is compromised or if traumatic injuries involve the thorax. All seriously traumatized patients require 100% inspired oxygen until it is certain that supplemental oxygen is not needed. Positive-pressure ventilation with a bag-valve-mask apparatus should be provided for any patient with inadequate respiratory effort. Unless there is increased ICP, the clinician should aim to achieve normocarbia (P_{CO_2} 35 to 40), and oxygen saturations of 100%. If there is evidence of increased ICP, therapeutic hyperventilation may be indicated (see the following).

Endotracheal intubation should be performed for any patient making inadequate or labored respiratory effort, or for patients who have a blunted gag reflex, cannot manage their oral secretions, or are comatose. Orotracheal intubation is generally safer than nasotracheal intubation if there is any concern about injuries to the midface. Care must be taken to minimize manipulation of the cervical spine during the process of intubation. Rapid sequence intubation is indicated for most patients with head injuries to ensure that the patients are comfortable and that intubation can be safely achieved. However, adequate oxygenation, ventilation, and protection of the airway are always the first priority. If intravenous (IV) access cannot be rapidly achieved, attempts at intubation may need to proceed without the use of adjunct medications.

Premedication for rapid sequence intubation begins with atropine 0.02 mg/kg (maximum dose 0.5 mg) for children less than 8 years of age, to lessen the vagal response to intubation. Lidocaine may also be useful, at a dosage of 1 to 2 mg/kg, as premedication to blunt the airway reflexes, thereby reducing the risk of coughing or choking, which may increase ICP.

If possible, a sedative drug should be used, both to make the patient comfortable, and to decrease the patient's responsiveness to airway manipulation. Thiopental at a dosage of 4 to 7 mg/kg is an ideal drug for patients with head injuries who have increased ICP because it decreases cerebral metabolism, thereby reducing the risk of ischemia. It must be used cautiously, however, in patients with any hemodynamic instability because it may reduce vasomotor tone and cardiac contractility, thereby leading to a decrease in blood pressure. If thiopental cannot be used, an alternative regimen is fentanyl (2 to 3 µg/kg) and midazolam (0.1 mg/kg), which provide sedation and analgesia with minimal effect on

cardiac contractility or vasomotor tone. In cases in which intubation must proceed but IV access cannot be achieved, midazolam may be given intramuscularly (0.1 mg/kg), with onset of action in about 3 minutes. Ketamine should be avoided in patients with head injuries because it can increase ICP.

For neuromuscular blockade, succinylcholine (1 to 1.5 mg/kg intravenously) offers the advantage of rapid action, with intubating conditions developing within 45 to 60 seconds. In theory, the diffuse fasciculations caused by succinylcholine may serve to increase resistance to venous drainage from the head and increase ICP. This potential risk can be avoided with the use of nondepolarizing agents, such as vecuronium (0.1 mg/kg intravenously) or rocuronium (0.6 to 1.2 mg/kg intravenously), which do not cause fasciculations. Although the onset of action for these agents is slower than for succinylcholine, rocuronium, at the higher end of its dosage range, can be expected to provide intubating conditions in approximately 60 to 90 seconds.

The circulatory status of patients with isolated head trauma is generally not compromised, although the potential for other organ system trauma, with associated hemodynamic compromise, must be immediately recognized. Intravenous access should be obtained immediately in all patients with moderate or severe head injuries.

Isotonic crystalloid solutions—normal saline or lactated Ringer's solution—should be given as needed to restore normal intravascular volume (see Chapter 104). The clinician should remember that the patient will only have an adequate cerebral perfusion pressure if the MAP is maintained in a normal range. On the other hand, for patients with adequate intravascular volume, excess fluid administration should be avoided. These patients can be treated with normal saline or Lactated Ringer's solution, running at one-half to two-thirds the maintenance fluid rate, while evaluation and treatment of the head injuries are being performed.

Brain-Specific Therapies

Once the ABCs of resuscitation have been addressed and the patient has been stabilized, attention can be given to the neurologic status. The neurologic assessment of the patient with a head injury, and the criteria for deciding which patients need neuroimaging are described in detail in Chapter 38.

In any patient with signs of increased ICP on examination (i.e., a progressively deteriorating neurologic status and/or signs of impending herniation), a computed tomography (CT) scan of the head should be performed immediately, with the goal of identifying any mass lesions that require evacuation. If a head CT scan is not available on site, emergent transfer to a facility where CT can be performed is usually the most appropriate course.

There is a long history in emergency neurotrauma care of empiric “blind” trephination (drilling of burr holes) for patients with signs of impending herniation. The goal of such therapy is to provide immediate decompression for patients who are clinically suspected of having an intracranial hematoma. As emergency CT imaging of the head has become more readily available, the role for empiric trephination is more limited. In most cases, the benefits of the information provided by head CT outweigh the costs of waiting a few extra minutes to have the scan performed, even in cases where herniation is impending. Especially for the pediatric age group, in which most cases of increased ICP result from diffuse brain swelling rather than intracranial hematoma, empiric trephination is likely to be of limited benefit. Nonetheless, empiric trephination may still have a role in select patients who are too unstable to be transported to the radiology suite, or in cases where the nearest CT scanner is too far away.

Medical treatment for increased ICP should be undertaken immediately when increased ICP is suspected. This treatment includes elevation of the head of the bed to an angle of 30 degrees, which promotes venous drainage from the head (thereby decreasing the volume of the intracranial vasculature). The head should be maintained in a midline position, which helps maintain venous outflow through the jugular system as well. Furthermore, sedating medications may be needed to prevent the patient from coughing and choking or from becoming agitated, both of which might be associated with increased intrathoracic pressure and, therefore, impaired venous drainage. Sedation should be used as sparingly as possible, however, so that the neurologic status can be monitored. Paralytic agents should be used only when sedating agents cannot be tolerated or when maximal therapy with sedation fails to control the patient's behavior.

Hyperventilation also decreases ICP by decreasing the volume of the intracranial vasculature. The cerebral arteriolar circulation responds to hypocarbia with reflex vasoconstriction. The therapeutic use of hyperventilation requires a delicate balance: too little ventilation leads to vasodilation and increased ICP, but too much ventilation leads to excess vasoconstriction and decreased CBF. The optimal balance for therapeutic hyperventilation appears to be achieved at a P_{CO_2} of 30 to 35 mm Hg. The gold standard for monitoring P_{CO_2} is the arterial blood gas. Therefore, an arterial line is invaluable in monitoring the progress of hyperventilation in the patient with a head injury. In the short run, however, useful information about trends in P_{CO_2} can be obtained readily with the use of end-tidal CO_2 monitoring.

Mannitol can also be given to lower the ICP, at an IV dose of 0.5 to 1 g/kg, which increases the serum osmolarity. The increased serum osmolarity draws free water into the vasculature, thereby leading to a decrease in the viscosity of blood. Because of the lower viscosity of blood, CBF is improved. The improvement in CBF leads to improved cerebral oxygenation, which helps prevent cerebral ischemia. Furthermore, the autoregulatory system responds to the improved cerebral oxygenation with reflex vasoconstriction, thereby lowering intracerebral volume (and ICP) without compromising CBF. The effect of mannitol on ICP is seen within a few minutes of administration. Over the ensuing hour or so, mannitol also leads to some degree of intravascular volume depletion because of its action as an osmotic diuretic. Clinicians should be cautious about the use of mannitol in any patient with possible hemodynamic compromise because the administration of a diuretic may exacerbate hypovolemia and worsen perfusion.

There is disagreement in the literature about the optimal use of hyperventilation and mannitol in the management of the patient with a head injury. The clearest indication for these maneuvers is to “buy time” for several minutes in a patient with clinical signs of impending herniation. Stabilization of the patient with impending herniation by using hyperventilation and/or mannitol may allow enough time for the patient to be safely transferred to the radiology suite, for emergency head

CT imaging. If an evacuable hematoma is discovered on CT, these maneuvers can be used to stabilize the patient en route to the operating suite, where the increased ICP will be more definitively relieved.

It is less clear that sustained hyperventilation or repeated doses of mannitol are useful in patients who have diffuse brain swelling but who do not have surgical mass lesions. In particular, numerous studies in recent years have documented a clear relationship between even mild degrees of hyperventilation (P CO_2 30 to 35) and decreased CBF. Because the overall goal of resuscitation in the patient with a head injury is to optimize CBF, prolonged hyperventilation may be counterprotective in that it may actually worsen cerebral ischemia. Therefore, hyperventilation is most useful as a transient therapy for acute changes in neurologic condition or as a second-line therapy after other methods of managing ICP have failed.

Some concern has also been raised that repeated doses of mannitol may be counterproductive in the ongoing care of patients with brain swelling because mannitol can leak across the injured blood–brain barrier, with its osmotic pull serving to worsen cerebral edema. Although some experimental models have documented this phenomenon, most data from clinical studies indicate lasting improvements in CBF with repeated doses of mannitol. Therefore, many authors consider mannitol to be a useful adjunct in the management of increased ICP in patients with brain swelling.

No evidence exists that hyperventilation or mannitol prevents the development of brain swelling. Therefore, the prophylactic use of these therapies is not recommended.

Many studies over the years have evaluated the utility of corticosteroids in the patient with a head injury. Theoretically, corticosteroids might blunt the inflammatory response to brain injury, thereby decreasing brain swelling. However, clinical researchers have been unable to show any improvement in outcome for patients with head injuries treated with corticosteroids. The use of corticosteroids for the treatment of head injury is therefore not recommended.

Anticonvulsant medications are clearly indicated for patients who are having ongoing seizure activity. Short-acting benzodiazepines (lorazepam or diazepam) may be used acutely in the management of ongoing seizures, and phenytoin or fosphenytoin may be used for maintenance anticonvulsant effect.

Phenytoin is also commonly used as prophylaxis for patients who have intracranial lesions associated with an increased risk of seizure activity, such as cerebral contusion, intraparenchymal hemorrhage, subarachnoid hemorrhage, or subdural hemorrhage. Patients who have epidural hematoma but no associated parenchymal injuries usually are not treated prophylactically.

Disposition

Generally, all patients with intracranial hematomas or brain injuries noted on head CT imaging should be hospitalized, no matter how mild or severe their symptoms. In addition, any patient with an abnormal mental status or neurologic examination should be hospitalized, even if head CT findings are normal. Patients with neurologic compromise and sizable intracranial hematomas require emergency operative intervention, and they are monitored postoperatively in the intensive care unit (ICU) for development of cerebral edema or recurrence of bleeding. Patients with neurologic compromise but no surgical lesions also need intensive care monitoring, often with the placement of a device for the measurement of ICP. In general, ICP monitors are indicated for any patient with a head injury who is comatose and who has an abnormal head CT. Although numerous different types of ICP monitors have been used, the intraventricular catheter has the advantage of being useful both for monitoring and for therapy because CSF can be drained through the catheter if needed to lower the ICP acutely. The goal of ICU management for patients with ICP monitors is to maintain an adequate CPP, which generally entails maintaining ICP at 20 mm Hg or less. More mildly symptomatic patients with a normal neurologic status and small cerebral contusions or intracranial hematomas may be candidates for observation in a ward setting.

Well-appearing patients with head injuries who either required no head CT scan (see Chapter 38) or who have no intracranial lesions on head CT imaging may be suitable for discharge to home with careful instructions. A discussion of the management of these patients follows.

Blunt Trauma: Specific Lesions

Concussion

Concussion is defined by the Centers for Disease Control and Prevention as a head trauma–induced alteration in mental status that may or may not involve loss of consciousness. Most clinicians use the term concussion to refer to mild head injuries, with no or minor depression in the level of consciousness (Glasgow Coma Scale scores of 13 to 15), and with no associated focal neurologic deficits. Concussion most commonly results from falls in infants and toddlers and from motor vehicle collisions or sports-related injuries in older children and adolescents.

Common symptoms of concussion include initial loss of consciousness, amnesia, confusion, headache, nausea, vomiting, and dizziness. For the most part, clinicians diagnose concussion in those patients with minor head trauma who have no brain imaging performed, or as a diagnosis of exclusion for patients with minor head injury who have no evidence of intracranial pathology on a head CT scan.

Increasingly in recent years, it has been recognized that many concussed patients with normal head CT findings do have subtle evidence of brain contusion or diffuse axonal injury noted on magnetic resonance imaging (MRI) of the brain. Researchers have also found abnormalities in cerebrovascular autoregulation in some patients with concussion who have normal CT scans.

In general, patients with concussion can be expected to do well, and no specific therapy is required. A number of large studies have shown that patients with minor head injury who have normal head CT scans are at low risk for clinical deterioration. In general, these patients may be safely discharged to home if no other issues require inpatient care.

There are several case reports in the literature of the second impact syndrome, in which patients have experienced an initial minor head injury during a sporting event, with some associated concussive symptoms, and then have had serious neurologic deterioration or died after a second seemingly minor head impact occurred. It is presumed that in these cases, the athletes had a relatively asymptomatic contusion or diffuse axonal injury, perhaps associated with impaired autoregulation, that was exacerbated by the second impact. Based on these frightening reports, the American Academy of Neurology has published recommendations for the management of athletes who have sustained a concussion. A summary of the recommendations has been published in *Morbidity and Mortality Weekly Report* and is presented in Table 105.1.

Grade 1 Concussion
Definition: transient confusion, no loss of consciousness, mental status abnormalities for ≤ 15 minutes.
Management: return to sports activities same day only if all symptoms resolve within 15 minutes; if a second grade 1 concussion occurs, no sports activity until asymptomatic for 1 week
Grade 2 Concussion
Definition: transient confusion, no loss of consciousness, mental status abnormalities for >15 minutes.
Management: no sports activity until asymptomatic for 1 full week; if a grade 2 concussion occurs on the same day as a previous grade 1 concussion, no sports activity for 2 weeks
Grade 3 Concussion
Definition: concussion involving loss of consciousness
Management: no sports activity until asymptomatic for 1 week if loss of consciousness was brief (seconds), or for 2 weeks if loss of consciousness was prolonged (minutes or longer)
Second grade 3 concussion, no sports activity until asymptomatic for 1 month
Any abnormality on computed tomography or magnetic resonance imaging, no sports activity for remainder of season; patient should be discouraged from any future return to contact sports
Modified from Morbidity and Mortality Weekly Report, Centers for Disease Control and Prevention.

Table 105.1. Recommendations for Return to Sports Activity after Concussion

In addition to these recommendations, the patient should be instructed to rest until symptoms improve. Acetaminophen may be prescribed for headache, but more potent analgesics should probably be avoided so that any progression of symptoms can be detected. The warning signs of progressing intracranial injury should be reviewed with the patient before discharge, with instructions to return immediately if any of these new symptoms or signs appear.

Symptoms after a concussion generally resolve quickly, often within minutes or hours after the injury. However, some patients develop the postconcussion syndrome, in which symptoms of confusion, amnesia, headaches, or dizziness may persist for days or even weeks after the injury.

Skull Fracture

Clinical Findings and Diagnosis

Skull fractures occur at a rate of approximately 2 per 1000 infants per year, and in approximately 0.5 to 1 per 1000 older children and adolescents. Skull fractures result mainly from falls in infants, but they may also result from motor vehicle collisions, child abuse, or other mechanisms. In older children and adolescents, skull fractures usually result from motor vehicle collisions or sports-related injuries.

Infants are clearly at a higher risk for skull fracture than older children, probably because their skulls are thinner. Many skull fractures in infants result from short distance falls; generally, about 50% of infants with skull fracture have fallen less than 4 or 5 feet. As the child matures beyond the first year of life, the propensity to sustain skull fracture disappears quickly.

Fractures may occur in any bone of the skull, although fractures of the parietal bone constitute about 70% of cases. The occipital and temporal bones are next most commonly involved, with the frontal bone least likely to fracture. Basilar skull fractures commonly occur in pediatrics as well, although less commonly in infants, and more often in older children and adolescents.

Most cases of skull fracture are associated with soft-tissue swelling or hematoma overlying the fracture site. Skull fracture may occur in the absence of recognized soft-tissue findings as well, perhaps because subtle swelling is missed beneath the patient's hair, or because it may take several hours after the injury before the swelling becomes clinically evident. Palpable bony abnormalities are rarely detected in cases of linear or minimally depressed skull fracture but may be evident in cases with more severe depression. Other symptoms and signs of head injury, such as loss of consciousness, vomiting, lethargy, seizures, or irritability, may be present, especially if intracranial lesions are associated, but they are often absent in cases of isolated skull fracture.

Signs of basilar skull fracture may include hemotympanum, Battle sign (hematoma or discoloration overlying the mastoid bone), “raccoon eyes” (blue or purple discoloration of the periorbital tissue), or CSF rhinorrhea or otorrhea. There may be no abnormalities on examination of the scalp, and there may be no signs or symptoms of intracranial injury.

Skull fracture may be diagnosed by plain radiographs of the skull or by head CT imaging (Fig. 105.2,Fig. 105.3 and Fig. 105.4). A head CT scan is usually the preferred imaging modality for evaluating children with head injuries because it provides information not only about the skull but also about the intracranial contents. Skull radiographs are somewhat

more sensitive for detecting skull fracture, especially for those horizontal fractures that run parallel to and between adjacent “cuts” on the CT scan. Head CT imaging is better than skull radiography, however, for detecting subtle degrees of depression of the bony fragments. Head CT imaging is also preferred in cases in which a diagnosis of basilar skull fracture is considered because it allows better imaging of the basilar skull and for visualization of pneumocephaly or fluid in the mastoid air cells, which are common associated findings (Fig. 105.5 and Fig. 105.6).

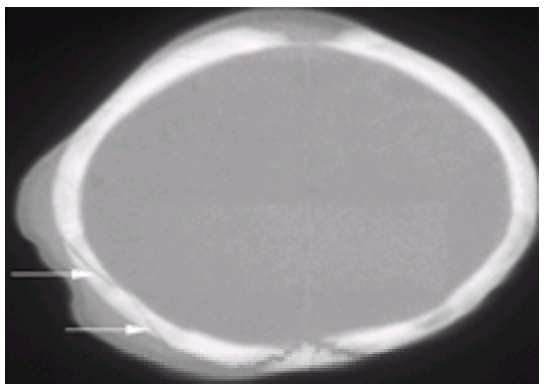

FIGURE 105.2. Linear skull fracture. This is a head computed tomography (CT) scan performed on a 6-month-old girl who fell down 20 steps. The *arrows* indicate a comminuted linear right parietal skull fracture. Note also the extensive soft-tissue swelling of the right parietal scalp. No intracranial abnormalities were identified.

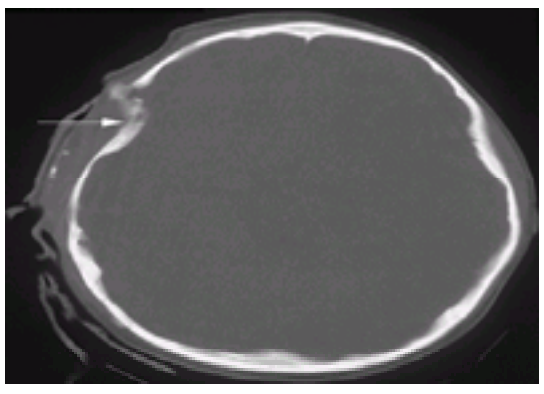

FIGURE 105.3. Depressed skull fracture. This head computed tomography (CT) scan was performed on a 6-year-old boy who was unresponsive and apneic after being a passenger in a high-speed motor vehicle collision. The *arrow* indicates a depressed skull fracture involving the right temporal bone. He also had an associated right temporal contusion (see Fig. 105.7).

FIGURE 105.4. Depressed skull fracture. This 3-month-old boy fell out of bed and was noted to have a palpable depression of the skull. Head computed tomography (CT) imaging shows a “ping-pong ball” type depressed skull fracture. No intracranial abnormalities were identified.

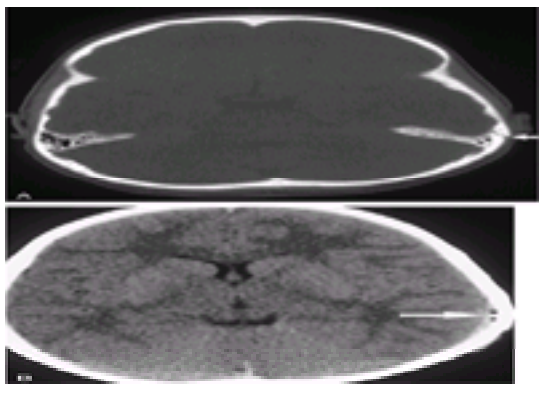

FIGURE 105.5. Basilar skull fracture. This 5-year-old girl was a pedestrian struck by a bicycle. Hemotympanum was noted on examination. **A.** The arrow indicates a fracture of the left temporal bone. The adjacent mastoid air cells are somewhat opacified. **B.** A small extraaxial hematoma with associated pneumocephaly is seen *(arrow)*.

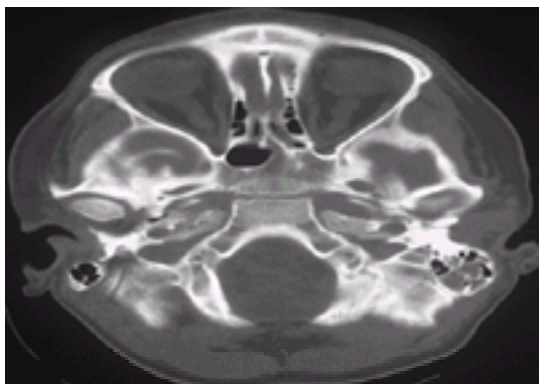

FIGURE 105.6. Basilar skull fracture. This 10-year-old male fell 10 feet to the ground. He had left hemotympanum. A head computed tomography (CT) scan shows a fracture through the petrous portion of the temporal bone (*thin arrow*), extending toward the internal carotid canal *(thick arrow).* The left mastoid air cells are somewhat opacified. Other “cuts” of the CT confirm that the fracture involves the wall of the carotid canal. A cerebral angiogram was performed, which showed normal vascular integrity.

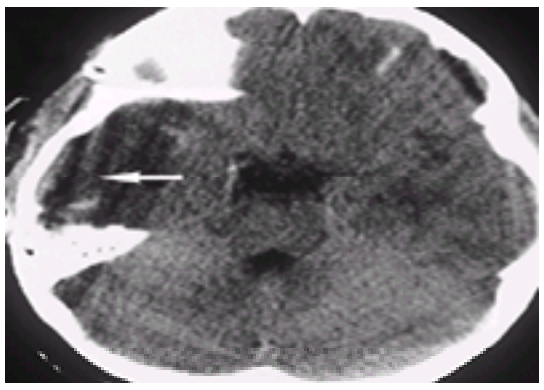

FIGURE 105.7. Cerebral contusion. This head computed tomography (CT) scan was performed on a 6-year-old boy who was found unresponsive and apneic after being a passenger in a high-speed motor vehicle collision. The *arrow* indicates a large area of hypodense nonhemorrhagic contusion in the right temporal lobe.

Management

Linear Skull Fracture. The main significance of linear skull fractures is that they indicate that the risk of intracranial injury is increased by as much as tenfold to twentyfold. In cases in which acute linear skull fracture is diagnosed, therefore, a head CT scan is essential to evaluate for possible intracranial injury. In addition, any diagnosis of skull fracture should lead the clinician to consider the possibility of child abuse. If the history provided is not a plausible explanation for the injuries observed, further evaluation for possible child abuse should be initiated.

Many clinicians routinely admit children with skull fracture to the hospital for a period (e.g., 24 hours) of observation to exclude the small possibility of late complications. For well-appearing children with a linear skull fracture and no associated intracranial injuries, however, the risk of late complications is low. Therefore, if a child with skull fracture but no intracranial lesions remains well over a short period of observation in the ED and child abuse is not suspected, the child may be considered for discharge to home. The warning signs of advancing intracranial injury should be carefully reviewed, with advice to return immediately if any of these signs are noticed. The family should also be counseled to expect the possible development of a subgaleal hematoma, which becomes more evident as the clotted blood overlying the fracture site begins to liquefy, and which presents as a large boggy swelling of the scalp, usually between 5 and 7 days after the injury. Unless the hematoma develops signs of infection, it will resolve gradually on its own and should not be aspirated.

Linear skull fractures generally heal well, with no specific therapy required. Fewer than 1% of patients will develop a growing skull fracture; that is, a fracture that fails to heal and becomes wider over time. All patients should have a follow-up examination 1 month after the initial injury to ensure that there is no evidence of development of a growing fracture.

Depressed Skull Fracture. Skull fractures may be associated with depression of the fracture fragments, which may range from barely detectable depressions to more obvious, palpable deformities in the skull (Fig. 105.3 and Fig. 105.4). If there are no other complicating features, isolated skull fractures with minimal depressions can be managed in a fashion similar to that previously described for linear skull fractures.

More significant depressions of the skull, however, are more serious because they may be associated with contusion or laceration to underlying brain. In cases in which injury to underlying brain is noted on head CT imaging, especially if there are seizures or focal neurologic findings referable to the brain injury, prompt surgical elevation of the fracture fragments may be required. Surgical intervention is also usually necessary for compound, or open, depressed skull fractures, in which early debridement and closure is performed, especially for those patients who have lacerations of the dura mater. Penetrating injuries of the skull are a special case of open, depressed skull fracture and are discussed later in the chapter.

Surgical elevation is generally necessary (although not necessarily emergently) for patients with depressed skull fracture who have associated compression to underlying brain parenchyma or intraparenchymal bone fragments. Patients with significant cosmetic deformity are candidates for surgical repair as well. Most neurosurgeons would recommend operative repair for any skull fracture with a 1-cm or greater depression, or for depressions with a depth greater than the thickness of the skull.

Basilar Skull Fracture. Fractures through the skull base are unique in that they may involve disruption of the mastoid air cells or the paranasal sinuses, raising the possibility of intracranial infection. Most recent studies suggest that the risk of meningitis after basilar skull fracture is low, with rates between 0.4 and 5%. The highest risk is in patients with evident CSF rhinorrhea or otorrhea. Although some controversy exists in the literature, it appears that prophylactic antibiotics reduce the risk of meningitis in high-risk patients. Many would recommend, therefore, that patients with basilar skull fracture and CSF leak be admitted to the hospital for IV antibiotics. Neurosurgical management of CSF leaks may also include several maneuvers, such as external CSF drainage to lower pressure and allow the leak to heal, packing of the sinuses, or operative repair of dural lacerations.

Although it has been traditional management for all patients with basilar skull fracture to be admitted to the hospital for observation, recent authors have suggested that if patients with basilar skull fracture are neurologically normal, have no intracranial pathology on head CT, and have no CSF leak, they may be safely discharged to home. If they are to be discharged, instructions about the management of head injury, as previously described for linear skull fractures, should be discussed in detail. Furthermore, the family should be warned to watch closely for fever, stiff neck, photophobia, or any other signs of developing intracranial infection.

Growing Skull Fracture. Growing skull fractures are seen almost exclusively in patients who sustain the initial injury in the first year of life. Growing skull fractures result from a tear in the dura underlying the fracture, allowing subsequent herniation of meningeal tissue into the fracture line. There is almost always associated injury to the underlying brain parenchyma, sometimes with herniation of parenchyma into the fracture line, and often with a porencephalic enlargement of the adjacent ventricle. Although the pathogenesis of growing skull fracture is not fully understood, it is believed that the constant pressure exerted by the herniated tissue leads to erosion of the fractured edges of bone.

Growing fractures are more likely in patients who had larger, more widely diastatic fractures on presentation, especially if damage to the underlying parenchyma was significant. Growing fractures present weeks or months after the initial injury, usually as persistent swelling overlying the fracture site, sometimes with the development of a boggy or pulsatile soft-tissue mass. Associated neurologic symptoms such as developmental delay, focal neurologic deficits, or seizures may be evident as well, probably reflecting injury and abnormal development of the brain tissue adjacent to the fracture.

Surgical repair is required in cases of growing skull fracture, with repair of the rent in the dura (often involving placement of a synthetic graft) and autologous bone graft over the fracture site.

Parenchymal Injuries

Cerebral Contusion and Intraparenchymal Hematoma

Pathophysiology. Cerebral contusion refers to bruising of the cerebral cortex. On a microscopic level, there is focal injury to neurons, glial cells, and blood vessels, with some extravasation of blood noted, and some swelling of neural cells.

Cerebral contusion occurs after blunt trauma because of the impact of the relatively mobile brain tissue against a relatively fixed skull. Injuries may occur at the point of traumatic impact (coup injuries) or at a site opposite the point of impact (contrecoup injuries). Contusions are most likely to occur in those locations where the brain is less cushioned by CSF and is more able to come into direct contact with the bony skull. Most commonly, contusions are seen in the undersurface of the frontal lobe or in the poles of the temporal lobes.

The presence of cerebral contusion indicates primary brain injury to the tissue involved. Focal neurologic deficits associated with dysfunction of the contused tissue should be expected. In addition, cerebral contusion leads to a risk for secondary brain injury because the contusion may exert some mass effect on surrounding tissue, with resulting cerebral dysfunction and risk for further ischemia. Finally, contusions are associated with a risk of late intraparenchymal hematoma.

Intraparenchymal hematoma may occur as a late complication of an initially nonhemorrhagic contusion, or it may be evident from the initial time of injury. These hemorrhages usually result from severe traumatic forces. The presence of hemorrhage may cause impaired blood flow to the adjacent parenchyma. If the hemorrhage becomes large enough, it may exert mass effect and even lead to cerebral herniation.

Clinical Manifestations. The severity of the clinical manifestations associated with cerebral contusion can vary widely. Often, there is history of loss of consciousness and/or some disturbance in the mental status. Focal neurologic deficits related to the contusion may be noted. Frontal contusions, for instance, may be associated with behavioral alterations, and temporal contusions may be associated with disturbances of memory. Seizures are relatively common as well.

With the increasing use of CT and MRI for patients with mild head trauma, an increasing number of contusions are being discovered in patients with no or mild symptoms (headache, nausea and vomiting, lethargy).

Most patients with intraparenchymal hematomas are comatose, and they may have focal neurologic deficits. Rare patients with intraparenchymal hematoma may initially be alert, but they have a high risk for deterioration over the

ensuing hours.

Diagnosis. Cerebral contusions are generally evident on a head CT scan as hypodense areas of edema, sometimes intermingled with hyperdense areas of hemorrhage (Fig. 105.7 and Fig. 105.8). Intraparenchymal hematomas are more uniformly hyperdense, although areas of active bleeding may be isodense (Fig. 105.9).

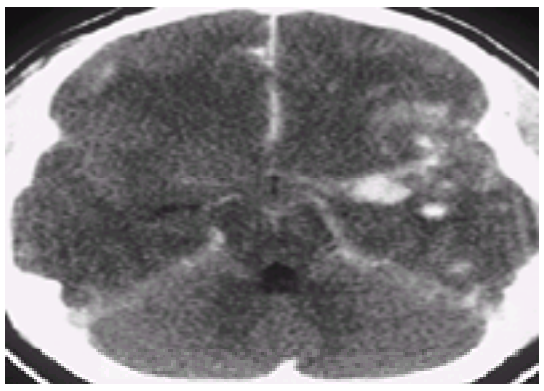

FIGURE 105.8. Cerebral contusion. This 16-year-old girl was comatose after being a passenger in a high-speed motor vehicle collision. A head computed tomography (CT) scan shows hemorrhagic contusion of the left temporal lobe, subdural hematoma along the tentorial margins, and effacement of the sulci throughout. The patient expired despite intensive medical management for increased intracranial pressure.

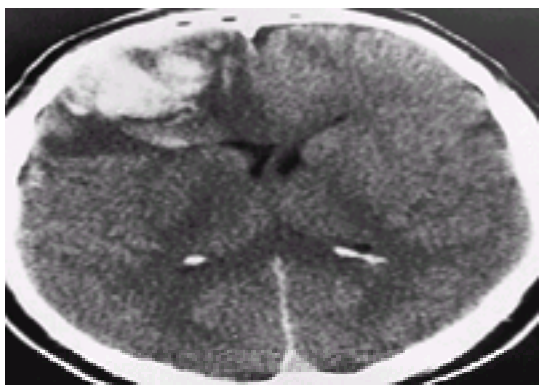

FIGURE 105.9. Intraparenchymal hemorrhage. This adolescent was an unrestrained passenger in a high-speed motor vehicle collision. He was comatose on presentation. A head computed tomography (CT) scan shows a large area of intraparenchymal hemorrhage in the right frontal region. Note also, the surrounding area of low-density cerebral contusion.

Management. All patients with acute cerebral contusion and intraparenchymal hematoma should be admitted to the hospital for observation. Patients with a smaller contusion, a normal neurologic status, and no other lesions noted on head CT imaging may be appropriately managed on the inpatient ward. More seriously ill patients with an abnormal neurologic status generally require ICU monitoring. Patients with intraparenchymal hemorrhage should be admitted to the ICU as well.

Management of cerebral contusions focuses on efforts to prevent secondary brain injury, with the recognition that the contused tissue and surrounding areas are at especially high risk for ischemia. For patients in coma, ICP monitoring is generally indicated, and maneuvers for managing increased ICP may be required. The clinician must be especially alert for the possibility that an initially nonhemorrhagic contusion will undergo late hemorrhage, which would manifest as a sudden increase in ICP and/or deterioration in clinical status.

Patients with large contusions exerting a significant mass effect may require surgical resection. Surgical resection should be avoided when possible, however, because the contused tissue may actually be viable and regain function over the long-term.

The management of intraparenchymal hematomas is problematic in that operative drainage of these lesions risks injury to adjacent brain tissue. Clearly, large hematomas exerting a significant mass effect require operative drainage. Smaller lesions may initially be managed nonoperatively, but with the clinician recognizing the high risk for sudden deterioration. ICP monitoring is necessary for most patients with intraparenchymal hematoma in order to detect early signs of a growing lesion. Prophylactic anticonvulsants are often given for patients with cerebral contusions or intraparenchymal hematoma.

The prognosis for patients with cerebral contusion can vary widely, depending mainly on the patient's neurologic status on presentation, and on the presence or absence of other lesions. Patients with more significant cerebral contusion often have some residual neurologic disability. Follow-up CT scans on such patients show areas of encephalomalacia at the site of injury. Other patients may have essentially full recovery, with no residual neurologic deficits evident. Patients with intraparenchymal hematomas tend to have incurred severe brain injury, and they often have a poor outcome.

Diffuse Axonal Injury

Pathophyisology. Diffuse axonal injury (DAI) is characterized pathologically by injury to the white matter tracts of the brain, often at the junction of gray and white matter or sometimes deeper at the level of the corpus callosum, brainstem, or cerebellum. Pathologically, degeneration of the axons themselves is noted, with the presence of axonal retraction balls, microglial proliferation, and demyelination. In addition, there is usually accompanying endothelial damage to the capillaries, with some punctate areas of hemorrhage. Edema is sometimes, but not always, associated.

DAI results from the application of severe acceleration/deceleration or angular rotational forces to the brain, which lead to shear injuries of the axons and associated vasculature.

DAI reflects diffuse primary injury to the white matter. In addition, DAI is often associated with other focal lesions, or with global brain swelling.

Clinical Manifestations. The clinical manifestations of DAI can vary greatly, ranging from those patients who have symptoms of concussion to those who present in coma. Loss of consciousness is common. In one study of patients with DAI, 82% developed coma. In general, patients with more diffuse areas of DAI noted on radiographic imaging (especially if involving the brainstem) have more severe symptoms.

Diagnosis. DAI is evident on CT scan as small nonexpansive hemorrhagic lesions of the white matter, most typically seen at the gray–white junction of the cerebral hemispheres, or in the corpus callosum, brainstem, or cerebellum. Cerebral swelling sometimes accompanies DAI, but it need not be present for a diagnosis of DAI to be made. Intraventricular hemorrhage is sometimes noted (Fig. 105.10). Intraparenchymal hemorrhages and cerebral contusions are commonly noted in patients with DAI as well.

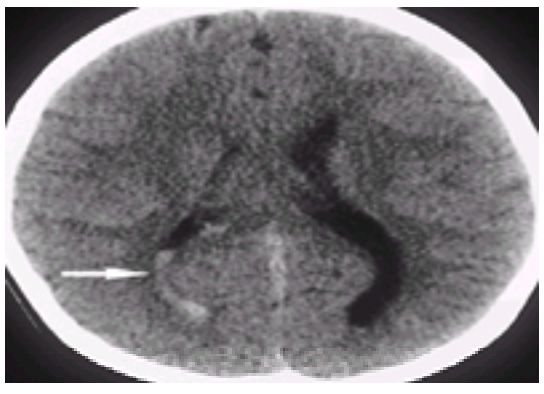

FIGURE 105.10. Intraventricular hemorrhage. This head computed tomography (CT) scan was performed on an 8-year-old girl who was involved in a sledding accident. She presented in coma. The *arrow* indicates hemorrhage in the right lateral ventricle. Note the layering of blood inferiorly in this supine patient. Other “cuts” of the CT showed areas of punctate hemorrhage consistent with diffuse axonal injury. The patient made an excellent recovery, with minimal neurologic deficits.

Management. Patients with a radiographic diagnosis of DAI should be admitted to the hospital for observation. Patients with DAI who have a normal neurologic examination and no other lesions evident on CT scan may be able to be managed on a general inpatient unit. Those with an abnormal neurologic status require ICU level monitoring. Management of patients with DAI is supportive, with efforts directed mainly at preventing secondary brain injury. ICP monitoring is generally indicated for patients who present in coma. Specific therapies may be required for the management of increased ICP, as outlined previously.

In some large series of patients with DAI, mortality rates range from 10 to 15%. Of those who survive, persistent neurologic dysfunction is common, occurring in 30 to 40% of patients. In general, however, children with DAI have a better prognosis than adults. A good functional outcome can be expected for patients with DAI who have mild symptoms of head injury (Glasgow Coma Scale score of 13 to 15).

Diffuse Brain Swelling

Pathophysiology

Diffuse brain swelling (DBS) is a common manifestation of head trauma, especially in pediatrics. In one large series of children with severe brain injury, 41% had DBS. The origin of DBS is probably multifactorial. One component of DBS appears to be an increase in intracerebral vascular volume, probably caused by a loss of normal autoregulatory reflexes and diffuse vasodilation.

Many authors argue that the time course of DBS, occurring within minutes after a traumatic insult, is most consistent with this process of vasodilation, rather than the development of edema, which might be expected to take longer. Nonetheless the development of edema likely plays at least some role in many, if not all, cases of DBS as well. Edema may be vasogenic (extravasated from injured or inflamed blood vessels), cytotoxic (representing intracellular swelling of injured brain cells), or interstitial (from inadequate drainage of CSF).

DBS is probably a final common manifestation of brain injury caused by a number of different mechanisms. It may, in some cases, be a manifestation of primary brain injury, as when it accompanies large areas of brain contusion or DAI. In

other cases, it probably represents secondary brain injury, caused by hypoxia or hypoperfusion. If left unchecked, the development of DBS can lead to a vicious cycle. That is, the presence of DBS causes an increase in ICP, and then the resulting ischemia leads to the development of more DBS.

Clinical Manifestations

Most patients with DBS are comatose on initial evaluation, sometimes with associated focal neurologic deficits. Rarely, patients with DBS have less impressive symptoms, with more minor neurologic deficits. These patients often experience neurologic deterioration over the ensuing several hours.

Diagnosis

DBS is diagnosed by a head CT scan when there is evidence of smaller ventricles, effacement of the sulci, or obliterated basal cisterns, in the absence of other intracranial pathology that may be exerting significant mass effect (Fig. 105.11). Signs of cerebral edema per se, such as loss of gray–white differentiation, may be present. Other accompanying intracranial lesions, such as diffuse axonal injury, subdural hemorrhage, cerebral contusion, or subarachnoid hemorrhage are often diagnosed as well. In one large study of children with DBS, however, 60% had no other intracranial lesions identified.

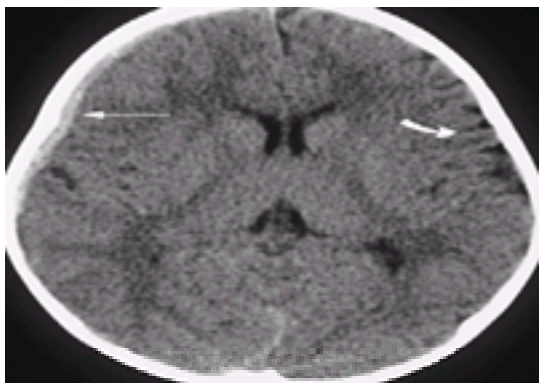

FIGURE 105.11. Brain swelling. This 1-year-old boy fell from a second-story window. The neurologic examination was normal. A head computed tomography (CT) scan shows a small, right-sided subdural hematoma in the temporal region. Note also the effacement of the sulci on the right *(straight arrow*), which can be compared with the normal sulci on the left *(curved arrow).* There is also a mild shift of the midline toward the left.

Management

Patients with DBS need to be admitted to the hospital. Generally, admission to the ICU is required for careful monitoring of hemodynamics, oxygenation and ventilation, and ICP. ICP monitors are indicated for any patient with DBS in coma.

Management of the patient with DBS focuses on optimizing cerebral perfusion and minimizing any stressors that may lead to worsening of the DBS. If the ICP is elevated, measures to control the ICP are required. DBS is often worse between 1 and 3 days after the primary injury occurred, so patients who initially have well-controlled ICP may have more serious difficulties later.

Numerous studies have shown that the outcome of DBS after head trauma is better for children than it is for adults. In one large study, 78% of children with DBS had a functional outcome. Patients with more severe neurologic symptoms on presentation clearly have worse outcomes, as do those with accompanying intracranial lesions, especially subarachnoid hemorrhage or intraventricular hemorrhage. Patients who experience secondary systemic insults (e.g., hypotension, hypoxia) clearly have a worse prognosis as well.

Epidural Hematoma

Pathophysiology

Most epidural hematomas (EDHs) result from blunt impact to the cranium. In most cases of EDH, the skull is fractured, with an associated laceration to the epidural vessels underlying the fracture site. In other cases, there is no fracture, but the deformation of the skull and associated linear deceleration from impact leads to shearing of the epidural arteries or veins.

Many patients with EDH have experienced relatively low-energy mechanisms of injury. In pediatrics, most cases of EDH result from falls, although a minority result from motor vehicle collisions, child abuse, or other mechanisms. About half of pediatric cases of EDH result from falls from heights of 6 feet or less.

Other mechanisms of injury, such as the shaking often implicated in cases of child abuse, are less likely to be associated with EDH because they do not lead to deformation of the skull. On the other hand, because the low impact falls that lead to EDH rarely involve high energy being applied to the brain itself, about 90% of cases EDH have no associated parenchymal injuries.

A small EDH may be asymptomatic. As the EDH expands, however, it begins to occupy an increasingly large intracranial

volume. This increasing mass effect leads to an increase in ICP and, if left unchecked, may result in diffuse secondary brain injury. If the EDH continues to expand, it may lead ultimately to cerebral herniation.

If an EDH can be recognized and surgically drained before this process occurs, secondary brain injury can be prevented. In many cases, the patient has a completely normal neurologic status after the EDH is drained. If the EDH is not drained in time, however, persistent neurologic deficits may result.

Approximately 18 to 36% of patients with EDH have an arterial source of bleeding identified. In most cases, the middle meningeal artery is involved. Another 10 to 20% have bleeding from meningeal veins, the emissary veins, the diploic veins, or from the dural sinuses. Finally, about 30 to 40% have no recognized source of bleeding identified and are probably oozing from small venous sites in the dura. In general, the more severe symptoms are seen in those patients with arterial bleeding, with an intermediate course in those patients with venous sources, and the most benign course in patients with no discrete source identified. Occasionally, patients with venous or oozing EDH are first diagnosed days or even weeks after the injury.

EDH in the occipital and frontal regions may be fairly well tolerated. In contrast, bleeding in the temporal region is more likely to cause symptoms early, because a temporal EDH will more quickly lead to tentorial herniation. Posterior fossa bleeding may also lead to earlier symptoms because of the potential for early compression of vital brainstem structures, as well as the potential for obstruction to CSF outflow and the development of hydrocephalus.

Clinical Symptoms/Signs

The classic presentation of EDH involves an initial loss of consciousness at the moment of impact, the “lucid interval” of several hours after the trauma when the patient is awake and relatively asymptomatic, and then neurologic deterioration as the enlarging hematoma begins to exert its mass effect.

In fact, however, pediatric patients with EDH rarely present with these classic symptoms. In one large series of pediatric patients with EDH, only 20% had an initial loss of consciousness, and 38% were alert with normal neurologic examinations at the time of diagnosis. The most common symptoms of EDH in pediatrics are headache, vomiting, and lethargy. In addition, ataxia may be noted in cases of posterior fossa EDH. Seizures are relatively rare, occurring in less than 10% of cases.

A small number of patients with EDH may not have any symptoms indicative of brain injury. Skull fracture may be a particularly important indicator of the risk of EDH, especially in patients with few other symptoms because skull fracture is noted in 70 to 80% of cases of EDH. Temporal or parietal skull fractures are particularly associated with a risk of EDH.

Diagnosis

EDH can be readily diagnosed by noncontrast CT of the head. The classic appearance on CT is that of a high-density biconvex lesion just subjacent to the skull (Fig. 105.12). The EDH is usually bounded by suture lines but may rarely cross these lines if diastasis of the suture has occurred. EDH is most commonly noted in the parietal, temporal, or temporoparietal regions (approximately 78%), and rarely in the frontal (16%) or occipital (6%) regions. The classic high density appearance on CT indicates clotted blood. Occasionally, an adjacent or intermixed, swirled isodense lesion is noted, which represents ongoing acute bleeding that has yet to clot (Fig. 105.13).

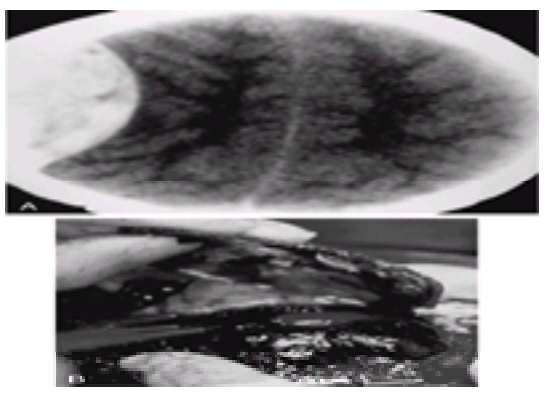

FIGURE 105.12. Epidural hematoma. This 8-year-old boy presented after a sledding accident. He had no loss of consciousness, but he complained of headache and vomiting. A head computed tomography (CT) scan **(A)** shows the classic biconvex hyperdensity of an epidural hematoma. He proceeded to the operating room, where a large mass of clotted blood **(B)** was removed.

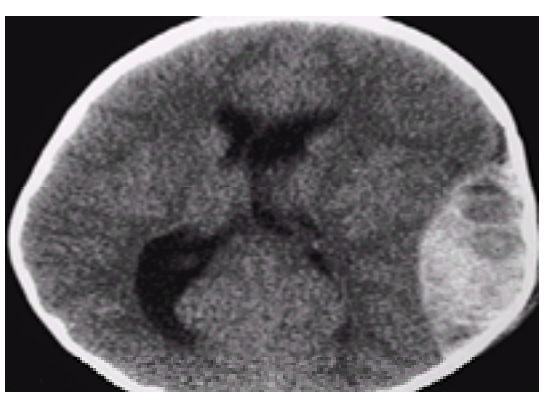

FIGURE 105.13. Epidural hematoma. This 11-month-old boy presented with progressive lethargy and vomiting after falling 2 feet off of a bed. Several hours after the injury, he became unresponsive with dilated, nonreactive pupils. This head computed tomography (CT) scan shows a large epidural hematoma, with intermingled hypodense areas representing active bleeding. Note also the midline shift and the compression of the ipsilateral ventricle. After an emergency craniotomy, the patient made a full recovery.

On CT midline shift, small ventricles, and loss of patency of the basal cisterns indicate a mass effect from the EDH. Signs of herniation may be seen. A careful survey for other associated intradural hematomas or parenchymal injuries should also be undertaken.

Management

The mainstay of treatment for EDH is craniotomy, with drainage of the hematoma and repair of the lacerated epidural vessels. Patients with EDH who have any depression in their level of consciousness, focal neurologic findings, pupillary abnormalities, and/or signs of increased ICP should proceed immediately to surgical intervention.

Increasingly in recent years, however, neurosurgeons have recognized that some patients with EDH may safely be managed with observation. Conservative management is only acceptable for patients with a small EDH (generally less than 30 mL in volume, and with a thickness of less than 2 cm), no focal neurologic deficits, and a normal level of consciousness.

In all cases, however, a neurosurgeon must be consulted immediately because even children who are initially well may experience rapid neurologic deterioration in the first several hours after diagnosis. In one series, 32% of patients who were initially managed conservatively ultimately required surgical drainage of the EDH. Particular caution must be exercised in dealing with those patients who had initial head CT within 6 hours of the trauma because these patients are more likely to have further progression in the size of the EDH. Patients with a temporal EDH, especially if noted to have a fracture overlying the middle meningeal vessels, are at particularly high risk because of the arterial source of their EDH. In addition, patients with posterior fossa EDH are generally not candidates for conservative management because of the high risk of medullary compression and hydrocephalus.

Mortality rates in EDH range from 0 to 10%. Approximately 85% of surviving children with EDH have a good neurologic outcome.

The most important predictor of outcome is the patient's neurologic status on presentation. Patients who present with coma and pupillary abnormalities are much more likely to have sustained secondary brain injury. But even among these patients, a significant percentage will have a good outcome. In one series, 64% of children presenting with an initial Glasgow Coma Scale score of 5 or less had a good neurologic outcome. In another study, 82% of patients with EDH who presented with bilateral nonreactive pupils survived, and 55% had either a good outcome or moderate disability. In general, those patients with EDH who are able to be managed conservatively do well, with a normal neurologic outcome in approximately 95% of cases.

Subdural Hematoma

Pathophysiology

Subdural hematomas (SDHs) result from tearing of the bridging veins that traverse the subdural space. Mechanisms of injury that are associated with shear forces being applied to these veins are especially likely to lead to SDHs. In particular, SDHs result from injuries associated with significant acceleration/deceleration forces.

In older children and adolescents, SDHs most commonly result from motor vehicle collisions. In infants, SDHs are commonly a result of the shaking impact syndrome of child abuse. SDHs may result from falls as well, especially if the fall is from a significant height. Because of the more significant forces applied to the brain in most injuries leading to SDHs, the SDH is often associated with other intracranial lesions. However, infants appear to be prone to the development of small SDHs even after more minor head injury, perhaps because they have wider subarachnoid spaces, which cause the bridging veins to be under more tension and more prone to shear injury.

For some patients with SDH, there is a “lucid interval” of several hours after the initial trauma before the neurologic status begins to deteriorate. For these patients, the mass effect of the accumulating SDH is the cause of significant neurologic morbidity and mortality.

In other patients with SDH, however, serious neurologic symptoms begin at the moment of impact. Many of these patients have cerebral contusions or diffuse brain swelling. For these patients, the SDH may be more of a marker of a high force mechanism of injury than a cause of neurologic injury in itself. Even if the SDH is drained in a timely fashion, therefore, serious brain injury may persist.

Clinical Manifestations

SDHs are often associated with an initial loss of consciousness and with a depressed mental status. Approximately 50% of patients with SDH present in coma. Pupillary abnormalities may be noted as well, indicating impending herniation. In less severely ill patients, headaches, vomiting, lethargy, irritability, visual difficulties, or seizures may be noted. In

patients with SDHs involving the posterior fossa, cerebellar signs such as ataxia or nystagmus may be noted. Because CT scanning is being used more routinely in cases of minor head injuries, many cases of asymptomatic SDH are being discovered as well.

Although most cases of SDH present within hours after the trauma, occasional cases of chronic SDH are diagnosed, days or even weeks after a head trauma. In pediatrics, chronic traumatic SDH is most commonly seen in infants, usually as a consequence of child abuse. Presenting symptoms in these infants may include tense fontanel, macrocephaly, psychomotor retardation, depressed level of consciousness, seizures, vomiting, irritability, or focal neurologic deficits.

Diagnosis

On head CT, acute SDH is seen as a hyperdense crescentic collection of extra-axial fluid (Fig. 105.14). There may be areas of hypodense fluid intermingled, which represent active bleeding, sometimes termed a hyperacute SDH (Fig. 105.14). Because the subdural space is continuous around each hemisphere, subdural blood flows freely through this space, while respecting the midline and tentorial margins (Fig. 105.15 and Fig. 105.16). SDH is usually noted unilaterally, although cases of child abuse are often associated with bilateral SDH. The CT should be evaluated for evidence of mass effect, and for associated intracranial lesions, such as brain swelling, cerebral contusions, or subarachnoid hemorrhage.

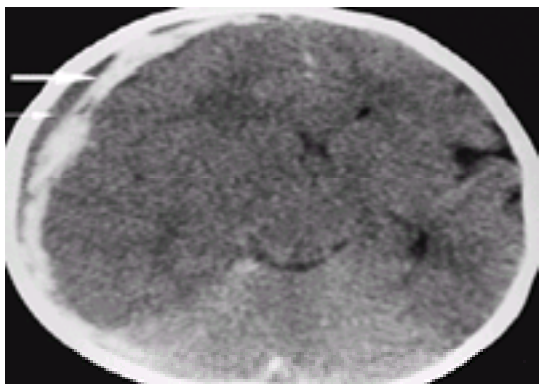

FIGURE 105.14. Subdural hematoma and brain swelling. This 9-month-old boy reportedly became suddenly unresponsive. There was no history of trauma. On examination, he was comatose with fixed dilated pupils and extensor posturing. Massive bilateral retinal hemorrhages were seen. This head computed tomography (CT) scan shows a right-sided subdural hematoma, with acute hyperdense *(thick arrow)* and hyperacute isodense *(thin arrow)* components. Midline shift to the left is noted. There is also evidence of brain swelling, with effacement of the sulci and poor gray–white differentiation. The diffuse hypodensity of the cerebral cortex can be contrasted with the normal density of the cerebellum, which appears whiter in this view. A diagnosis of nonaccidental trauma was made.

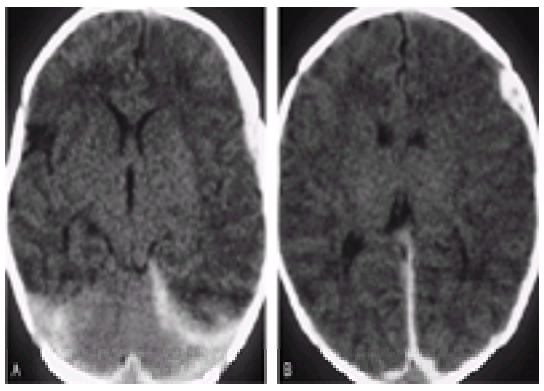

FIGURE 105.15. Subdural hematoma. This 14-day-old patient presented with a complaint of lethargy and vomiting. There was no history of trauma. This head computed tomography (CT) scan shows subdural blood tracking along the tentorium **(A)**, in the interhemispheric fissure **(B)**, and in the left frontoparietal region (**A** and **B**). There is also some effacement of the sulci and loss of gray–white differentiation. Nonaccidental trauma was suspected.

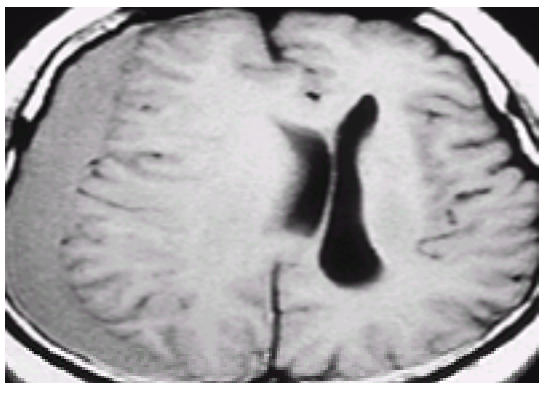

FIGURE 105.16. Subdural hematoma. This T_1-weighted magnetic resonance imaging image shows the classic appearance of a large acute subdural hematoma, with a crescent-shaped extra-axial hematoma that covers the entire right cerebral convexity. There is significant midline shift with compression of the right lateral ventricle and enlargement

of the contralateral ventricle, which probably represents obstruction to cerebrospinal fluid outflow. An emergency operative procedure was performed.

The density of the subdural fluid collection varies over time. With older chronic hematomas, the collection may be almost isodense with CSF.

Management

In the early 1980s a number of researchers were able to show remarkable decreases in mortality from SDH if patients underwent timely surgical drainage. Not surprisingly, this effect was most evident in those patients with large SDHs, associated with midline shift and coma. In this subgroup, the neurologic prognosis is optimized when surgery is performed within 4 hours of the trauma. It appears that early relief of the mass effect in these patients prevents secondary brain injury.

Many patients with SDH who are not so severely ill can be managed nonoperatively. Some authors have proposed nonoperative management for patients with SDH who are not comatose, who have small SDHs with no mass effect, and who have patent basal cisterns. Generally, even well-appearing patients with posterior fossa SDHs are not candidates for nonoperative management because of the high risk of brainstem compression if the lesion expands. Even if a patient appears to be a candidate for nonoperative management, however, immediate consultation with a neurosurgeon is essential because of the potential for rapid clinical deterioration over the first several hours of observation.

Patients presenting with chronic SDH generally do not require craniotomy and decompression. Instead, most centers now manage these patients with a strategy of serial subdural taps or continuous external drainage. In some cases, the SDH is successfully drained in this manner, and no further therapy is necessary. In most cases, eventual placement of a subduroperitoneal shunt for ongoing drainage of the lesion is necessary.

The clinician must recognize the strong association of SDH with child abuse, especially in infants with no clear mechanism of injury reported, and in those patients with associated retinal hemorrhages. If the circumstances of the injury cannot be clearly explained, further evaluation for nonaccidental trauma should be pursued.

In several studies, mortality rates for children with acute SDH range from 10 to 20%. Of those who survive, persistent neurologic sequelae are common. Those patients who are comatose on admission or who have pupillary abnormalities clearly have poorer prognoses. In addition, those patients with CT evidence of more significant brain injury or increased ICP have a worse prognosis. Even the patients who appear to be at high risk, however, may do relatively well. In one study, 14 of 21 patients with a Glasgow Coma Scale score of 8 or less who were less than 40 years of age were functional survivors.

In general, patients who are managed nonoperatively do well, with few if any neurologic sequelae, and with full resolution of the SDH. A small percentage (6% in one series), however, will develop chronic SDH. Chronic SDH is seldom lethal, with a greater than 99% survival rate reported in one large series. Nonetheless, many patients with chronic SDH have ongoing neurologic problems. In one pediatric series, 40% had seizures and 30% had developmental delay.

Subarachnoid Hemorrhage

Pathophysiology

Subarachnoid hemorrhage (SAH) is a common complication of head trauma, especially in more severely injured patients. In one large study SAH occurred in approximately 25% of patients who were comatose on initial evaluation.

SAH results from tearing of the small vessels of the pia mater. SAH generally occurs after relatively severe blunt trauma to the head, or as a result of significant shear forces.

Because the cerebral subarachnoid space is large and freely communicates with the basal cisterns and the spinal subarachnoid space, the blood in an SAH can be distributed widely. As a consequence of this wide distribution and because it is generally smaller pial vessels that bleed, SAH rarely accumulates to the extent that it causes clinically important mass effect.

SAH appears to exert its main pathophysiologic effect by causing cerebral vasospasm. A number of studies have shown that SAH is associated with increased cerebrovascular resistance, and consequently, with an increased risk of cerebral ischemia or infarction. In general, SAH is seen in association with other intracranial injuries, especially SDH, cerebral contusion, and intraparenchymal hemorrhage. Because SAH is often seen in association with other intracranial injuries, its presence may be most important as a marker for severe primary brain injury, rather than as a cause of secondary injury in itself.

Because of the presence of the blood in the subarachnoid space, and probably because of associated release of inflammatory mediators from adjacent cells of the pia and arachnoid mater, SAH causes meningeal irritation and clinically mimics many of the symptoms and signs of meningitis.

Clinical Manifestations

Patients with posttraumatic SAH often have other intracranial hemorrhage or parenchymal injuries as well, so they may

present with a wide range of symptoms, from those who have minimal if any symptoms to those who are comatose with signs of impending cerebral herniation. As an isolated intracranial lesion, SAH most commonly causes headache and other signs of meningeal irritation, such as nausea and vomiting, nuchal rigidity, and photophobia. Patients with isolated SAH often have a history of loss of consciousness, and sometimes present with depressed mental status or even coma. Seizures are reported in 2 to 10% of cases. Subhyaloid or preretinal hemorrhages, located just adjacent to the optic nerve head, may be seen with SAH as well.

Diagnosis

SAH can usually be detected on noncontrast head CT as a collection of hyperdense fluid in the CSF spaces, either in the subarachnoid space overlying the cerebral convexity or in the basal cisterns. Subarachnoid blood overlying the cerebral hemisphere can be distinguished from SDH in that the subarachnoid blood may flow into the depths of the brain sulci, fissures, and cisterns, whereas the subdural space does not penetrate into these depths. Head CT imaging has a sensitivity of only about 90% for detecting SAH, with a lower sensitivity for patients seen later than 24 hours after the SAH began. However, lumbar puncture is not recommended and may be dangerous in the setting of focal intracranial pathology or brain swelling because it may increase the risk of cerebral herniation.

Management

All patients with traumatic SAH should be admitted to the hospital for observation. In most cases, specific therapy directed at the SAH is not required. The general principles of management for patients with head injuries should, of course, be followed. Prophylactic anticonvulsants are often used for patients with SAH.

In one study, 24% of patients with traumatic SAH died, and another 24% had a poor neurologic outcome. Patients with associated intracranial injuries and those with a larger SAH (especially if it involves both the hemispheric convexities and the basal cisterns) have the worst outcome. Patients with minor or no symptoms and small SAH generally do well.

Penetrating Trauma

Penetrating trauma includes injury from sharp objects, such as knives, darts, or animal bites, and from missiles, usually bullets. Penetrating head trauma is far less common than blunt head trauma in pediatrics, but the incidence of penetrating injuries, especially from gun shots, has increased at an alarming rate in the past decade. Several urban hospitals have reported that the rate of hospital admissions for teenagers shot in the head increased by as much as tenfold between the 1980s and 1990s.

Pathophysiology

Penetrating head trauma leads to brain injury by several mechanisms. First, there is direct injury along the path of the penetrating object, with laceration or contusion of neural tissue, and hemorrhage from injured vessels. For low-velocity penetrating injuries (e.g., knife wounds), this may be the primary source of brain injury.

Higher-velocity injuries (from bullets) also cause significant damage because of the shock waves created by the impact of the penetrating object. These shock waves can cause contusions or vascular injury at sites that had no contact with the penetrating object itself.

Vascular injury may result from direct laceration, or from percussion-related damage. Dissection of an intimal flap, thrombosis of the vessel lumen, or aneurysm formation may result. Ischemia or infarction resulting from these vascular injuries may occur immediately after impact, or they may occur days or even weeks later.

Because the skull and dura are violated by the penetrating object, there is direct communication from the CSF spaces or brain to the outside world, and consequently, a risk for intracranial infection. Penetrating objects that pass through the paranasal sinuses or mastoid air cells particularly increase the risk of intracranial infection.

Clinical Manifestations

Patients with penetrating injuries sometimes may present without a clear history, either because the injury was not witnessed or because the patient and/or witnesses fear the repercussions of full disclosure.

The local signs of penetrating injury can sometimes be subtle, especially if the penetrated area is covered by hair or by a dressing. Without careful exploration, the entrance wound might be mistaken for a superficial scalp laceration. In more obvious cases of penetrating injury, meningeal or parenchymal tissue may be visualized in the wound, or CSF may be oozing.

Signs of neurologic injury are often severe for patients with high-velocity injuries, but they may be more subtle in patients with low-velocity injuries. Patients with progressively enlarging intracranial hematomas or worsening brain swelling may deteriorate quickly over the several hours after presentation.

Management

The initial management of the patient who has sustained penetrating head trauma focuses on the ABCs of resuscitation, as outlined previously. Once the ABCs have been addressed, attention can be focused on the possible need for brain-specific therapies, as described.

In addition to the management priorities already outlined, some unique aspects of caring for the patient with penetrating

head trauma should be discussed. The patient should be examined carefully for evidence of entrance or exit wounds. The clinician should recognize the potential for multiple penetrating wounds, or for exit wounds in unpredicted locations because of a complicated migratory path of the penetrating object.

Any bleeding lacerations should be occluded with direct pressure. Occasionally, immediate suturing of the laceration may be necessary to achieve hemostasis. Other penetrating wounds may simply be covered with a sterile gauze dressing, until more definitive debridement and repair can be achieved. Any penetrating objects still in place should not be removed, because of the potential for serious hemorrhage if the object is tamponading a lacerated vessel.

As for all patients with head injuries, cervical spine precautions should be maintained until the clinician is certain that no cervical spine injury has occurred, usually requiring cervical-spine radiographs in addition to a thorough physical examination. In particular, the clinician should recognize the possibility that the bullet or penetrating object may have directly penetrated the neck and injured the vertebral column.

Patients with penetrating head injuries generally require prophylactic antibiotic therapy. A first-generation cephalosporin such as cefazolin (30 mg/kg intravenously; maximum dose 2 g) is usually appropriate.

Most patients with penetrating head injuries are started on prophylactic anticonvulsant therapy (usually with phenytoin), especially if there is any concern about parenchymal injury or subarachnoid hemorrhage.

Patients with penetrating injuries to the head require immediate head CT to delineate the extent of brain injury and to assess for associated intracranial hematomas, brain swelling, and/or mass effect. The head CT is also able to demonstrate the presence of intracranial foreign material.

As with blunt trauma patients, all patients with intracranial hematomas exerting any mass effect will need an immediate operation to evacuate the hematoma. Patients with large areas of contusion exerting a significant mass effect may require surgical resection as well.

In addition, most patients with penetrating injuries to the head require prompt operation to debride the infected or contused brain tissue at the entry site. Controversy exists about the need for extensive debridement of deeper tissues along the path of the penetrating object. Increasingly, published reports have shown equally high success rates with more limited debridement. After debridement, the dura must be repaired to achieve a watertight seal.

In general, it is unnecessary to remove deeply embedded foreign material (e.g., bullets), because the risk of infection does not seem to increase when these objects are left in place. Bullets that are lodged in the ventricular system usually are removed, however, because of the potential for outflow obstruction and hydrocephalus if the bullet migrates.

All patients with penetrating head injury require hospitalization. Except in circumstances of superficial penetration with no underlying brain injury, ICU level monitoring is indicated. Intracranial ICP monitoring is indicated for all patients in coma.

Most patients with penetrating head injuries need to have angiography performed (either conventional or MRI angiography) to exclude the possibility of traumatic injuries to the cerebral vasculature. Although some controversy exists about the optimal timing of angiography, most authors recommend angiography as soon as it can safely be performed.

The prognosis after penetrating head injury depends most on the level of neurologic function at the time of presentation. For patients with severe neurologic dysfunction (Glasgow Coma Scale scores in the range of 3 to 5), the likelihood of a good functional outcome is low, although occasional patients will do well. Most neurosurgeons will not operate on patients who present with an absence of neurologic function (Glasgow Coma Scale score of 3 and nonreactive pupils) because the prognosis is so dismal for this subgroup.

For patients with a better neurologic status, the prognosis is influenced by the degree of tissue damage seen on a head CT scan. Tracks of injury that cross the midline or that involve the ventricles are associated with a worse prognosis, presumably because more brain tissue is injured. Patients with subarachnoid hemorrhage also have a worse prognosis.

SPINAL CORD TRAUMA

Spinal cord injury is rare in pediatrics. Most case series in the literature, from pediatric trauma centers, report only 1 to 2 patients with spinal cord injuries each year. Nonetheless, spinal cord injuries, when they do occur, are associated with significant morbidity and mortality, and the consequences of missing patients with early signs of spinal cord injury can be devastating. Furthermore, recent clinical evidence suggests that prompt diagnosis and therapy for spinal cord injuries may improve the prognosis.

Anatomy

The spinal cord runs from the foramen magnum, where it extends from the base of the medulla, to its distal tip, in the upper lumbar region of the spine. The cord consists of an H-shaped central section of gray matter, and surrounding white matter which is divided by this “H” into a ventral (motor) compartment, and dorsal and lateral (sensory) compartments.

Upper motor neurons originate on one side of the cerebral cortex, but then cross to the other side at the level of the medulla before entering the spinal cord. Axons from motor neurons in the left cerebral cortex, therefore, run in the right-sided white matter of the spinal cord and serve the right side of the body. Sensory neurons, in contrast, originate on one side at the level of the dorsal root ganglion and then cross immediately to the other side (at the level of the spinal nerve roots) before entering the cord. Sensory impulses from the right side of the body, therefore, run in the left-sided

white matter of the spinal cord, before reaching the left cerebral cortex.

The spinal cord is surrounded by three layers of meningeal tissue, which are continuous with the meninges surrounding the brain. As with the cerebral meninges, blood could potentially accumulate in the spaces between these layers of tissue, leading to spinal subdural or epidural hematomas.

The spinal cord runs in the vertebral canal, protected by the bones of the spinal column, with the vertebral bodies anteriorly and the vertebral arches laterally and posteriorly. Injuries to the spinal cord generally involve some injury to the surrounding spinal column. Because the spinal cord is significantly shorter than the spinal column, injuries at a certain level of the spinal column are associated with spinal cord injuries corresponding to a lower level. A low thoracic spinal column injury, for instance, may be associated with neurologic deficits corresponding to the lumbar cord.

Some unique features of the pediatric spine make spinal cord injury more likely in children. Specifically, the ligaments of the pediatric spine are especially lax, allowing more movement, or subluxation, of vertebrae on one another. In addition, the paraspinous musculature, which provides stabilization and support to the adult spine, is less developed in children. Decreased ossification of the spine, which progresses throughout childhood, also makes the cervical spine more pliable. Furthermore, the facet joints between adjacent vertebral bodies are more flattened or horizontal in pediatric patients, which makes subluxation more likely.

Finally, it should be noted that there is excess strain on the cervical spine, especially the upper cervical spine, in young children, because the head is a proportionately larger component of total body weight. The fulcrum of movement of the cervical spine is at the level of C2–C3 in young children, as opposed to C5–C7 in older children and adults. This feature makes the upper cervical spine especially prone to injury in young children. These “immature” features of the pediatric spine persist to around the age of 8 years. For older children, the anatomy of the spine is fairly similar to that seen in adult patients.

Pathophysiology

As with the pathophysiology of brain injury, injuries to the spinal cord can be considered to occur in two phases: primary and secondary cord injury. Primary spinal cord injury refers to the irreversible neural damage initiated at the time of traumatic impact. Secondary spinal cord injury refers to the associated pathophysiologic processes that occur hours to days later, damaging neurons not necessarily injured by the primary impact itself.

Primary injuries to the spinal cord may result from several different mechanisms. Rarely, direct transection of the cord can occur, as a result of penetrating injuries to the spine or, more commonly, from bone fragments displaced after fracture or subluxation. More common are bruises or contusions of the spinal cord, which result from compression of the cord by subluxated bone or herniated intervertebral disks. Spinal cord injuries may also result from the application of shear forces to the cord, as when the spine is hyperflexed, hyperextended, or distracted during blunt trauma. The flexible spinal column of the young child makes these types of shear injuries especially common. Finally the spinal cord can be injured if its vascular supply is disrupted, leading to ischemia and infarction of the cord, sometimes in the absence of any direct traumatic force being applied to the cord itself.

Secondary pathophysiologic changes are thought to cause much of the clinical disease noted after spinal cord injury. One important source of secondary injury is lipid peroxidation, which leads to autodestruction of cell membranes in areas of injured spinal cord. This lipid peroxidation process may be initiated by the mechanical trauma itself, or by free radicals liberated during the inflammatory response. Once this process begins, it leads to the development of more inflammation, with further cell injury resulting. Much of the research on acute spinal cord injury in the past decade has focused on efforts to interrupt this injury cascade.

Secondary pathophysiologic changes may also result from the local mass effect caused by subdural or epidural hematoma, or from the edema associated with a contused area of cord. Secondary spinal cord injury can also be initiated or exacerbated by any systemic process that leads to hypoxia or ischemia. Areas of spinal cord that are already contused, or partially compressed by local bleeding or edema, may be at especially high risk for ischemic injury.

Clinical Manifestations

Spinal cord injuries are generally associated with significant mechanisms of injury, such as motor vehicle collisions, falls from significant heights, or child abuse. Patients with injuries to the spinal cord often have evidence of injuries to other organ systems. In particular, there is a high association between head injuries and injuries to the cervical spine and/or spinal cord. Injuries to the thoracic or lumbar spine may also be associated with head injuries, but they are seen more often in the setting of chest or abdominal trauma.

Patients with high cervical cord injuries may sometimes have abnormal vital signs, reflecting an interruption of autonomic impulses to the heart and the vasculature. These patients demonstrate bradycardia and hypotension, along with peripheral vasodilation, a syndrome known as spinal shock. They may also have abnormal or absent respiratory effort. Because most trauma patients with hypotension are hypovolemic and have a reflex tachycardia, those with bradycardia should be strongly suspected of having spinal shock.

Spinal cord injuries should also be suspected in any traumatized patient who complains of decreased motor strength, or in whom focal deficits in strength or tone are noted on examination. In the acute setting, severe spinal cord injuries are usually associated with decreased or absent reflexes. Partial injuries to the cord, on the other hand, may be associated with initial hypertonia and hyperreflexia. Abnormalities of bladder control and rectal tone may also be noted.

Motor deficits correspond to the spinal roots whose neural impulses are compromised by the spinal cord injury. Most typically, all motor impulses that originate from spinal nerve roots at or below the level of the spinal cord injury are

affected. An understanding of the motor deficits after spinal cord injury requires knowledge of the innervation of the important muscle groups of the body. A list of the important muscle groups and the spinal roots that serve them is presented in Table 105.2.

Muscle	Segmental Innervation	Peripheral Nerve
Diaphragm	C3-C5	Phrenic nerve
Trapezius	C3-C4	Spinal accessory nerve
Deltoid	C5-C6	Axillary nerve
Supraspinatus	C5-C6	Suprascapular nerve
Biceps brachii	C5-C6	Musculocutaneous nerve
Triceps brachii	C6-C8	Radial nerve
Wrist extensors	C6-C7	Radial nerve
Finger extensors	C6-C8	Radial nerve
Wrist flexors	C6, C7-T1	Ulnar, median nerve
Intrinsic hand muscles	C8-T1	Ulnar nerve
Psoas	L1-L2	Psoas nerve
Quadriceps femoris	L2-L4	Femoral nerve
Gastrocnemius	L5-S1	Deep peroneal nerve
Urinary bladder	S2-S4	—

Table 105.2. Major Muscle Groups, Listed with the Spinal Roots and Peripheral Nerves That Supply Them

Sensory deficits may be noted as well, and these may range from paresthesias to complete loss of sensation. Because sensory impulses are carried in both the dorsal columns and the lateral compartments of the spinal cord, injuries to one of these compartments may lead to partial sensory deficits (e.g., loss of pain and temperature sense from the lateral compartment or loss of joint position sense and vibration sense from the dorsal column), but with other forms of sensation intact for the same body part. A diagram of the body's sensory dermatomes is shown in the Appendix (see Appendix D, Sensory Nerve Dermatomes). Often, a well-demarcated sensory “level” of the spinal cord can be identified, below which sensory impulses are absent, and above which sensation is intact.

Many injuries to the spinal cord involve solely or predominantly one of the two lateral sides of the cord. Because of the distribution of sensory and motor neurons in the spinal cord, a lesion to the left spinal cord affects left-sided motor strength but right-sided sensation. This classic crossed pattern of sensory and motor deficits is known as the Brown-Sequard syndrome.

Other patterns of neurologic deficits may also be noted. Partial injuries to the spinal cord may result in partial deficits. In some cases of ventral cord injury, for instance, only motor deficits may be observed. Cases of hyperextension injury may cause more severe injury to the central, or deep regions of the cord (the gray matter) while sparing the more superficial white matter. This leads to a “paradoxical” pattern of symptoms known as the central cord syndrome, in which the more distal function (served by the white matter) is spared, but more proximal function (served by gray matter) is compromised. Finally, occasional patients with more minor injuries to the spinal cord report transient symptoms of paresthesias, numbness, or weakness that may have resolved by the time of evaluation.

Patients with associated head injury may be obtunded and therefore unable to report symptoms of spinal cord injury. However, even in comatose patients, asymmetric motor tone, strength, or reflexes may be noted. Abnormalities of posture or tone may be a clue to the presence of spinal cord injury in these patients. In patients with injuries at the level of C-6, for instance, biceps function (with impulses from nerve root C-5) is intact, but triceps function (impulses from C-6) is not, and the elbow is held in tonic flexion.

Patients with injury to the spinal column are at risk for spinal cord injury even if no such injury has occurred at the time of evaluation. Therefore, children with signs of spinal injury, such as pain, tenderness, decreased range of motion, or deformity of the back or neck must be treated with the utmost caution.

Management

As with all trauma patients, care for the patient with spinal cord injury begins with the ABCs of resuscitation. This initial resuscitation must be accomplished with meticulous attention to the stabilization of the spine. For older children and adolescents, a semirigid cervical collar or manual in-line stabilization should be used. For infants, a semirigid cervical collar might actually be too large and may lead to distraction or hyperextension, which could be deleterious. For some infants, therefore, it may be preferable to immobilize with sandbags on the sides of the head, without using a cervical collar. In addition, the patient should be maintained in a supine position on a backboard so that no undue manipulation of the spine occurs. Because infants have a relatively large occiput, supine positioning on a flat surface may result in flexion of the neck. Proper neutral positioning for these young patients may require that the occiput be allowed to rest at a level slightly lower than the shoulders. Any transfers of the patient from one bed to another or “log-rolling” of the patient to examine the back should be done with careful attention to maintaining neutral positioning at all times.

The patient with spinal cord injury requires adequate oxygenation, ventilation, and perfusion. Careful attention should be given to positioning of the airway (using the jaw-thrust, rather than the chin-lift maneuver), suctioning if needed, and supplemental oxygenation. The head should never be turned in efforts to clear secretions from the oropharynx; if the patient needs to be turned, a log-roll maneuver should be used. Patients with inadequate respiratory effort require positive pressure ventilation. Patients with coma, inadequate respiratory effort, or inadequate airway protective reflexes need intubation with mechanical ventilation. When possible, rapid sequence intubation (as previously described; also see Chapter 5) should be performed. If sedating or paralytic agents are to be used, a brief neurologic assessment (see the following) should precede the administration of the drug if time allows.

The circulatory status of patients with spinal cord injury may be impaired if there are other organ system injuries leading

to hemorrhage and hypovolemia. Fluid and blood product resuscitation should be initiated in the usual fashion (see Chapter 104), and the definitive treatment of the hemorrhagic injuries should be pursued. Rare patients with spinal cord injuries have signs of spinal shock, with bradycardia and hypotension, and signs of peripheral vasodilation. These patients may require the use of pressor agents, such as dopamine, epinephrine, or phenylephrine, to maintain adequate vascular tone.

As soon as possible after an injury, the patient's neurologic status should be assessed and recorded so that any early progression of neurologic symptoms can be noted and so that no undue concerns are raised that the injuries were a result of the emergency care provided. For conscious, cooperative patients, the neurologic examination should include a test of motor strength in all four extremities, tone in all extremities, and deep tendon reflexes. Rectal tone should also be assessed. The sensory examination, assessing for light touch, pain sensation (as from a pinprick), and joint position sense (of fingers and toes) should be assessed as well. For patients with depressed consciousness, an assessment of tone and reflexes may be all that is possible.

Patients with suspected spinal cord injury should have plain radiographs of the spine to evaluate for fractures or subluxations. However, the absence of abnormalities on radiographic imaging does not eliminate the possibility of spinal cord injury. The syndrome of spinal cord injury without radiographic abnormalities (SCIWORA) is well reported in the literature. Several series of children with spinal cord injury report that 15 to 20% of cases may be classified as SCIWORA. Children have an especially high risk for SCIWORA because of the flexibility of the pediatric spinal column, which allows the spinal cord to withstand shear or compressive forces without necessarily causing a fracture. In some cases, the flexible and lax spinal column may sublux transiently, causing a compressive injury to the cord and then reduce back into normal position before radiographs are obtained.

In recent years, MRI has become a mainstay for the diagnostic evaluation of patients with suspected spinal cord injury. MRI offers the advantage of providing good detail of the soft tissue of the spinal cord itself, which cannot be well imaged by plain radiographs or by CT. A number of studies in the literature have reported evidence of spinal cord contusion, transection, or hemorrhage seen on MRI in most patients with spinal cord injury, and especially in those with more severe and lasting deficits. Increasingly, MRI has been used to document spinal cord abnormalities in cases that would be classified as SCIWORA by plain radiographs and CT.

MRI should be performed as soon as possible for patients with progressive neurologic deficits, who may have extra-axial mass lesions compressing the spinal cord, such as subdural or epidural hemorrhage, or a herniated intervertebral disk. In other cases, MRI may provide useful diagnostic and perhaps prognostic information, but it is less likely to alter the acute management.

Specific therapy directed at the spinal cord focuses on the prevention of secondary cord injury. The mainstay of this therapy is careful immobilization, as outlined previously. In all cases of spinal cord injury, a neurosurgeon should be consulted immediately. If compressive spinal cord lesions are noted, especially with incomplete but progressing neurologic injury, emergent laminectomy with surgical evacuation of the lesion may be necessary. Displaced fractures or subluxations of the spinal column require immobilization and generally some form of traction (e.g., a halo brace, skull tongs) to reduce them and maintain stability (see Chapter 106 for more details). Some patients with irreducible subluxations or unstable fractures require urgent surgery to achieve reduction. Patients with SCIWORA usually require long-term immobilization as well because they should be presumed to have some ligamentous instability of the spine, even if the initial radiographic studies show normal alignment.

High-dose corticosteroid therapy is widely used in the setting of spinal cord injury. It is believed that corticosteroid therapy decreases the extent of secondary cord injury by interfering with the lipid peroxidation pathway. The use of corticosteroids is based on the results of a multicenter prospective, randomized trial known as NASCIS (the National Acute Spinal Cord Injury Study). The NASCIS found that high-dose corticosteroid therapy significantly improved the outcome for those patients treated within 8 hours of injury, but not in those patients treated later. On the basis of these findings, the following regimen is recommended for patients with neurologic deficits attributable to spinal cord injury who are seen within 8 hours of injury: methylprednisolone at an initial IV bolus dose of 30 mg/kg followed by an infusion of methylprednisolone at 5.4 mg/kg per hr for the subsequent 23 hours.

All patients with spinal cord injury need to be admitted to the hospital for careful observation, immobilization, and specific therapy as outlined already. For patients with persistent deficits, a long-term plan for rehabilitation and ongoing medical care will need to be developed.

The prognosis after spinal cord injury depends most on the severity of disease at presentation. Patients with complete loss of function below the injured level clearly have the worse prognosis. On the other hand, patients with partial injuries often have significant improvements. MRI findings are of useful prognostic value as well. Those patients with documented transection of the cord clearly have little hope of recovery. In contrast, patients with minor or no abnormalities on MRI generally do well.

Pediatric patients with spinal cord injuries tend to fare better than their adult counterparts. In a few large studies, most pediatric patients with some useful motor function at the time of presentation regained full function of the compromised motor groups. Those patients with no motor function distal to the site of injury usually have some permanent disability, although they still often have some improvement after their initial presentation.

Penetrating Spinal Cord Trauma

Penetrating spinal cord trauma is an especially rare phenomenon in pediatrics. It may occur as a result of violent injuries, from stabbing or from gunshot wounds. Accidental injuries may occur, most commonly from shards of glass that penetrate into the spinal column.

Stab wounds or penetrating sharp foreign bodies usually involve penetration from the posterolateral aspects of the neck. These injuries generally lead to hemisection of the cord, with only one side of the cord affected. This predilection for unilateral injury probably reflects the fact that the posterior spinous processes and lateral transverse processes form an anatomic “gutter” through which the penetrating object is guided, thereby offering some protection to the opposite side of the cord. Bullets, on the other hand, propelled by high energy forces, may penetrate the bones of the spinal column and cause less predictable patterns of injury.

Some cases of penetrating spinal cord injury may not be obvious on presentation. Gunshot wounds to the head, for instance, may involve migration of the bullet to the level of the spinal cord, even if the initial trajectory of the bullet might not have suggested cord involvement. Some cases of stab wounds or penetrating glass present with what appear to be innocent lacerations of the neck or back.

When penetrating spinal cord injury is suspected, the principles of management, as outlined previously, should be followed. Plain radiographs demonstrate the presence of many radiopaque foreign bodies; in some cases, CT may be required to demonstrate less radiodense materials. In cases in which no foreign body is left in the wound, plain radiographs and CT may be normal. MRI is the best imaging modality for delineating injury to the cord itself.

A neurosurgeon should be consulted in all cases of penetrating cord injury. In some cases, surgical removal of intraspinal foreign bodies, bone fragments, or expanding hematomas is indicated. Surgical repair of ongoing CSF leak from the site of injury may also be indicated. In all cases, appropriate immobilization is necessary. Prophylactic antibiotic therapy is recommended for penetrating spinal cord injury as well. Generally, a first-generation cephalosporin such as cefazolin (30 mg/kg intravenously; maximum dose 2 g) is preferred.

PERIPHERAL NERVE INJURIES

Pathophysiology

Peripheral nerve injuries in pediatrics usually involve the extremities, most commonly the hand and upper extremity. Most peripheral nerve injuries in pediatrics result from acute traumatic insults. Transection of the nerve may result from deep soft-tissue lacerations or from severe crush injuries. Rarely, transection of a nerve may also result from a fracture, with laceration of the nerve by a displaced bony fragment.

More commonly, however, displaced fractures or dislocations lead to reversible compression injuries to the nerve. Nerve compression may also occur in the absence of acute trauma, usually because of tight anatomic compartments that exert constant pressure on the nerve (carpal tunnel syndrome is one common example).

Peripheral nerve injuries may be graded in terms of the severity of the clinical course. The mildest form of nerve injury is known as neurapraxia, which refers to nerve conduction impairment without structural injury to the axon itself. Neurapraxia commonly results from a situation of transient compression or ischemia, as when a patient complains that a limb has “fallen asleep.” The numbness and paresthesias reflect a rapidly reversible physiologic conduction block. If biopsy of the nerve were performed in this situation, no histologic abnormalities would be expected.

More severe cases of neurapraxia may be associated with symptoms that persist for as long as several months. In these cases, histologic examination reveals focal demyelination in the injured area of the nerve, but with no injury to the axon itself. In general, as long as the axon itself is not injured, full recovery can be expected.

Axonotmesis is a more severe injury to peripheral nerve, involving injury to the axon itself, but with preservation of the surrounding connective tissue of the nerve sheath. Axonotmesis generally results from crush injuries to the nerve. Good recovery of peripheral nerve function is likely to occur, although it will progress slowly, with lengthening of the nerve axon from its proximal stump progressing at a rate of approximately 1 to 4 mm/day. Distal nerve lesions, in close proximity to the target muscles or sensory regions, are associated with earlier and more complete recovery of function than in more proximal lesions.

The most severe form of nerve injury is known as neurotmesis, which involves injury both to the nerve axon and to the surrounding connective tissue. Neurotmesis usually results from direct laceration to the nerve or, rarely, from severe crush injuries. Because there is no intact nerve sheath to guide the development of the regenerating proximal nerve, spontaneous recovery of function is unlikely, and surgical repair is required.

Clinical Manifestations

Significant peripheral nerve injuries usually are seen in association with other obvious signs of traumatic injury, such as a soft-tissue laceration, crush injuries to the extremity, or fracture. In some cases, however, such as with repetitive microtrauma or with anatomic compressive lesions (as seen in carpal tunnel syndrome), the symptoms of peripheral nerve injury may be the primary complaint.

The cardinal symptoms of peripheral nerve injury are disturbances of sensory or motor function in the distribution of the nerve. Appropriate diagnosis of peripheral nerve injury requires an understanding of the anatomic distribution of sensory and motor function of the major peripheral nerves. A full description of this clinical anatomy is beyond the scope of this discussion, but a summary of the motor functions of the major peripheral nerves is presented in Table 105.2.

Disturbances of sensation may include paresthesias, pain (which may be described as sharp, burning, or stabbing), or numbness. In some cases, the patient may not report a sensory deficit, but sensory abnormalities are noted on examination. A gross assessment of sensory function can be obtained simply by testing the patient's ability to recognize

light touch stimuli in the distribution of the nerve in question.

A more sensitive test for identifying disturbances in sensory function is two-point discrimination. Although instruments for assessing two-point discrimination are commercially available, in common practice, a paper clip is often used. The paper clip should be unfolded so that the two ends are in close proximity, with a distance of approximately 1 cm in between. The patient should then be briefly trained on an uninjured part of the body to differentiate between being touched with one or two points of the paper clip simultaneously. Once it is determined that the patient can reliably perform the task, the injured area should be assessed.

The two points must touch the skin simultaneously, and they must occur in the same axial line. If the patient is able to successfully discriminate one- and two-point stimuli, the distance between the two ends of the paper clip can be decreased successively to find the patient's threshold for discrimination. A hand with normal sensation should be able to distinguish between two points that are 2 to 5 mm apart at the fingertips, 7 to 10 mm at the base of the palm, and 7 to 12 mm on the dorsum of the hand. More proximal parts of the upper extremity may have even less sensitive discriminatory abilities.

A problem occasionally arises in trying to assess sensory function in a patient who is unresponsive or who cannot communicate with the examiner. In these cases, a test of sympathetic innervation, such as the O'Riain wrinkle test, may be useful. To perform this test, the patient's hand is immersed in a warm water bath for approximately 20 minutes. Normal digital pulps will wrinkle; fingers with disrupted sympathetic innervation will not wrinkle.

Appropriate motor testing of the peripheral nerves depends on careful isolation of muscle activity that reflects the peripheral nerve in question. The clinician must be careful to recognize that a patient will compensate for a motor deficit by using other motor groups to accomplish the same task. Motor function can be assessed by examining not only active motor strength but also resting tone and, for more chronic injuries, muscle bulk.

Management

Clean lacerations to a primary nerve are often repaired primarily. There is some evidence to indicate that recovery of function is better if the nerve is repaired within 48 hours of injury. Crush injuries to peripheral nerves are often repaired secondarily, several days or weeks after the injury. For these cases, many surgeons believe that delayed repair allows better debridement of devitalized tissue and easier identification of injured nerve tissue that needs to be resected. In all cases in which transection of a peripheral nerve is suspected, prompt consultation with an appropriate surgical consultant is indicated so that a decision can be made about the appropriate timing of repair.

Injuries to peripheral nerve associated with fracture or dislocation generally improve after the orthopedic injury is reduced. If there is any question about the recovery after reduction, nerve function should be carefully followed.

Nerve compression syndromes not associated with acute traumatic injuries can generally be treated with rest and nonsteroidal antiinflammatory medications. Splinting may be indicated for some syndromes as well. In all cases, appropriate follow-up should be arranged so that the patient can be referred for further interventions if necessary.

The prognosis after peripheral nerve injuries clearly depends on the severity of the injury. Compressive lesions without transection of the nerve have a better prognosis, with full recovery expected in all but the most severe or most chronic cases. With transections of the nerve, however, there is often some degree of permanent disability. In general, those lesions resulting from clean lacerations that can be repaired primarily have a somewhat better prognosis. Children, in general, have a better prognosis after peripheral nerve injury than do adults. In some cases, remarkable recoveries after severe crush injuries to the nerve have been reported.

Suggested Readings

Bezircioglu H, Ersahin Y, Demircivi F, et al. Nonoperative treatment of acute extradural hematomas: analysis of 80 cases. J Trauma 1996;41:696–698.

Bracken M, Sheaprd M, Collins W, et al. Methylprednisolone or naloxone treatment after acute spinal cord injury: 1-year follow-up data. J Neurosurg 1992;76:26–31.

Bullock R, Chesnut R, Clifton G, et al. Guidelines for the management of severe head injury. New York: Brain Trauma Foundation, 1996.

Bullock R, Golek J, Blake G. Traumatic intracerebral hematoma—which patients should undergo surgical evacuation? CT scan features and ICP monitoring as a basis for decision making. Surg Neurol 1989;32:181–187.

Centers for Disease Control and Prevention. Sports-related recurrent brain injuries—United States. MMWR 1997;46:224–227.

Domenicucci M, Signorini P, Strzelecki J, et al. Delayed post-traumatic epidural hematoma. A review. Neurosurg Rev 1995;18:109–122.

Duhaime A. Exciting your neurons to death: can we prevent cell loss after brain injury? Pediatr Neurosurg 1994;21:117–123.

Duhaime AC AJ, Lewander J, Schut L, et al. Head injury in very young children: mechanisms, injury types, and ophthalmologic findings in 100 hospitalized patients younger than 2 years of age. Pediatrics 1992;1992:179–185.

Eckstein M. The prehospital and emergency department management of penetrating head injuries. Neurosurg Clin North Am 1995;6:741–751.

Ersahin Y, Mutluer S, Mirzai H, et al. Pediatric depressed skull fractures: analysis of 530 cases. Childs Nerv Syst 1996;12:323–331.

Fortune J, Feustel P, Graca L, et al. Effect of hyperventilation, mannitol, and ventriculostomy drainage on cerebral blood flow after head injury. J Trauma 1995;39:1091–1099.

Greene K, Marciano F, Johnson B, et al. Impact of traumatic subarachnoid hemorrhage on outcome in nonpenetrating head injury. J Neurosurg

1995;83:45–52.

Greenes DS, Schutzman SA. Isolated skull fractures in infants: what are their clinical characteristics, and do they require hospitalization? Ann Emerg Med 1997;30:253–259.

Greenwald B, Ghajar J, Notterman D. Critical care of children with acute brain injury. Adv Pediatr 1995;42:47–89.

Gruen J, Weiss M. Management of complicated neurologic injuries. Surg Clin North Am 1996;76:905–922.

Hadley M, Zabramski J, Browner C, et al. Pediatric spinal trauma: review of 122 cases of spinal cord and vertebral column injuries. J Neurosurg 1988;68:18–24.

Hainline B. Nerve injuries. Med Clin North Am 1994;78:327–343.

Kadish H, Schunk J. Pediatric basilar skull fracture: do children with normal neurologic findings and no intracranial injury require hospitalization? Ann Emerg Med 1995;26:37–41.

Kaufman H, Levy M, Stone J, et al. Patients with Glasgow Coma Scale scores 3, 4, 5 after gunshot wounds to the brain. Neurosurg Clin North Am 1995;6:701–714.

Kriss V, Kriss T. SCIWORA (Spinal cord injury without radiographic abnormality) in infants and children. Clin Pediatr 1996;35:119–124.

Lang D, Teasdale G, Macpherson P, et al. Diffuse brain swelling after head injury: more often malignant in adults than children? J Neurosurg 1994;80:675–680.

Levi L, Guilburd J, Lemberger A, et al. Diffuse axonal injury: analysis of 100 patients with radiologic signs. Neurosurgery 1990;27:429–432.

Lloyd DA, Carty H, Patterson M. Predictive value of skull radiography for intracranial injury in children with blunt head injury. Lancet 1997;349: 821–824.

Macciocchi S, Barth J, Alves W, et al. Neuropsychological functioning and recovery after mild head injury in collegiate athletes. Neurosurgery 1996;39:510–514.

McLaughlin M, Marion D. Cerebral blood flow and vasoresponsivity within and around cerebral contusions. J Neurosurg 1996;85:871–876.

Mitti R, Grossman R, Hiehle J, et al. Prevalence of MR evidence of diffuse axonal injury in patients with mild head injury and normal head CT findings. Am J Neuroradiol 1994;15:1583–1589.

Mohanty A, Kolluri VS, Subbakrishna D, et al. Prognosis of extradural haematomas in children. Pediatr Neurosurg 1995;23:57–63.

Muhonen M, Piper J, Menezes A. Pathogenesis and treatment of growing skull fractures. Surg Neurol 1995;43:367–373.

Osenbach T, Menezes A. Pediatric spinal cord and vertebral column injury. Neurosurgery 1992;30:385–390.

Rosner M, Rosner S, Johnson A. Cerebral perfusion pressure: management protocol and clinical results. J Neurosurg 1995;83:949–962.

Sambasivan M. An overview of chronic subdural hematoma: experience with 2300 cases. Surg Neurol 1997;47:418–422.

Schutzman S, Barnes P, Mantello M, et al. Epidural hematomas in children. Ann Emerg Med 1993;22:535–541.

Shugerman R, Paez A, Grossman D, et al. Epidural hemorrhage: is it abuse? Pediatrics 1996;97:664–668.

Seelig J, Becker D, Miller J, et al. Traumatic acute subdural hematoma: major mortality reduction in comatose patients treated within 4 hours. N Engl J Med 1981;304:1511–1518.

Sloan E. Nerve injuries in the hand. Emerg Med Clin North Am 1993;11:651–670.

Vadasz A, Torres C, Chang J. Accidental penetrating cervical cord injury in a young child. Pediatr Emerg Care 1996;12:428–431.

CHAPTER 106

Neck Trauma

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Pediatric neck injuries, fortunately, are uncommon. Many children will be evaluated for cervical spine injuries secondary to trauma, but few will have injuries identified. Even fewer children will need to be evaluated for penetrating or direct blunt trauma to the neck. However, neck injuries can be life-threatening and need to be assessed in a timely and orderly manner. It is imperative to appreciate how apparently minor or innocuous neck injuries can progress rapidly to more serious and life-threatening events. Subtle neck injuries can be easily overlooked in a patient with obvious head or chest trauma. This chapter initially discusses evaluation and management of penetrating and direct blunt injuries and then concentrates on evaluation of the cervical spine.

When considering injuries to the neck in a child, initial management must include immediate assessment of airway breathing circulation (ABCs) and treatment of abnormalities with care not to allow an injury to progress to a more significant event. Airway abnormality may be subtle but progressive, with the precipitating injury not obvious on initial examination. Patients should be monitored closely and physically observed during their emergency department (ED) stay. A listing of common mechanisms of neck injury is given in Table 106.1.

Penetrating Trauma	Blunt Trauma
High-velocity missiles	Motor vehicle accidents
Low-velocity missiles	Sports
Knives	Fights
Windshields	Falls
Sharp objects	Clothesline injuries
Explosions	Handlebars
Iatrogenic (intubation, endoscopy, gastric tubes)	Barotrauma (bottlecap under pressure or compressed air source)
	Nonaccidental (abuse)
	Exposures (fires, caustics)

Table 106.1. Common Mechanisms of Blunt and Penetrating Neck Injuries

There are several differences in neck anatomy between children and adults. The child's relatively large head and mandible and short neck make the anterior neck less accessible to direct trauma but may increase the possibility of acceleration/deceleration injuries to the cervical spine. The increased potential for acceleration/deceleration injuries is partially offset by the elasticity of the pediatric cervical spine and the child's light weight. Internal neck anatomy also differs from that of an adult and may influence the types of injuries seen. The cricoid ring is the narrowest portion of the airway and is located at the C-4 level as opposed to the adult location at C-7. The arytenoids are proportionately larger and the child's cartilage is more pliable and easily damaged.

The soft tissues and visceral components of the neck are protected by the spine posteriorly, the mandible anteriorly and superiorly, the shoulders and clavicles anteriorly and inferiorly, and the neck muscles. The head and chest of the child protrude more anteriorly than the neck and will often absorb most blunt traumatic force, lessening the chance of a direct neck injury. Most injuries to the neck involve forces with relatively large masses and slow velocity, which partially accounts for the low incidence of serious direct trauma in this population. If the neck is hyperextended, however, the structures of the anterior neck, including the larynx, trachea, and esophagus, are more susceptible to direct trauma. The large number of vital organs and structures in the relatively small neck area enhances the potential severity of direct penetrating or blunt injuries (Table 106.2).

Musculoskeletal	Vascular	Venous	Gastrointestinal	Glandular
Cervical spine	Arterial	Jugular: internal, external	Esophagus	Thyroid
Cervical muscles	Carotids: common, internal,	Lymphatics	Neurologic	Parathyroid
Ligaments	external	Thoracic duct	Spinal cord	Parotid
Clavicle	Vertebral	Airway	Cranial nerves IX–XII	Submandibular
Pectoralis	Innominate	Larynx	Cervical nerves	
Hyoid	Subclavian	Trachea	Cervical sympathetics	
		Aspiration of lung	Brachial plexus	

Table 106.2. Neck Contents and Closely Approximated Structures

The neck can be divided into three anatomic zones (Fig. 106.1, Fig. 106.2 and Fig. 106.3). Zone I encompasses the area between the thoracic inlet and the cricoid (the lower boundary of zone I is the thoracic inlet, the upper boundary is most often classified as the cricoid; most authors use the clavicle or sternal notch as the upper boundary as well); zone II is the area between the cricoid (clavicle or sternal notch) and the angle of the mandible; and zone III is the area above the angle of the mandible. Knowledge of the divisions and structures they contain is useful in evaluation and management of neck trauma (Fig. 106.1, Fig. 106.2 and Fig. 106.3). Lesions in zones I and III are often occult and difficult to diagnose by physical examination alone. Operative exploration is more difficult in zones I and III than in zone II, where injury presentation and surgical exploration are often more straightforward. The neck can also be divided into anterior and posterior elements, with the dividing line being the palpable transverse processes of the cervical spine. The posterior neck contains muscles with their individual nerve supplies and the posterior elements of the cervical spine, and the anterior neck houses most vital organs and structures. No major vascular components are contained in the posterior area of the neck. Morbidity and mortality with neck injuries result from central nervous system (CNS) trauma, airway compromise, exsanguination, vascular disruption or thrombosis, venous embolism, sepsis, or mediastinitis.

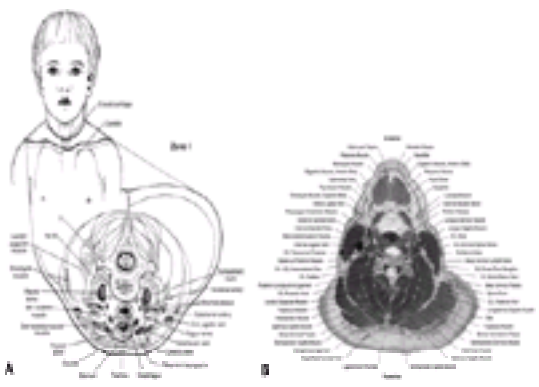

FIGURE 106.1. A. Anatomic neck divisions and contents of zone I. Zone I encompasses area between the thoracic inlet and the cricoid (some authors use the clavicle or sternal notch as the upper boundary). **B.** Anatomical specimen demonstrating zone I relationships. (106.1B reprinted with permission from Atlas of the Visible Human Male. Reverse Engineering of the Human Body, Spitzer and Whitlock. Jones and Bartlett, 1998.)

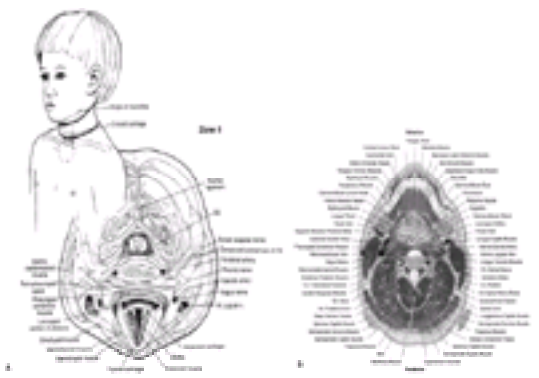

FIGURE 106.2. A. Anatomic neck divisions and contents of zone II located between the upper boundary of zone I and the angle of the mandible. **B.** Anatomical specimen demonstrating zone II relationships. (106.2B reprinted with permission from Atlas of the Visible Human Male. Reverse Engineering of the Human Body, Spitzer and Whitlock. Jones and Bartlett, 1998.)

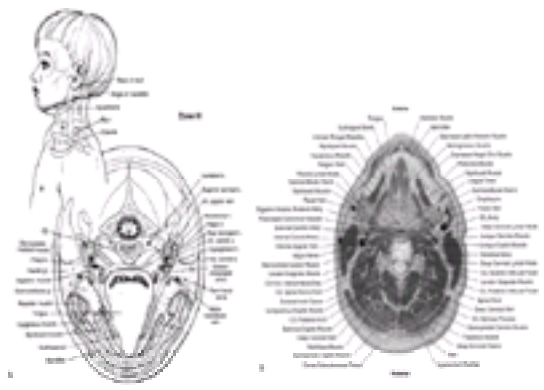

FIGURE 106.3. A. Anatomic neck divisions and contents of zone III. Zone III includes the area above the upper boundary of zone II. **B.** Anatomical specimen demonstrating zone III relationships. (106.3B reprinted with permission from Atlas of the Visible Human Male. Reverse Engineering of the Human Body, Spitzer and Whitlock. Jones and Bartlett, 1998.)

PENETRATING TRAUMA

Penetrating neck trauma is uncommon in children. Penetrating trauma may be associated with extracervical injuries and may involve multiple organ systems within the neck. Most pediatric penetrating trauma is the result of a wound from a gunshot (usually low velocity), knife, broken windshield, other sharp object, or explosion (Table 106.1). The history is important in evaluation of penetrating neck trauma. Inquiries about mechanism of injury, time of incident, events before arrival in the ED, amount of blood loss, history of pulsatile lesions, neurologic dysfunction (including transient ischemic attack, limb paresthesias, hemiplegia, blindness, Horner's syndrome, and aphasia), and airway compromise should all be noted. In particular, knowledge of the mechanism of injury can help direct the management of both the stable and unstable patient. Mortality with penetrating neck trauma is between 3 and 6% with or without surgical exploration. Common causes of death include vascular, neurologic, and airway injury.

Many gunshot wounds seen in pediatric patients involve low-velocity weapons, including handguns (90 m/sec) or shotguns (300 m/sec) at ranges of greater than 5 meters as opposed to shotguns at close range or military-style weapons (760 m/sec) (Fig. 106.4). Unlike higher-velocity missiles, low-velocity missiles tend to be redirected when they encounter vascular or other structures. Visceral injuries may be anticipated but not completely predicted by the path of the missile. Internal injuries may be more predictable with an isolated knife wound. Low-velocity neck wounds are associated with major pathology in approximately 50% of cases compared with more than 90% with high-velocity missiles.

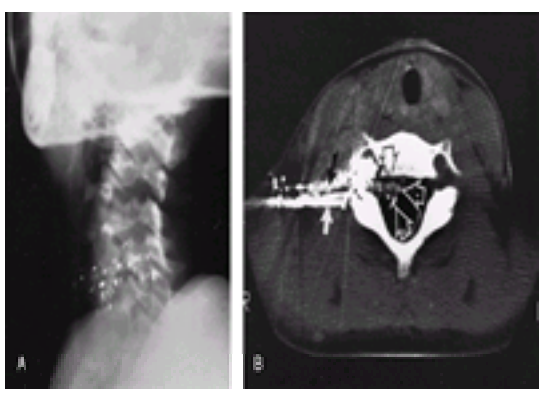

FIGURE 106.4. Gunshot wound (0.22 caliber) to the neck in a 5-year-old girl. **A.** Lateral neck radiograph showing fragmentation of bullet along path. **B.** Computed tomography scan of same patient demonstrating bullet fragments in and around spinal canal, as well as cerebrospinal fluid and contrast leak from disruption of the dura.

Vascular injury is the most common complication of penetrating trauma and is the second most common cause of death. History of large blood loss, pulsatile lesion, rapidly expanding hematoma, hypovolemic shock, or neurologic deficits (paresis, visual loss or aphasia, altered level of consciousness) indicates the possibility of cervical arterial injury. Major vessels that can be injured in the neck include the common, internal, and external carotid arteries; vertebral arteries; internal and external jugular veins; and nearby innominate and subclavian vessels (Table 106.2). Injury to the vessels can be dramatic, with exsanguination, rapidly expanding hematoma causing airway compromise, acute neurologic deficits from ischemia or hypoperfusion, or venous air embolism, or it may be subtle with an initially normal examination. Approximately one-third of arterial injuries present with neurologic deficits, whereas the remaining two-thirds are often more challenging to diagnose. The symptoms and signs suggestive of vascular and other neck injuries are presented in Table 106.3. Completely transected arteries often retract and contract with minimal bleeding. Vessels that are partially severed may continue to bleed significantly with normal pulses because blood flow may not be totally interrupted. Vascular abnormalities can be assessed partially by evaluating the carotid (external), superficial temporal, and brachial pulses, although no pulses are easily accessible to evaluate for the internal carotid or vertebral arteries. Abnormal pulses suggest vascular injury, whereas normal pulses do not guarantee vascular integrity.

Table 106.3. Symptoms and Signs of Neck Injuries

Auscultation of the neck is useful to identify bruits. Although a carotid bruit may be normal in children, a continuous bruit suggests a traumatic arteriovenous fistula, whereas a systolic bruit suggests a partial arterial tear. Bleeding from a posterior neck wound, neurologic deficits in areas supplied by the vertebral arteries (brainstem, cerebellum), bleeding not controlled by carotid compression, a posterior bruit, or bleeding that accompanies a cervical spine transverse process fracture suggests a vertebral artery injury. Carotid artery trauma should be suspected if presentation involves an anterior triangle hematoma, Horner's syndrome (ptosis, miosis, enophthalmos, loss of sweating on the ipsilateral side of the face), transient ischemic attacks, loss of consciousness after a lucid interval, or hemiplegia. Evaluation of the chest for signs of major vessel injury, including hemothorax, widened mediastinum, and cardiac tamponade, should accompany the neck examination.

Neck vessels may be injured indirectly as a result of shock waves from a missile. These patients may have clinically inapparent vascular intimal damage that can progress to vascular thrombus or occlusion. Venous or lymphatic (thoracic duct) injuries also occur with penetrating trauma. These injuries are rarely severe and usually present as an expanding hematoma or less often with a venous air embolism. Pulmonary embolus secondary to a venous thrombus is a rare event.

Injuries to the aerodigestive tract (pharynx, larynx, trachea, esophagus) are also seen in cases of penetrating trauma, although these relatively mobile structures are often spared. The esophagus is usually collapsed as it courses through the neck but may be injured by direct penetrating objects. Penetrating injury of the larynx and trachea occur, although blunt trauma to these areas is more common and can be associated with significant morbidity and mortality (see Chapter 112).

Direct nervous system injury (brachial plexus, spinal cord, cervical nerves, cervical sympathetics) is possible with penetrating neck trauma and evaluation of the patient should assess these structures. Symptoms will correspond to the injured structure, which may or may not require primary surgical repair (Fig. 106.5). Primary injury to the cervical cord often results from bony or foreign body penetration or impingement or cord distraction. Secondary cord injury can occur from vascular compromise, edema, lipid peroxidation, ischemia, and ligamentous damage. When assessing neurologic findings or predicting location of injury, it is important to remember that spinal cord and vertebral levels are not the same. In the cervical area, the cord level lies one segment higher than the corresponding vertebral level (C4 cord level lies opposite the C3 vertebral body) In the lower cervical area, a disparity of up to two levels may be present.

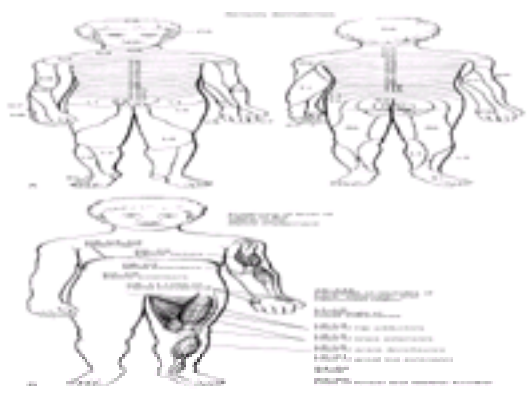

FIGURE 106.5. A. Sensory dermatomes. **B.** Motor dermatomes. Knowledge of sensory and motor dermatomes can be invaluable in description of neurologic findings during initial and subsequent evaluations.

BLUNT TRAUMA

Blunt trauma is often the result of a motor vehicle accident, although it can also be seen from sports; clothesline-type and handlebar injuries from bicycles, motorcycles, all-terrain vehicles, and snowmobiles; strangulation; hanging; blows from fists or feet; and the battered child syndrome (Table 106.1). Blunt trauma is often associated with extracervical injuries, especially maxillofacial, head, and chest, but is less likely than penetrating trauma to involve multiple structures within the neck. Blunt trauma is less likely than penetrating trauma to cause vascular damage, but the incidence of aerodigestive tract injuries is increased. The airway is often injured with direct blunt trauma in part as a result of the anterior and relatively fixed position of the larynx and trachea. As mentioned, the anterior neck is relatively well protected by bony structures unless the neck is extended. With neck extension, the larynx, trachea, and esophagus are exposed to direct trauma, and a blunt force may crush these structures against the posterior spinal column. A tracheal tear or rupture

may occur from a sudden increase in intratracheal pressure against a closed glottis, crush injury, or acceleration/deceleration injury. Shearing forces can cause edema, submucosal hematoma, laceration, perforation, vocal cord injury, and less commonly, partial or complete airway transection. A prime target for airway fracture is the cricoid ring, which is the only complete tracheal ring. The triad of dyspnea, stridor, and hemoptysis suggests laryngeal injury, although any or all of the symptoms and signs listed in Table 106.3 may be present.

Approximately 85% of patients with blunt tracheal injury will reportedly have subcutaneous emphysema, although the onset may be delayed (Fig. 106.6). However, airway injuries may be subtle and not apparent with initial history or physical examination. Unfortunately, these subtle injuries may progress to severe abnormalities. The same percentage of airway narrowing from edema or hematoma may lead to significantly more distress in a child compared with an adult. Airway obstruction from tracheal edema has been reported as late as 48 hours after the injury. If a laryngeal injury is noted, the patient should be evaluated carefully for commonly associated injuries, which include cervical spine, chest, facial, pharyngoesophageal, and recurrent laryngeal nerve. Airway injuries can be seen as a result of endoscopy, thermal trauma, or caustic ingestions. There are numerous reports in the literature of laryngeal and tracheal trauma secondary to intubation attempts.

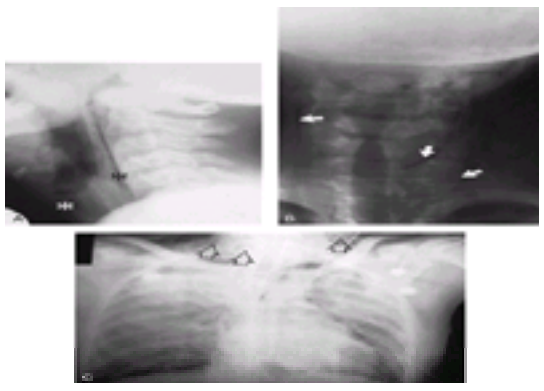

FIGURE 106.6. A–C. Subcutaneous emphysema of neck and chest in 11-year-old patient from barotrauma sustained when opening carbonated beverage container with teeth.

As mentioned, the esophagus is mobile and usually collapsed as it courses through the neck but may be dilated while eating. This mobility helps protect the esophagus, but its delicate mucosal walls can be damaged easily by blunt or penetrating traumatic events. Iatrogenic esophageal injuries can result from endoscopy, passage of a nasogastric or orogastric tube, vigorous suction, and difficult intubations. Esophageal injuries can also be seen with ingested foreign bodies and caustic chemicals. The symptoms and signs associated with esophageal injury are listed in Table 106.3 and include neck tenderness and pain, dysphagia, odynophagia, drooling, crepitus, subcutaneous emphysema, hematemesis, fever, and mediastinitis (see Chapter 29 and Chapter 93). The injuries, which can be subtle, occult, and difficult to diagnose, can lead to increased morbidity and mortality if not suspected and discovered.

Isolated or concurrent hyoid bone injuries are also possible. The hyoid is mobile and fairly well protected, which explains the paucity of isolated injury. In a 1991 review by Szeremeta and Morovati, only four children 16 years of age or younger had been reported with an isolated hyoid fracture, three of whom sustained the injury in motor vehicle accidents. Symptoms and signs of hyoid injury include pain in the throat that worsens with swallowing or coughing, tenderness to palpation, neck crepitus, pain on head rotation, dysphagia, dyspnea, or dysphonia. As with other injuries, these symptoms and signs can be subtle initially, with progressive edema and airway obstruction.

Although vascular injuries are rare with blunt trauma, they do occur. These injuries are often unsuspected and undiagnosed on routine examination. The clinician must consider subclavian or innominate vessel injuries if a fracture of the clavicle or first rib is identified. The most common vascular structure injured with blunt trauma is the common carotid artery. The vertebral arteries are rarely injured by blunt forces unless a concurrent transverse process or other fracture of the cervical spine or atlantooccipital dislocation occurs. Many patients with atlantooccipital dislocation and arterial injury die in the field, but some survive and may recover with appropriate therapy. Vascular contusions with intimal damage may also be seen with blunt neck trauma.

The glandular structures in the neck, including the thyroid, parathyroid, parotid, and submandibular glands, may also be injured. Although these organs may be traumatized, they are rarely completely destroyed.

Burn management is covered in Chapter 114, but the physician should be aware of special considerations involving the neck. The airway must be evaluated and protected as indicated by the severity of the burn, realizing that initial symptoms may be subtle. Circumferential burns may become edematous and require an escharotomy for respiratory or vascular sufficiency. Escharotomy in the neck involves a vertical incision from the chin to the superior aspect of the sternal notch.

EVALUATION AND MANAGEMENT

The goals of management are to ensure airway patency and respiratory sufficiency, control hemorrhage, maintain osseous stability, and identify and prevent progression of all injuries. Methodical and timely acquisition of historical and physical findings is mandatory. The patient must be managed with strict adherence to the ABCS, with consideration of possible rapid or gradual deterioration. Penetrating objects that are in the neck on evaluation should remain until removed under surgical care, preferably in an operating room. All patients, other than those with minor injuries such as contusions, abrasions, or superficial lacerations (not through the platysma muscle), should receive supplemental humidified oxygen, correct airway positioning, suctioning, vigilant observation, and monitoring. The patient should be

maintained in a supine or Trendelenburg position to avoid the possibility of venous air embolism. If venous air embolism is suspected because of an unexplained decrease in cardiac output and blood pressure, increase in central venous pressure, cyanosis, arrhythmias, or air in the heart on chest radiograph, the patient should be placed in the left lateral decubitus and Trendelenburg positions. A decision tree for evaluation of direct blunt and penetrating neck trauma is presented in Figure 106.7.

FIGURE 106.7. Evaluation of blunt and penetrating neck trauma.

Airway assessment is the initial step in the evaluation of all traumatized patients. Any airway manipulation should be accomplished with consideration and prevention of possible cervical spine injury. Potential indications for an artificial airway with neck trauma include stridor, dyspnea, hypoxia, rapidly expanding hematoma, expanding crepitus, pneumothorax, hemothorax, tracheal deviation, altered mental status, quadriplegia, hemiparesis, and other signs of vascular or airway insufficiency. Orotracheal intubation is the preferred method in children. Intubation should be attempted only after preparation for placement of a surgical airway, if time allows. Fiberoptic intubation, if available, may be useful if time and patient condition permit. The physician must be especially careful with the use of blind nasotracheal intubation in the patient with blunt or penetrating neck trauma because the airway anatomy may be distorted. Passage of the nasotracheal or orotracheal tube into a false or blind passage may make subsequent airway control attempts difficult if not impossible. Therefore, along with the difficulty of emergent surgical airway placement in children, elective intubation is not recommended outside of a setting where a surgical airway can be efficiently and skillfully placed.

If there is evidence of crepitus over the larynx, laryngeal or tracheal tenderness, a flattened thyroid prominence, anterior neck deformity, severe respiratory distress, an abnormal neck radiograph, or other evidence suggestive of a laryngotracheal fracture or disruption, a tracheostomy may be preferable. Intubation should be attempted only if the airway is completely obstructed. Attempts at intubation from above may separate a tenuously attached trachea and larynx and result in a total loss of the airway, with the trachea commonly retracting substernally into the chest (Fig. 106.8). Attempts at cricothyrotomy in patients with direct laryngeal trauma may result in retrotracheal placement of the airway. Cricothyrotomy is helpful in patients who have severe facial or other neck injuries that preclude intubation from above. Intubation may be attempted through an open laryngeal wound if present, although if possible a tracheostomy should not be placed through injured tissue. The flexible fiberoptic bronchoscope may be helpful in evaluating the patency of the airway and in establishing the artificial airway. If patient condition allows, rigid bronchoscopy can also be useful in securing an airway in these patients.

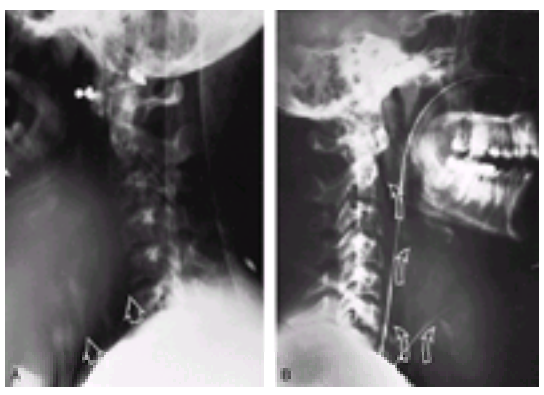

FIGURE 106.8. Tracheal injury. **A.** Initial lateral neck radiograph showing subcutaneous emphysema in 14-year-old girl kicked in the neck by a horse. Patient's airway clinically stable at the time of this radiograph (more than 1 hour after the injury). **B.** Postcricothyrotomy radiograph in same patient demonstrating significant subcutaneous emphysema and an artificial airway in place. Initial attempt at orotracheal intubation separated tenuously attached trachea, completing traumatic disruption of the trachea and requiring immediate placement of surgical airway.

Breathing should be evaluated for associated injuries. Missiles to the neck may also pass through or lodge in the chest. Zone I injuries of the neck can easily involve the lung apices and result in hemothorax, pneumothorax, or pneumomediastinum. Further penetration may lead to cardiac tamponade. A chest radiograph is helpful in the assessment.

In addition to the usual assessment of hypovolemia, the patient should be examined for expanding hematomas or other obvious external bleeding. External bleeding should be treated with gentle compression. Attempts to clamp bleeding

vessels in the neck can injure the vessels and surrounding structures and jeopardize subsequent repair attempts. Two large-bore intravenous (IV) catheters should be inserted. If a subclavian vein injury is suspected, one of the IV lines should be placed in the lower extremity. Type-specific or crossmatched blood should be made available and used with volume expanders as necessary.

The subtle presentation of vascular injuries has led many authors to suggest mandatory exploration of all neck injuries when the outermost muscle layer (platysma) is penetrated. Controversy exists in the literature regarding surgical exploration with low-velocity penetrating neck trauma (Table 106.4). Proponents of mandatory exploration of penetrating neck wounds report that overall morbidity and mortality has decreased with routine neck exploration and surgical repair of vascular and other abnormalities. However, recent literature suggests that careful evaluation with ancillary studies, including arteriography, and the use of selective exploration will identify most significant injuries, and the potential delay in operative evaluation and repair will not increase morbidity and mortality. Surgical exploration is practiced uniformly for high-velocity or multiple low-velocity wounds. With pediatric penetrating injuries, routine neck exploration, even in the relatively straightforward zone II, is not always a benign procedure.

Arguments Favoring Mandatory Surgical Exploration with Penetrating Trauma
Most patients have injuries to important structures.
Morbidity and mortality increase with delay in surgery.
Morbidity of exploration is relatively low.
Negative physical examination does not preclude injury.
Morbidity of missed injuries is high.
Specific radiologic tests and expertise are needed to selectively manage.
More skill is needed to observe appropriately.
Length of hospitalization is similar with or without exploration.
Arguments Favoring Selective Surgical Exploration with Penetrating Trauma
Most injuries are not secondary to high-velocity weapons.
There are high negative rates of injuries with exploration in asymptomatic patients.
It is unclear whether morbidity and mortality increase with delay in surgery.
Zone II is involved most commonly and injuries are rarely occult.
Skill is needed to explore neck.
Morbidity of the more difficult zone I and III explorations is seen.
Ancillary tests are helpful in zone I and III evaluation.
Exploration may miss occult injuries.

Table 106.4. Surgical Exploration with Penetrating Trauma

Repair of vascular injuries depends on the vessel injured, type of injury, and the patient's clinical status. Arterial injuries with neurologic deficits often are not repaired but are ligated to avoid the chance of reperfusion injuries to the brain. Other authors suggest that primary arterial repair may be indicated, despite the possibility of reperfusion injury. Vascular repair in zones I and III is especially difficult and not without operative morbidity. Venous injuries may not need repair unless persistent bleeding or associated morbidity is demonstrated. The need for surgical repair of vascular intimal injuries is controversial.

A neurologic examination for signs of cerebral injury secondary to vascular insufficiency, direct spinal cord, cranial or cervical nerve, or brachial plexus injury should be completed. An abnormal neurologic examination may indicate progressive vascular insufficiency and the need for rapid surgical evaluation. Direct neurologic injuries may not necessitate surgical repair.

Tetanus status should be assessed in all patients with penetrating trauma. The clinician should consider a broad-spectrum antibiotic for a patient with evidence of neck trauma, especially if esophageal or pharyngeal injury seems likely. Placement of a nasogastric or an orogastric tube is controversial for the patient with cervical injury because it may worsen a preexisting esophageal injury or dislodge clots in zone I of the neck. When placed, these tubes should be well lubricated, inserted gently and slowly, and withdrawn if difficulty in passage or evidence of obstruction occurs.

Superficial abrasions, lacerations, and puncture wounds are common in children. Wounds superficial to the platysma can be cleaned and sutured in the normal fashion under local anesthesia in the ED. Clean wounds can be sutured as late as 12 to 18 hours after the injury because of the excellent blood flow in the neck. In wounds older than 12 to 18 hours, closure after 72 hours is recommended. Penetration of the external muscle layer in the neck, the platysma, is an indication for surgical referral and, in some cases, surgical exploration. When neck wounds that penetrate the platysma are evaluated, exploration in the ED is discouraged because of the risk of clot dislodgment and venous air embolism. Rapid surgical exploration and repair is indicated in patients with unstable vital signs, uncontrollable bleeding, rapidly expanding hematomas, progressive airway compromise, worsening neurologic symptoms, those struck by a high-velocity missile, increasing subcutaneous emphysema, or bubbling wounds (Table 106.5).

Unstable vital signs
Expanding or massive hematoma
Pulsatile or active bleeding
Hemorrhagic shock
Vascular deficits in the upper extremities
Abnormal distal pulses (brachial, superficial temporal, ophthalmologic, fundi)
Hematemesis, hemoptysis, epistaxis
Hemothorax
Progressive respiratory distress
Airway obstruction
Expanding subcutaneous emphysema
Bubbling or sucking wound
Pneumothorax
Progressive neurologic deficits
Hemiparesis
Horner's syndrome
Cranial or cervical nerve dysfunction
Diaphragm paralysis
Decreased sensorium
Neurologic deficits in upper extremity
Increasing dysphagia
Odynophagia or dysphonia
Hoarseness
Severe neck pain or tenderness
High-velocity wounds (rifles, explosions)
Multiple low-velocity wounds
Anxiety (allegiance) studies not available
Equivocal observation personnel not available

Table 106.5. Indications for Surgical Evaluation in the Patient with Neck Trauma

Surgical evaluation may be falsely negative with esophageal tears, small vessel lacerations, pharyngeal tears, or tracheal injuries. The patient who has apparently stable vital signs, no symptoms of impaired neurologic or cardiovascular status, an intact airway, and mechanisms of injury with a low-velocity bullet or single knife wound may be managed expectantly with the use of ancillary diagnostic tests and close, experienced observation, preferably for at least 48 hours. These decisions should be made in conjunction with experienced surgical staff.

Adjuncts to the history and physical examination are presented in Table 106.6. Initial evaluation should include cervical spine radiographs to detect bony or structural abnormalities and a soft-tissue lateral neck radiograph to assess for blood, edema, subcutaneous air, foreign bodies, and airway impingement or disruption. A chest radiograph should be evaluated for evidence of hemothorax or pneumothorax, mediastinal emphysema or widening, and heart size. If a serious injury is at all likely, these radiographs should be performed in the ED or the patient should be accompanied to the radiology department by someone skilled in airway management. Fluoroscopy may be helpful in airway evaluation. If the patient is stable and a vascular injury is suspected, an arteriogram should be performed (Fig. 106.9). Arteriography has excellent sensitivity, specificity, and accuracy, as well as low morbidity. Some authors suggest that an arteriogram is not needed in penetrating zone II injuries if an exploration is to be performed because this can be done fairly easily without significant complications. Others suggest that, even with zone II injuries, the stable patient should receive an arteriogram before the operative procedure.

Cervical spine radiographs	Xeroradiography
Soft-tissue neck radiograph	Tomography
Chest radiograph	
Computed tomography scan	Indirect (mirror) laryngoscopy
Arteriography	Direct laryngoscopy
Doppler	Flexible bronchoscopy
Esophagram	Direct broncho/esophagoscopy
Contrast laryngotracheography	Surgical exploration
Oculoplethysmography	

Table 106.6. Adjuncts to History and Physical Examination

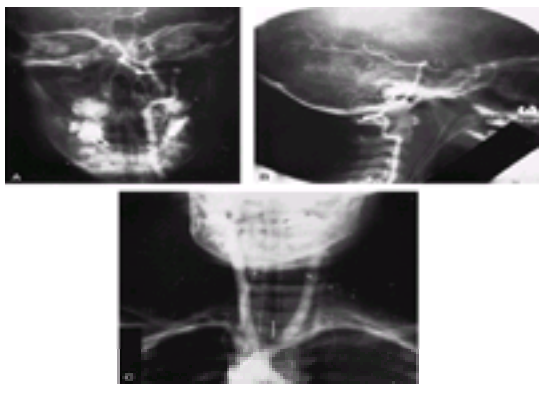

FIGURE 106.9. Angiograms in 5-year-old child shot in neck, demonstrating normal vascular integrity. **A.** Vertebral artery angiogram (anteroposterior view). **B.** Vertebral artery angiogram (lateral view). **C.** Carotid angiogram.

Noninvasive Doppler studies and oculoplethysmography may be useful in evaluating vascular injuries. Contrast laryngography, tomography, and xeroradiography have been used for further evaluation; however, these methods have generally been replaced by the computed tomography (CT) scan. The noninvasive CT scan provides excellent bone and soft-tissue detail and can be obtained easily with a stable, immobilized patient. The addition of IV contrast allows identification and initial evaluation of the cervical vasculature. The advent of spiral CT technology allows rapid scans and illustrative coronal, sagittal, and three-dimensional reconstruction of the neck anatomy. CT may not be accurate for detection of mucosal degloving injuries, mucosal perforation in the presence of subcutaneous emphysema, endolaryngeal edema or hematoma, and partial laryngotracheal separation.

Barium or Gastrografin esophagram is helpful in evaluating the esophagus for tears or perforations, but false-negative rates of up to 50% have been reported. Evaluation can also include indirect mirror laryngoscopy to assess the larynx, vocal cord mobility, presence of mucosal edema, ecchymosis, and mucosal tears and direct endoscopy to examine for tracheal, bronchial, and esophageal damage. Flexible endoscopy may be less invasive and easier to accomplish, but rigid endoscopy offers the most complete examination. Even rigid endoscopy, however, is not 100% sensitive in detecting tracheal and esophageal injuries. As mentioned, operative evaluation is mandatory for some patients and optional for others. Determinants of specific management direction include mechanism of injury, wound size and type, patient signs and symptoms, and relative stability. The clinician must maintain a high index of suspicion for potential injury to the structures contained in the neck. Consequences of missed injuries include airway obstruction, delayed hemorrhage, neurologic compromise, and deep neck infection, with potentially significant morbidity and mortality.

CERVICAL SPINE EVALUATION

Cervical spine injuries are also uncommon in children, occurring in an estimated 1 to 2% of patients with multiple trauma. However, the clinician must assume that all children who sustain multiple trauma, have head or neck injuries, or have symptoms of neurologic impairment, including altered level of consciousness, have a cervical spine injury until proven otherwise. Goals in the care of these children include effectively stabilizing the primary injury that has occurred and preventing progression to a more severe or significant injury. The devastating nature of a cervical cord injury makes it imperative to not inadvertently miss a potentially unstable cervical spine injury. While attending to the basic ABCS of trauma resuscitation, the clinician should stabilize the cervical spine. Caution must be exercised when applying airway maneuvers to a child with a possible cervical spine injury. Airway interventions, however, often cannot wait until the cervical spine is “cleared.” The clinician must prioritize and proceed with lifesaving airway maneuvers, while minimizing motion of the potentially unstable cervical spine.

Hyperextension of the neck to facilitate intubation should be avoided. A vigorous chin lift or jaw thrust may also inadvertently hyperextend the unstable cervical spine. Gentle cricoid pressure should not cause excessive movement to the cervical spine; however, if applied vigorously, it may cause flexion of the spine. When in-line neck immobilization is used to assist with airway maneuvers, the clinician should be careful to avoid applying significant traction to the spine as this pressure can also stress the unstable cervical column. Tracheal intubation in a patient with a potential cervical spine injury ideally requires at least two participants to perform the procedure safely and efficiently. One provider should maintain in-line immobilization of the neck, while another performs the intubation. The immobilization is often best accomplished from below, allowing the intubator as much room as possible to maneuver (Fig. 106.10). The hard cervical collar should be opened anteriorly, or removed, while this process in being performed. It is difficult to intubate a child with the anterior trachea unless the jaw immobilization afforded by the collar is temporarily removed. As usual, oral intubation is often the preferred method because of the child's anterior airway and the usual experience of providers. The collar should be resecured after the airway intervention is complete.

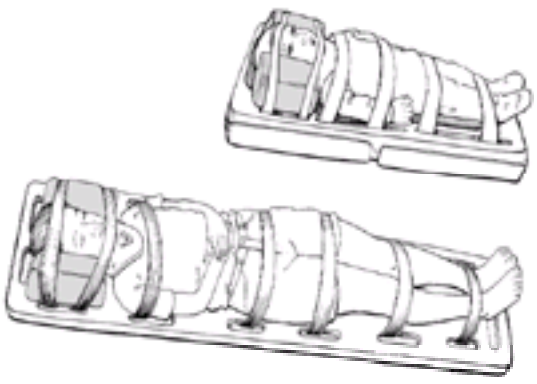

FIGURE 106.10. A. Manual immobilization from below. **B.** Manual immobilization from above. Adequate and expert manual cervical spine immobilization is required during airway maneuvers. The head and neck can be adequately secured from above or below. Immobilization by the second provider from below allows the airway maneuver to be accomplished without requiring a change in preferred positioning of the professional performing the maneuver.

Several concepts should be kept in mind concerning cervical immobilization in children. Soft cervical collars offer no protection to an unstable spine and hard (Philadelphia, Stifneck) collars alone still allow a fair amount of flexion, extension, and lateral movement of the cervical spine. Ideal immobilization involves a hard cervical collar in conjunction with a full spine board, soft spacing devices, and securing straps (Fig. 106.11). An appropriately sized hard cervical collar should be chosen. The tallest collar that does not hyperextend the neck is the correct choice. The choice between a one-piece (Stifneck) or two-piece (Philadelphia) collar is important only in that correct fit must be ensured and the provider must understand how to apply the specific brand of collar. It is helpful to fold over the Velcro connectors on the collar before sliding it under the patient's neck to avoid Velcro attachment to the child's hair or clothing. If a patient is seated and needs to have a collar placed, this maneuver should be accomplished by positioning the collar's chin portion first, followed by placement of the posterior portion. If the patient is wearing a helmet, it should be carefully removed. Helmet removal, if possible, should involve at least two people to avoid potential neck motion. In-line stabilization is ensured by one provider while the helmet is spread and gently removed. Occasionally, mechanical bivalving of the helmet may be required for safe removal.

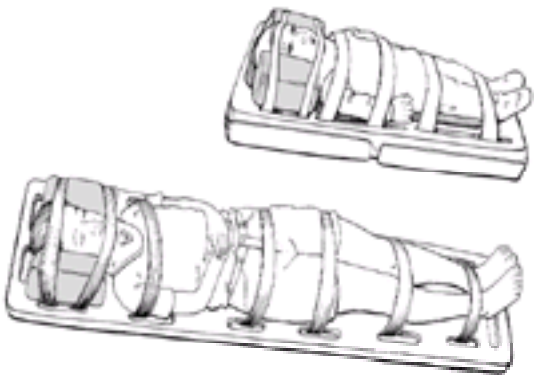

FIGURE 106.11. Cervical spine immobilization should not place the patient at an increased risk for morbidity. Securing straps should be placed around bony prominences, and strap location reassessed after any movement of the patient. A

neutral position of the neck should be ensured, and if necessary (younger child), a spacer can be placed underneath the child's torso and lower extremities to achieve the desired position.

The clinician must be prepared to log-roll the patient if vomiting occurs. This reaction may happen at any stage of the immobilization process and should be anticipated. Adequate personnel to safely log-roll the vomiting patient are required to avoid potential gagging, aspiration, or secondary spinal or cord injury. The patient should be secured to a long spine board by tape or straps that cross the forehead, chin area of the cervical collar, and bony prominences of the shoulders and pelvis. Incorrect immobilization may impede respiration by obstructing chest rise or contributing to secondary spinal injury by hyperextending the neck. The securing straps should be assessed periodically to ensure adequate and safe attachment of the patient to the spine board. When a child is immobilized on a spine board, the clinician must consider that the child's head is disproportionately large compared with the adult's. A child's head reaches 50% of postnatal growth by approximately age 2 years, whereas chest circumference reaches 50% of postnatal growth by about age 8 years. This disparate growth of the head and trunk causes the neck to be forced into relative kyphotic position when a child is placed on a hard spine board (Fig. 106.12). This is distinctly different from the adult patient whose neck is in 30 degrees of lordosis, the neutral position, when immobilized on a hard spine board. Suggestions have been made to allow a recess in the head area of the spine board to accommodate the child's large occiput or to place a spacing device such as a blanket underneath the torso to allow the neck to rest in a neutral position (Fig. 106.12). Figure 106.13 demonstrates that cervical spine alignment can be greatly affected and improved by this technique.

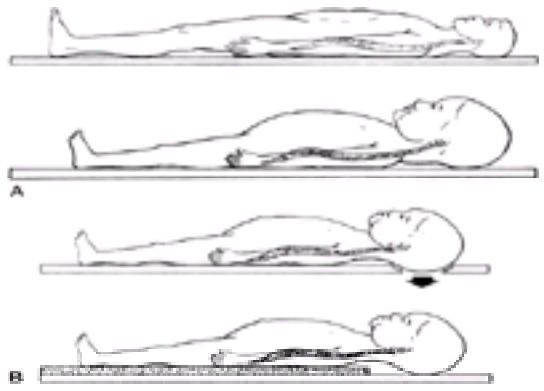

FIGURE 106.12. Effects of backboard on cervical spine position. **A.** Adult and child immobilized on standard backboard. **B.** Backboards modified with occipital recess and mattress pad to allow neutral positioning of the cervical spin in a young child. (Reprinted with permission from Herzenberg J, Hensinger R, Dedrick D, et al. Emergency transport and positioning of young children who have an injury of the cervical spine: the standard backboard may be hazardous. J Bone Joint Surg 1989;71-A:15–21.)

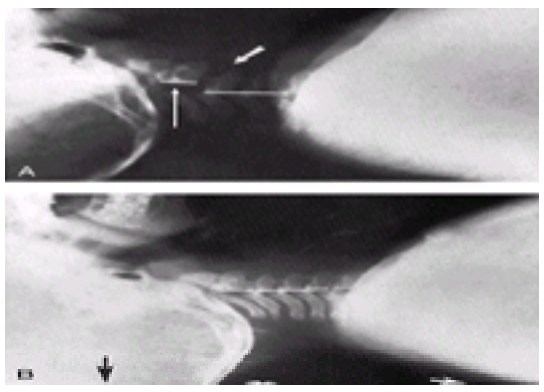

FIGURE 106.13. Effects of backboard on cervical spine position in 6-month-old child with a hangman fracture (traumatic spondylolisthesis of posterior elements of C-2) indicated by *thin arrow.* **A.** Large occiput contributes to anterior subluxation *(thick arrow)* of unstable cervical spine. **B.** Same child on backboard with occipital recess. Anterior subluxation is decreased. (Reprinted with permission from Herzenberg J, Hensiger R, Dedrick D, et al. Emergency transport and positioning of young children who have an injury of the cervical spine: the standard backboard may be hazardous. J Bone Joint Surg 1989;71-A:18.)

Patients often arrive in the ED with full or partial cervical spine immobilization already in place. An immediate assessment of that immobilization is imperative. Several important issues should be examined: 1) Is the patient appropriately and fully immobilized? 2) Is the cervical collar of the correct size and type for that patient? 3) Is the patient's neck in a neutral position? 4) Is the patient securely strapped to a long spine board? 5) Has there been a shift in the patient or the immobilization during the prehospital or interfacility transport that might diminish effective immobilization, cause hyperflexion or hyperextension of the cervical spine, or compromise excursion of the chest with respiration? and 6) Does the immobilization interfere with assessment or management of the ABCs? If these or other immobilization difficulties are identified, they should be immediately addressed.

Occasionally, the use of a semipermanent immobilization device (tongs, halo) may be indicated (Fig. 106.14). This should be accomplished after neurosurgical consultation. Attempts to rapidly reduce a cervical fracture are usually

discouraged to avoid potential further cord injury. Frequent reassessment is necessary for the patient in cervical traction. Transport of the patient in cervical traction has the potential to further damage the injured cervical spine.

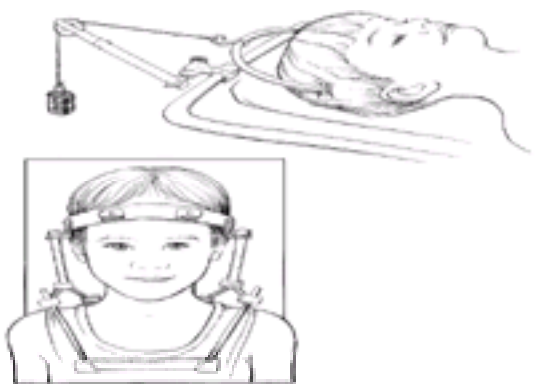

FIGURE 106.14. A. Gardner-Wells tongs. **B.** Halo traction brace. Unstable cervical spine injuries may require immediate placement of semipermanent immobilization devices. These should be administered only by those experienced in appropriate usage and application.

The pediatric cervical spine and its evaluation differ in many ways from the adult spine. The fulcrum of the cervical spine of an infant is at approximately C2–C3 and reaches C3–C4 by the age of 5 to 6 years. At about the age of 8, the fulcrum (C5–C6) and other characteristics of the cervical spine approximate that of an adult. The higher fulcrum of a child's spine in combination with relatively weak neck muscles and poor protective reflexes account for young children often having fractures that involve the upper cervical spine, whereas older children and adults have fractures that more often involve the lower cervical spine. Neurologic disability can occur from cervical lesions at all levels, but high cervical cord injuries are more likely to be fatal than are lower cervical cord injuries.

The large amount of cartilage present in a pediatric cervical spine cushions forces that are transmitted to the spine but can also make radiographic evaluation somewhat challenging. The radiolucent nature of cartilage makes the ability to appreciate soft-tissue changes on the radiograph extremely important. The pediatric cervical spine seems to have more anterior and posterior movement than its adult counterpart as a result not only of radiolucent cartilage, but also of ligamentous laxity and relatively horizontal facet joints. The pediatric cervical spine also has the ability to revert to a relatively normal appearance after a significant distortion, which can hinder the radiographic search for abnormalities. It is important to realize that any persistent distortion demonstrated on the radiographs was probably more exaggerated during the actual precipitating event. There is more room around the spinal cord within the spinal column in a young child than in an adult, which means that compressive problems such as tumors or bleeds may be slower to manifest neurologic symptoms.

Evaluation of the traumatized child begins with a focused history and complete physical exam. The history (if reliable) can be invaluable in helping identify the potential for cervical spine or cord injury. The following questions should be answered: 1) Was the child involved in a high-speed motor vehicle accident? If so, was he or she restrained and at what angle did the car(s) collide? 2) Was there a sports injury? If so, did it involve a spearing motion? 3) Did the child fall? If so, how high was the fall and how did the child land? A neurologic history is imperative to assess whether there was any evidence of abnormal findings such as paresthesias, paralysis, or paresis at any time after the injury. These symptoms may have been transient and may not be present at the time of the examination or volunteered by the patient during gathering of the history, yet they are important because they may suggest a cervical contusion, a concussion, or a spinal cord injury without radiographic abnormality (SCIWORA). The answers to these and other historical questions can often be obtained from the patient, parents, bystanders and Emergency Medical Services personnel and can help rule in or rule out significant possibilities of cervical injury. A plethora of clues can aid in the diagnosis of a cervical cord injury (Table 106.7). The symptoms and signs may be obvious or they may be masked by other abnormalities, such as altered level of consciousness, hypovolemic shock, or concurrent head injury. Head and neck injuries may present with overlapping abnormal neurologic signs and differentiation of causation may be difficult.

Abnormal motor exam (paresis, paralysis, flaccidity, ataxia, spasticity, rectal tone)	Diaphragmatic breathing without retractions
Abnormal sensory examination (pain, sensation, temperature, paresthesias, anal wink)	Spinal (neurogenic) shock (hypotension with bradycardia)
Altered mental status	Priapism
Neck pain	Decreased bladder function
Torticollis	Fecal retention
Limitation of motion	Unexplained ileus
Neck muscle spasm	Autonomic hyperreflexia
Abnormal or absent reflexes	Blood pressure variability with flushing and sweating
Clonus without rigidity	Poikilothermia
	Hypothermia or hyperthermia

Table 106.7. Symptoms and Signs of Cervical Spine Injury

Consideration of cervical spine radiographic evaluation is the next step in assessment. Radiographic options include radiographs, CT, and magnetic resonance imaging (MRI). MRI scans are more appropriate when evaluating the subacute

or chronic stages of injury or when looking for an acute problem with cord impingement by blood or soft tissues such as tumors or intervertebral discs. MRI does not image cortical bone well and should not be used to evaluate the cervical spine for fractures, whereas the CT scan demonstrates fractures clearly. A CT scan is often used as a secondary screen when adequate plain radiographs cannot be obtained or to substantiate suspected fractures. A common scenario is the use of CT to supplement viewing the C1/C2 region in young, traumatized children. The CT scan images soft tissue well; however, it does not demonstrate the intrathecal, ligamentous, disc, or vascular detail that can be obtained with an MRI scan.

The plain radiograph remains the preferred initial test for acutely traumatized patients. Several authors have attempted to devise criteria to limit the use of cervical spine radiographs because the number of positive studies constitutes a small proportion of the total number of radiograph studies completed. The perception of unnecessary tests should be balanced against the severity of consequences that may occur with a missed cervical spine injury. The literature suggests that if the patient does not have a high-risk mechanism of injury (motor vehicle accident, fall, dive, or sports injury), is awake and alert, can have an interactive conversation (not inebriated, no altered level of consciousness, older than 4 to 5 years of age), does not complain of cervical spine pain, has no tenderness on palpation (especially in the midline), has normal neck mobility, has a completely normal neurologic examination without history of abnormal neurologic symptoms or signs at any time after the injury, and has no other painful injuries (which may distract the patient and mask neck pain), the patient probably does not need radiographic evaluation of the cervical spine. The clinician must be sure, however, to never “clear” the cervical spine, regardless of studies performed, in an unconscious patient in the ED.

When radiographs are obtained, a normal lateral radiograph does not “clear” the cervical spine. The sensitivity of a lateral cervical spine radiograph varies between 82 and 98% in the literature. When evaluating a lateral cervical spine radiograph, the clinician must ensure that C-1 through C-7 are included as well as the C7–T1 junction. Additional films, which include an anteroposterior (AP) view of C-3 through C-7, and an AP open-mouth (odontoid) view of C1–C2, increase the sensitivity of initial radiographic evaluation to more than 95%. An adequate open-mouth (odontoid) view is often technically difficult to obtain in young children and those who are intubated. If further information is required, a CT scan of C1/C2 can be useful to augment or replace the open-mouth view. A CT scan is more expensive than a plain film of C1/C2, but it is easier to obtain, offers better and more consistent information, and avoids the risk of missing a subtle injury in that critical area. The advent of the spiral CT scan allows this study to be completed in 1 to 2 minutes and to be reconstructed by the computer to demonstrate vivid detail of the region (Fig. 106.15). An algorithm for considering radiographic evaluation is presented in Figure 106.16. An approach to ordering cervical spine imaging studies is shown in Figure 106.17.

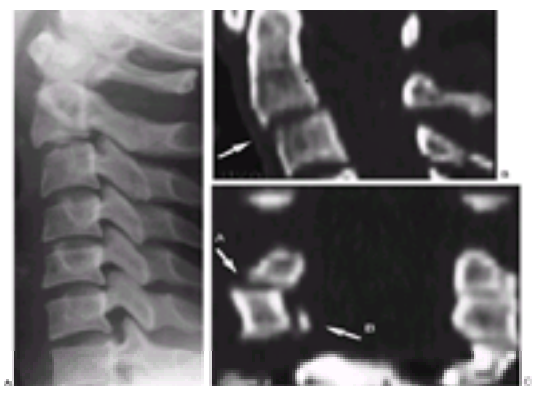

FIGURE 106.15. A. Apparently “normal” lateral cervical spine radiograph (16-year-old patient after motor vehicle accident). **B.** Spiral computed tomography (CT) scan demonstrating dens fracture *(arrow).* **C.** Sagittal view of spiral CT scan demonstrating dens fracture *(arrow A)* and vertebral body avulsion fracture *(arrow B).* The detail demonstrated by the spiral CT scan can help the clinician quickly identify lesions not easily visible or appreciated on conventional radiographs.

FIGURE 106.16. Decision tree for radiographic and clinical evaluation of patient with possible neck injury.

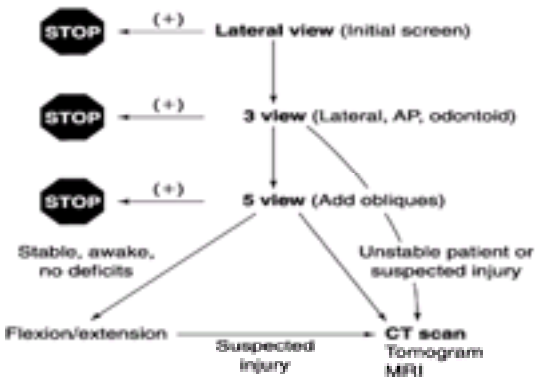

FIGURE 106.17. Approach to ordering cervical spine radiographs. Additional films may not be needed in the acute situation if a fracture or other abnormality is identified. If an abnormality is suspected, but not demonstrated, continue with algorithm as presented.

The cervical spine has anterior (vertebral bodies, intervertebral discs, ligaments) and posterior (lamina, pedicles, neural foramen, spinous processes, ligaments) components (Fig. 106.18). The initial three-view series evaluates the anterior cervical spine well; however, it is not ideal for evaluating the posterior cervical spine. Oblique (pillar) views are helpful in imaging those posterior elements. In practice, however, oblique films rarely add significant information to the initial radiographic assessment. Flexion and extension films are accomplished in an awake patient by having the patient flex and extend the neck as far as possible without discomfort. As the end point involves the sensation of pain, flexion/extension films should not be performed in a patient who has preexisting neck pain. Dynamic fluoroscopy could be substituted in specific instances for flexion/extension films. These studies can help evaluate underlying soft-tissue or ligamentous injury that was not evident on the initial films. These films are often inadequate because the neck muscles have splinted the cervical column into a position of comfort and stability, and alignment does not change with flexion and extension. If a question remains concerning the integrity of the cervical spine after following this radiographic scheme, a CT scan should be considered. Tomograms can also be performed but require patient movement, are time-consuming, and are not performed easily in the acutely ill patient. An MRI should be considered to detect ligamentous, soft-tissue, or subtle cord injuries.

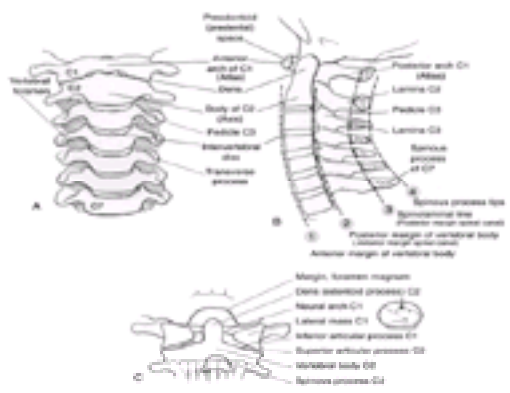

FIGURE 106.18. A–C. Knowledge of normal cervical spine anatomy is useful when evaluating cervical spine radiographs.

A systematic approach should be used when evaluating radiographs of the cervical spine. The ABCS method is a useful approach (Fig. 106.19). Alignment is assessed as demonstrated in Figure 106.20, keeping in mind that the spinal cord lies between the posterior spinal line and the spinolaminal line. These lordotic curves may not be present in children less than 6 years of age, those on hard spine boards or in cervical collars, or those with cervical neck muscle spasm. Gross malalignment should be detectable with this assessment.

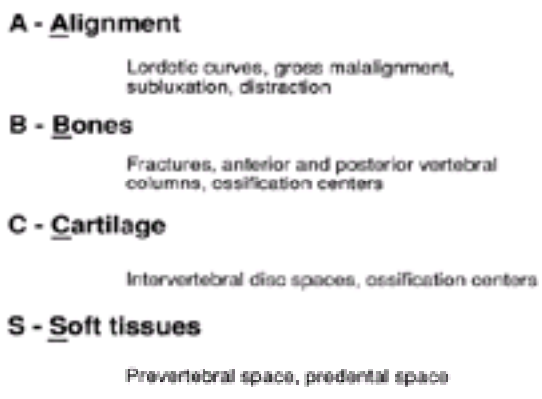

FIGURE 106.19. The ABCS of radiographic cervical spine interpretation.

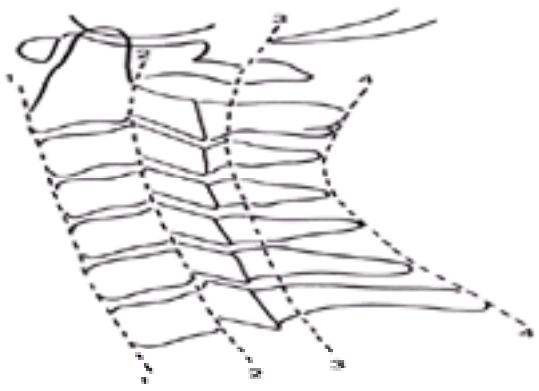

FIGURE 106.20. Four contour lines of alignment with normal cervical spine lordosis: *1,* anterior vertebral bodies; *2,* posterior vertebral bodies (anterior spinal canal); *3,* spinolaminal line (posterior spinal canal); *4,* spinous process tips. (Reprinted with permission from Gerlock A, Kirchner S, Heller R, et al. Advanced Exercises in Diagnostic Radiology: The Cervical Spine in Trauma. Philadelphia: WB Saunders, 1978:6.)

The bones should be evaluated for typical abnormalities, realizing that these may be subtle. The clinician should be aware that structures that overlaythe spine, including the skull and the teeth, may simulate fractures.

The next area of evaluation involves assessing the cartilage. Cartilage is radiolucent on plain radiographs. Children's spinal columns contain significant cartilage that may buffer a traumatic force and help prevent some injuries but that can also make radiographic evaluation challenging. The cartilaginous areas include the synchondroses or growth plates and intervertebral disc spaces (Table 106.8). The growth plates may mimic fractures and may be confusing to those who are unaware of their presence. Growth centers in the anterior-superior vertebral bodies cause a sloped appearance that may appear as a compression fracture to the untrained eye. Vertebral disc abnormalities may indicate specific types of injuries. A vertebral disc space that is narrowed anteriorly may indicate disc extrusion, whereas a widened space suggests a hyperextension injury with posterior ligamentous disruption.

Cartilage artifact
Tapered anterior vertebrae
Apparently absent ring of C-1
Atlas (C-1) body not ossified at birth and may fail to close
Axis (C-2) has four ossification centers
Apex of odontoid ossifies between ages 12 and 15 yr
Spinous process ossification centers
Increased mobility
Pseudosubluxation
C-1 override on dens
Increased predental space (5 mm maximum)
Ligament laxity
Facet joints shallow
Growth plates (synchondrosis)
Dens ossifies between ages 3 and 6 yr (may persist into young adults)
Posterior arch of C-1 ossifies at age 3 yr
Anterior arch of C-1 ossifies at age 6–9 yr
C-1 reaches adult size at 3–4 yr
C-2 through C-7 reach adult size at ages 5–6 yr
Lack of cervical lordosis
Fulcrum varies with age
Soft-tissue variability with respiration
Congenital clefts or other bony abnormalities (ex odontoideum, spondylolisthesis, spina bifida, dysraphism)

Table 106.8. Radiographic Characteristics of the Pediatric Cervical Spine

Soft-tissue evaluation is extremely important. Abnormal soft-tissue spaces may be the only clue to the underlying ligament, cartilage, or subtle bone injury, which may not be obvious on the radiograph. The soft-tissue widening may represent blood or edema, which suggests an underlying injury. The prevertebral space at C-3 should be less than one-half to two-thirds of the AP width of the adjacent vertebral body (Fig. 106.21). This space will double, to approximately the width of the adjacent vertebral body, below C-4 (the level of the glottis) as the usually non–air-filled esophagus is present at this area. Care must be taken when evaluating the prevertebral soft-tissue space because crying, neck flexion, or the expiratory phase of respiration may produce a pseudothickening in the prevertebral space (Fig. 106.22). Soft-tissue abnormality should be reproducible on repeated radiographs if an actual underlying injury exists.

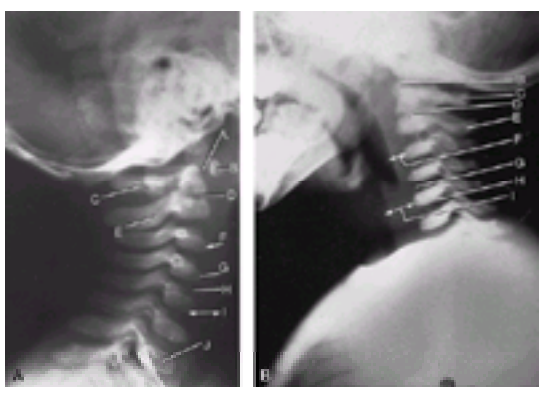

FIGURE 106.21. Normal pediatric lateral cervical spine radiographs. **A.** 3-month-old child. **B.** 20-month-old child. *A,* predental (predontoid) space; *B,* anterior ring of C-1 (note apparent override of C-1 over dens); *C,* posterior ring of C-1; *D,* dens synchondrosis (growth plate); *E,* posterior elements C-2; *F,* normal prevertebral space (C-3 level); *G,* wedged vertebral appearance caused by cartilage artifact; *H,* intervertebral disc space; *I,* Normal prevertebral space below the

glottitis (thickened because of radiopaque collapsed esophagus); *J,* vertebral body C-7.

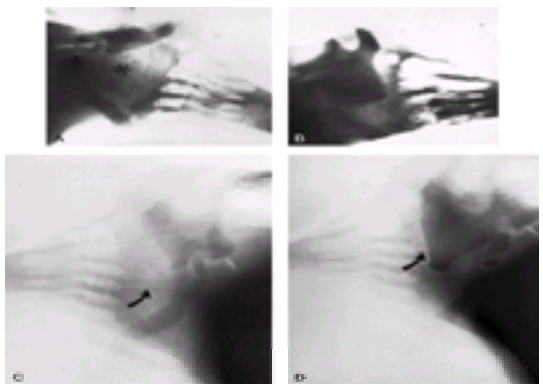

FIGURE 106.22. Effects of inspiration and positioning on prevertebral (retropharyngeal) soft tissues. **A.** Increased prevertebral space with expiration. **B.** Repeat radiograph in same patient during inspiration reveals normal prevertebral space with no suggestion of cervical spine abnormality. **C.** Increased soft-tissue space with expiratory phase of respiration. **D.** Normal soft-tissue space with inspiration in patient **C.** (**A** and **B** reprinted with permission from Harris J, Edeiken-Monroe B. The Radiology of Acute Cervical Spine Trauma. 2nd ed. Baltimore: Williams & Wilkins, 1987:6.)

SPECIFIC INJURIES

The Jefferson fracture is a bursting fracture of the ring of C-1 as a result of an axial load. The axial force compresses the ring of C-1 between the occipital condyles of the skull and the lateral masses of C-2. This reaction can cause an outward burst of C-2, but it rarely causes immediate neurologic impairment because the fracture does not physically impinge on the spinal cord. The radiographic criteria for diagnosis of a Jefferson fracture is lateral offset of the lateral mass of C-1 of greater than 1 mm from the vertebral body of C-2 (Fig. 106.23). Neck rotation may give a false-positive radiograph. These fractures are unstable, however, and require adequate immobilization. Approximately one-third of Jefferson fractures are associated with other cervical spine fractures, most often involving C-2. The clinician must be aware of the pseudo-Jefferson fracture of childhood, which is present in 90% of children at the age of 2 years and usually normalizes by age 4 to 6 years. The pseudo-Jefferson fracture has the radiographic appearance of a Jefferson fracture because of increased growth of the atlas (C-1) compared with the axis (C-2) and radiolucent cartilage artifact. This disorder can present with unilateral or bilateral lateral mass offset. If a Jefferson fracture is suspected by radiographic findings and mechanism of injury in a child less than 4 years of age, a CT scan may be necessary to further elucidate the suspected injury (Fig. 106.15).

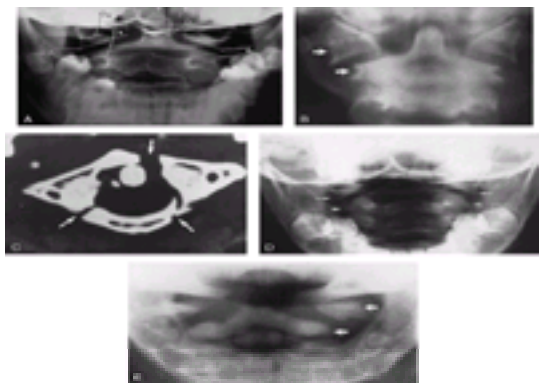

FIGURE 106.23. A. Normal anteroposterior (AP) (open-mouth, odontoid) view of C-1 and C-2. *C_1,* first cervical vertebra (lateral mass); *C_2,* second cervical vertebra; *T,* central incisors overlying dens *(D);A,* normal relationship between lateral mass of C-1 and vertebral body of C-2. **B.** Jefferson fracture in AP view. Note lateral offset of C-1 on C-2. **C.** Jefferson fracture. Computed tomography coronal view. Note three distinct fractures and bursting nature of injury. **D.** Pseudo-Jefferson fracture of childhood in a 3-year-old child because of disparate growth of C-1 and C-2 and cartilage artifact. **E.** Pseudo-Jefferson fracture demonstrating marked offset of the lateral masses of C-1 on C-2. (**B** and **C** reprinted with permission from Swischuk L. Emergency Radiology of the Acutely Ill or Injury Child. 2nd ed. Baltimore: Williams & Wilkins, 1986:591; **D** reprinted with permission from Aslamy W, Danielson K, Hessel S, et al. A 3-year old boy with neck pain after motor-vehicle accident. West Med J 1991;155:301–302.)

The hangman's fracture is a traumatic spondylolisthesis of C-2. This injury occurs as a result of hyperextension, which fractures the posterior elements of C-2. Hyperflexion, with resultant ligamentous damage, may follow the hyperextension or may lead to anterior subluxation of C-2 on C-3 and subsequent damage of the cervical cord (Fig. 106.24). The subluxation associated with a hangman's fracture can sometimes be mistaken for the normal or physiologic subluxation that exists in the C2–C3 or C3–C4 region in approximately 25% of children less than the age of 8 years; it also may be seen up to the age of 16. This pseudosubluxation is caused by ligamentous laxity, relatively horizontal facet joints, weak neck muscles, and cartilage artifact. Distinguishing between a subtle hangman's fracture and pseudosubluxation can be accomplished using Swischuk's “posterior cervical line” as described in Figure 106.25. A value of more than 1.5 to 2 mm suggests an occult hangman's fracture as the source of the anterior subluxation of C-2 on C-3. The increase in

magnitude of the distance between the cortex of the spinous process of C-2 and the posterior cervical line in a hangman's fracture is the result of anterior displacement of the skull, C-1 and the anterior portion of C-2 on the remainder of the lower cervical spine.

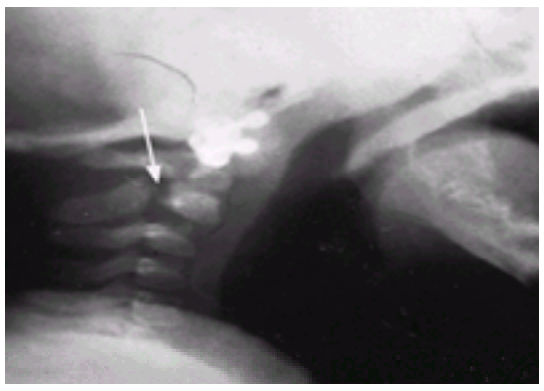

FIGURE 106.24. Hangman's fracture. A 7-week-old infant with fracture through the posterior elements of C-2 as indicated by the *arrow.* (Reprinted with permission from Sumchai A, Sternback G. Hangman's fracture in a 7-week-old infant. Ann Emerg Med 1991;20:87.)

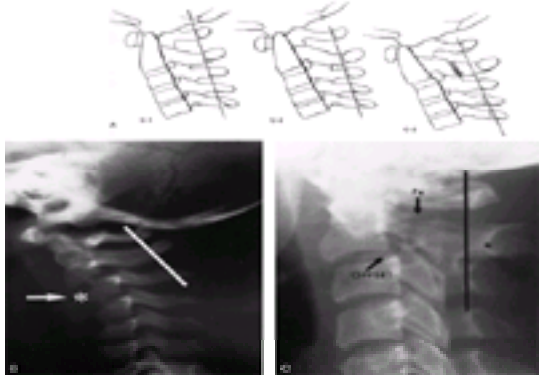

FIGURE 106.25. A. Posterior cervical line of Swischuk. Line is drawn from the cortex of the spinous process of C-1 to the cortex of the spinous process of C-3. Relationship of the line to cortex of the spinous process of C-2 is noted. If the line is situated more than 2.0 mm anterior to the cortex of the spinous process of C-2, underlying cervical pathology should be present. This line should only be used with anterior displacement of C-2 on C-3. **B.** Pseudosubluxation of C-2 on C-3 with normal posterior cervical line in a 2-year-old child. Note apparent widening of prevertebral soft tissue. **C.** Abnormal posterior cervical line with underlying hangman's fracture. Actual offset is 4 mm (**C** reprinted with permission from Swischuk L. Emergency Radiology of the Acutely Ill or Injured Child. 2nd ed. Baltimore: Williams & Wilkins, 1986:562–563.)

Atlantoaxial (AA) subluxation is a result of movement between C-1 and C-2 secondary to transverse ligament rupture or a fractured dens (Fig. 106.26). Ligament instability may be precipitated by tonsillitis, cervical adenitis, pharyngitis, arthritis, or connective tissue disorders. It is also well described in patients with Down syndrome. Approximately 15% of patients with Down syndrome have radiographically demonstrated AA subluxation and therefore should be discouraged from contact sports. The presence or absence of AA subluxation in patients with Down syndrome, once thought to be a static phenomenon, may actually be transient and/or progressive. This ligament instability may progress to ligament rupture with minor trauma. Subluxation caused by a transverse ligament disruption is evidenced by a widened predental (preodontoid, atlantodental interval) space on a lateral radiograph (Fig. 106.26). Normal predental measurement in children is less than 5 mm, compared with less than 3 mm in adults. This space is wider in children than in adults for the same reasons described with pseudosubluxation. Steele's rule of three states that the area within the ring of C-1 consists of one-third odontoid, one-third spinal cord, and one-third connective tissue (Fig. 106.27). Therefore, limited space is available for dens movement or predental space widening without neurologic compromise. Neurologic symptoms often are not seen until the predental space exceeds 7 to 10 mm. A dens fracture is the cause of AA subluxation more often than ligamentous disruption in a young child because the weakest part of the musculoskeletal system in a child is the osseus component (Fig. 106.26). Several case reports describe odontoid fractures in children facing forward in car seats as a result of rapid stops. These fractures may traverse the growth plate in the young child, although the clinician must be careful not to overcall fractures because of the presence of a growth plate. Neurologic damage can occur from direct spinal cord injury or secondarily from vertebral artery damage.

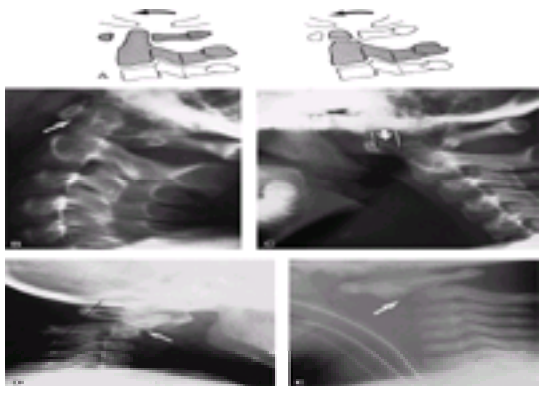

FIGURE 106.26. A. Diagrammatic representation of transverse ligament disruption *(left)* and dens fracture *(right).* **B.** Widened predental space on initial lateral radiograph in 15-year-old girl (actual measurement was 4 mm). **C.** Flexion radiograph in same patient demonstrating increased predental space with evidence of transverse ligament disruption. (**A** used with permission from Swischuk L. Emergency Radiology of the Acutely Ill or Injured Child. 2nd ed. Baltimore: Williams & Wilkins, 1986:572.) **D.** Dens fracture with anterior subluxation of C-1 and the dens on the remainder of the spinal column. *Arrow* indicates fracture. Abnormal posterior cervical line is also shown. **E.** Dens fracture *(arrow)* with anterior subluxation of the dens on the body of C-2.

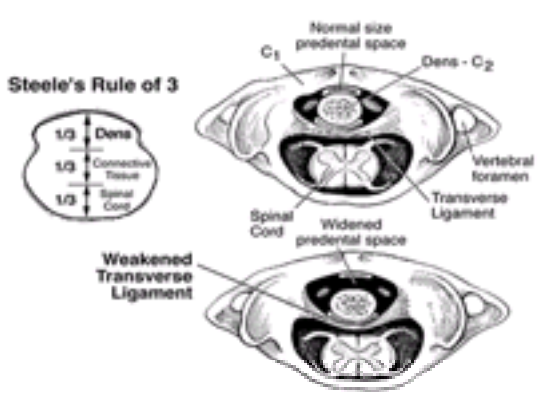

FIGURE 106.27. A cross section through the ring of C1 demonstrates Steele's rule of three. The space between the cervical cord and dens allows limited movement between C1 and C2 without immediate neurologic compromise.

Cervical distraction injuries may result from rapid acceleration- or deceleration-type incidents, such as high-speed motor vehicle or pedestrian accidents (Fig. 106.28). An injury that was incompatible with long-term survival, but for which initial cardiopulmonary resuscitation was successful, is shown in Figure 106.28A. These injuries may be obvious or subtle on the lateral radiograph. Measurements for potential distraction injuries include the atlanto-occipital and C1/C2 interspinous distances. The atlanto-occipital distance should not exceed 5 mm. The C1/C2 interspinous distance should not exceed 10 mm. A ratio of measurements of the basion to the posterior arch of C1 (BC) and the opisthion to the anterior arch of C1(OA) is demonstrated in Figure 106.29. If the BC:OA ratio is greater than 1, it signifies atlanto-occipital dislocation. Atlanto-occipital dislocation is often fatal, but there are reports of survivors. Neurologic deficits may develop from direct spinal damage or associated carotid or vertebral artery injury. Distraction injuries may also be seen with difficult newborn deliveries. These injuries may not be visible on a plain radiograph because the pediatric cervical spine can transiently distract 2 inches before residual radiographic evidence of spinal column separation is present. However, the spinal cord can distract only one-quarter inch before permanent neurologic damage occurs. An MRI scan is useful in assessing an infant with diminished motor activity who is suspected of having a distraction injury.

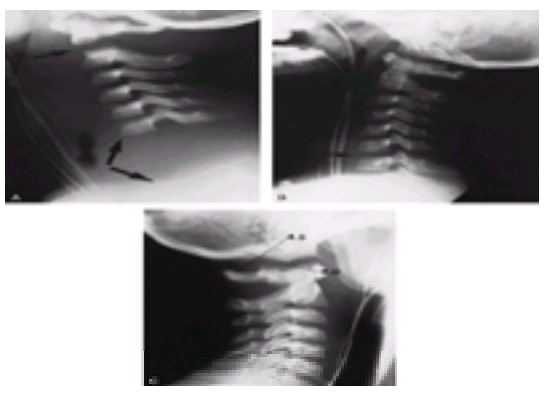

FIGURE 106.28. Distraction injuries. **A.** Dens fractures with distraction and accompanying C6–C7 distraction injury in 3-year-old child. Injury was fatal. **B.** C6–C7 distraction injury in 8-year-old child. **C.** BC:AO ratio is greater than 1.0, suggesting atlanto-occipital dislocation (see also Fig. 106.29).

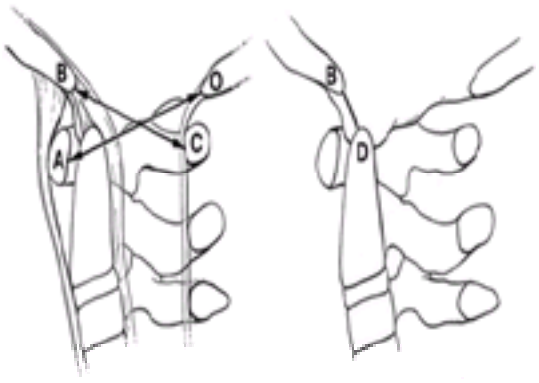

FIGURE 106.29. Examples of methods to assess occipatal/C1 relationships. *A,* C1 anterior arch; *B,* basion (anterior margin of foramen magnum); *C,* anterior portion of the posterior ring of C1; *O,* opisthion (posterior margin of foramen magnum); *D,* tip of the dens (odontoid process). These landmarks may not be easily visible on all radiographs. A BC:AO ratio of greater than 0.9 to 1.0 suggests anterior dislocation or subluxation of the atlanto-occipital joint. A BD distance of greater than 10 to 12.5 mm should be viewed as suspicious for atlanto-occipital dislocation.

Vertebral compression injuries are suggested by isolated anterior wedging, teardrop fractures, or burst vertebral bodies (Fig. 106.30). The vertebral bodies should be regular, cuboid, and consistent between adjacent cervical levels (Fig. 106.30). A flexion/rotation stress can lead to anterior subluxation of one vertebral body on another with facet dislocation ("locked" or "jumped" facet) (Fig. 106.31). If the anterior displacement is less than 50% of the vertebral body width, it is consistent with a unilateral facet dislocation (Fig. 106.31). More than a 50% anterior subluxation suggests a bilateral facet dislocation (Fig. 106.31). These injuries are often accompanied by widened interspinous and interlaminar spaces, anterior soft-tissue swelling, and a narrowed disc space.

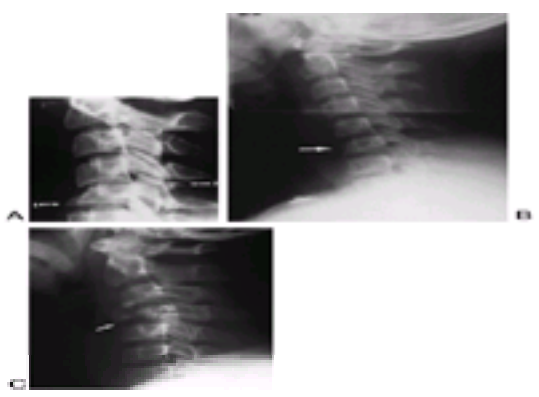

FIGURE 106.30. Examples of cervical compression injuries. **A.** Teardrop fracture. This patient sustained a whiplash injury with resultant flexion injury. A typical flexion teardrop fracture is demonstrated at *(1). (2)* demonstrates an increased interspinous distance and an associated avulsion fracture of the posterior elements of C5. **B.** Anterior C6 vertebral wedge fracture. (**A** reprinted with permission from Swischuk L. Emergency Radiology of the Acutely Ill or Injured Child. 2nd ed. Baltimore: Williams & Wilkins, 1986:674.) **C.** Burst fracture of C4 vertebral body.

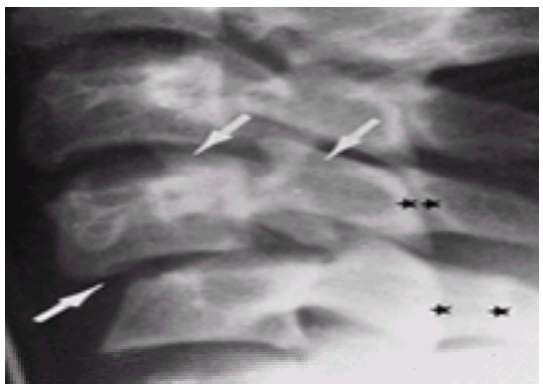

FIGURE 106.31. Unilateral facet dislocation. C4 is offset anteriorly on C5 less than 50% of the width of the vertebral body. *Arrows* denote the offset of vertebral body and apophyseal joints. The disc space between C4 and C5 is narrowed. Note that the distance between the posterior cortex of the apophyseal joint facet and the anterior cortex of the spinous process tip is wider below the level of dislocation than above the level *(stars).* Anterior vertebral offset of more than 50% would denote a bilateral facet dislocation. (Reprinted with permission from Swischuk L. Emergency Radiology of the Acutely Ill or Injured Child. 2nd ed. Baltimore: Williams & Wilkins, 1986:697.)

SCIWORA has been described as occurring in up to 67% of children with cervical cord injuries (Fig. 106.32). SCIWORA probably accounts for about 25% of cervical cord injuries in children less than 8 years of age. These injuries mainly occur in children less than 8 years of age who present with or develop symptoms consistent with cervical cord injuries without any radiographic or tomographic evidence of bony abnormality. SCIWORA is not often seen in the older population (older than age 8) because the forces necessary to injury the spinal cord also cause persistent spinal abnormalities. The young

child's elastic spinal column allows the spine to deform beyond physiologic extremes, injuring the cord, then reducing spontaneously without any persistent (radiographic) evidence of bony injury. The causes of the neurologic compromise can include vascular injury (occlusion, spasm, infarction), ligamentous injury, disc impingement, or incomplete neuronal destruction. A subset of patients have initial transient neurologic symptoms as previously described, apparently recover, and then return an average of 1 day later with significant neurologic abnormalities. Therefore, many authors recommend hospitalization and immobilization for young patients who have a history of transient neurologic symptoms. At the least, neurologic consultation is recommended if the history suggests a SCIWORA-type injury in a child less than 8 years of age.

FIGURE 106.32. Magnetic resonance imaging (MRI) of SCIWORA patient. Accompanying cervical spine radiographs were normal. The MRI demonstrates an area of cord contusion in the midcervical area. This patient had physical evidence of a central cord syndrome. (Reprinted with permission from Swischuk L. Emergency Radiology of the Acutely Ill or Injured Child. 2nd ed. Baltimore: Williams & Wilkins, 1986:710.)

Torticollis (wry neck) is a common complaint in the pediatric ED. The clinician should always inquire about traumatic causes because an underlying bone injury may be present. Often, however, torticollis is caused by spasm of the sternocleidomastoid (SCM) muscle. The patient with muscular torticollis has muscle spasm of the SCM on the opposite side that the chin points because the cause of torticollis is muscular spasm. This condition is opposite from rotary subluxation. Rotary subluxation is a cervical spine injury that is often misdiagnosed or undiagnosed because of difficulty in interpreting these patient's radiographs. Rotary subluxation or displacement may be spontaneous or may follow an upper respiratory infection or minor or major trauma. These patients rarely present with abnormal neurologic findings. They will assume the typical (cockrobin) position with the muscle spasm of the SCM on the same side as the chin points. This reaction is logical considering that the SCM is attempting to reestablish normal neck position. Radiographs may be useful to help distinguish between muscular torticollis and rotary subluxation, although the radiographs may be normal in both cases (Fig. 106.33 and Fig. 106.34). Rotary subluxation should be suspected if, on an open-mouth radiograph, one of the lateral masses of C-1 appears forward and closer to the midline, while the opposite lateral mass appears narrow and away from the midline (lateral offset), although a normal film does not rule out rotary subluxation. Cineradiography can demonstrate that C1-2 moves as a unit; however, the CT scan appears to be the most useful diagnostic tool in rotary subluxation (Fig. 106.34). Patients with mild rotary subluxation should be treated with a cervical collar and analgesia for comfort, whereas those with moderate or resilient rotary displacement may need immobilization and traction. If anterior displacement of C-2 on C-1 is present, longer immobilization may be needed to allow injured ligaments to heal.

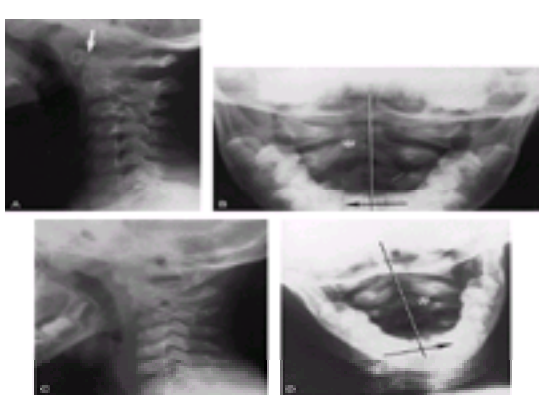

FIGURE 106.33. Torticollis (wryneck). **A.** Lateral cervical radiograph with C-2 cocked forward on C-3 and normal predental space *(arrow).* **B.** Anteroposterior (AP) view demonstrating spinous process of C-2 *(*)* on the same side of the midline as the mandible points. **C.** Difficult to interpret lateral cervical spine because of the rotation effect of torticollis. **D.** AP view demonstrating spinous process of C-2 *(*)* on the same side of the midline as the mandible points. (Reprinted with permission from Swischuk L. Emergency Radiology of the Acutely Ill or Injured Child. 2nd ed. Baltimore: Williams & Wilkins, 1986:588.)

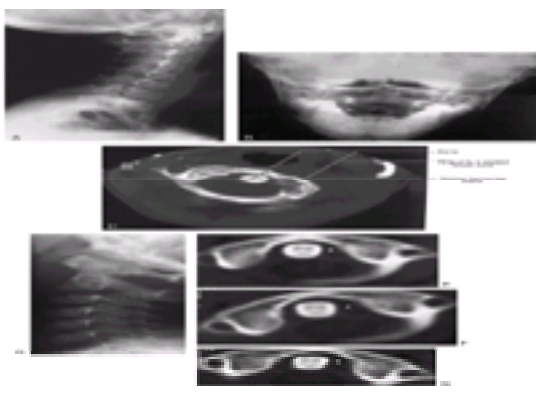

FIGURE 106.34. Rotary subluxation of C-1 and C-2. **A.** Grossly normal lateral neck radiograph in an 8-year-old child with rotary subluxation. **B.** Grossly normal open-mouth (odontoid) radiograph in an 8-year-old child with rotary subluxation. **C.** Compute tomography (CT) scan demonstrating marked rotary subluxation of C-1 clockwise around dens. Actual measurement was 22 degrees of rotation. **D–G.** CT evidence of fixed rotary subluxation in a 6-year-old child. **D.** Lateral radiograph demonstrating mild increased distance of predental space. **E.** Axial CT scan demonstrating increased distance between dens and patient's left side of C1 (asymmetry between right and left sides). **F.** Axial CT scan with patient's head turned to the right, demonstrating asymmetry between the dens and ring of C1. **G.** Axial CT scan with patient's head turned to the left, again demonstrating fixed asymmetry between the dens and the ring of C1.

Several specific spinal cord syndromes may be encountered in the ED (Fig. 106.35). A spinal cord concussion (transient traumatic paralysis) involves neurologic symptoms that completely resolve over a short period. This condition can occur with or without associated fracture or dislocation. A complete cord transection (either mechanical or physiologic) results in immediate and permanent loss of all neurologic function distal to that level (Fig. 106.35). The anterior spinal artery (anterior cord) syndrome results from loss of neurologic function in those areas supplied by the anterior spinal artery (Fig. 106.35). Motor function is lost below the level of the lesion. Touch and proprioceptive functions, carried by the dorsal (posterior) columns, are preserved. The posterior cord syndrome is rare (Fig. 106.35). It involves loss of proprioceptive functions, deep pressure and pain and vibratory sense, with preservation of motor and temperature sensation. This can occur with direct posterior cord trauma or posterior spinal artery involvement. The Brown-Sequard syndrome (hemisection of the cord) involves contralateral loss of pain and temperature sensation with ipsilateral motor findings (weakness or paralysis) below the lesion (Fig. 106.35). The central cord syndrome signifies an injury that is most severe in the center of the cord and less so toward the periphery (Fig. 106.35). The resultant physical examination demonstrates motor strength that is more severely affected in the arms than in the legs. These designations are useful in suggesting prognosis. Approximately two-thirds of those patients with central cord syndrome and one-third of those with Brown-Sequard recover, whereas complete transections and anterior spinal artery syndrome usually signify nonreversible lesions. Patients with posterior cord syndrome usually recover but may demonstrate some degree of ataxia.

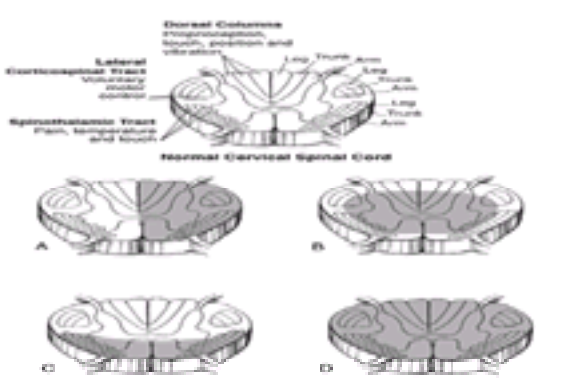

FIGURE 106.35. Graphic illustrations of a normal cervical spinal cord and specific postinjury syndromes. **A.** Brown-Sequard syndrome. **B.** Central cord syndrome. **C.** Anterior artery syndrome. **D.** Complete transection.

The os odontoideum is an abnormality that may be the result of an occult flexion injury with a subsequent incomplete healing and bone resorption (Fig. 106.36). It may also represent an overgrowth of the ossiculum terminale, often associated with a hypoplastic dens. This leads to a risk of increased mobility and cord injury at the C1/C2 level and may require surgical stabilization. This condition can be confused with a fracture at the base of the odontoid. The ossiculum terminale is a small ossicle at the tip of the dens (Fig. 106.37). It is seen in most children, fusing with the rest of the dens by adolescence. This ossicle can be large and associated with a hypoplastic dens, as previously described.

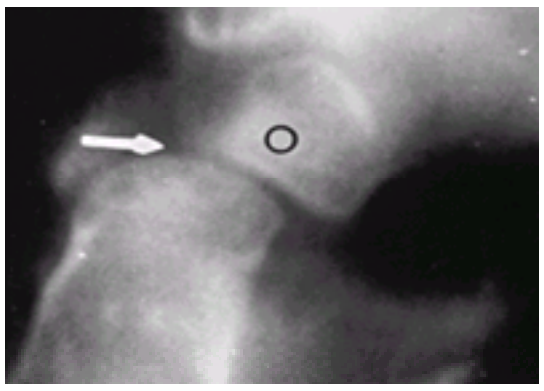

FIGURE 106.36. Example of os odontoideum. Note the hypoplastic dens and overgrown ossiculum terminale or ossiculum odontoideum *(O).* The *arrow* indicates posterior displacement, attesting to instability of the lesions. (Reprinted with permission from Swischuk L. Emergency Radiology of the Acutely Ill or Injured Child. 2nd ed. Baltimore: Williams & Wilkins, 1986:717.)

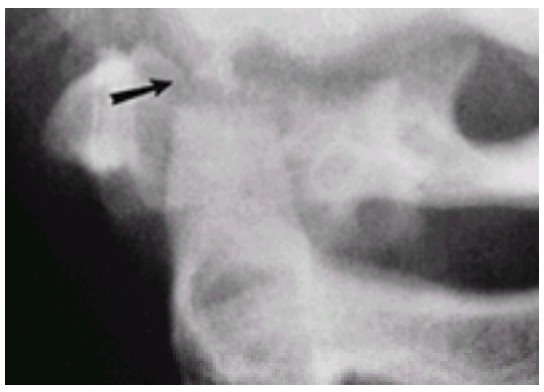

FIGURE 106.37. Normal ossiculum terminale at the tip of the dens *(arrow).* (Reprinted with permission from Swischuk L. Emergency Radiology of the Acutely Ill or Injured Child. 2nd ed. Baltimore: Williams & Wilkins, 1986:717.)

Spinal epidural hematomas are also seen in the pediatric population. These hematomas are venous bleeds that compress the adjacent spinal cord and present hours or days after apparently minor trauma, with ascending neurologic symptoms as the bleed progresses. The MRI scan can be helpful in evaluating these patients (Fig. 106.38). Rapid evaluation and surgical decompression are mandatory.

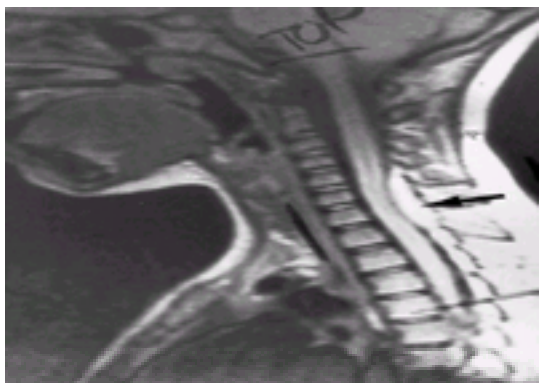

FIGURE 106.38. Magnetic resonance imaging (MRI) of cervical spine demonstrating epidural hematoma *(arrow)* from C-5 to T-1. Note excellent soft tissue, intervertebral disc, and fluid detail afforded by the MRI scan.

Treatment of children with suspected cervical spine injuries involves basic and advanced life support measures, initiation and/or maintenance of immobilization, neurosurgical consultation, and consideration of pharmacologic treatment. Airway support for patients with traumatic quadriplegia should be considered because they will develop respiratory embarrassment as they tire. Children may present in spinal shock (hypotension, bradycardia, peripheral flush) from the loss of sympathetic input to the vascular system. The physical examination may be misleading in that these patients are bradycardic (unable to mount tachycardic response to relative hypovolemia) and demonstrate warm, flushed skin (loss of vasomotor tone). These symptoms may also be superimposed upon traumatic (hypovolemic) shock.

These patients need fluid resuscitation and may require inotropic support such as dopamine to maintain adequate perfusion and to avoid fluid overload. Appropriate fluid management is important in preventing hypoperfusion of the already injured spinal cord. Recent investigations suggests that methylprednisolone (Solu-Medrol) in a dosage of 30 mg/kg over 15 minutes followed by 5.4 mg/kg per hour for 24 to 48 hours may improve functional outcome in patients with spinal cord injury. Although these studies specifically excluded children less than 13 years of age, the medication may be helpful for younger patients. Methylprednisolone appears to be most effective if administered as soon as possible after the injury and maintained for 24 to 48 hours depending on the time of initiation (Table 106.9). If started within 3 hours, it should be maintained for 24 hours. If started between 3 to 8 hours after the injury, continuation for 48 hours is

recommended. Methylprednisolone is not recommended in conjunction with penetrating neck injuries.

Time after Injury	0–3 hrs	3–8 hrs	>8 hrs
Initial IV dosage	30 mg/kg (over 15 mins)	30 mg/kg (over 15 mins)	Efficacy not demonstrated
Maintenance IV dosage	5.4 mg/kg/hr	5.4 mg/kg/hr	
Suggested duration	24 hrs	48 hrs	

Table 106.9. Suggested Methylprednisolone Administration Schedule for Blunt Cervical Spinal Cord Trauma

Suggested Readings

BLUNT AND PENETRATING NECK TRAUMA

Adams V, Hirsch C. Venous air embolism from head and neck wounds. Arch Pathol Lab Med 1989;113:498–502.

Back MR, Baumgartner FJ, Klein SR. Detection and evaluation of aerodigestive tract injuries caused by cervical and transmediastinal gunshot wounds. J Trauma 1997;42:680–686.

Balkany T, Rutherford R, Narrod J, Jafek B. The management of neck injuries. In: Zuidema G, Rutherford R, Ballinger W, eds. The Management of Trauma. 4th ed. Philadelphia: WB Saunders, 1985:359–378.

Biller H, Lawson W. Management of acute laryngeal trauma. In: Bailey B, Biller H, eds. Surgery of the Larynx. Philadelphia: WB Saunders, 1985:149–154.

Birney J, Kulig K. Laryngeal emergencies. In: Kravis T, Warner C, eds. Emergency Medicine: A Comprehensive Review. Rockville, MD: Aspen, 1987:1265–1269.

Brennan J, Meyers A, Jafek B. Penetrating neck trauma: a five year review of the literature, 1983–1988. Am J Otolaryngol 1990;11:191–197.

Cassisi N, Isaacs J Jr. Trauma. In: Cummings C, ed. Otolaryngology—Head and Neck Surgery. St Louis: Mosby, 1986:1943–1964.

Conlan A, Wessels A, Hammond C, et al. Pharyngoesophageal barotrauma in children: a report of six cases. J Thorac Cardiovasc Surg 1984;88: 452–456.

Cooper A, Barlow B, Niemirska M, Gandhi R. Fifteen years' experience with penetrating trauma to the head and neck in children. J Pediatr Surg 1987;22:24–27.

Frykberg E, Vines F, Alexander R. The natural history of clinically occult arterial injuries: a prospective evaluation. J Trauma 1989;29:577–583.

Gallia L. Laryngotracheal trauma. In: Blaisdell F, Trunkey D, eds. Trauma Management. Vol III: Cervicothoracic Trauma. New York: Thieme, 1986:117–128.

Gold SM, Gerber ME, Shott SR, Myer CM III. Blunt laryngotracheal trauma in children. Archives of Otolaryngology—Head and Neck Surgery 1997;123:83–87.

Goodnight J. Cervical injury. In: Blaisdell F, Trunkey D, eds. Trauma Management. Vol III: Cervicothoracic Trauma. New York, Thieme, 1986:94–102.

Grewal H, Rao PM, Mukerji S, Ivatury RR. Management of penetrating laryngotracheal injuries. Head and Neck 1995;17:494–502.

Hall J, Reyes H, Meller J. Penetrating zone-II neck injuries in children. J Trauma 1991;31:1614–1617.

Hancock B, Wiseman N. Tracheobronchial injuries in children. J Pediatr Surg 1991;26:1316–1319.

Humar A, Pitters C. Emergency department management of blunt cervical tracheal trauma in children. Pediatr Emerg Care 1991;7:291–293.

Hurst J. Common Problems in Trauma. Chicago: Year Book Medical Publishers, 1987:130–138.

Jeffrey R. CT of laryngeal trauma. In: Federle M, Brant-Zawadzki M, eds. Computed Tomography in the Evaluation of Trauma. 2nd ed. Baltimore: Williams & Wilkins, 1986:108–115.

Kadish H, Schunk J, Woodward GA. Blunt pediatric laryngotracheal trauma: case reports and review of the literature. Am J Emerg Med 1994;12:207–211.

Lee C, Woodring J, Walsh J. Carotid and vertebral artery injury in survivors of atlanto-occipital dislocation: case reports and literature review. J Trauma 1991;31:401–407.

Lowry L. Trauma. In: Cummings C, ed. Otolaryngology—Head and Neck Surgery. St Louis: Mosby, 1986:1621–1630.

Lupetin AR. Computed tomographic evaluation of laryngotracheal trauma. Current Problems in Diag Radiol 1997;26:185–206.

Lusk R. The evaluation of minor cervical blunt trauma in the pediatric patient. Clin Pediatr 1986;25:445–447.

Martin W, Gussack G. Pediatric penetrating head and neck trauma. Laryngoscope 1990;100:1288–1291.

Reece G, Shatney C. Blunt injuries of the cervical trachea: review of 51 patients. South Med J 1988;81:1542–1548.

Shearer VE, Giesecke AH. Airway management for patients with penetrating neck trauma: a retrospective study. Anesth Analg 1993;77(6):1135–1138.

Simpson RK, Venger BH, Narayan RK. Treatment of acute penetrating injuries of the spine: a retrospective analysis. J Trauma 1989;29(1):42–46.

Splavski B, Vrankovic D, Blagus G, et al. Spinal stability after war missile injuries of the spine. J Trauma: Injury, Infection, and Critical Care 1996;41(5):850–853.

Stanley RB Jr., Armstrong WB, Fetterman BC, Shindo MC. Management of external penetrating injuries into the hypopharyngeal-cervical esophageal funnel. J Trauma 1997;42:675–679.

Swor R. Penetrating and blunt neck trauma. In: Tintinalli J, Krome R, Ruiz E, eds. Emergency Medicine: A Comprehensive Study Guide. 2nd ed. New York: McGraw-Hill, 1988:846–849.

Szeremeta W, Morovati S. Isolated hyoid bone fracture: a case report and review of the literature. J Trauma 1991;31:268–271.

Vadasz AG, Torres CF, Chang JK. Accidental penetrating cervical cord injury in a young child. Pediatr Emerg Care 1996;12(6):428–431.

Walt A, ed. Early Care of the Injured Patient. 3rd ed. Philadelphia: WB Saunders, 1982:222–234.

Ward R. Injury to the cervical cerebral vessels. In: Blaisdell F, Trunkey D, eds. Trauma Management. Vol III: Cervicothoracic Trauma. New York: Thieme, 1986:262–281.

Watson JM, Goldstein LJ. Golf club shaft impalement: case report of a zone III neck injury. J Trauma 1996;41(6):1036–1038.

Way L. Current Surgical Diagnosis and Treatment. 8th ed. Norwalk, CT: Appleton & Lange, 1988:195–197.

Wiener S, Barrett J, eds. Trauma Management for Civilian and Military Physicians. Philadelphia: WB Saunders, 1986:172–191.

CERVICAL SPINE

Apple J, Kirks D, Merten D, Martinez S. Cervical spine fractures and dislocations in children. Pediatr Radiol 1987;17:45–49.

Aslamy W, Danielson K, Hessel S, Levy J. A 3-year-old boy with neck pain after motor-vehicle accident. West J Med 1991;155:301–302.

Bachulis B, Long W, Hynes G, Johnson M. Clinical indications for cervical spine radiographs in the traumatized patient. Am J Surg 1987;153:473–478.

Bivins H, Ford S, Bezmalinovic Z, et al. The effects of axial traction during orotracheal intubation of the trauma victim with an unstable cervical spine. Ann Emerg Med 1988;17:53–57.

Bohn D, Armstrong D, Becker L, Humphreys R. Cervical spine injuries in children. J Trauma 1990;30:463–469.

Bracken M, Shepard MJ, Collins W, et al. A randomized, controlled trial of methylprednisolone or naloxone in the treatment of acute spinal-cord injury: results of the second national acute spinal-cord injury study. New Engl J Med 1990;322:1405–1411.

Bracken M, Shepard MJ, Holford TR, et al. Administration of Methylprednisolone for 24 or 48 hours or Tirilazad Mesylate for 48 hours in the treatment of acute spinal cord injury: results of the third national acute spinal cord injury randomized controlled trial. JAMA 1997;277:1597–1604.

Bulas D, Fitz C, Johnson D. Traumatic atlanto-occipital dislocation in children. Radiology 1993;188:155–158.

Cohen A, Hirsch M, Katz M, Sofer S. Traumatic atlanto-occipital dislocation in children: review and report of five cases. Pediatr Emerg Care 1991; 7:24–27.

Criswell JC, Parr MJ, Nolan JP. Emergency airway management in patients with cervical spine injuries. Anaesthesia 1994;49(10):900–903.

Curran C, Dietrich AM, Bowman MJ, et al. Pediatric cervical-spine immobilization: achieving neutral position? J Trauma 1995;39(4):729–732.

David S, Salluzzo RF, Bartfield JM, Dickinson ET. Spontaneous cervicothoracic epidural hematoma following prolonged Valsalva secondary to trumpet playing. Am J Emerg Med 1997;15(1):73–75.

Davis JW, Parks SN, Detlefs CL, Williams GG, Williams JL, Smith RW. Clearing the cervical spine in obtunded patients: the use of dynamic fluoroscopy. J Trauma 1995;39(3):435–438.

Dickman C, Rekate H, Sonntag V, Zabramski J. Pediatric spinal trauma: vertebral column and spinal cord injuries in children. Pediatr Neurosci 1989;15:237–256.

Doris P, Wilson R. The next logical step in the emergency radiographic evaluation of cervical spine trauma: the five-view trauma series. J Emerg Med 1985;3:371–385.

Elliot S, Morton R, Whitelaw R. Atlantoaxial instability and abnormalities of the odontoid in Down's syndrome. 1988;63:1484–1489.

Evans D, Bethem D. Cervical spine injuries in children. J Pediatr Orthop 1989;9:563–568.

Fesmire F, Luten R. The pediatric cervical spine: developmental anatomy and clinical aspects. J Emerg Med 1989;7:133–142.

Fuchs S, Barthel M, Flannery A, Christoffel K. Cervical spine fractures sustained by young children in forward-facing car seats. Pediatrics 1989;84:348–354.

Gerlock A, Kirchner S, Heller R, Kaye J. Advanced Exercises in Diagnostic Radiology: The Cervical Spine in Trauma. Philadelphia: WB Saunders, 1978.

Harris J. Radiographic evaluation of spinal trauma. Orthop Clin North Am 1986;17:75–86.

Harris J, Edeiken-Monroe B. The Radiology of Acute Cervical Spine Trauma. 2nd ed. Baltimore: Williams & Wilkins, 1987.

Herzenberg J, Hensinger R, Dedrick D, Phillips W. Emergency transport and positioning of young children who have an injury of the cervical

spine: the standard backboard may be hazardous. J Bone Joint Surg 1989; 71-A:15–22.

Huerta C, Griffith R, Joyce S. Cervical spine stabilization in pediatric patients: evaluation of current techniques. Ann Emerg Med 1987;16:55–60.

Ivy ME, Cohn SM. Addressing the myths of cervical spine injury management. Am J Emerg Med 1997;15:591–595.

Jaffe D, Binns H, Radkowski M, et al. Developing a clinical algorithm for early management of cervical spine injury in child trauma victims. Ann Emerg Med 1987;16:57–63.

Jones E, Hensinger R. The x-ray look of young spines. Emerg Med 1982; 7:74–87.

Kriss VM, Kriss TC. Imaging of the cervical spine in infants. Pediatr Emerg Care 1996;13(1):44–49.

Kriss VM, Kriss TC. SCIWORA (spinal cord injury without radiographic abnormality) in infants and children. Clin Pediatr 1996;35(3):119–124.

Lasker M, Torres-Torres M, Green R. Neonatal diagnosis of spinal cord transection. Clin Pediatr 1991;30:322–324.

Levine A, Edwards C. The management of traumatic spondylolisthesis of the axis. J Bone Joint Surg 1985;67-A:217–226.

Levitt M, Flanders A. Diagnostic capabilities of magnetic resonance imaging and computed tomography in acute cervical spinal column injury. Am J Emerg Med 1991;9:131–135.

MacDonald R, Schwartz M, Mirich D, et al. Diagnosis of cervical spine injury in motor vehicle crash victims: how many x-rays are enough? J Trauma 1990;30:392–397.

McKee T, Tinkoff G, Rhodes M. Asymptomatic occult cervical spine fracture: case report and review of the literature. J Trauma 1990;30:623–626.

Morell RC, Colonna DM, Mathes DD, Wilson JA. Fluoroscopy-assisted intubation of a child with an unstable subluxation of C1/C2. J Neurosurg Anesthesiol 1997;9(1):25–28.

Nitecki S, Moir CR. Predictive factors of the outcome of traumatic cervical spine fracture in children. J Pediatr Surg 1994;29(11):1409–1411.

Orenstein JB, Klein BL, Gotschall CS, et al. Age and outcome in pediatric cervical spine injury: 11-year experience. Pediatr Emerg Care 1994;19(3):132–137.

Pang D, Pollack I. Spinal cord injury without radiographic abnormality in children—the SCIWORA syndrome. J Trauma 1989;29:654–664.

Pang D, Wilberger J. Spinal cord injury without radiographic abnormality in children. J Neurosurg 1982;57:114–129.

Pennecot G, Leonard P, Peyrot Des Gachons S, et al. Traumatic ligamentous instability of the cervical spine in children. J Pediatr Orthop 1984;4:339–345.

Prendergast MR, Saxe JM, Ledgerwood AM, et al. Massive steroids do not reduce the zone of injury after penetrating spinal cord injury. J Trauma 1994;37:576–580.

Proudfoot J. Pediatric cervical spine injury: navigating the nuances and minimizing complications. Pediatr Emerg Med Reports 1996;1:83–94.

Rachesky I, Boyce W, Duncan B, et al. Clinical prediction of cervical spine injuries in children: radiographic abnormalities. Am J Dis Child 1987;141:199–201.

Ringenberg B, Fisher A, Urdaneta L, Midthum M. Rational ordering of cervical spine radiographs following trauma. Ann Emerg Med 1988; 17:792–796.

Ross S, Schwab C, David E, et al. Clearing the cervical spine: initial radiographic evaluation. J Trauma 1987;27:1055–1060.

Schafermeyer R, Ribbeck B, Gaskins J, et al. Respiratory effects of spinal immobilization in children. Ann Emerg Med 1991;20:1017–1019.

Schwartz GR, Wright SW, Fein JA, et al. Pediatric cervical spine injury sustained in falls from low heights. Ann Emerg Med 1997:30(3)249–252.

Schwarz N, Buchinger W, Gaudernak T, et al. Injuries to the cervical spine causing vertebral artery trauma: case reports. J Trauma 1991;31: 127–133.

Smith D. Atlanto-occipital dislocation. J Emerg Med 1992;10:699–703.

Stauffer E, Mazur J. Cervical spine injuries in children. Pediatr Ann 1982;11:502–511.

Streitweiser D, Knopp R, Wales L, et al. Accuracy of standard radiographic views in detecting cervical spine fractures. Ann Emerg Med 1983;12:35–39.

Sumchai A, Sternbach G. Hangman's fracture in a 7-week-old infant. Ann Emerg Med 1991;20:86–89.

Suss R, Zimmerman R, Leeds N. Pseudospread of the atlas: false sign of Jefferson fracture in young children. Am J Roentgenol 1983;140: 1079–1082.

Swischuk L. Emergency Radiology of the Acutely Ill or Injured Child. 3rd ed. Baltimore: Williams & Wilkins, 1994:653–735.

Treloar DJ, Nypaver M. Angulation of the pediatric cervical spine with and without cervical collar. Pediatr Emerg Care 1997;13(1):5–8.

Wilberger J. Spinal Cord Injuries in Children. New York: Futura, 1986.

Woodring J, Selke A, Duff D. Traumatic atlantooccipital dislocation with survival. Am J Roentgenol 1981;137:21–24.

Woodward GA, Kunkel CN. Cervical spine immobilization and imaging. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:329–341.

Vanden Hoek T, Propp D. Cervicothoracic junction injury. Am J Emerg Med 1990;8:30–33.

Zwimpfer T, Bernstein M. Spinal cord concussion. J Neurosurg 1990; 72:894–900.

CHAPTER 107

Thoracic Trauma

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Thoracic trauma in the pediatric population is relatively uncommon and only in the last two decades has it received careful scrutiny. Included in thoracic trauma are injuries to the chest wall, trachea, bronchi, lungs, heart, thoracic aorta and great vessels, esophagus, and diaphragm. The report of the National Pediatric Trauma Registry contains a detailed analysis of major thoracic trauma in children. In the United States, only 4 to 6% of children admitted to pediatric trauma centers have thoracic injuries, but because severe injuries to vital thoracic organs produce profound physiologic disturbances, many patients die at the scene and never reach a hospital. In patients who reach the hospital, most thoracic injuries do not require operative intervention other than tube thoracostomy.

When thoracic trauma occurs in isolation, the mortality rate is relatively low. The mortality rate triples when thoracic trauma occurs concurrently with head or abdominal trauma. In one study, 82% of patients with thoracic trauma had a multisystem injury and 58% of those patients had a concomitant head injury. In the same study, children with thoracic trauma had a lower mean trauma score and a higher mean injury severity score. Mortality rates were 20 times higher for children with thoracic involvement than for those without. Most scene fatalities result from lacerations of the lung, heart, blood vessels, and bronchi. In the hospital, cardiac tamponade, injuries to the aorta and great vessels, and tension hemothorax or pneumothorax have the greatest potential for death.

Similar to other types of pediatric trauma, blunt thoracic injuries are more common than penetrating. Motor vehicle–related accidents account for at least 75% of all blunt thoracic injuries. Other common mechanisms include falls, assaults, and bicycle accidents. Penetrating wounds occur in approximately 15% of children sustaining major thoracic trauma. Gunshot wounds followed by stab wounds are the most common mechanism of penetrating thoracic trauma, although the mechanism varies with geographic location.

The most common injuries in blunt thoracic trauma include lung contusions (53%), pneumothorax or hemothorax (38%), and fractures (38%). Pneumothorax or hemothorax (64%) occur more often in penetrating thoracic trauma, and diaphragmatic (15%), cardiac (13%), and vascular injuries (10%) are also common in penetrating thoracic trauma.

PATHOPHYSIOLOGY

Children with thoracic trauma may present in respiratory or circulatory failure. Children in respiratory failure, which is more common than circulatory failure, may present with tachypnea, chest wall retractions, and agitation secondary to hypoxia.

Respiratory Failure

Because approximately 80% of thoracic trauma occurs as part of a multisystem injury, the most common cause of respiratory failure is neurologic compromise. If left untreated, patients develop anoxic brain injury and respiratory

acidosis caused by hypercarbia. Immediate control of the airway with positive-pressure ventilation followed by treatment of the neurologic emergency is indicated.

Airway obstruction, external compression of the pulmonary structures, direct injury to the pulmonary parenchyma, and chest wall injuries, all caused by thoracic trauma, also affect oxygenation and ventilation if left untreated.

Airway Obstruction

Blood, vomitus, or foreign bodies (teeth) may obstruct the airway. Failure to remove or bypass the foreign body leads to hypoxia and anoxic brain injury. Initial treatment includes repositioning and suctioning of the airway along with cervical spine immobilization. If initial treatment fails, endotracheal intubation is indicated. If the endotracheal tube cannot bypass the obstruction, a cricothyroidotomy or surgical tracheostomy should be performed.

External Compression of the Pulmonary Structures

External compression of the lungs, most commonly caused by air or blood within the pleural cavity, causes respiratory distress. Patients may initially present with tachypnea, retractions, and hypoxia. As more lung segments are involved, ventilation is affected and the carbon dioxide begins to rise. Tube thoracostomy is the preferred treatment for a pneumothorax or hemothorax.

Diaphragmatic hernia and gastric dilation can also cause respiratory distress by external compression of the lungs. Patients with a diaphragmatic hernia need prompt surgical intervention. A nasogastric or oral gastric tube helps decompress the stomach and decreases the likelihood of the patient aspirating vomit or swallowing blood.

Pulmonary Parenchyma Injury

Aspiration of blood or vomitus into the terminal bronchi and alveoli causes chemical pneumonitis. Direct trauma to the lung parenchyma (pulmonary contusion) also causes a leakage of blood and fluid into the alveolar space. Both of these injuries cause shunting of deoxygenated blood into the systemic circulation. Endotracheal intubation with positive-pressure ventilation is indicated if the patient's respiratory status worsens. Patients with severe chemical pneumonitis or pulmonary contusion may require high inflation pressures to maintain adequate oxygenation. These patients are at risk for pneumothorax because of high inflation pressures coupled with an already injured lung.

Although not as common as chemical pneumonitis or pulmonary contusion, penetrating thoracic trauma causes direct pulmonary parenchymal damage but is usually associated with a hemothorax or pneumothorax.

Chest Wall Injuries

Chest wall injuries, such as rib fractures or a flail chest, may cause hypoventilation as well as hypoxia secondary to pain and inadequate air exchange. Endotracheal intubation with positive-pressure ventilation is the preferred treatment in those patients with rising carbon dioxide levels or severe hypoxia.

Circulatory Failure

Thoracic hemorrhage, obstruction of venous return to the heart, or direct cardiac injury can cause circulatory compromise and shock.

Thoracic Hemorrhage

Laceration of the hilum of the lung, a great vessel, or the heart itself causes a significant amount of bleeding. Patients can hemorrhage more than 50% of total blood volume into the pleural cavity. The compensatory mechanisms to the blood loss include an increase in both the heart rate and total peripheral vascular resistance. Relying solely on the systemic blood pressure in children may be deceiving because children may lose up to 25% of their total blood volume before their systemic blood pressure decreases. Children may have a normal blood pressure but be tachycardic and poorly perfused with a prolonged capillary refill time. Therapy should be initiated before the occurrence of hypotension. Treatment includes fluid resuscitation, blood transfusion, and surgical repair if the patient continues to require multiple blood transfusions.

Obstruction of Venous Return to the Heart

A tension pneumothorax or hemothorax occurs when air or blood progressively accumulates within the pleural cavity, which causes a shift of the mediastinal structures to the contralateral side. Venous return to the heart is reduced because the inferior vena cava is relatively fixed and becomes obstructed. As diastolic filling decreases, the stroke volume of the heart drops. Patients with a tension pneumothorax or hemothorax are tachycardic and peripherally vasoconstricted, and if left untreated, develop shock. Initial treatment consists of needle decompression. If a tension pneumothorax is present, an immediate release of air should be noted and the patient's hemodynamic status should improve. Needle decompression is only a temporizing measure and must be followed by tube thoracotomy.

Direct Injury to the Heart

Myocardial contusion, ventricular or atrial rupture, and valvular disruption all may produce cardiogenic shock. Circulatory compromise results from a reduction in cardiac output, usually from a decrease in myocardial contractility. Patients may present in congestive heart failure with an enlarged liver, gallop heard on cardiac examination, and rales. Transesophageal echocardiography is helpful in identifying the type of injury. Positive inotropic agents are the preferred

drugs for improving myocardial contractility.

Pericardial tamponade, resulting from air or blood inside the pericardium, also decreases cardiac output and causes circulatory collapse. If a patient is decompensating and a pericardial tamponade is suspected, a pericardiocentesis should be performed.

DIFFERENCES BETWEEN CHILDREN AND ADULTS

Pediatric thoracic trauma differs from adult thoracic trauma in the mechanism, type of injury, and extent of involvement of other organ systems.

Falls are the most common mechanism in the infant and child. Older children are often injured as pedestrians or as unrestrained passengers in motor vehicle accidents. Adolescents are more likely to be involved as occupants in motor vehicle–related accidents. Penetrating injuries secondary to violence are more common in the adolescent population.

Lung contusion is the most common pediatric thoracic injury, with intrapleural injury second. Lacerations of the heart, great vessels, and lungs are relatively uncommon, occurring in less than 10% of thoracic trauma cases. A multicenter study of adults sustaining thoracic trauma showed that 50% had a chest wall injury and that a flail chest injury occurred in 5% of those with chest wall injuries. Injuries to the lung parenchyma occurred in 26% of patients. Simple rib fractures are the most common type of thoracic injury in adults. Only 30% of pediatric patients, compared with 50 to 75% of adults, sustain rib fractures because of increased compliance in the pediatric thoracic cage secondary to the increased cartilage content and the greater elasticity of the bones. Because of this increased compliance, kinetic energy is transferred more readily to the underlying organs. Thus, a pediatric patient may have an internal injury (lung contusion) without external evidence of trauma (e.g., rib fracture, laceration, bruising). Air or fluid within the pleural space (pneumothorax or hemothorax) more easily displace the mediastinum compromising venous return and cardiac output in children (Fig. 107.1). Adults tolerate greater mediastinal shift and compromised venous return than do pediatric patients. The internal diameter of the pediatric trachea is smaller than the adult trachea (Fig. 107.2). At rest, resistance to air flow is inversely proportional to the fourth power of the airway radius; therefore, any small amount of obstruction secondary to blood, secretions, or edema in children can cause significant respiratory distress and hypoxia. With agitation, air flow becomes turbulent and resistance to air flow increases is inversely proportional to the fifth power of the airway radius. Thus, pediatric patients with respiratory distress must be kept calm and quiet. Younger children are also more sensitive to hypoxia and may develop a reflex bradycardia or asystole.

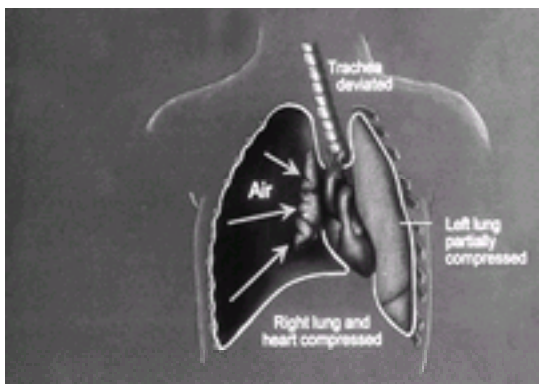

FIGURE 107.1. Tension pneumothorax with a mediastinal shift. *Please see the color-tip insert (Color Plate 107.1).*

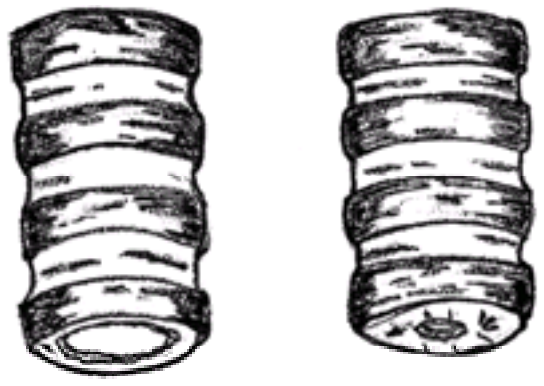

FIGURE 107.2. With swelling or edema, the internal diameter of a pediatric trachea **(A)** is much more compromised than the internal diameter of an adult trachea **(B).**

Because approximately 80% of thoracic trauma occurs as part of a multisystem injury, the physician must also consider head, neck, and intra-abdominal injuries when treating children with chest trauma. Thoracic trauma is routinely associated with abdominal trauma in children because the chest and abdominal cavities lie in close proximity (Fig. 107.3). Gastric distension may also impede pulmonary function, and a nasal or oral gastric tube may relieve this distension (Fig. 107.4).

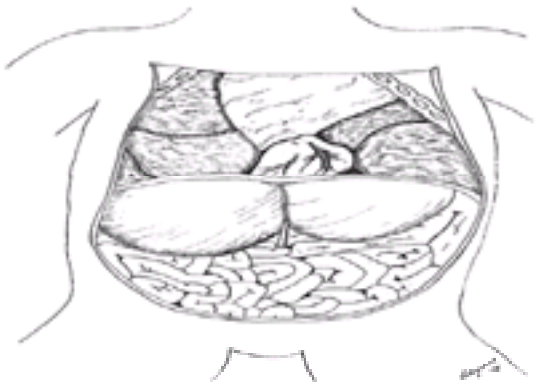

FIGURE 107.3. Drawing demonstrating the close proximity of the chest and abdominal cavities in the pediatric patient.

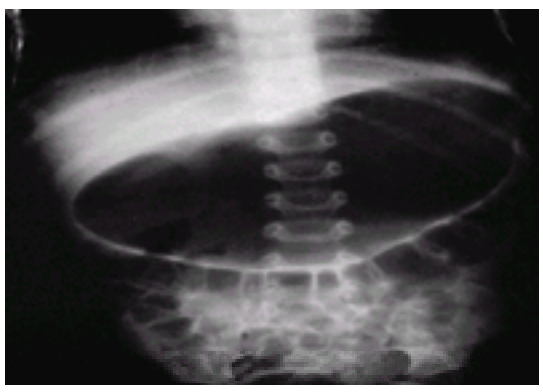

FIGURE 107.4. Radiograph of a patient with gastric distension. The patient's respiratory distress resolved once a nasogastric tube was inserted.

Mortality in thoracic trauma significantly increases with each organ system involved. Children with an isolated chest injury have a mortality rate of 5%. Children with chest and abdominal injuries have a mortality rate of 20%. The mortality rate increases to 35% in children with head and chest trauma, with more than 50% of those patients dying from their head injuries. In an adult study, 43% of patients with thoracic trauma had a concomitant head injury, but the overall mortality was only 15.5%. Children have a higher incidence of head trauma; therefore, the probability of survival in a pediatric patient with thoracic trauma is lower compared with an adult thoracic trauma patient.

Thermoregulation is a concern in the small child. The pediatric patient can lose significant heat and become hypothermic secondary to large surface area compared to small mass. Because most trauma patients need to be undressed and exposed, heating lights, warm fluids, and warming mattresses can all help stabilize the patient's body temperature.

Finally, the developmental stages of the pediatric patient must be taken into account in the evaluation of thoracic trauma. Younger children are unable to communicate effectively. They may be tachycardic and tachypneic from the injury itself or because they are frightened. Agitation may worsen an already compromised respiratory status. Inclusion of parents during evaluation and treatment is helpful in calming and reassuring children.

CLINICAL MANIFESTATIONS AND EVALUATION

Injuries to the chest can be divided into four main categories: 1) pulmonary, 2) cardiac, 3) vascular (great vessel), and 4) other intrathoracic injuries. Pulmonary injuries are the most common. Cardiac and vascular injuries are less common but have a higher mortality rate.

Pulmonary Injuries

Pulmonary injuries consist of contusion, laceration, pneumothoraces, hemothoraces, or pneumohemothoraces. Contusions are more common in blunt trauma, whereas lacerations occur more often in penetrating trauma.

Patients may complain of chest pain or difficulty breathing or have no symptoms. Physical findings may include tachypnea, asymmetric breath sounds, and/or chest wall tenderness. Patients with a tension pneumothorax may develop tachycardia, muffled heart sounds, hypotension, and/or distended neck veins. In the stable patient, chest radiography and thoracic computed tomography (CT) scan are helpful in the evaluation of pulmonary injuries.

Pulmonary injuries may initially cause hypoxia and respiratory distress, or the patient may present with minimal symptoms but progress to severe respiratory distress. Therefore, all patients with a pulmonary injury need to be observed for worsening of the disease process.

Cardiac Injuries

Cardiac injury in blunt thoracic trauma is rare, occurring in less than 5% of pediatric patients. Most patients with structural damage to the heart secondary to blunt trauma die at the scene and never reach a hospital. Cardiac contusions far outnumber lacerations. Contusions are usually self-limited unless ventricular fibrillation, which is rare, develops. On the

other hand, there are case reports of sudden death following a forceful precordial blow (commotio cordis).

Patients may complain of chest or sternal pain. Physical examination may reveal tachycardia, an irregular heart rhythm, a new heart murmur, acute onset of congestive heart failure, or in the case of cardiac tamponade, muffled heart tones. Evaluation for suspected cardiac contusion should include a 12-lead electrocardiogram (ECG), which may show ST-T wave changes or arrhythmias, and short-term observation. Creatine phosphokinase (CPK-MB) is not a useful screening tool because the test has a high false-positive rate. Symptomatic patients should be further evaluated by either a transesophageal or transthoracic echocardiogram.

Vascular (Great Vessel) Injuries

Life-threatening injuries to the great vessels of the thorax are rare and carry a high incidence of mortality. In blunt and penetrating trauma, the aorta is most commonly involved. Early detection of such injuries is vital for survival. Clinical signs and symptoms may include hypotension, paraplegia, anuria, absent or diminished femoral pulses, or excessive chest tube bleeding. Radiographic findings may include a widened mediastinum, blurred aortic knob, pleural cap, or tracheal or nasogastric tube deviation. The preferred method of diagnosis is angiography, although in the unstable patient a transesophageal echocardiogram often suffices.

Other Intrathoracic Injuries

Diaphragmatic, esophageal, and tracheobronchial disruptions are rare but are often overlooked in the initial evaluation of thoracic trauma. The chest radiograph may initially appear normal in 30 to 50% of diaphragmatic hernias. When abnormal, the chest radiograph may show a bowel gas pattern in the lungs, a displaced nasogastric tube, or an elevated hemidiaphragm. The patient may complain of chest pain or difficulty breathing. The examination may be normal or show decreased breath sounds, respiratory distress, or a scaphoid abdomen. Surgical exploration is indicated in all suspected cases because a diaphragmatic hernia does not improve without surgical correction.

Patients with esophageal and tracheobronchial disruptions may present with a continuous air leak from a chest tube, pneumomediastinum, subcutaneous emphysema, and for those patients with esophageal disruption, gastric contents from the chest tube. Bronchoscopy or esophagoscopy is indicated in suspected cases.

INITIAL MANAGEMENT

The ABCs (airway, breathing, and circulation) of trauma management apply regardless of the organ system injured. A top priority in any patient with respiratory or circulatory failure should be airway stabilization (see Chapter 5) and identification and treatment of shock (see Chapter 3). The injured child should be evaluated according to the primary survey of trauma management. The first priority in trauma patients with or without thoracic injury is establishing a secure, patent airway. Indications for endotracheal intubation in the thoracic trauma patient include depressed neurologic status, inadequate oxygenation or ventilation, compromised circulatory status, or an unstable airway including burns.

After the airway is secured, breathing is assessed. Inspection (symmetry, adequate chest rise, neck veins, displaced trachea) and auscultation (equal breath sounds, heart tones) of the chest provide information about ventilation. The ideal site for auscultation of the lungs is in the midaxillary line. An oxygen saturation by oximetry provides information on oxygenation.

If a patient has an abnormal examination but appears to be oxygenating and ventilating well and is not in shock, chest radiography is indicated. If breathing is inadequate after endotracheal intubation and breath sounds are asymmetric, intervention is required before a chest radiograph. The patient with absent breath sounds on one side and tracheal shift to the opposite side requires immediate needle decompression and subsequent tube thoracostomy. Only after the patient is stabilized should a chest radiograph be obtained.

The patient's circulatory status is evaluated after airway and breathing have been stabilized. Pericardial tamponade and a tension pneumothorax or hemothorax should be considered in the poorly perfused, shocky patient, especially when sources of blood loss have been excluded and volume resuscitation has not improved the patient's status. Physical examination may reveal muffled heart or breath sounds with decreased or absent pulses. Pericardiocentesis or thoracentesis and subsequent tube thoracostomy are lifesaving procedures that usually need to be performed in the unstable trauma patient before going to the operating room for definitive treatment.

Once the patient is stabilized and the immediate life-threatening injuries such as airway obstruction, tension pneumothorax, hemothorax, and pericardial tamponade are treated, a chest radiograph and thoracic CT scan should be obtained to provide valuable information regarding other potentially life-threatening and operative injuries. Thoracic injuries requiring operative intervention are described in Table 107.1 and in Figure 107.5. The use of ultrasound is rapidly becoming a standard diagnostic modality in the evaluation of the adult trauma patient. One adult study demonstrated that thoracic ultrasound was as sensitive and specific in identifying a hemothorax as a chest radiograph. The utility of ultrasound in the pediatric trauma patient has yet to be determined. No study has been conducted to specifically ascertain its usefulness except in the evaluation of cardiac injuries. Numerous studies have shown thoracic CT to be superior to routine chest radiograph in identifying pulmonary contusions, pneumothoraces, and hemothoraces. Thoracic CT should be part of the initial evaluation of pediatric trauma patients if a lung contusion, pneumothorax, or a hemothorax is suspected or if the cause of the patient's respiratory distress is unknown. Thoracic CT is also useful in the asymptomatic patient with chest radiograph findings suggestive of a traumatic rupture of the thoracic aorta.

Injury	Signs and Symptoms
Tracheal/bronchial rupture	Active chest tube air leak
Lung parenchyma, internal mammary artery laceration, intercostal artery laceration	Chest tube bleeding greater than 2–3 mL/kg/hr or hypotension unresponsive to transfusions
Esophageal disruption	Abnormal esophagogram (leak) or esophagoscopy Gastric contents in the chest tube
Diaphragmatic hernia	Abnormal gas pattern in the hemithorax Displaced nasogastric tube in the hemithorax
Pericardial tamponade	Positive pericardiocentesis
Great vessel laceration	Widened mediastinum Tracheal or nasogastric tube deviation Blurred aortic knob Abnormal aortogram (gold standard)

Table 107.1. Thoracic Trauma Injuries Requiring Operative Intervention

FIGURE 107.5. Indications for surgery in thoracic trauma.

Chest Wall Injuries

The elasticity and flexibility of a child's thoracic cage makes chest wall injuries less common than internal organ injuries, such as a pulmonary contusion. When chest wall injuries occur, the patient is at increased risk for intrathoracic injuries. Included in chest wall injuries are rib, sternal, and scapular fractures and flail chest.

Rib Fractures

Rib fractures may occur from either a direct blow to the rib or compression of the chest in an anterior-posterior direction. With a direct blow, the rib will fracture inward and may puncture the pleural cavity, causing a pneumothorax (Fig 107.6A and Fig 107.6B). A hemothorax is caused by a rib lacerating an intercostal artery, an internal mammary artery, or the lung parenchyma. Compression of the chest wall can cause the lateral portions of the ribs to fracture outward. Intrathoracic injury is seen less commonly with this type of fracture.

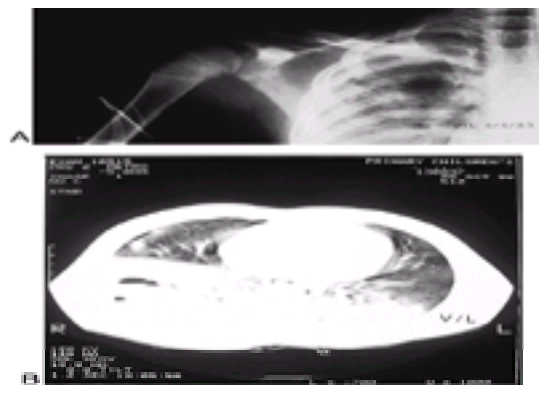

FIGURE 107.6. A 7-year-old involved in an auto–pedestrian accident. Crepitus and decreased breath sounds were noted on the right side. Chest radiograph **(A)** shows rib fractures, clavicle fracture, and a pulmonary contusion. A pulmonary contusion and a pneumothorax are seen on the thoracic computed tomography **(B).**

In one study, rib fractures occurred in 32% of all children admitted with thoracic trauma. Motor vehicle accidents were the most common mechanism of injury, similar to adult studies. Single rib fractures did not correlate with the severity of injury, but as the number of fractures increased, so did the likelihood of multisystem and intrathoracic injuries. Children with rib fractures and both head and thoracic injuries had a doubled mortality rate compared with children with rib fractures and an isolated thoracic injury.

Because of the relatively protected nature of the first rib and the amount of force required to fracture it, first rib fractures should be approached with a high index of suspicion for other serious injuries, such as vascular disruption or tracheal laceration. Patients with these lesions are usually symptomatic (hypotension, anuria, pulse differences or deficits). In one study, all patients with a first rib fracture and injury to the great vessels exhibited physical examination abnormalities such

as loss of the radial pulse on the involved side, discrepancy in blood pressure between the upper extremities, a flail chest, and/or hypotension. No patients with an isolated first rib fracture and a completely normal physical examination had an injury to a great vessel. In this and other studies, no correlation existed between level of rib fracture and associated vascular injury in the otherwise asymptomatic patient.

The pediatric patient with a rib fracture may splint and hypoventilate secondary to pain. Physical examination may reveal point tenderness and, if the pleura has been involved, crepitus. If the patient has any respiratory or circulatory compromise, a tube thoracotomy is indicated for a pneumothorax or hemothorax. The tube should be placed at a separate site from the area of the fracture. If the patient is stable, then relief of pain, monitoring the respiratory status, and further evaluation (chest radiography, thoracic CT) for underlying injury is indicated. Wrapping or binding the chest wall are contraindicated since these measures may impair ventilatory function. Analgesics are helpful but should be used with caution because they can also cause respiratory depression. Epidural analgesia is helpful, especially for lower rib fracture. Intercostal nerve block is also useful but should be performed carefully to avoid puncturing the pleura.

Patients with multiple rib fractures should be admitted to the hospital for pain control, pulmonary physiotherapy, and observation for worsening respiratory status. Prognosis for isolated rib fractures is excellent with most healing within 6 weeks. The chest wall will remodel leaving no permanent disability.

Sternal and Scapular Fractures

Sternal and scapular fractures are uncommon in children, secondary to the marked compliance of the chest wall (Fig. 107.7). Although they require a thorough evaluation for other thoracic injuries because of the significant force required to fracture these bones, sternal and scapular fractures are rarely associated with vascular or brachial plexus injuries. In one adult study, scapular fracture alone was not a significant marker for mortality or neurovascular injury. In another study evaluating patients with blunt cardiac injury, only 2% had an associated sternal fracture.

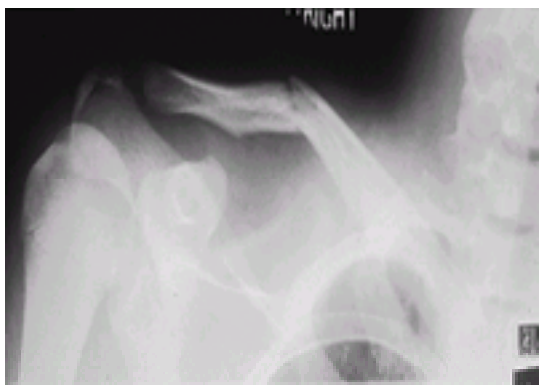

FIGURE 107.7. A 10-year-old child evaluated after a fall of more than 10 feet. The patient was asymptomatic except for shoulder pain. The only abnormalities noted were a scapular and clavicle fracture.

Flail Chest

Fracturing two or more ribs on the same side may result in that particular chest wall segment losing continuity with the thoracic cage. This condition is called a flail chest. Direct impact to the rib, as in a crush injury, is the most common mechanism for a flail chest. Flail chest is uncommon in children because of the significant compliance of the chest wall. In adult thoracic trauma series, flail chest occurred in approximately 10% of patients compared with less than 1% in the pediatric population. When a flail chest occurs, it is usually associated with an intrathoracic injury, most often pulmonary contusion, because of the force involved.

The pediatric patient may develop respiratory distress and failure as a result of a flail chest caused by numerous mechanisms. In the early 1900s, Bauer described the pendelluft theory, which attributes inefficient ventilation and oxygenation to a pendulumlike movement of air from the injured lung to the uninjured lung. The harder and faster a patient works to breathe, the more shifting of air from one side to the other occurs. The paradoxical movement of the chest also impairs the normal inspiratory/expiratory function of the lung (Fig. 107.8). Another mechanism for respiratory distress is the association of underlying pulmonary injury with a flail chest. Edema within the airways alters alveolar ventilation-perfusion ratios and produces pulmonary arteriovenous shunting with hypoxemia and subsequent respiratory distress. Finally, the pain associated with rib fractures causes voluntary and involuntary splinting. These patients are at increased risk for atelectasis and pneumonia secondary to poor pulmonary function.

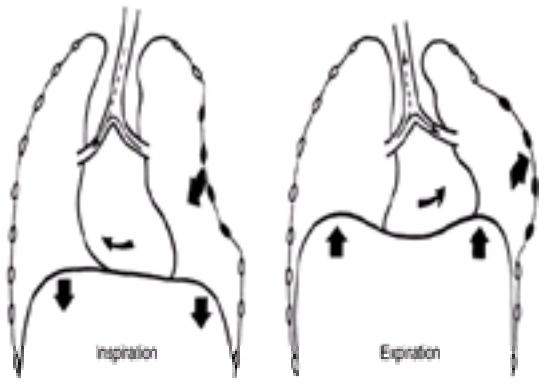

FIGURE 107.8. Pathophysiologic consequence of flail chest with paradoxical motion. (Reprinted with permission from Textbook of Pediatric Emergency Medicine. 3rd ed. Baltimore: Williams & Wilkins, Fig. 101.4.)

The goal of treatment should be to stabilize the involved portion of the thoracic cage. At the scene of an accident, the patient can be placed with the injured side down, thus improving tidal volume and ventilation. Any patient with respiratory distress should be intubated and placed on positive-pressure ventilation. This treatment serves two purposes: 1) the patient's airway is well protected and the effectiveness of breathing is maximized; 2) the positive pressure provides optimal expansion and splinting of the injured segment. Unfortunately, high inflating pressures can cause a pneumothorax and care must be taken when delivering positive pressure to the injured child. If the patient does not need to be intubated, aggressive pulmonary physiotherapy along with pain control is the preferred treatment. For patients with an underlying pulmonary contusion, fluids must be carefully monitored. Fluid may leak out of the injured capillary bed, worsening the pulmonary contusion.

Pulmonary Contusions and Lacerations

Pulmonary contusion is the most common thoracic injury in children. Pulmonary contusion occurs when a blunt force, such as a crush injury, is applied to the lung parenchyma. As in any contusion or bruise, the capillary network becomes damaged, leaking fluid into the surrounding tissues. A ventilation-perfusion mismatch occurs because of the extravasation of fluid, interfering with oxygenation. As the edema and swelling worsens, the patient's respiratory status also deteriorates. A pulmonary contusion may initially be invisible on a chest radiograph. Chest CT is more sensitive in detecting pulmonary contusion (Fig. 107.9A and Fig. 107.9B). Lung parenchymal injuries are often noted when a few cuts of the thoracic cavity are imaged while obtaining an abdominal CT.

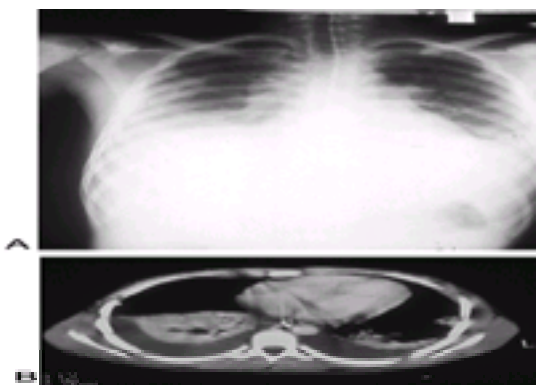

FIGURE 107.9. A 15-year-old child involved in an auto–pedestrian accident. Vital signs were stable at the scene, but the child had decreased breath sounds bilaterally. **(A)** Chest radiograph showed bilateral hemothorax and pulmonary contusions. **(B)** Computed tomography confirmed and better delineated the hemothorax and pulmonary contusions.

In one study, tachypnea, abnormal breath sounds, external thoracic wall contusion, and fracture of the bony thorax were each absent in more than 50% of patients with a pulmonary contusion. Interestingly, the chest radiograph in these patients did not dramatically worsen from time of admission. Nonetheless, mild contusions require close observation in the hospital for worsening respiratory status and supportive care.

Patients with moderate to severe pulmonary contusions may be tachypneic and have an oxygen requirement secondary to shunting within the lung. If the patient can no longer maintain oxygenation, endotracheal intubation and mechanical ventilation with positive pressure is the preferred treatment. Fluid restriction is helpful to avoid exacerbation of pulmonary edema. Many of these patients have associated injuries, making fluid restriction difficult; intensive care management with measurement of central venous and pulmonary arterial pressure may be helpful for those patients with major multisystem trauma. Double-lumen endotracheal–endobronchial tubes can be used in patients with severe lung contusions refractory to normal ventilatory management.

Pulmonary lacerations occur more often in penetrating trauma but can occur in rapid deceleration injuries. Rib fractures secondary to blunt trauma may also puncture the lung. Patients are usually tachypneic and have abnormal breath sounds. Large lacerations may cause hemoptysis. Chest radiograph shows pneumothorax or hemothorax. Treatment includes endotracheal intubation for patients in respiratory distress and tube thoracotomy for pneumothorax or hemothorax. Adequate intravenous access and blood for transfusion should be available before chest tube placement. Insertion of the chest tube can disrupt hemostasis in the chest cavity, and the patient may exsanguinate. Indications for surgery include continuous hemorrhage or air leak through the chest tube, massive hemoptysis, or air embolism.

Air embolism is usually fatal but it should be considered when a patient deteriorates suddenly after endotracheal intubation, when focal neurologic findings develop without evidence of a craniospinal injury, or when frothy blood is withdrawn from an arterial puncture. Treatment includes open thoracotomy with either occlusion of the hilar structure on the affected side or direct aspiration of the air. Neither of these treatment options is usually successful.

Intrapleural Injuries

In one study, intrapleural injury occurred in 40% of children with a thoracic injury. Hemopneumothorax, hemothorax, and

pneumothorax were evenly distributed. Pneumothorax was associated with the lowest mortality rate (15%) and hemothorax had the highest incidence (57%). Auscultation is helpful but not 100% accurate in diagnosing a hemothorax, pneumothorax, or hemopneumothorax. A recent report found that auscultation to detect hemothorax, pneumothorax, or hemopneumothorax had a sensitivity of 58% and a specificity of 98%. Most intrapleural injuries do not need surgical intervention and can be managed either by hospital observation or tube thoracostomy (Fig. 107.10).

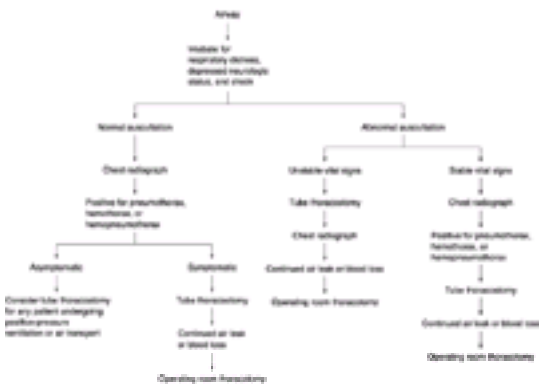

FIGURE 107.10. Algorithm for the management of intrapleural injuries.

Pneumothorax

Pneumothorax is the second most commonly encountered entity in blunt thoracic trauma and the most common in penetrating thoracic trauma. Air within the pleural cavity can arise from chest wall penetration, lung parenchyma disruption, a tear of the tracheobronchial structures, or esophageal rupture.

Patients may be asymptomatic, complain of pleuritic chest pain, have tachypnea, or be in severe respiratory distress. Physical examination may be normal or it may reveal diminished or absent breath sounds, crepitus, or hyperresonance to percussion on the side of the pneumothorax. In the asymptomatic or mildly symptomatic patient, a chest radiograph is helpful in diagnosing and determining the type of treatment necessary (Fig. 107.11). If the pneumothorax is small and the patient asymptomatic, observation in the hospital and administration of 100% oxygen is all that is necessary. A small pneumothorax is classically described as being less than 15%, although it is common to underestimate the size of a pneumothorax on plain films, only to find a much more extensive lesion on CT scan. Tube thoracostomy is indicated in symptomatic patients, patients undergoing positive-pressure ventilation, and those requiring air transport. An asymptomatic patient may rapidly become symptomatic if a small, simple pneumothorax progresses to a tension pneumothorax; therefore, even asymptomatic children with a traumatic pneumothorax should be admitted to the hospital for observation.

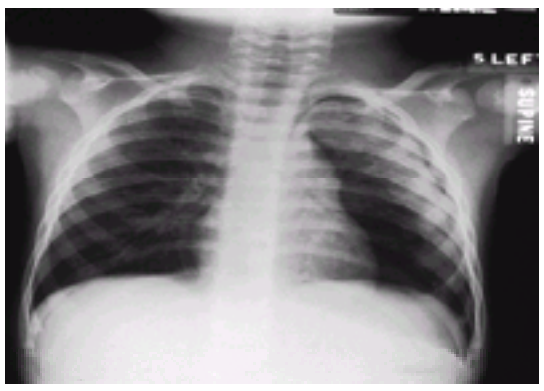

FIGURE 107.11. This 3-year-old child was an unrestrained passenger in a motor vehicle accident. The patient was tachypneic and had decreased breath sounds on the left side, but was otherwise asymptomatic. Chest radiograph revealed a left pneumothorax with a pulmonary contusion.

Tension Pneumothorax

A tension pneumothorax is the most common complicated intrapleural injury. Tension pneumothorax develops in up to 20% of children after simple pneumothorax. A tension pneumothorax occurs when air progressively accumulates within the pleural cavity. A laceration to the chest wall, pulmonary parenchyma, or bronchial wall may function as a one-way valve, allowing air to enter but not leave the pleural space. The progressive accumulation of air within the pleural cavity not only collapses the ipsilateral lung but compresses the contralateral lung as well. These patients may present in severe respiratory distress with decreased breath sounds on the side of the pneumothorax. The mediastinal structures also shift to the contralateral side (Fig 107.12). Two-thirds of the blood supply to the body is returned to the heart via the inferior vena cava. Because the inferior vena cava is relatively fixed and cannot shift as much as the superior vena cava, venous return to the heart is reduced and the patient may appear tachycardic, peripherally vasoconstricted, and in hypotensive shock. This underscores the importance that whenever a trauma patient suddenly deteriorates, the treating physician must return to airway and breathing, before jumping to circulation.

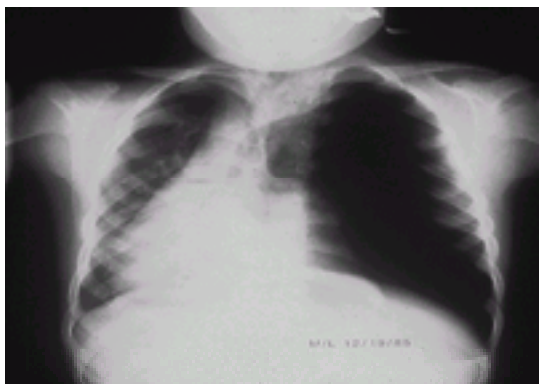

FIGURE 107.12. A 5-year-old girl fell off of and was then kicked in the chest by a horse. Upon arrival of the life-flight team, the patient was found to be in both respiratory and cardiovascular distress. Chest radiograph demonstrated a left-sided tension pneumothorax. The patient was intubated, and a chest tube was placed before the patient was transported. After the intubation and chest tube insertion, both the patient's respiratory and cardiovascular status improved.

Initial treatment consists of needle decompression performed in the midclavicular second intercostal space of the ipsilateral side. If a tension pneumothorax is present, an immediate release of air should be noted. If positive, the needle decompression is only a temporizing measure and must be followed by tube thoracotomy. Tube thoracotomy is usually done in the midaxillary line at the level of the fifth intercostal space (nipple level). Chest radiograph is performed only after the insertion of the chest tube and should not be used to diagnose a tension pneumothorax in the symptomatic patient. If a significant air leak continues after chest tube placement, a tracheobronchial rupture must be considered.

Open Pneumothorax

An open pneumothorax is the result of penetrating trauma. A direct connection exists between the pleural space and the outside atmosphere. As in a bronchial tear or lung parenchymal injury, air may enter but not leave the pleural space.

Initial treatment includes placement of an occlusive dressing at the wound site. This is best done when the patient is in full expiration. A chest tube should be placed immediately to prevent development of a tension pneumothorax. The chest tube should be inserted at a site different than the open wound. Larger open wounds may need surgical closure. Any patient in respiratory distress should be intubated and receive positive-pressure ventilation.

Hemothorax

Hemothorax is much more common in penetrating than in blunt thoracic trauma. In blunt thoracic trauma, a hemothorax can occur from rib fractures lacerating the lung, pulmonary parenchymal injuries without rib fractures, lacerations of the internal mammary arteries or intercostal arteries, or disruption of the major vascular structures in the mediastinum or hilum. A hemothorax secondary to a major injury of the great vessels usually results in death. Liver and spleen injuries can also cause a hemothorax with disruption of the diaphragm. The most common cause of a hemothorax is injury to the intercostal or internal mammary arteries, whereas injuries to the lung or great vessels causing a hemothorax are much less common but more serious.

Patients may present in respiratory distress or in profound shock secondary to obstruction of venous return or massive blood loss. Decreased breath sounds are noted on the affected side and tracheal or mediastinal deviation may occur. Thirty to forty percent of the patient's blood volume may be rapidly lost into the pleural cavity. This reaction usually occurs with major vessel lacerations. Bleeding from the intercostal or internal mammary arteries stops secondary to low systemic pressures. Also, when the lung is reexpanded, effective tamponade of the bleeding occurs. A chest radiograph should be obtained to confirm the diagnosis. If a hemothorax is suspected clinically and the patient is in severe respiratory or circulatory distress, immediate tube thoracostomy should be performed before a chest radiograph is taken.

Treatment of a major hemothorax should include aggressive airway and circulatory management and evacuation of the pleural blood. Endotracheal intubation and positive-pressure ventilation should be initiated in any unstable airway. Patients should be typed and crossmatched for packed red blood cells and adequately volume resuscitated, preferably with two large intravenous lines in place. O-negative blood, if type-specific blood is not available, should be available at the patient's bedside as soon as possible.

Tube thoracotomy is performed to evacuate blood within the pleural cavity, reexpand the lung, and prevent or treat any mediastinal shift. The chest tube is placed in the midaxillary line at the level of the fifth intercostal space (nipple level). This location is the same as in a pneumothorax. Many hemothoraxes may actually represent hemopneumothoraxes. After placement of a chest tube, blood should be slowly evacuated from the pleural space. Blood within the pleural cavity may tamponade a significant bleeding source within the chest and evacuating that blood may cause new bleeding to occur. Patients can exsanguinate rapidly; therefore, intravenous access, adequate volume resuscitation, and blood available for transfusion should be a priority. Blood removed by thoracostomy can be administered to the patient as an autotransfusion. Thoracostomy drainage needs to be closely monitored. Large ongoing blood loss from a chest tube should be collected in a system that allows autotransfusion. Thoracotomy is indicated for continued bleeding (greater than 1 to 2 mL/kg per hour), inability to expand the lung, or retained blood within the pleural cavity. Failing to adequately drain a hemothorax may result in restrictive lung disease from a fibrothorax or an empyema from the clotted material becoming infected.

Chylothorax

Chylothorax is rare in thoracic trauma and most commonly occurs secondary to iatrogenic complications. Chylothorax can result from penetrating injuries or from a hyperextension injury to the spine. Disruption of the thoracic duct will lead to chyle draining into the mediastinum and pleural space. Diagnosis is confirmed when chyle is aspirated from the pleural cavity. Infection is rare because chyle is bacteriostatic, and treatment consists of tube thoracostomy, dietary manipulation, and if all else fails, thoracic duct ligation.

Tracheobronchial Injuries

Injury to the tracheobronchial tree in children occurs rarely, with an incidence of less than 1%. This injury is most commonly caused by acceleration or deceleration forces. Major vessels or pulmonary parenchyma are more likely to be injured in penetrating trauma than the tracheobronchial tree or esophagus. Cervical tracheal rupture may be caused by a direct blow to the trachea or from the patient's head violently traveling forward and backward. This whiplash effect can cause a tear between two cartilaginous rings. Lower tracheobronchial injury usually occurs from a sudden increase in intrabronchial pressure. Because the child's chest wall is elastic, the trachea and main bronchi can be compressed between the chest wall and the vertebral spine. Compression of the chest with a closed glottis can cause a sudden increase in intrabronchial pressure, resulting in a tracheobronchial tear. Shear forces, traction, and crushing the airway between the chest and vertebral column may also cause a tracheobronchial injury. Approximately 80% of tracheobronchial injuries occur near the origin of the main stem bronchus.

The diagnosis of tracheobronchial injury may be difficult to make in the pediatric population. Mechanism of injury (e.g., fall, crush, direct blow) provides an important clue. Symptoms such as chest pain and dyspnea are common but nonspecific. Unlike the adult population, rib fractures are rare because of the elastic nature of the child's chest. Clinical signs include cyanosis, hemoptysis, tachypnea, and subcutaneous emphysema (cervical, mediastinal, or both). Pneumomediastinum and cervical emphysema are seen commonly in airway rupture (Fig. 107.13). If a pneumothorax is present with these findings, a bronchial rupture should be suspected. A continued air leak after insertion of a thoracostomy tube should also alert the physician to the possibility of a bronchial tear. Because of anatomic differences, ruptures of the bronchi occur on the right side more often than on the left. In the absence of a pneumothorax, tracheal rupture should be suspected if a pneumomediastinum or cervical emphysema is present.

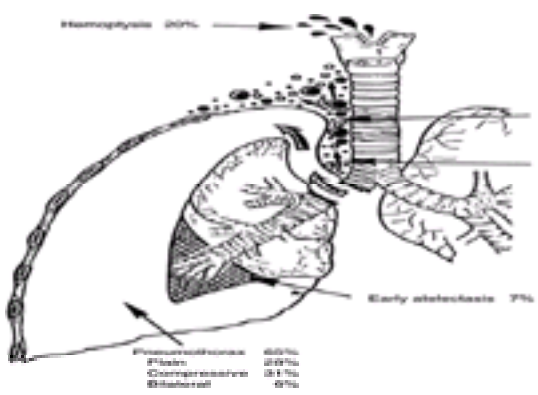

FIGURE 107.13. Initial signs of bronchial rupture. (Reprinted with permission from Textbook of Pediatric Emergency Medicine. 3rd ed. Baltimore: Williams & Wilkins, Fig. 101.9.)

Treatment includes initial airway stabilization and then bronchoscopic evaluation of the airway. Numerous reports in the literature record a partial tracheal tear becoming complete after endotracheal intubation. Therefore, if the airway is stable, oral tracheal intubation should be performed in the operating room under bronchoscopic guidance. This procedure prevents further trauma to the airway, and if a complication arises, emergency surgical access to the airway is readily available. If the airway is unstable and emergent endotracheal intubation needs to be performed, efforts should be made to prepare for backup measures such as cricothyroidotomy, tracheostomy, or fiberoptic bronchoscopy. An advantage of early bronchoscopy is exact identification and location of the lesion. The best surgical results are achieved when operative exploration is performed early. In the stable patient, CT of the chest can also help confirm the diagnosis and identify other injuries.

Esophageal Injuries

Esophageal injury is rare in children but presents a diagnostic challenge when it does occur. Timely and accurate diagnosis of an esophageal injury is paramount. The complications include mediastinal sepsis and death. The most common cause for esophageal perforation in the pediatric population is iatrogenic, followed by penetrating trauma (e.g., gunshot wound, stab wound). Esophageal perforation can occur in blunt trauma if a significant amount of chest compression occurs. The cervical and thoracic regions are more commonly affected, with the thoracic region having the highest mortality rate (35%).

The patient's signs and symptoms depend on the region injured. Patients with an esophageal rupture in the cervical region may complain of neck stiffness or neck pain. They may regurgitate bloody material and have cervical subcutaneous emphysema or odynophagia. A lateral neck radiograph may show retroesophageal emphysema. In the thoracic region, patients may present with abdominal pain and guarding, chest pain, subcutaneous emphysema, tachycardia, or dyspnea. A chest radiograph may show a pneumothorax, pneumomediastinum, or an air–fluid level in the

mediastinum. Perforation of the intra-abdominal esophagus may cause retrosternal, epigastric, or shoulder pain.

Patients with suspected esophageal perforation should be adequately volume resuscitated, have a nasogastric tube placed, and receive antibiotics covering Gram-positive, Gram-negative, and anaerobic organisms. The diagnosis of an esophageal perforation can be made by either esophagography, esophagoscopy, or both. In one study, flexible esophagoscopy had a sensitivity of 100% and specificity of 96%. Depending on the expertise at each institution and the patient's stability, these studies may be paired to lessen the chance of a misdiagnosis. Once the diagnosis is made, prompt surgical correction is mandatory. If the diagnosis is made within 24 hours, mortality is approximately 5%. Delayed diagnosis for more than 24 hours after injury is associated with a mortality of 70%.

Diaphragmatic Injuries

In the 16th century, diaphragmatic rupture was described by Ambrose Paré. He noted that “the stomach and intestines are sometimes drawn into the thoracic cavity” after diaphragmatic injury. Diaphragmatic injuries are more common in blunt trauma. A crushing force will produce a sudden increase in the intrathoracic and intra-abdominal pressure against the fixed diaphragm. Because of the flexible nature of the child's chest wall, rib fractures are rare. Even though penetrating thoracoabdominal trauma is uncommon in children, a diaphragmatic injury should be suspected in any thoracic or abdominal penetrating injury. The level of the diaphragm fluctuates greatly with respirations, and injuries of the diaphragm have been reported with penetrating wounds as high as the third and as low as the twelfth rib. Early reports of blunt traumatic diaphragmatic rupture were mostly left-sided. Because of a greater awareness of diaphragmatic injuries, right and bilateral diaphragmatic injuries have been reported more recently. Approximately 80% of diaphragmatic injuries still occur on the left, and 20% occur on the right. The left diaphragm is relatively unprotected, whereas the liver protects the right side. Right-sided diaphragmatic injuries are associated with increased mortality; patients usually have a greater physiologic insult and more numerous associated injuries.

Motor vehicle accidents are the most common mechanism of injury, and some authors believe the direction of impact may play a role in the side and type of diaphragmatic rupture. A lateral torso impact has been shown to be three times more likely than a frontal impact to result in a ruptured diaphragm. The rupture tends to be on the same side as the impact. Right-sided diaphragmatic ruptures may be associated with right-sided impact to the passenger side of the vehicle. Associated injuries, such as pulmonary contusions, hepatic or splenic lacerations, and fractures of the extremities, are present in more than 75% of patients. Thoracic aortic injuries have been reported in up to 10% of adults with diaphragmatic injury and should be considered in children with diaphragmatic trauma.

Patients may present in respiratory distress and have a scaphoid abdomen, although they are more likely to be symptomatic from associated injuries than from the diaphragmatic rupture. The verbal child may complain of chest pain or ipsilateral shoulder pain. The presence of bowel sounds within the thoracic cavity is nonspecific because in children bowel sounds can be transmitted from the abdominal cavity. More commonly, bowel sounds are absent because of an associated ileus. A nasogastric tube may be difficult to pass in patients with a diaphragmatic injury and gastric herniation. Even though the diagnosis is usually made upon initial review of the chest radiograph (Fig. 107.14), some series reported that up to 30 to 50% of initial chest radiographs were normal with a diaphragmatic injury. This finding emphasizes the importance of serial evaluations and chest radiographs in patients suspected of having a diaphragmatic injury. Other diagnostic studies, such as chest and abdominal CT with contrast or upper and lower gastrointestinal tract series, can help confirm the diagnosis.

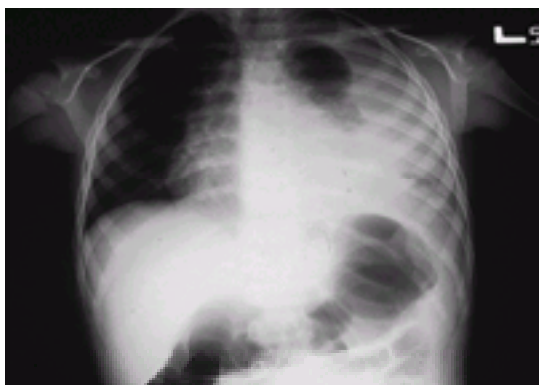

FIGURE 107.14. This 5-year-old boy was on a snowmobile when it crashed into a tree. Initially there was no respiratory distress, but upon arrival at the emergency department, the patient became tachypneic and required oxygen. Breath sounds were reportedly normal. Chest radiograph showed a left-sided diaphragmatic hernia. This injury was surgically repaired in the operating room and the patient did well postoperatively.

Before performing a tube thoracostomy for a pneumothorax or hemothorax, the physician should consider diaphragmatic injury to avoid injury to herniated intra-abdominal organs. In patients who clinically appear to have a diaphragmatic injury (scaphoid abdomen, bowel sounds auscultated in the thoracic cavity), a finger should be inserted in the chest tube incision site and the diaphragm should be palpated before placing a chest tube.

Herniation and strangulation of bowel may result from a delayed diagnosis. Diaphragmatic defects do not spontaneously heal because of motion associated with respirations and cyclical tension. Exploratory laparotomy or laparoscopy should be performed in cases in which a diaphragmatic hernia is strongly suspected.

Traumatic Asphyxia

Traumatic asphyxia results from direct compression of the chest or abdomen. The most common mechanism is a child being run over by a motor vehicle or pinned underneath a heavy object. In anticipation of impending injury, the child may inspire, tensing the thoracoabdominal muscles and closing the glottis. Traumatic asphyxia also occurs in patients with asthma, seizures, persistent vomiting, and pertussis.

Positive pressure is transmitted to the mediastinum and blood is forced out of the right atrium into the valveless venous and capillary system. The clinical manifestations occur because the increase in pressure dilates the capillary and venous system. Areas drained by the superior vena cava are particularly affected, explaining the significant difference between the patient's head and neck as opposed to the lower body. Patients with traumatic asphyxia usually present with the clinical picture of subconjunctival and upper body petechial hemorrhages, cyanosis, periorbital edema, respiratory distress, altered mental status, and associated injuries.

The primary goal of treatment is to stabilize the patient and identify associated injuries. The external appearance of a patient with traumatic asphyxia is impressive, but initial attention should be paid to the airway, breathing, and circulatory status. Pulmonary contusions and hepatic injuries are commonly seen with traumatic asphyxia and CT is helpful in identifying head, chest, and abdominal injuries. Because the most severe injuries cause immediate death, the prognosis is good for any patient surviving the first few hours after the incident. Cutaneous manifestations resolve with time, and neurologic sequela are rare. Neurologic injury usually results from hypoxia, not intracranial hemorrhage.

Aortic and Other Vascular Injuries

Traumatic rupture of the thoracic aorta (TRA) is uncommon in children but carries a high mortality rate (75 to 95%). TRA is associated with sudden deceleration forces, commonly from automobile accidents, causing a sheering stress. The aortic arch remains fixed, but the descending aorta is mobile. With deceleration, bending or sheering take place at the level of the ligamentum arteriosum, which is the most common site of aortic tears in adults and children.

TRA occurs in approximately 10 to 30% of adults sustaining blunt trauma but is much less common in the pediatric population. In one study, TRA occurred in only 2.1% of pediatric patients with thoracic trauma. The overall mortality rate was 93%. Why pediatric patients have a lower incidence of TRA than adults is unclear. One reason may be the mechanism of injury. In adults, most TRAs occur when the driver of an automobile forcibly strikes the steering wheel. The sudden deceleration force is isolated to the chest. Children who are passengers in a motor vehicle are less likely to strike an object that can deliver deceleration forces centrally to the chest. In children, one of the most common causes of blunt trauma is auto–pedestrian accidents, which produce forces distributed over a much wider area.

Children are usually symptomatic from associated injuries, and TRA can easily be missed. Clinical signs may include difference in pulses between the arms or arms and legs, thoracic ecchymosis, thoracic and back tenderness, paraplegia, and anuria. Patients with paraplegia and back pain may be initially diagnosed with a spinal cord injury. Unfortunately, 50% of patients may have no signs pertaining directly to TRA. More than 90% of patients have an abnormal chest radiograph (Fig. 107.15). Widened mediastinum, loss of the aortic knob, left-sided pleural cap, tracheal deviation, and nasogastric tube deviation may all be seen on a chest radiograph. Much has been written in the adult literature about the association of TRA with first rib fractures. More recent studies have shown that isolated first rib fractures without any other signs or symptoms do not correlate with TRA.

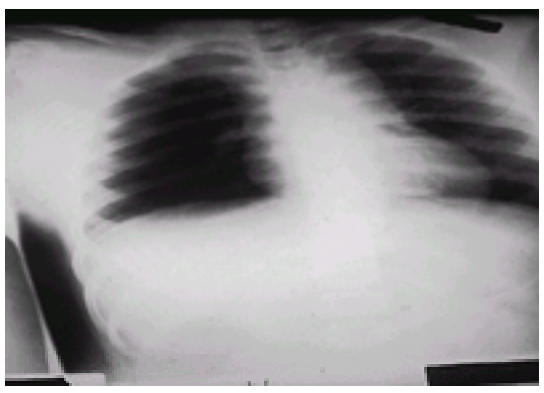

FIGURE 107.15. This 12-year-old girl was an unrestrained passenger involved in a motor vehicle accident. The patient was hypotensive at the scene and could not move her legs. In the emergency department, she had no motor or sensory function to her lower extremities and was anuric. Chest radiograph showed a widened mediastinum from traumatic rupture of the aorta.

Early diagnosis is imperative in patients with TRA (Fig. 107.16). Morbidity and mortality increase threefold if operative intervention is delayed more than 12 hours. The preferred method for diagnosing TRA is aortography (Fig. 107.17). Thoracic CT is only 55 to 65% accurate but helpful in diagnosing associated injuries. In one study, transesophageal echocardiography was shown to be a highly sensitive and specific method of detecting injury to the thoracic aorta. In contrast, another study showed transesophageal echocardiography to be only 63% sensitive and 84% specific in identifying patients with TRA. Pediatric patients were not included in either of these studies. If the patient is stable and TRA is suspected, aortography should be performed. Life-threatening intracranial, thoracic, or intra-abdominal injuries must first be evaluated and stabilized before aortography. If the patient is unstable, a transesophageal echocardiography can be performed while the patient's other life-threatening injuries are being treated.

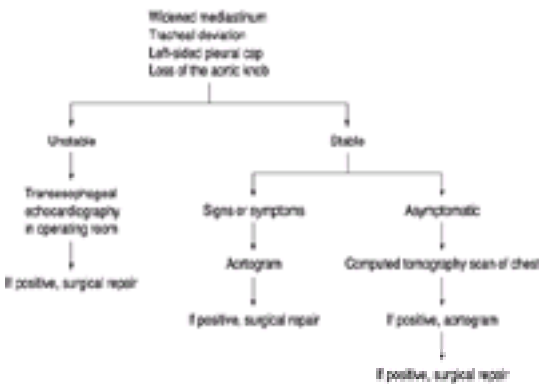

FIGURE 107.16. Algorithm for the evaluation and diagnosis of traumatic rupture of the thoracic aorta.

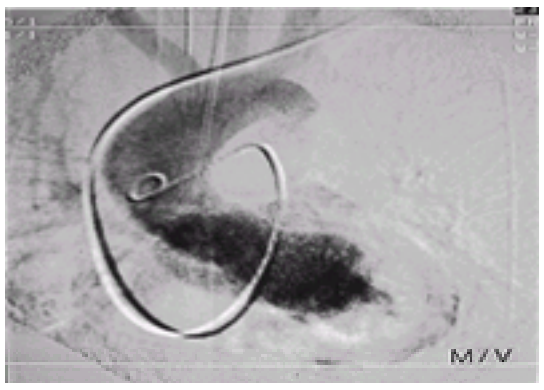

FIGURE 107.17. Emergency pulmonary angiography confirmed an aortic transection distal to the left subclavian artery with a pseudoaneurysm and narrowing of the proximal descending thoracic aorta in a 12-year-old girl.

Pericardial Tamponade

Pericardial tamponade occurs when the myocardium is injured and blood accumulates in the pericardial sac. Because of the nondistendible pericardium, pressure is exerted on the heart. Cardiac output decreases secondary to a decrease in venous return and stroke volume. The body initially tries to compensate with an increase in the pulse rate and peripheral vascular resistance. As the pressure within the pericardial sac increases, the systolic blood pressure decreases, causing a narrowing of the pulse pressure and subsequent hypotension and cardiogenic shock.

Pericardial tamponade may initially be difficult to diagnose because of associated injuries obscuring the clinical signs and symptoms. Patients may present with distant heart sounds, low blood pressure, poor perfusion, a narrow pulse pressure, or electromechanical dissociation (Fig. 107.18). Pulsus paradoxus, blood pressure falling more than 10 mm Hg during inspiration, occurs in less than one-half of patients with pericardial tamponade and should not be relied on to make the diagnosis of pericardial tamponade. Chest radiograph may show an enlarged heart (Fig. 107.19) and an ECG may show low-voltage QRS waves. Neither of these tests is diagnostic for pericardial tamponade, and performing these tests should not delay treatment in the unstable patient. In the stable patient, an echocardiogram can demonstrate fluid within the pericardial sac.

FIGURE 107.18. Algorithm for the evaluation and diagnosis of pericardial tamponade.

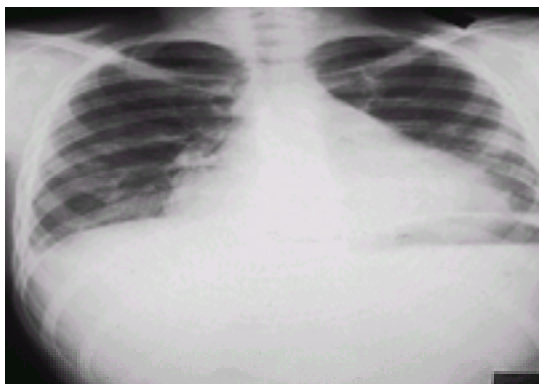

FIGURE 107.19. A 16-year-old boy had a steel bar strike him in the chest. Initially, he was hemodynamically stable but had muffled heart sounds. The patient quickly decompensated and required emergent pericardiocentesis after the chest radiograph. In the operating room, the patient was noted to have a small epicardial laceration on the surface of the heart.

In the unstable patient in whom pericardial tamponade is suspected, treatment includes control of the airway, intravascular volume resuscitation, and pericardiocentesis (Fig. 107.20). Pericardiocentesis is performed by inserting a 20-gauge spinal needle below the xiphoid process at a 45-degree angle toward the left shoulder. If time permits, an ECG monitor can be attached to the spinal needle. If the needle touches the heart, a current will be noted on the ECG monitor. Blood aspirated from the pericardial sac can be differentiated from intracardiac blood because pericardial blood is defibrinated and does not clot. Even though patients may show transient improvement after removal of blood from the pericardial sac, the patient should be taken to the operating room immediately for a pericardial window or other surgical intervention. A catheter should be placed in the pericardial sac over a wire guide for continual drainage of blood until surgical correction can be performed.

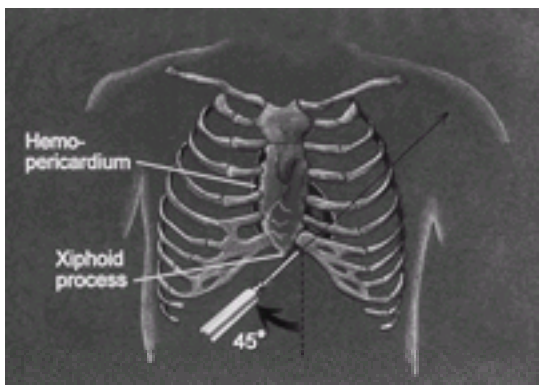

FIGURE 107.20. Pericardiocentesis is performed by inserting a 20-gauge spinal needle below the xiphoid process at a 45-degree angle toward the left shoulder. *Please see the color-tip insert (Color Plate 107.20).*

Blunt Cardiac Injuries

Blunt cardiac injury (BCI) occurs more commonly with associated injuries than in isolation and represents a spectrum of injuries. Myocardial contusion, ventricular or atrial rupture, and valvular disruption are all considered BCIs. Myocardial contusion is the most common and ventricular rupture the most lethal of injuries. In one study of 1288 patients with blunt thoracic trauma, 60 (4.6%) had a diagnosis of BCI. Other series have reported the incidence of BCI to range from 0 to 43%. Complications of BCI include arrhythmias, pump failure, congestive heart failure, and shock.

Cardiac rupture is the most common cause of death in blunt cardiac trauma. The right ventricle is the chamber most commonly ruptured because of its location directly beneath the sternum. Septal rupture can also occur, with the condition of the patient correlating with the size of the rupture. Patients with valvar injury may present in congestive heart failure with a new regurgitation murmur. Coronary artery injury is rare but should be considered in patients with persistent ECG changes consistent with ischemia following blunt thoracic trauma.

Unlike adults, pediatric patients with BCI often have few presenting signs or symptoms. Approximately 70% of adults with BCI complain of chest pain, whereas in one pediatric study less than half of the awake patients with BCI complained of chest pain, and external evidence of thoracic injury was present in only 60% of these patients. In the same study, cardiac examination was abnormal in less than one-quarter of the patients. BCI should be considered in any patient with thoracic trauma who develops a cardiac arrhythmia or a new murmur, or who is in congestive heart failure.

Evaluation of suspected BCI remains controversial. In one study, all children who developed heart failure or serious cardiac arrhythmias during their hospital course initially presented to the emergency department (ED) either in shock or with a serious arrhythmia. Patients with suspected BCI can be monitored in the ED or hospital and, if no arrhythmias develop on ECG, can be safely sent home. CPK-MB ratios have a high false-positive rate and are not a helpful screening tool. Transesophageal echocardiography should be performed in thoracic trauma patients with an abnormal ECG or arrhythmia, or a new heart murmur. Transesophageal echocardiography is more sensitive than transthoracic echocardiography in detecting myocardial injury.

Some general guidelines regarding patients with suspected BCI include the following: 1) If a pediatric patient with

suspected BCI is hemodynamically stable and has not experienced any arrhythmias, a serious life-threatening arrhythmia or pump failure is unlikely. 2) Any patient with suspected BCI who is hemodynamically unstable or has arrhythmias should undergo a thorough evaluation, such as with transesophageal echocardiography, and be admitted to the intensive care unit. 3) All patients with suspected BCI need close follow-up.

Penetrating Thoracic Trauma

Although not as common as blunt thoracic trauma, penetrating thoracic trauma is becoming more common in the pediatric population. In one study, penetrating thoracic trauma occurred in 20% of pediatric patients evaluated for a thoracic injury. The most common mechanism of injury was gunshot wounds followed by stab wounds (Fig. 107.21). Pediatric patients with blunt thoracic trauma are more likely to die from associated intracranial and intra-abdominal injuries. In contrast, penetrating thoracic trauma is usually a single-system disease and more than 95% of deaths are caused by the thoracic wound.

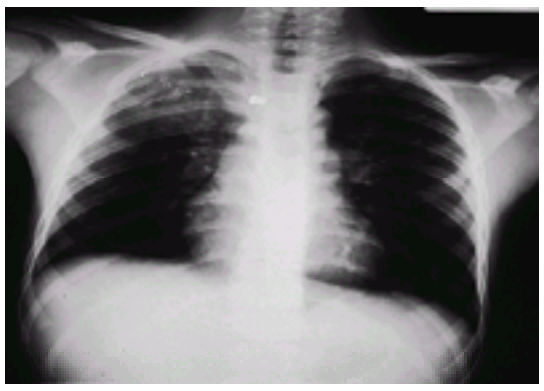

FIGURE 107.21. A 12-year-old boy playing with his father's loaded gun when it accidentally discharged. There was no cardiac or great vessel involvement and the patient did well postoperatively.

The most common penetrating thoracic injuries are hemothorax and pneumothorax, which almost always require tube thoracostomy. Intra-abdominal injuries should always be suspected because of the close proximity of abdominal contents to the thoracic cavity. In one study, intra-abdominal injuries occurred in 20% of patients with penetrating thoracic injury. More than half of children with penetrating thoracic injury require operative intervention. This percentage is higher than the 15% reported in the adult literature. Why children have this higher rate of operative intervention is unclear, but it may be a result of the close proximity of the vital organs in the thoracic cavity compared with adults.

Evaluation and treatment includes airway stabilization, fluid resuscitation, and management of the chest wound. Radiopaque markers (paper clips) may be placed by the entry and exit sites to help determine the course of the missile. Penetrating injuries near the mediastinum may be critical, especially if the patient is hemodynamically unstable. Pericardial tamponade should be considered and treated in the unstable patient. In the stable patient, transesophageal or transthoracic echocardiogram is helpful in evaluating the heart and determining whether fluid has entered the pericardial sac. Diaphragmatic lacerations are difficult to diagnose and sometimes require exploratory laparotomy or laparoscopy for diagnosis and treatment.

Emergency Department Thoracotomy

Emergency department thoracotomy (EDT) is one of the most aggressive resuscitative measures for patients with thoracic trauma. With the advancement of transport systems and the regionalization of trauma centers, patients who would have died at the scene are arriving at trauma centers for evaluation and treatment. EDT allows the physician to evaluate and evacuate the pericardial sac, perform open cardiac massage, and temporarily control bleeding from the heart, hilum, or lung. Catheters can also be placed directly into the right atrium, helping with fluid resuscitation, and the thoracic aorta can be compressed, improving central circulation to the brain and heart.

Anecdotal reports that suggest that EDT may be useful in the pediatric trauma patient who has vital signs but loses them during transport or resuscitation have been published. Other studies have shown the outcome of EDT in pediatric blunt trauma victims has been poor. The more recent literature has reevaluated the need for EDT and tried to select a more specific population. In one study, none of the 17 pediatric patients undergoing EDT after thoracic trauma survived, although 15 of 17 patients had blunt trauma and only two had isolated penetrating thoracic trauma. The authors concluded that EDT was not indicated in the blunt thoracic trauma patient arriving in the ED without any vital signs or ECG tracing. Another study of thoracic gunshot wounds in the pediatric population found no survivors in patients undergoing EDT and advocated a reappraisal of the indications for EDT among pulseless pediatric victims of thoracic gunshot wounds.

The one accepted indication for EDT is the patient with penetrating thoracic trauma who had and then lost vital signs before arrival or during ED resuscitation. In addition, EDT may be useful for the patient with blunt thoracic trauma who acutely deteriorates in the ED during resuscitation, but the chance of survival is dismal. Lifesaving interventions, such as airway management, fluid resuscitation, and pericardiocentesis, should not be delayed while waiting for EDT to be performed. The pediatric patient with vital signs but not responding to initial treatment, such as tube thoracotomy and pericardiocentesis, is a candidate for thoracotomy in the operating room, rather than the ED.

The outcome is directly dependent on the patient's status before arrival in the ED and mechanism of injury. In blunt thoracic trauma, 100% of patients who present to the ED without vital signs have a fatal injury, regardless whether EDT is

performed. Pediatric patients who present with cardiac arrest or tamponade caused by penetrating trauma, and in whom vital signs were present either in the field or in the ED, have the best chance of survival, although small, if EDT is performed.

Suggested Readings

GENERAL

Allshouse M, Eichelberger M. Patterns of thoracic injury. In: Eichelberger M, ed. Pediatric Trauma. Prevention, Acute Care, Rehabilitation. St Louis: Mosby, 1993:437–448.

Cooper A. Thoracic Injuries. Semin Pediatr Surg 1995;4:109–116.

Cooper A, Barlow B, DiScala C, et al. Mortality and truncal injury: the pediatric perspective. J Pediatr Surg 1994;29:33.

Jones K. Thoracic trauma. In: Mayer T, ed. Emergency Management of Pediatric Trauma. Philadelphia: WB Saunders, 1985:254–271.

LoCicero J, Mattox KL. Epidemiology of chest trauma. Surg Clin North Am 1989;69:15–18.

Peclet M, Newman K, Eichelberger M, et al. Patterns of injury in children. J Pediatr Surg 1990;25:85–91.

Peclet M, Newman K, Eichelberger M, et al. Thoracic trauma in children: an indicator of increased mortality. J Pediatr Surg 1990;25:961–966.

Shorr R, Grittenden M, Index M. Blunt thoracic trauma: analysis of 515 patients. Ann Surg 1987;306:200–205.

Trupka A, Waydhas C, Hallfeldt KJ. Value of thoracic computed tomography in the first assessment of severely injured patients with blunt chest trauma: results of a prospective study. J Trauma 1997;43;405–412.

Welch KJ. Thoracic injuries. In: Randolph JG, et al, eds. The Injured Child. Chicago: Yearbook Medical Publishers, 1980:215–231.

CHEST WALL INJURY

Freedland M, Wilson R, Bender J, et al. The management of flail chest injury: factors affecting outcome. J Trauma 1990;30:1460–1468.

Garcia V, Gotschall M, Eichelberger M, et al. Rib fractures in children: a marker of severe trauma. J Trauma 1990;30:695–700.

Harris G, Soper R. Pediatric first rib fractures. J Trauma 1990;30:343–345.

Landercasper J, Cogbill T, Strutt P. Delayed diagnosis of flail chest. Crit Care Med 1990;18:611–613.

Rankin A, Comber R. Management of fifty cases of chest injury with a regimen of epidural bupivacaine and morphine. Anaesth Intens Care 1984;12:311–314.

Schweich P, Fleisher G. Rib fractures in children. Pediatr Emerg Care 1985;1:187–189.

Shorr R, Crittenden M, Index M. Blunt thoracic trauma: analysis of 515 patients. Ann Surg 1987;306:200–205.

Stephens N, Morgan A, Corvo P, et al. Significance of scapular fracture in the blunt-trauma patient. Ann Emerg Med 1995;26:439–442.

Ziegler D, Agarwal N. The morbidity and mortality of rib fractures. J Trauma 1994;37:975–979.

LUNG INJURY

Blostein P, Hodgman C. Computed tomography of the chest in blunt thoracic trauma: results of a prospective study. J Trauma 1997;43:13–18.

Bonadio W, Hellmich T. Post-traumatic pulmonary contusion in children. Ann Emerg Med 1989;18:1050–1052.

Frame S, Marshall W, Clifford T. Synchronized independent lung ventilation in the management of pediatric unilateral pulmonary contusion: case report. J Trauma 1989;29:395–397.

Johnson J, Cogbill T, Winga E. Determinants of outcome after pulmonary contusion. J Trauma 1986;26:695–697.

PNEUMOTHORAX/HEMOTHORAX

Blaisdell F. Pneumothorax and hemothorax. In: Blaisdell F, Trunkey D, eds. Cervicothoracic Trauma. New York: Thieme, 1986.

Chen S, Markmann J, Kauder D, et al. Hemopneumothorax missed by auscultation in penetrating chest injury. J Trauma 1997;42:86–89.

Ma O, Mateer J. Trauma ultrasound examination versus chest radiography in the detection of hemothorax. Ann Emerg Med 1997;29:312–315.

Nakayama D, Ramenofsky M, Rowe M. Chest injuries in childhood. Ann Surg 1989;210:770–775.

Symbas P. Cardiothoracic trauma. Curr Probl Surg 1991;28:742–797.

TRACHEAL AND BRONCHIAL INJURY

Baumgartner F, Sheppard B, Virgilio C, et al. Tracheal and main bronchial disruptions after blunt chest trauma: presentation and management. Ann Thorac Surg 1990;50:569–574.

Gaebler C, Mueller M, Schramm W, et al. Tracheobronchial ruptures in children. Am J Emerg Med 1996;14:279–285.

Hancock B, Wiseman N. Tracheobronchial injuries in children. J Pediatr Surg 1991;26:1316–1319.

Kadish H, Schunk J, Woodward G. Blunt pediatric laryngotracheal trauma: case report and review of the literature. Am J Emerg Med 1994; 12:207–211.

Panagiotis N, Alexander G, Richard R. Rupture of the airways from blunt trauma: treatment of complex injuries. Ann Thorac Surg 1992;

54:177–183.

Taskinen S, Salo J, Halttunnen P. Tracheobronchial rupture due to blunt chest trauma. Ann Thorac Surg 1989;48:846–849.

ESOPHAGEAL INJURY

Backer C. Computed tomography in patients with esophageal perforation. Chest 1990;98:1078–1080.

Flowers J, Graham S, Ugarte M, et al. Flexible endoscopy for the diagnosis of esophageal trauma. J Trauma 1996;40:261–266.

Jones W, Ginsber R. Esophageal perforation: a continuing challenge. Ann Thorac Surg 1992;53:534–543.

DIAPHRAGMATIC INJURY

Beauchamp G, Khalfallah A, Girard R, et al. Blunt diaphragmatic rupture. Am J Surg 1984;148:292–296.

Boulanger B, Milzman D, Rosati C, et al. A comparison of right and left blunt traumatic diaphragmatic rupture. J Trauma 1993;35:255–260.

Brandt M, Luks F, Spigland N. Diaphragmatic injury in children. J Trauma 1992;32:298–301.

Guth A, Pachter L, Kim U. Pitfalls in the diagnosis of blunt diaphragmatic injury. Am J Surg 1995;170:5–9.

Murray J, Demetriades D, Cornwell E, et al. Penetrating left thoracoabdominal trauma: the incidence and clinical presentation of diaphragm injuries. J Trauma 1997;43:624–626.

Pagliarello G, Carter J. Traumatic injury to the diaphragm: timely diagnosis and treatment. J Trauma 1992;33:194–197.

TRAUMATIC ASPHYXIA

Gorenstein L, Blair G, Shandling B. The prognosis of traumatic asphyxia in childhood. J Trauma 1986;21:753–756.

Newquist M, Sobel R. Traumatic asphyxia: an indicator of significant pulmonary injury. Am J Emerg Med 1990;8:212–215.

Sklar D, Baack B, McFeeley P, et al. Traumatic asphyxia in New Mexico: a five-year experience. Am J Emerg Med 1988;5:219–223.

AORTIC AND GREAT VESSEL INJURY

Dart C, Braitman H. Traumatic rupture of the thoracic aorta. Arch Surg 1976;111:687–690.

Eddy A, Rusch V, Fligner C, et al. The epidemiology of traumatic rupture of the thoracic aorta in children: a 13-year review. J Trauma 1990; 30:989–992.

Lee J, Harris J, Duke J, et al. Noncorrelation between thoracic skeletal injuries and acute traumatic aortic tear. J Trauma 1997;43:400–404.

Saletta S, Lederman E, Fein S. Transesophageal echocardiography for the initial evaluation of the widened mediastinum in trauma patients. J Trauma 1995;39:137–141.

Smith M, Cassidy M, Souther S, et al. Transesophageal echocardiography in the diagnosis of traumatic rupture of the aorta. N Engl J Med 1995;332:356–362.

BLUNT CARDIAC INJURY

Biffl W, Moore F, Moore E, et al. Cardiac enzymes are irrelevant in the patient with suspected myocardial contusion. Am J Surg 1994;169:523–528.

Dowd M, Krug S. Pediatric blunt cardiac injury: epidemiology, clinical features, and diagnosis. J Trauma 1996;40:61–67.

Karalis D, Victor M, Davis G, et al. The role of echocardiography in blunt chest trauma: a transthoracic and transesophageal echocardiographic study. J Trauma 1994;36:53–58.

Maron B, Poliac L, Kaplan J, et al. Blunt impact to the chest leading to sudden death from cardiac arrest during sports activities. N Engl J Med 1995;333:337–42.

Tellez D, Hardin W, Takahashi M, et al. Blunt cardiac injury in children. J Pediatr Surg 1987;22:1123–1128.

PENETRATING THORACIC INJURY

Fernandez L, Radhakrishnan J, Gordon R, et al. Thoracic BB injuries in pediatric patients. J Trauma 1995;38:384–389.

Inci I, Ozcelik C, Nizam O. Penetrating chest injuries in children: a review of 94 cases. J Pediatr Surg 1996;31:673–676.

Nance M, Sing R, Reilly P. Thoracic gunshot wounds in children under 17 years of age. J Pediatr Surg 1996;31:931–935.

Peterson R, Tiwary A, Kissoon N, et al. Pediatric penetrating thoracic trauma: a five-year experience. Pediatr Emerg Care 1994;10:129–131.

EMERGENCY DEPARTMENT THORACOTOMY

Beaver B, Colombani P, Buck J, et al. Efficacy of emergency room thoracotomy in pediatric trauma. J Pediatr Surg 1987;22:19–23.

Boyd M, Vanek V, Bourguet C. Emergency room resuscitative thoracotomy: when is it indicated? J Trauma 1992;33:714–721.

Durham L, Richardson R, Wall M, et al. Emergency center thoracotomy: impact of prehospital resuscitation. J Trauma 1992;32:775–779.

Esposito T, Jurkovich G, Rice C, et al. Reappraisal of emergency room thoracotomy in a changing environment. J Trauma 1991;31:881–887.

Langer J, Hoffman M, Pearl R, et al. Survival after emergency department thoracotomy in a child with blunt multisystem trauma. Pediatr Emerg Care 1989;5:255–261.

Lorenz H, Steinmetz B, Lieberman J. Emergency thoracotomy: survival correlates with physiologic status. J Trauma 1992;32:780–788.

Powell R, Gill E, Jurkovich G, et al. Resuscitative thoracotomy in children and adolescents. Am Surg 1988;54:188–191.

Sheikh A, Brogan T. Outcome and cost of open- and closed-chest cardiopulmonary resuscitation in pediatric cardiac arrest. Pediatrics 1994; 93:392–398.

CHAPTER 108

Abdominal Trauma

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Childhood injuries are the leading cause of death after the age of 1 year. In particular, trauma causes nearly 70% of deaths from age 5 to 19 years of age, with head and thoracic injuries accounting for most. Abdominal injuries occur in approximately 20% of children with fatal polytrauma. Importantly though, abdominal trauma is the most common unrecognized cause of fatal injuries.

Recognition of intra-abdominal injuries may be difficult, especially in the context of multisystem trauma. The physician should approach any child with a high index of suspicion for intra-abdominal injury. As well, intra-abdominal injury may be occult in cases of child battering, and the examining physician must exclude abdominal trauma in these instances.

APPROACH

The assessment of any trauma patient must begin with the ABCs: airway, breathing, and circulation. Priorities in evaluation and treatment include recognition and relief of airway obstruction, appropriate protection of the cervical spine, and management of life-threatening chest injuries and shock. Once resuscitation and cervical spine stabilization have begun, evaluation of the abdomen is included in both the primary and secondary surveys.

The evaluation for intra-abdominal injuries in children begins with an elicitation of the mechanism of trauma. Blunt injuries account for most morbidity and mortality of childhood trauma, although the frequency with which penetrating injuries occur is increasing. Penetrating trauma is usually evident on careful inspection of both the anterior and posterior torso. In contrast, blunt abdominal trauma must be suspected from historical information and careful physical examination. Children with severe multiple trauma are obviously at risk for intra-abdominal injuries, but sufficient energy to injure also may be present in apparently minor falls and direct blows to the abdomen from balls, bats, bicycle handlebars, and countless toys.

Life-threatening abdominal injuries may be occult or manifest in several ways: abdominal distension, shock, and/or external hemorrhage (e.g., from a penetrating injury). Historical information or physical examination findings are often lacking or subtle. Children have the capacity to maintain a normal blood pressure in the face of significant blood loss, and hence mask major intra-abdominal bleeding. The examining physician must always keep in mind that the abdomen is a large potential reservoir.

Physical Examination

A traumatized child is often difficult to examine; extra-abdominal injuries may obscure abdominal findings. In addition, the results of physical examination may be subtle or unreliable in the unconscious, intoxicated, agitated, or fearful child. Vital signs, including blood pressure and pulse, are often normal for age, especially in children with isolated injuries of the liver and spleen. Furthermore, external signs of injury, abdominal tenderness, and absent bowel sounds seldom differentiate pediatric patients who require laparotomy from those who do not.

However, careful serial examinations are critically important in maintaining the index of suspicion necessary to proceed with more sophisticated testing when appropriate. Inspection should note abrasions, lacerations, ecchymoses, penetrating wounds (including missile entry and exit sites), and telltale markings (e.g., tire tracks, seat belt marks). Attention should be paid to the anterior and posterior abdomen and to both flanks, as well as to the lower thorax when considering abdominal injuries. Abdominal distension may be caused by hemoperitoneum or peritonitis but most often results from gastric distension from air swallowed by the crying child. Early gastric decompression may assist the

abdominal examination and prevent vomiting with aspiration of gastric contents. The presence or absence of bowel sounds is generally not of much significance in the initial evaluation, but prolonged ileus may be a sign of intra-abdominal pathology. Tenderness upon palpation, percussion, or shaking may all be caused by abdominal wall contusion but may also indicate intra-abdominal injuries. Pelvic stability is evaluated by gently compressing the ileac wings. Digital rectal examination should be performed; the presence of blood may indicate perforation of the bowel. A boggy or high-riding prostate, blood at the urethral meatus, or a distended bladder may be present with urethral disruption, and preclude bladder catheterization until a retrograde urethrogram has been performed (see Chapter 109). Diminished rectal sphincter tone may indicate a spinal cord injury.

Laboratory Data

Blood should be obtained and sent for immediate typing and crossmatching not only in all instances of multiple trauma but also if isolated intra-abdominal injury is suspected. The hospital's blood bank must have O-negative blood ready for resuscitation if needed. Additional laboratory studies should include serum amylase, liver transaminases, and urinalysis. Hyperamylasemia may be present with pancreatic injury, but its absence does not preclude injury. Elevated serum liver transaminases may also be associated with intra-abdominal injuries, especially hepatic injuries. Screening for intra-abdominal injuries by evaluating transaminase levels is not universally accepted because sensitivity and specificity varies widely in the literature. In general, though, significantly elevated transaminase levels (AST greater than 450 IU/L and ALT greater than 250 IU/L) correlate well with hepatic injuries, and the evaluation of patients with such elevations should always include abdominal computed tomography (CT) scan. Urine should be tested for the presence of blood. Grossly bloody urine or a microscopic examination of urine that reveals more than 20 red blood cells per high-powered field (RBC/hpf) suggests injury to the kidneys and potentially the adjacent organs (see Chapter 109).

Arterial blood gas determinations may be helpful in the evaluation of pulmonary injuries and may indicate persistent metabolic acidosis when volume resuscitation is inadequate. A decreasing hematocrit on serial determinations suggests ongoing blood loss.

INITIAL MANAGEMENT PRINCIPLES

Basic Principles of Management

Treatment of the seriously injured patient requires a team approach, which includes a designated leader who directs team members who have specific responsibilities during the initial evaluation and management. Airway management and cervical spine stabilization are first priorities (Fig. 108.1). Any child with significant injuries should receive supplemental oxygen, especially if signs of shock are present. Intravenous or intraosseous access should be obtained while the primary survey is completed; immediate life-threatening injuries should be managed and hemorrhagic shock should be treated with infusion of isotonic crystalloid solution, when necessary, starting at 20 mL/kg. Large-bore catheters should be used whether in the upper or lower extremities to allow rapid infusion of large volumes of fluid during resuscitation. Accessing the femoral vein is acceptable and in fact is a preferred site in the child.

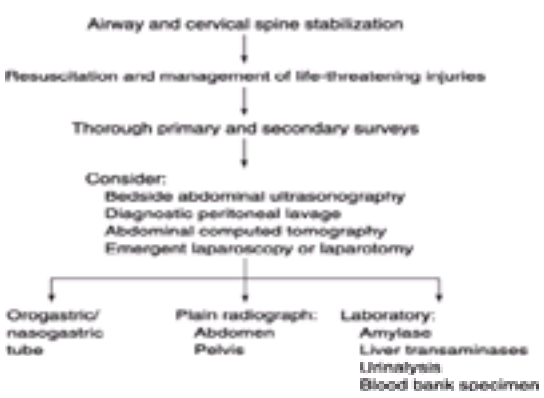

FIGURE 108.1. Initial evaluation and treatment of the child with abdominal trauma.

The current recommendation by the American College of Surgeons remains that aggressive fluid resuscitation be pursued. Although animal data suggest that limited (hypotensive) fluid resuscitation may improve survival by limiting hemorrhage into the peritoneal space, application to the management of children is still controversial.

As the initial evaluation proceeds, the priorities of management depend on the extent of multisystem injuries and the stability of the patient (Fig. 108.2). Patients who are unstable as a result of ongoing blood loss or an expanding intracranial hemorrhage require intervention early in the evaluation phase.

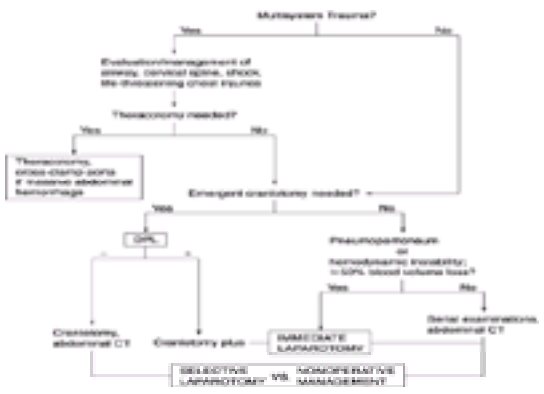

FIGURE 108.2. Management of blunt abdominal trauma. *DPL,* diagnostic peritoneal lavage; *CT,* computed tomography (see Table 108.2).

1. Mechanism of injury suggesting abdominal trauma
2. Slowly declining hematocrit
3. Unaccountable fluid or blood requirements
4. Neurologic injury precluding accurate abdominal examination
5. Hematuria
6. Acute "need to know" (e.g., before general anesthesia)

Table 108.1. Indications for Abdominal Computed Tomography Scan in the Pediatric Trauma Patient

The Unstable Patient

Immediate life-threatening injuries, such as airway obstruction, tension pneumothorax, pericardial tamponade, and obvious sources of external blood loss, must be treated promptly. The role of emergency department (ED) thoracotomy is controversial in children; its use should be confined to situations in which control of intrathoracic bleeding is needed (e.g., with lung or heart lacerations) or in situations in which previously detected vital signs are lost. If emergent thoracotomy is performed in the latter instance for presumptive intra-abdominal hemorrhage, the aorta is cross-clamped at a level just above the diaphragm.

If significant head trauma has occurred, a determination must be made with regard to the need for immediate neurosurgical intervention. A rapidly performed CT scan of the head is usually sufficient to determine the presence of a hematoma, which can be drained by the neurosurgeon. If hemodynamic instability or the need for immediate craniotomy exists and does not allow for CT evaluation of the abdomen (Table 108.1), a diagnostic peritoneal lavage (DPL) should be performed either in the ED or in the operating suite. If the peritoneal lavage is positive (Table 108.2), laparotomy and craniotomy proceed simultaneously. Finally, if neither thoracotomy nor craniotomy is indicated, emergent laparotomy is performed when pneumoperitoneum is noted on a plain radiograph or when the patient remains hemodynamically unstable in the face of historical or physical evidence of abdominal trauma (Fig. 108.2).

1. >5 mL of gross blood
2. Obvious enteric contents (e.g., bile)
3. Peritoneal lavage fluid exiting from chest tube, urinary bladder catheter
4. Positive laboratory analysis of peritoneal lavage fluid
   a. >100,000 RBC/mm^3
   b. >500 WBC/mm^3
5. Elevated amylase in effluent

RBC, red blood cells; WBC, white blood cells.

Table 108.2. Positive Diagnostic Peritoneal Lavage Criteria

The Stable Patient

Commonly, the injured child can be stabilized in the ED with proper airway and cervical spine management, and with intravenous fluid therapy and blood transfusion. A careful secondary survey should then be performed. Based on history and careful and serial abdominal examinations, CT is indicated when intra-abdominal injuries are suspected (Table 108.1). Children who have had even minor injuries should be examined serially and monitored in the ED. At times, an abdominal CT scan is merited based solely on severe force inherent in a particular mechanism of injury despite an

unremarkable physical examination. The utility of abdominal ultrasonography in trauma is now well documented in the adult literature, and the stable patient may benefit from an early ultrasound, especially when an abdominal CT is not immediately available or if evaluation of the abdomen will be delayed during head or chest CT. In addition, the emergence of the utility of emergent laparoscopy for adults with trauma may preclude the need for DPL or laparotomy in many instances. Importantly, ultrasonography and laparoscopy may eventually allow diagnosis and management of intra-abdominal injuries in the stable pediatric patient.

Additional Management

Children with abdominal trauma often need decompression of the stomach; this procedure facilitates examination, may provide information concerning gastric or diaphragmatic injury (bloody aspirate; radiographic evidence of the nasogastric tube in the thoracic cavity), and relieves the discomfort of an ileus. Maxillofacial trauma precludes nasogastric tube placement, but an orogastric tube suffices in these instances. Urinary bladder catheterization may provide evidence of genitourinary system injury and is helpful in monitoring urinary output. Bladder catheterization is contraindicated when urethral disruption is suspected.

Diagnostic Imaging

Radiographic evaluation of children with abdominal trauma includes plain radiographs, contrast studies, radionuclide scans, ultrasound, and CT. The stable child with abdominal trauma is best evaluated with abdominal CT using intravenous contrast. If a nasogastric or orogastric tube is in place, it should be withdrawn temporarily into the esophagus to avoid an artifact from its radiopaque marker. Abdominal CT has its lowest sensitivity for small gastrointestinal perforations and pancreatic injury. Although CT is the most common technique used in childhood trauma, the surgeon's decision to proceed to laparotomy may be based more on the clinical status of the child than on the radiologic findings. Although abdominal CT is considered the most sensitive diagnostic tool, abdominal ultrasonography may provide important data early in the course of the management of a child with suspected intra-abdominal injuries. Data from the adult literature show that the sensitivity for the detection of intraperitoneal fluid ranges from 85 to 98%. Although currently not a universal component of the evaluation of the child with blunt abdominal trauma, the value of ultrasound as a rapid diagnostic tool is evident, and will likely become a part of trauma evaluation protocols.

Diagnostic Peritoneal Lavage

DPL is occasionally a helpful adjunct to the management of children with abdominal trauma. The disadvantages of DPL include the introduction of air and fluid into the abdomen (subsequent radiologic evaluations are less helpful) and peritoneal irritation caused by the procedure (subsequent physical examinations are less reliable).

It is rarely necessary to perform laparotomy on children with free intraperitoneal blood; thus, DPL, which effectively detects small volumes of blood, is often too sensitive in children. The primary indication for DPL in children is an urgent “need to know” with regard to the status of the peritoneal cavity, such as in the child who is hemodynamically unstable or requires immediate craniotomy and cannot delay for abdominal CT.

The technique for DPL in children is similar to that in adults, although in young children a small supraumbilical incision is preferred over the usual infraumbilical approach to avoid the bladder. If a nasogastric tube and urinary bladder catheter have not been placed, they should be inserted before peritoneal lavage is performed. Ringer's lactate solution (10 mL/kg, maximum 1000 mL) is instilled into the peritoneal cavity over 10 minutes and then removed for analysis. Criteria for a positive lavage are shown in Table 108.2.

Emergent versus Selective Laparotomy

The indications for immediate laparotomy are limited in blunt abdominal trauma (Table 108.3). In most cases of childhood trauma (Fig. 108.2), emergency laparotomy is not necessary and further diagnostic studies direct either elective (selective) laparotomy or observation and monitoring. Most children with blunt abdominal trauma require only in-hospital observation and monitoring after delineation of the site and extent of their injury by abdominal CT.

1. Multisystem injuries with indications for craniotomy in the presence of a positive diagnostic peritoneal lavage, free peritoneal fluid on ultrasonography, or strong historical, physical, or radiographic evidence of abdominal injury
2. Persistent and significant hemodynamic instability with evidence of abdominal injury in the absence of extra-abdominal injury
3. Penetrating wounds to the abdomen
4. Pneumoperitoneum
5. Significant abdominal distension associated with hypotension

Table 108.3. Indications for Immediate Laparotomy for Children with Abdominal Trauma

The indications for emergent laparotomy in children with penetrating trauma are illustrated in Figure 108.3. Any gunshot wound to the abdomen mandates immediate exploration. Other types of penetrating wounds in the presence of unexplained hemodynamic compromise, evisceration, pneumoperitoneum, or any evidence of violation of the peritoneum

require prompt laparotomy.

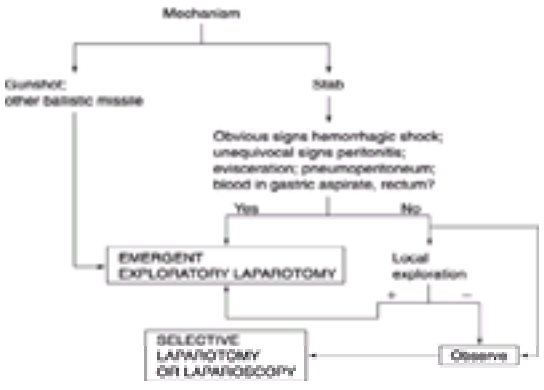

FIGURE 108.3. Management of penetrating abdominal trauma.

BLUNT ABDOMINAL TRAUMA

Abdominal Wall Contusions

Many children have minor trauma to their abdomen in the course of play and as a result of minor accidental events. Balls, bats, swings, toys, and rough play may cause contusions of the abdominal wall.

Children without signs of intra-abdominal pathology can be sent home. Those with a troubling history or any worrisome signs should receive a diagnostic laboratory evaluation and be observed in consultation with a surgeon. Suspicion for more serious intra-abdominal injuries is based on mechanism of injury and careful abdominal examination. Bilious or bloody vomiting, persistent vomiting, abdominal distension, any signs of peritoneal irritation, rectal blood or hematuria, hyperamylasemia, and elevated liver transaminase levels all suggest possible visceral injury. A low threshold for the use of abdominal CT should be maintained. Children with even minor contusions of the liver, spleen, pancreas, or hollow viscera should be hospitalized by the surgery service.

Solid Organ Injuries

The spleen is the most commonly injured intra-abdominal organ, followed by the liver. Most of these injuries are the result of automobile–pedestrian trauma, although falls and bicycle accidents are also common mechanisms. The potential morbidity and mortality result from the highly vascular anatomy of this organ, and hemorrhage into the large potential space of the peritoneal cavity.

Patients who have splenic injuries may present with either diffuse abdominal pain or localized tenderness. Subphrenic blood may cause referred left shoulder pain. Percussion and palpation tenderness is usually of greatest magnitude in the left upper quadrant of the abdomen. Abdominal radiographs occasionally reveal a medially displaced gastric bubble. CT or a radionuclide splenic scan will identify the extent of injury (Fig. 108.4 and Fig. 108.5).

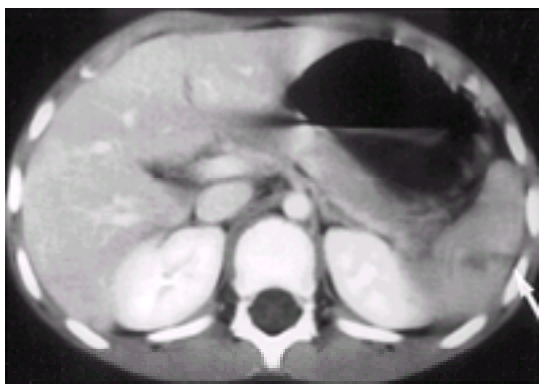

FIGURE 108.4. Abdominal computed tomography from a 13-year-old girl who fell from a horse onto her left side, showing a splenic laceration.

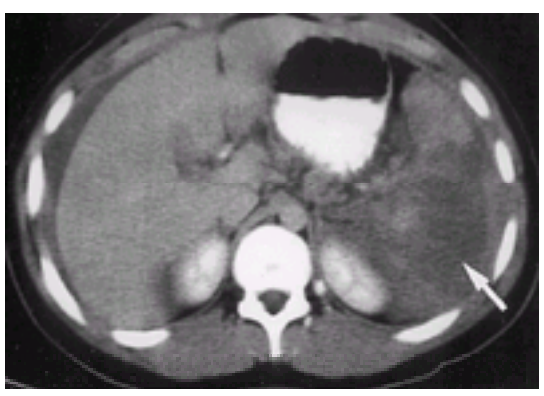

FIGURE 108.5. Abdominal computed tomography from a 10-year-old boy struck by a motor vehicle while crossing a street, showing massive splenic rupture and hemoperitoneum.

Management of splenic injuries has evolved over the last three decades since the recognition of the postsplenectomy sepsis syndrome, resulting from the influence of both clinical and diagnostic advances. Nonoperative management of splenic injuries has largely replaced the traditional treatment, which included splenectomy or splenorrhaphy. The safety of nonoperative management for most childhood spleen injuries has been well documented, and postsplenectomy sepsis has declined. The availability of noninvasive diagnostic CT also has allowed for greater confidence in the nonoperative approach to splenic trauma.

Blunt liver trauma is the most common fatal abdominal injury (Fig. 108.6). Mechanisms of injury are those common to splenic trauma. Diffuse abdominal tenderness may be a result of hemoperitoneum, but maximal tenderness is elicited in the right upper quadrant of the abdomen.

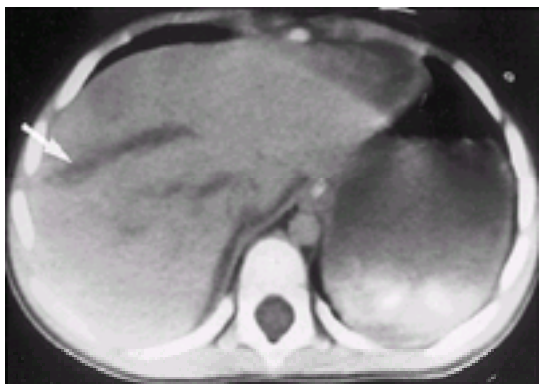

FIGURE 108.6. Abdominal computed tomography from a 9-year-old boy struck by a motor vehicle while riding his bicycle, showing a large liver laceration.

As with trauma to the spleen, nonoperative management of blunt hepatic injuries has become more common and is now the rule rather than the exception.

Blood transfusion therapy was necessary in 5.5% of patients in the National Pediatric Trauma Registry in 1997, and fewer than 1% of patients with blunt trauma admitted to Children's Hospital in Boston over the last year required transfusion therapy. The need for blood transfusion is based on careful monitoring of vital signs, clinical status, serial abdominal examinations, and analysis of hematocrit and fluid balance.

Pancreatic Injuries

Blunt abdominal injuries, particularly from bicycle handlebars, are the most common cause of pancreatic pseudocyst formation in children, although this injury is uncommon. Diagnosis is often delayed because of the nonspecific nature of subjective complaints and physical examination findings. The classic triad of epigastric pain, a palpable abdominal mass, and hyperamylasemia are detected only rarely in children and may develop slowly. The pancreas is relatively well protected, and associated trauma such as hepatic and intestinal injuries commonly are present when injury to the pancreas has occurred. Abdominal ultrasound and contrast CT (often serial examinations) are used to make the diagnosis (Fig. 108.7); however, acute pancreatic injuries may not be apparent on the initial CT scan.

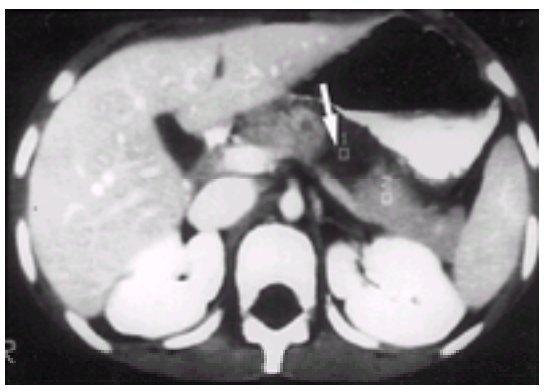

FIGURE 108.7. Abdominal computed tomography from a 6-year-old boy who fell onto the handlebar of his bicycle, showing a pancreatic hematoma and pseudocyst formation.

Severe injury of the pancreas is rare, but blood loss and leakage of enzyme-laden secretions may result in hypovolemia and peritonitis. Blunt abdominal trauma may also injure the ductal elements of the pancreas, and diagnosis depends on a

high index of suspicion, consideration of the mechanism of injury, physical examination, serum amylase determination, and diagnostic imaging. Of note, however, is that the absence of hyperamylasemia does not preclude pancreatic trauma because elevated serum amylase is detected in 16 to 80% of cases of blunt injury. Elevated serum amylase should suggest the possibility of pancreatic injury, but the absolute value does not correlate with the degree of injury. Elevation of the amylase in fluid returned from DPL suggests injury to bowel or pancreatic ductile elements.

Nasogastric decompression and bowel rest are indicated when pancreatic injury is suspected. Nonoperative therapy is normally used initially for children with isolated pancreatic pseudocyst caused by blunt trauma. Maturation of the pseudocyst may necessitate surgical drainage, although spontaneous resolution may occur in 25% of children. Experience with percutaneous drainage of pancreatic pseudocysts in children is increasing, but the traditional approach has been to use surgical internal drainage once a pseudocyst has persisted beyond 6 weeks. When severe pancreatic crush or transection is suspected, the surgeon may elect to perform immediate exploration and resection or drainage.

Hollow Abdominal Viscera Injuries

Intestinal perforation caused by blunt abdominal trauma is rare in the pediatric age group, but the most common causes of these injuries are automobile–pedestrian trauma, automobile lap belt injuries, and child abuse. The mechanisms of injury usually involve rapid acceleration or deceleration of a structure near a point of anatomic fixation (e.g., ligament of Treitz), or trapping of a piece of bowel between two unyielding structures such as a lap belt and the spine. Hollow viscera injury may be difficult to diagnose because physical findings may be minimal and/or nonspecific for the first few hours and abdominal CT is not particularly sensitive for this injury. However, succus entericus, bile, and activated pancreatic enzymes are extremely irritating to the peritoneum over time; the development of fever or worsening peritonitis on serial physical examinations should alert the examining physician to the possibility of bowel perforation.

Plain radiographs of the abdomen demonstrate free intra-abdominal air in only 30 to 50% of cases. Similarly, pneumoperitoneum or leakage of gastrointestinal contrast is only rarely seen on the CT scan. DPL may demonstrate bile or amylase in the effluent and is sensitive for bowel perforations. Most perforations or transections of bowel are found through laparotomy, which the surgeon has chosen to perform because of advancing peritonitis or unexplained persistent fever. Management depends on the site and extent of structural injury.

Late Presentations of Intra-Abdominal Trauma

Some children with abdominal trauma do not have evidence of intra-abdominal pathology on the initial evaluation but may return days or weeks later with abdominal distension and/or pain, persistent emesis, or hematochezia. In particular, three injuries are characterized by late presentations: 1) pancreatic pseudocyst (previously discussed), 2) duodenal hematoma, and 3) hematobilia.

Intramural duodenal hematoma is an uncommon injury that results from a direct blow to the epigastrium (blunt force delivered by a small-diameter instrument such as a broom handle or the toe of a boot) or from rapid deceleration and may cause partial or complete gastric outlet obstruction. Bleeding into the wall of the duodenum causes compression and therefore symptoms of intestinal obstruction, including pain, bilious vomiting, and gastric distension. Diagnosis is made by ultrasonography or a contrast upper gastrointestinal study, revealing the “coiled spring sign.” Injury of the pancreas must be suspected when duodenal hematoma is considered. Nonoperative management includes nasogastric decompression and parenteral nutrition for up to 3 weeks.

Rupture of the gallbladder is rare and is almost always associated with severe blunt trauma to the liver. Likewise, hematobilia is associated with hepatic trauma and is a result of pressure necrosis from an intrahepatic hematoma or direct injury to the biliary tree. Children with hematobilia present several days after a blunt abdominal trauma with abdominal pain and upper gastrointestinal bleeding. Cholangiography confirms the diagnosis. Embolization is used to achieve hemostasis, but hepatic resection is necessary when this treatment fails.

PENETRATING ABDOMINAL TRAUMA

Penetrating abdominal trauma is much less common than blunt trauma in the pediatric age group. However, the evolution of a more heavily armed society has resulted in a worrisome increase in the frequency with which children sustain penetrating injuries.

The high morbidity and mortality associated with penetrating trauma to the abdomen is a result of the destructive force of ballistic missiles and fragments, rapid hemorrhage of vascular structures and solid organs after missile and stab injuries, difficulty of surgical repair of grossly injured intra-abdominal organs, and postoperative complications. Intra-abdominal organs are at risk for penetrating trauma, depending on their size and location. The colon and small bowel are large in volume and are the most commonly injured structures, followed by the liver, spleen, and major vessels. Hypovolemia and/or signs of peritonitis are then the result of brisk hemorrhage and spillage of enteric contents into the peritoneal space.

The approach to these patients includes resuscitation of all life-threatening injuries and treatment of hemorrhagic shock. The need for laparotomy must be determined quickly, and broad-spectrum antibiotics, such as ampicillin (50 to 100 mg/kg), clindamycin (10 mg/kg), and gentamicin (2.5 mg/kg), should be given.

Gunshot Wounds

The destructive energy of ballistic missiles and fragments is related to mass and velocity (kinetic energy = $\frac{1}{2}MV^2$, where M is the mass, and V is the velocity), and more than 90% of gunshot wounds to the abdomen are associated with significant injuries. Hollow viscera and large vessels are often involved, and solid organs such as the liver and spleen

may demonstrate burst injuries. Therefore, laparotomy is mandated in all gunshot wounds to the abdomen.

Stab Wounds

Stab wounds to the abdomen carry potential for devastating injury depending on which intra-abdominal structures are involved. The extent of the injury also depends on the type, size, and length of the weapon and on the trajectory. Major vascular injuries pose the greatest threat; commonly injured vessels include the intra-abdominal aorta, the inferior vena cava, the portal vein, and the hepatic veins.

Anterior stab wounds are explored via laparotomy if hemodynamic instability or signs of peritonitis are present, if blood is noted in the gastric aspirate or on rectal examination, or if pneumoperitoneum or evisceration is noted (Fig. 108.3). Local exploration is needed to rule out penetration of the peritoneum even in minor stab wounds.

Stab wounds to the flank or back are less readily and less quickly diagnosed than anterior wounds; the retroperitoneal structures are more protected by paraspinal musculature, and bleeding is often tamponaded in this area. Dorsal stab wounds are sometimes managed nonoperatively unless hemodynamic instability or signs of peritonitis are present, although selective laparotomy is a common surgical strategy.

LAP AND SHOULDER BELT AND AIR BAG INJURIES

Children who are too large for child safety seats but too small for adult seat belts are at increased risk for injuries. In particular, children restrained only by lap belts in motor vehicles involved in rapid deceleration crashes are at risk to sustain Chance fractures (compression or flexion-distraction fractures of the lumbar spine) in association with intra-abdominal injuries (the lap belt complex). As many as half of the children with Chance fractures have intra-abdominal injuries; therefore, a high index of suspicion must be maintained to detect such injuries. The hallmark indicator of the lap belt complex is abdominal or flank ecchymosis in the pattern of a strap or belt. A normal abdominal CT does not rule out ruptured viscous, and laparoscopy or laparotomy should be considered for children in whom the lap belt complex is suspected. Carotid injuries caused by high-riding shoulder restraints in motor vehicle collisions are much less common.

Although it is well advertised that children younger than 12 years or less than 5 feet in height should not ride in the front seat of a vehicle that has functioning air bag restraints, significant injuries and deaths have occurred. Life-threatening injuries caused by air bag deployment are typically related to cervical spine injuries and closed head trauma. Less severe airbag injuries include abrasions to the face, neck, and chest; minor burns to the upper extremities; blunt ocular trauma; and chemical keratitis.

CHILD ABUSE

At least 1.6 million children are abused or neglected every year in the United States. Major blunt abdominal trauma resulting from physical child abuse is uncommon but highly fatal; mortality rates are as high as 50%. This high fatality rate is the result of the unfortunate but typical delay with which parents or caretakers who abuse children seek treatment.

Children who are seriously injured because of physical abuse commonly have more than one site of trauma; some of the injuries can be occult, and others may have been inflicted at different times. Abdominal injuries are usually inflicted by fists, feet, or small handheld objects and are rarely penetrating. The diagnosis of blunt abdominal injury caused by battering is difficult to make unless a high index of suspicion for child abuse is maintained. An important clue is often an implausible historical account for the seriousness of the injury. As with abdominal trauma caused by other mechanisms, physical examination findings may not be obvious. Laboratory analyses and abdominal CT may be necessary to confirm the diagnosis. However, severe injuries may present with obtundation and shock, abdominal distension, and tenderness. Intra-abdominal injuries most commonly involve the liver and spleen, as well as the pancreas–duodenum–jejunum region. In all such cases in which child battering is suspected, a child protection consultant should be involved early.

Suggested Readings

Agran PF, Dunkle DE, Winn DG. Injuries to a sample of seatbelted children evaluated and treated in a hospital emergency room. J Trauma 1987;27:58–64.

Akgur FM, Aktug T, Olguner M, et al. Prospective study investigating routine usage of ultrasonography as the initial diagnostic modality for the evaluation of children sustaining blunt abdominal trauma. J Trauma 1997;42:626–628.

Arkovitz MS, Johnson N, Garcia VF. Pancreatic trauma in children: mechanisms of injury. J Trauma 1997;42:49–53.

Bensard DD, Beaver BL, Besner GE, Cooney DR. Small bowel injury after blunt abdominal trauma: is diagnostic delay important? J Trauma 1996;41:476–483.

Bivens B, Jona J, Belin R. Diagnostic peritoneal lavage in pediatric trauma. J Trauma 1976;16:739–742.

Boyle EM, Maier RV, Salazar JD, et al. Diagnosis of injuries after stab wounds to the back and flank. J Trauma 1997;42:260–265.

Branney SW, Moore EE, Cantrill SV, et al. Ultrasound based key clinical pathway reduces the use of hospital resources for the evaluation of blunt abdominal trauma. J Trauma 1997;42:1086–1090.

Carillo EH, Bergamini TM, Miller FB, Richardson JD. Abdominal vascular injuries. J Trauma 1997;43:164–171.

Childhood injuries in the United States. Division of Injury Control, Centers for Disease Control. Am J Dis Child 1990;144:627–646.

Cobb L, Vinocur C, Wagner C, et al. Intestinal perforation due to blunt trauma in children in an era of increased non-operative treatment. J Trauma 1986;26:461–463.

Collins P, Golocovsky M, Salander J, et al. Intra-abdominal vascular injury secondary to penetrating trauma. J Trauma 1988;28(Suppl): S165–S170.

Cooper A, Floyd T, Barlow B, et al. Major blunt abdominal trauma due to child abuse. J Trauma 1988;28:1483–1487.

Ford E, Hardin W, Mahour G, et al. Pseudocyst of the pancreas in children. Am Surgeon 1990;56:384–387.

Galat J, Grisoni E, Gauderer M. Pediatric blunt liver injury: establishment of criteria for appropriate management. J Pediatr Surg 1990;11:1162–1165.

Georgi B, Massad M, Obeid M. Ballistic trauma to the abdomen: shell fragments versus bullets. J Trauma 1991;31:711–716.

Hennes H, Smith D, Schneider K, et al. Elevated liver transaminase levels in children with blunt abdominal trauma: a predictor of liver injury. Pediatrics 1990;86:87–90.

Jaffe D, Wesson D. Emergency management of blunt trauma in children. N Engl J Med 1991;324:1477–1482.

Kane N, Cronan J, Dorfman G, et al. Pediatric abdominal trauma: evaluation by computed tomography. Pediatrics 1988;82:11–15.

Karp M, Cooney D, Pros G, et al. The non-operative management of pediatric hepatic trauma. J Pediatr Surg 1988;18:512–518.

Keller MS, Stafford PW, Vane DW. Conservative management of pancreatic trauma in children. J Trauma 1997;42:1097–1100.

Lally K, Rosario V, Mahour G, et al. Evolution in the management of splenic injury in children. Surg Gynecol Obstet 1990;170:245–248.

Leppaniemi A, Haapiainen R, Kiviluoto T, et al. Pancreatic trauma: acute and late manifestations. Br J Surg 1988;75:165–167.

McKenney MG, Martin L, Lentz K, et al. 1,000 consecutive ultrasounds for blunt abdominal trauma. J Trauma 1996;40:607–612.

Meyer D, Thal E, Weigelt J. The role of abdominal computed tomography in the evaluation of stab wounds to the back. J Trauma 1989; 29:1226–1229.

Nagy KK, Krosner SM, Joseph KT. A method of determining peritoneal penetration in gunshot wounds to the abdomen. J Trauma 1997; 43:242–246.

Nance ML, Peden GW, Shapiro MB. Solid viscus injury predicts major hollow viscus injury in blunt abdominal trauma. J Trauma 1997;43:618–623.

Newman KD, Bowman LM, Eichelberger MR, et al. The lap belt complex: intestinal and lumbar spine injury in children. J Trauma 1990;30: 1133–1140.

Oldham K, Guice R, Kaufman R, et al. Blunt hepatic injury and elevated hepatic enzymes: a clinical correlation in children. J Pediatr Surg 1984;19:457–461.

Patton JH, Syden SP, Croce MA, et al. Pancreatic trauma: a simplified management guideline. J Trauma 1997;43:234–241.

Powell R, Green J, Oschner G, et al. Peritoneal lavage in pediatric patients sustaining blunt abdominal trauma: a reappraisal. J Trauma 1987; 27:6–10.

Rizzo M, Federle M, Griffiths B. Bowel and mesenteric injury following blunt abdominal trauma: evaluation with computed tomography. Radiology 1989;173:143–148.

Rothenburg S, Moore E, Marx J. Selective management of blunt abdominal trauma in children—the triage role of peritoneal lavage. J Trauma 1987;27:1101–1106.

Sahdev P, Garramone R, Schwartz R, et al. Evaluation of liver function tests in screening for intra-abdominal injuries. Ann Emerg Med 1991;20:830–841.

Saladino R, Lund D, Fleisher G. The spectrum of live and spleen injuries in childhood: failure of the pediatric trauma score and clinical signs to predict isolated injuries. Ann Emerg Med 1991;6:636–640.

Showers J, Apolo J, Thomas J, et al. Fatal child abuse: a two decade review. Pediatr Emerg Care 1985;1:66–70.

Stylianos S, Jacir NN, Hoffman MA, et al. Experimental volume replacement through lower extremity veins. J Trauma 1993;35:666–670.

Stylianos S, Wootten S, Lund D. Treatment of spleen and liver injuries in children without operation or transfusion (Abstract). Detroit: Eastern Association for Surgery of Trauma, 1991.

Subcommittee of Advanced Trauma Life Support of the American College of Surgeons Committee on Trauma. Advanced Trauma Life Support Instructor Manual. Chicago: American College of Surgeons, 1993.

Taylor GA, Eichelberger MR, Potter BM. Hematuria: a marker of abdominal injury in children after blunt abdominal trauma. Ann Surg 1988;208:688–693.

Taylor G, Fallat M, Potter B, et al. The role of computed tomography in blunt abdominal trauma in children. J Trauma 1988;28:1660–1664.

van der Sluis CK, Kingma J, Eisma WH, ten Duis HJ. Pediatric polytrauma: short-term and long-term outcomes. J Trauma 1997;43:501–506.

West K, Grosfeld J. Post-splenectomy sepsis: historical background and current concepts. World J Surg 1985;9:477–483.

Wisner D, Chun Y, Blaisdell W. Blunt intestinal injury. Arch Surg 1990;125:1319–1323.

Zantut LF, Ivatury RR, Smith RS, et al. Diagnostic and therapeutic laparoscopy for penetrating abdominal trauma: a multicenter experience. J Trauma 1997;42:825–831.

CHAPTER 109

Genitourinary Trauma

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In children who sustain multiple injuries, genitourinary trauma is second in frequency only to central nervous system trauma. Approximately 10% of trauma patients have urogenital injuries. Most injuries (90%) are the result of blunt trauma that involves crush injuries and acceleration/deceleration forces. Vehicular and pedestrian accidents account for a large percentage of blunt trauma. Other mechanisms of injury include falls and sports-related incidents. Penetrating injuries are less common in children than in adults. High-velocity bullet wounds produce direct tissue injury as well as damage to adjacent tissue because of the energy liberated by the missile. Low-velocity bullet wounds and stab wounds cause injury by penetrating the tissue directly. Iatrogenic trauma has been reported after operative procedures.

Injuries to other systems are often encountered in patients sustaining genitourinary trauma. Common associated problems include head injuries, fractures (extremities, pelvis, ribs, spine, skull), spinal cord injuries, and lacerations of the liver and spleen. Simultaneous upper and lower tract injuries are rare and are usually incompatible with survival. Isolated urologic injuries are rarely the cause of death.

The clinical approach to the injured child should strictly follow Advanced Trauma Life Support guidelines. Figure 109.1 provides an algorithm for diagnostic evaluation of the pediatric patient with genitourinary trauma. Urologic management may be temporized to permit urinary drainage in the initial phases; the patient may subsequently require operative procedures.

FIGURE 109.1. Algorithm for evaluation of the pediatric patient with genitourinary trauma. *IVP,* intravenous pyelogram; *CT,* computed tomography; *RBC,* red blood cells; *hpf,* high-powered field; *US,* ultrasound.

KIDNEY

The most common urinary tract injury encountered in children is injury to the kidney. More than 47% of genitourinary injuries involve the kidney. Blunt trauma accounts for up to 90% of renal injuries. Most pediatric renal trauma is sustained in motor vehicle accidents. Falls, sports incidents, and direct blows are also common mechanisms of injury.

Penetrating trauma accounts for a small percentage (4%) of cases. However, an increasing prevalence nationwide of gunshot and stab wounds may significantly alter these figures in the future. Approximately 10% of penetrating abdominal injuries involve the kidney. Penetrating renal trauma may occur as a complication of amniocentesis or percutaneous manipulation.

Associated injuries often occur, with head injuries being the most common. Associated intraperitoneal injuries occur in 80% of patients with penetrating renal trauma and 20% of patients with blunt renal trauma.

Children are more likely than adults to sustain renal injuries. In the child, the kidney is larger in proportion to the size of the abdomen than in the adult. The child's kidney may retain fetal lobations, which allow for easier parenchymal

disruption. Weaker abdominal musculature, a less well-ossified thoracic cage, and less developed perirenal fat and fascia fail to provide adequate protection for the kidney.

Coincidental congenital renal anomalies and intrarenal tumors have been documented in up to 10% of injuries. Symptoms and signs of a significant renal injury sustained with only minimal trauma should alert the clinician to the presence of preexisting anomalies, such as ureteropelvic junction obstruction, horseshoe kidney, pelvic kidney, or Wilms' tumor.

Classification

Renal injuries have been described using different classification systems based on the clinical and radiologic assessment of the patient. The Organ Injury Scaling Committee of the American Association for the Surgery of Trauma has devised an injury severity score, which represents an amalgamation of previous scales and was developed to facilitate clinical research. This classification system is illustrated in Figure 109.2. Grade I injuries include contusions or subcapsular, nonexpanding hematomas. Grade II injuries include nonexpanding hematomas confined to the retroperitoneum or lacerations less than 1 cm in depth without urinary extravasation. Grade III injuries include lacerations extending more than 1 cm into the renal cortex without collecting system rupture or urinary extravasation. Grade IV injuries include lacerations extending into the collecting system or renal vascular injuries with contained hemorrhage. Grade V injuries include completely shattered kidneys or avulsions of renal hilum with devascularized kidneys.

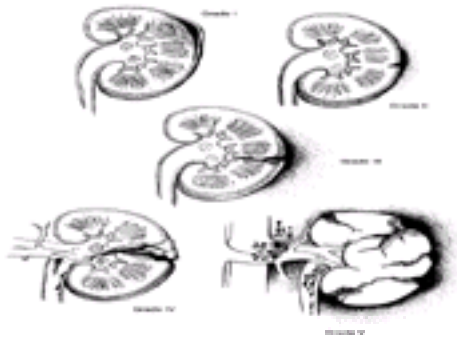

FIGURE 109.2. Classification of renal injuries as proposed by Organ Injury Scaling Committee.

Parenchymal contusions and hematomas are the most common renal injuries, accounting for 60 to 90% of all lesions from blunt trauma. Lacerations account for up to 10% of renal injuries and may involve disruption of the capsule, collecting system, or both. Severe injuries, such as shattered kidney or pedicle injuries, constitute approximately 3% of renal injuries. Pedicle injuries result from lateral displacement of the kidney with stretching of the tethered renal vessels.

Clinical Presentation

Children who sustain significant renal injuries usually present with localized signs such as flank tenderness, flank hematoma, or a palpable flank mass. Findings also include nonspecific symptoms and signs often associated with injury to other intra-abdominal organs. Generalized abdominal tenderness, rigidity of the abdominal wall, paralytic ileus, and hypovolemic shock may all be part of the clinical picture. Penetrating injuries to the chest, abdomen, flank, and lumbar regions should alert the clinician to the possibility of renal injury.

Hematuria has long been considered the cardinal marker of renal injury. Although controversy exists about the correlation between the degree of hematuria and the severity of the injury, the presence of any degree of hematuria should be regarded as potential indication of underlying renal injury or anomaly. It should be emphasized that hematuria may be absent in up to 50% of patients with vascular pedicle injuries and in 29% of patients with penetrating injuries. Hematuria out of proportion to the mechanism of injury should suggest a congenital anomaly or neoplasm.

Hematuria with abdominal symptoms has been associated with an increased risk of nonurologic intra-abdominal injuries. Injuries to other organs can be seen regardless of renal injury. Some series suggest that clinically significant liver and spleen injuries are more common in children with hematuria than are renal injuries.

Diagnostic Evaluation

Evaluation of the genitourinary system can be undertaken once life-threatening conditions have been identified and the child has been resuscitated. A urinalysis should be obtained in all patients with multisystem trauma or suspected isolated renal injury.

Radiographic evaluation of the genitourinary tract is necessary in patients with gross hematuria or microscopic hematuria associated with major mechanism, clinical findings indicative of renal injury, or other significant injuries (Fig. 109.1). Hematuria of more than 20 red blood cells per high-powered field (RBC/hpf) in patients without associated clinical findings has been recommended as a threshold for radiographic workup. Microscopic hematuria with shock is one of the clinical criteria used to guide radiographic evaluation in the adult population. Hypotension is an uncommon event in children with renal injuries and therefore is not a reliable indication of renal injury in the pediatric population.

Initial evaluation of suspected pediatric renal trauma should include radiographs of the chest, abdomen, and pelvis. Plain

films may show obliterated renal and psoas shadows, scoliosis with the concavity toward the injured site, intra-abdominal mass effect, or a coincident rib, spinous process, or pelvic fracture.

Traditionally, the intravenous pyelogram (IVP) has been the cornerstone of evaluation in renal trauma (Fig. 109.3). IVP is available in most institutions and provides information about both kidneys. It can be obtained on an unstable patient urgently in the emergency department (ED) or in the operating room before surgery. A scout radiograph should be made before the contrast study. IVP is performed by administering 1 to 4 mL/kg (maximum 100 mL) of nonionic contrast agent intravenously, followed by abdominal films in 1, 5, and 15 minutes. A one-shot IVP may be indicated for the unstable patient and is completed by obtaining a single film 3 to 5 minutes after injection of contrast material. Indications of renal injury include delayed excretion of contrast by the injured kidney, nonvisualization of the caliceal system, or extravasation of contrast into the perinephric tissues. The presence of a normally functioning kidney contralateral to the injured kidney should be specifically noted.

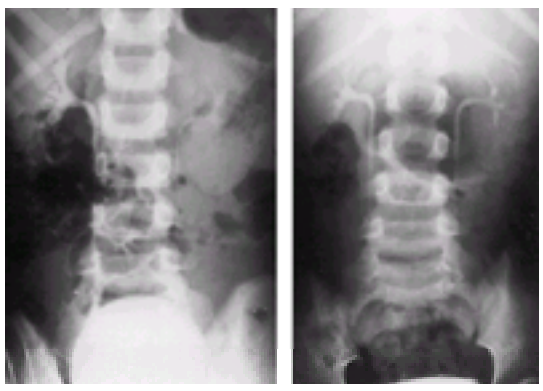

FIGURE 109.3. Renal contusion. Intravenous pyelogram (IVP) *(left panel)* shows decreased concentration of contrast material in the left kidney. Follow-up IVP *(right panel)* 1 month after the injury reveals normal renal function.

Even under optimal conditions, the IVP cannot always reliably identify and stage renal trauma. In one review, Cass reported the urogram to be definite in 5% of contusions, 50% of lacerations, and 29% of pedicle injuries. However, the importance of the IVP lies in rapidly assessing the overall functional and anatomic integrity of the kidney, as well as the presence of urinary extravasation and the presence of a congenital anomaly. More recently, contrast-enhanced computed tomography (CT) has become the preferred study for evaluation of major abdominal injury, including renal injury (Fig. 109.4). CT scan has certain advantages over the IVP, the most important of which is the detection of associated injuries. In addition, CT provides three-dimensional views and imaging independent of the vascularity of the kidney.

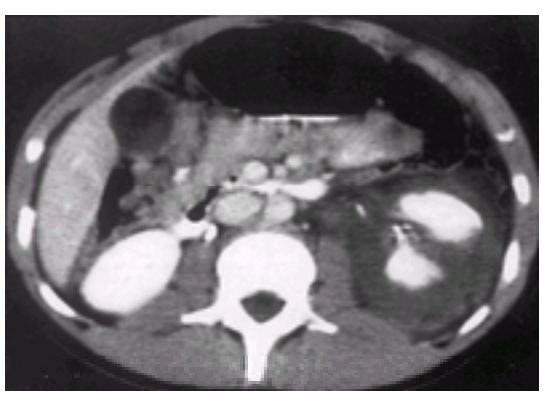

FIGURE 109.4. Renal fracture. Computed tomography section of the abdomen shows fracture of the left kidney with moderate subcapsular hematoma.

CT imaging can be used to determine the degree of renal parenchymal injury, to evaluate the presence of nonviable tissue, to demonstrate extravasation and perirenal collections, and to diagnose most pedicle injuries. The diagnostic accuracy of the CT scan has been reported as high as 98%, and CT scanning is becoming the standard imaging modality for renal trauma. CT scanning has also proven to be a useful tool for following patients after trauma.

Ultrasonography has been advocated for the staging of renal trauma. Its sensitivity in demonstrating renal injury is only 70% compared with CT scanning. However, ultrasound has certain advantages over CT scanning. It is readily available, can be performed at the bedside, and does not require the use of contrast material or sedation. Ultrasound can evaluate the size of the kidney and the integrity of the urinary drainage system. Pulsed flow duplex Doppler ultrasound can assess renal arterial and venous flow and may represent the most immediate means of screening for renal pedicle injury.

Ultrasound can be particularly helpful in patients with perirenal collections who require early follow-up imaging during the hospital stay. It may be an alternative modality for the evaluation of the pregnant trauma patient. It is also often used for long-term outpatient follow-up.

Angiography (Fig. 109.5) has been largely replaced by noninvasive modalities, especially in the pediatric patient in whom technical problems with vascular access result in a higher complication rate than in adults. Arteriography does not add

useful information to contrast CT scanning and may increase diagnostic delay during the preoperative workup. It is useful in patients who require therapeutic embolization of an active bleeding site.

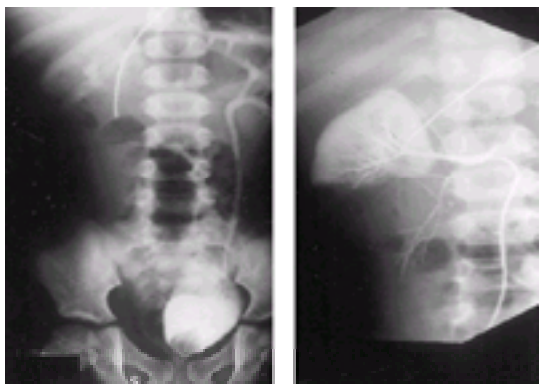

FIGURE 109.5. Renal fracture. Intravenous pyelogram *(left panel)* demonstrates nonvisualization of the lower pole of the right kidney. The arteriogram *(right panel)* confirms the diagnosis.

Radionuclide imaging can assess pedicle competence, parenchymal integrity, and renal function. A nuclear scan can be obtained instead of sonography to assess change in size of perinephric collections. Its most important role is in follow-up evaluation of renal injury (Fig. 109.6), particularly in the setting of new-onset hypertension.

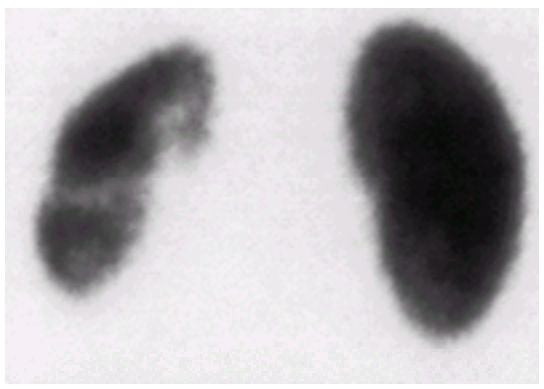

FIGURE 109.6. Follow-up renal scan of patient in Figure 109.5 obtained 4 months after the injury. The study reveals several areas of decreased radiotracer uptake in the left kidney.

In conclusion, hemodynamically stable patients who have experienced major mechanisms of injury, and who have associated injuries, clinical findings, or gross hematuria should undergo radiographic evaluation. These patients should be evaluated by CT scan. Children with suspected isolated renal injuries or microscopic hematuria of more than 20 RBC/hpf without associated findings can be evaluated by IVP or CT scan. Children who remain unstable despite resuscitative measures should undergo a one-shot IVP before emergency laparotomy. This procedure is particularly important to confirm the presence of a normal contralateral kidney. Patients with microscopic hematuria of less than 20 RBC/hpf, stable vital signs, no focal findings, and no signs of injury to other organ systems do not require emergent imaging. These patients may be discharged and can be evaluated on an outpatient basis with CT, IVP, or ultrasound if hematuria persists. However, in some centers, management of these patients involves hospitalization for observation followed by nonemergent radiographic evaluation.

Management

The principle underlying the management of pediatric renal trauma is preservation of renal tissue and function with minimal morbidity and mortality. Once the patient's general condition has been assessed and the presence of associated injuries established, treatment of renal trauma should proceed based on staging of the traumatic lesion.

In cases of blunt trauma, minor injuries, such as contusions and lacerations without urinary extravasation (grades I, II, and III), are treated conservatively. Current therapy involves strict bed rest, analgesia, and prophylactic antibiotics. Once gross hematuria has cleared, limited activity is instituted until microscopic hematuria resolves.

Management of the remaining patients (grades IV and V) evokes significant controversy. Treatment of deep lacerations and fragmentation depends on the child's hemodynamic status, degree of urinary extravasation and renal bleeding, and associated injuries. More patients are now being managed conservatively with close monitoring of vital signs, serial hematocrits, and broad-spectrum antibiotics. Patients treated with conservative nonoperative management have a complication rate of up to 50%. These complications include persistent or recurrent hemorrhage, extravasation and urinoma formation, infection, infarction, and segmental hydronephrosis. The nephrectomy rate with delayed surgery is usually less than 10%. Immediate surgical intervention results in a nephrectomy rate of 10 to 20% in most series. However, there is a significant decrease in the number of surgical procedures performed for subsequent complications.

Renal pedicle injuries are uncommon in the pediatric population and few revascularization attempts are successful. Reconstruction of unilateral pedicle injuries should be attempted only in children who are stable, who do not have major

associated injuries, who are less than 12 hours from initial injury, where the kidney is shown to be intact, and where contralateral function is good. If reconstruction fails, nephrectomy should then be performed. Whether the infarcted kidney should be removed electively in children treated nonoperatively is debatable. Bilateral renal vascular injury is associated with high morbidity and mortality. Surgery in these patients should be performed within 24 hours after the injury.

Penetrating renal injuries have traditionally been managed with operative intervention. Hemodynamically stable patients with isolated lower-grade injuries can be treated expectantly. Delayed bleeding may occur in up to 24% of patients with grades III and IV injuries who are managed nonoperatively. Therefore, these patients should undergo renal exploration when laparotomy is indicated for other injuries. Children with vascular injuries, urinary extravasation, or hemodynamic instability require surgery.

Complications

Short-term complications of renal trauma include delayed hemorrhage, urinary extravasation, abscess formation, and ureteral obstruction secondary to clot formation. Long-term complications include hypertension (probably less than 5%), hydronephrosis, arteriovenous fistula, renal intestinal fistula, and stone formation. The child with a history of renal trauma requires regular follow-up for at least 1 year, and longer in severe trauma, to ensure that complications are diagnosed and treated promptly.

URETER

Ureteral injuries are uncommon in the child, accounting for less than 1% of all urologic trauma. These injuries can be caused by blunt, penetrating, or iatrogenic trauma.

Blunt trauma usually involves the ureteropelvic junction. Disruption of the ureter from the pelvis results from stretching of the ureter by sudden hyperextension of the trunk. Traditionally, this injury has been described more often in children. The degree of hyperextension necessary to cause avulsion of the ureter was thought to be fatal in adults. However, an increased number of ureteropelvic junction injuries has recently been reported in adults. In the past, many of the injuries in adults may have been misdiagnosed as parenchymal lacerations involving the collecting system.

Trauma to the ureter should be suspected in patients presenting with fracture of the transverse process of a lumbar vertebra. Pelvic fracture, hip fracture, lower rib fracture, splenic laceration, liver laceration, and diaphragmatic rupture have also been reported in association with ureteral injuries.

Early diagnosis of blunt ureteral injuries is critical. The injury is often overlooked. Fewer than 50% of patients are diagnosed within 24 hours of presentation. The physical examination may be unremarkable. However, an enlarging flank mass in the absence of signs of retroperitoneal bleeding suggests urinary extravasation. Hematuria is an unreliable sign. The urinalysis may be normal in 30% of confirmed cases. When the diagnosis has been delayed, ureteral injury may manifest with fever, chills, lethargy, leukocytosis, pyuria, bacteriuria, flank mass or pain, fistulas, and ureteral strictures.

Avulsion of the ureter should be suspected when the IVP demonstrates extravasation of contrast material and nonfilling of the affected ureter. Contrast-enhanced CT is an appropriate alternative to the IVP. CT findings suggestive of ureteral injury include medial perirenal extravasation of contrast material, a circumrenal urinoma, and lack of opacification of the ureter distal to the injury. Delayed images must be obtained regardless of which diagnostic modality is used. Both CT scan and IVP have been shown to have a low sensitivity for ureteral injuries, identifying only 33% of cases. Retrograde pyelogram may be a more reliable examination, but it is rarely performed in the initial evaluation of the trauma patient.

The management of complete transection of the ureter depends on the level of the injury. Important elements include debridement of devitalized tissue and a watertight, tension-free anastomosis. Ureteroureterostomy, transureteroureterostomy, or pyeloureterostomy is recommended for upper and midureteral injuries. Mid- to lower ureteral injuries are best treated with ureteroneocystostomy. Placement of a ureteral stent is indicated in most cases. Conservative treatment with stent placement alone may be adequate for patients with lacerations.

If the ureteral lesion is identified within 5 to 10 days of the injury, prompt repair of the ureter is indicated. If diagnosis is delayed for more than 10 days, urinary diversion above the lesion should be performed with subsequent definitive repair 4 to 6 months later. The incidence of nephrectomy is approximately 5% when the injury is detected early, but it is as high as 32% when recognition is delayed.

Penetrating injuries may occur at any point along the length of the ureter and are associated with injuries to other intra-abdominal organs in up to 90% of cases. Stab wounds rarely cause ureteral injuries. However, up to 50% of patients with gunshot wounds to the abdomen have injury to the ureter. Occasionally, the ureter may be accidentally injured during pelvic operations or ureteroscopy. Repair of penetrating injuries to the ureter in the child follows the same guidelines as those observed in adults. Injuries caused by high-velocity missiles require wide debridement to ensure an adequate anastomosis and to preserve blood supply.

BLADDER

Bladder injuries may occur after blunt or penetrating trauma. Blunt trauma secondary to motor vehicle accidents is the leading cause of bladder injuries. Approximately 80% of bladder injuries are associated with pelvic fractures and penetration of the bladder by a bony fragment. However, only 10% of patients with pelvic fractures sustain lower urinary tract injury. The probability of having an associated bladder injury increases proportionally with the number of fractured pubic rami. Mortality rate associated with bladder rupture may be as high as 40%. Death is usually caused by associated

head injuries rather than by bladder injuries themselves.

During childhood, the bladder has a higher abdominal location, which renders the organ more susceptible to injury than in adults. The bladder can also be more easily damaged when full.

Bladder injuries are classified as extraperitoneal, intraperitoneal, or combined. Extraperitoneal injuries are more frequently associated with pelvic fractures. Intraperitoneal injuries are usually caused by blunt trauma to a distended bladder. Combined injuries are usually seen with gunshot wounds. Bladder injuries may range from contusions to rupture. Contusions are incomplete, nonperforating tears of the mucosa.

Hematuria and dysuria are symptoms commonly seen at presentation. More than 90% of patients with rupture of the bladder have hematuria. Microscopic hematuria is associated with less severe injuries such as contusions. Inability to void may be associated with large tears. Patients with intraperitoneal ruptures may develop a palpable fluid wave from extravasation of urine into the peritoneal cavity and peritoneal irritation. Elevated levels of blood urea nitrogen out of proportion to creatinine result from more rapid peritoneal reabsorption of urea.

Diagnostic evaluation is indicated in patients who sustain pelvic or lower abdominal trauma with hematuria and inability to void. Evaluation begins with a radiograph to exclude a pelvic fracture. If no pelvic fracture, no blood at the meatus, and no dislocation of the prostate on rectal examination are present, the urethra can be catheterized gently and a retrograde cystogram can be performed. A stress cystogram with the bladder full and anteroposterior and oblique views and a postdrainage film should be obtained. Observation of strict sterile technique is mandatory when performing the cystogram. Foley balloon catheters may result in false-negative cystograms because the inflated balloon may occlude a small tear. False-negative results may also occur if the bladder is not adequately distended with contrast material. If urethral catheterization is not successful or a urethral injury is suspected, a retrograde urethrogram should be completed. Once bladder or urethral injury is ruled out, imaging of the upper tract can be initiated.

Cystogram of a contused bladder may show a teardrop shape or elevation of the bladder out of the pelvis. No evidence of extravasation of contrast material will be apparent. Extraperitoneal perforation is demonstrated by the presence of extravasated medium in the area of the pubic symphysis and pelvic outlet. In cases of intraperitoneal rupture, contrast may outline intra-abdominal organs or paracolic gutters. Contrast-enhanced CT scan and CT cystography can also be used in the evaluation of bladder injuries.

Conservative management with or without urethral catheter drainage is the standard of care in patients with contusion. Extraperitoneal vesical rupture can be managed by urethral catheter or suprapubic drainage for 7 to 10 days. Treatment of large extraperitoneal tears or intraperitoneal tears involves transperitoneal exploration and repair with placement of a suprapubic cystostomy tube.

Iatrogenic bladder injuries may occur during herniorrhaphy, cystoscopy, and umbilical artery cutdown. Patients with myelodysplasia who have undergone bladder augmentation may experience spontaneous bladder rupture in the presence of infection, bacteremia, or overdistension. Symptoms and signs of sepsis, as well as shoulder pain, may be encountered at presentation. Emergent exploration is indicated after a cystogram is completed.

URETHRA

Motor vehicle accidents, straddle injuries, and instrumentation account for most urethral injuries sustained during childhood. Urethral injuries occur primarily in males. In boys, the urethra is divided by the urogenital diaphragm into an anterior (pendulous and bulbous) and posterior (membranous and prostatic) urethra (Fig. 109.7). Anterior and posterior urethral injuries differ from each other by mechanism of injury, clinical presentation, and treatment.

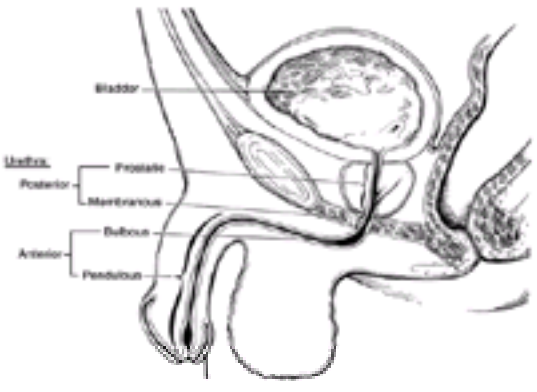

FIGURE 109.7. Sagittal section of male lower urinary tract illustrating levels of urethra.

Anterior urethral injuries result from direct trauma, are often isolated, and are associated with a low mortality rate. The pendulous urethra may be damaged by blunt or penetrating forces. Bulbar injuries are commonly caused by straddle injuries as the urethra is compressed between the symphysis pubis and a solid object. The major sign of acute anterior injury is bleeding from the urethra. Blood at the meatus has been reported in up to 90% of patients sustaining anterior urethral injuries. Other findings include hematuria, inability or difficulty voiding, and periurethral or perineal edema and ecchymosis. Perineal ecchymosis in the shape of a butterfly is typical for these injuries. Blind placement of a urethral catheter may convert a partial tear into a complete transection and therefore should be discouraged.

Diagnosis can be made by performing a retrograde urethrogram under sterile conditions. A Foley catheter appropriate for the size of the patient is inserted into the urethra to the fossa navicularis without inflating the balloon. Contrast material is injected via the catheter into the urethra and images are obtained in an oblique position. If a Foley catheter is already in place, the urethrogram can still be performed via a small feeding tube passed alongside the catheter. An optional technique involves simple retrograde syringe injection through the urethral meatus. Retrograde urethrography should be performed under fluoroscopy with minimal pressure. Gross extravasation of contrast agent at the site of the injury without visualization of the proximal urethra and bladder is diagnostic for complete rupture of the urethra. Partial rupture is represented by localized extravasation at the site of the injury with contrast passing into the proximal urethra and bladder. If no extravasation is noted, the urinary catheter can be gently advanced into the bladder.

Anterior urethral injuries can be managed by 7 to 10 days of urethral catheterization and antibacterial therapy. More severe injuries require urinary diversion by suprapubic cystostomy. Penetrating wounds demand surgical repair with exploration, debridement of devitalized tissue, and copious irrigation.

Posterior urethral injuries occur with severe trauma to the body and are usually associated with other injuries, particularly pelvic fractures. The mortality rate with fractured pelvis has been reported to be as high as 30%. The high death rate in these patients is attributed primarily to associated injuries.

The urogenital diaphragm located between the pubic rami fixes the membranous urethra and makes it vulnerable to rupture when the pubic arch is fractured. Tears may also result from shearing of the prostatic urethra at the superior border of the urogenital diaphragm. Injuries to the prostatic urethra may extend to the bladder neck. Posterior urethral injuries in men almost uniformly occur distal to the prostate. In adults, the mature prostate, puboprostatic ligament, and bladder stabilize the prostatic urethra making it less susceptible to trauma.

Signs of proximal urethral injury include blood at the meatus, hematuria, inability to void, displacement of the prostate on rectal examination, and perineal ecchymosis. Catheterization of the urethra is contraindicated. The diagnosis is best made by retrograde urethrography as described for anterior urethral injuries. CT scan is not adequate for diagnosing urethral injuries and is only presumptive if extravasation is detected at the bladder neck or urethra (Fig. 109.8). The IVP may demonstrate elevation of the bladder out of the pelvis.

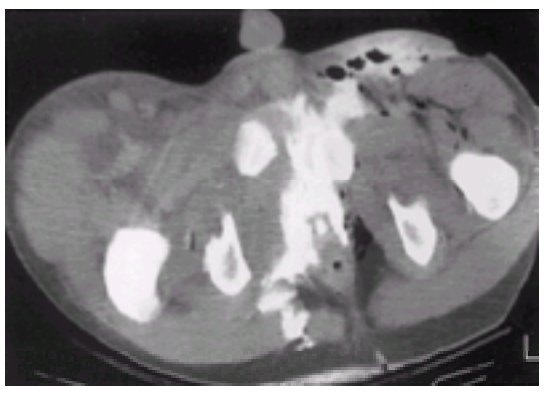

FIGURE 109.8. Posterior urethral disruption and pelvic fracture. Computed tomography of pelvis shows extravasation of contrast from posterior urethra into the surrounding tissues.

Initial management of posterior urethral injuries remains controversial. Therapeutic options vary from immediate exploration with primary repair to placement of a suprapubic tube with delayed urethroplasty. Urethral rupture may also be treated by realigning the urethra over an indwelling urethral catheter. Primary repair or operative realignment should be reserved for urethral injuries associated with rectal laceration, a high-riding bladder, or injury to the bladder neck.

Although the incidence of urethral strictures is higher in patients undergoing cystostomy with delayed repair, impotence and urinary incontinence are more prevalent when immediate urethral repair or realignment is performed and secondary repair of stricture is often needed. Some studies suggest that long-term outcome may be determined by the location of the injury, regardless of treatment method. Membranous urethral tears may have a more favorable outcome. Bladder neck injuries have the lowest rate of continence.

Trauma to the urethra in girls is rare because the female urethra is relatively mobile and short. Injuries may occur after surgical procedures or instrumentation. Most serious injuries involve the vesicourethral junction and result from blunt abdominal trauma in motor vehicle accidents. The lesion generally occurs in association with pelvic fractures. The injury often extends to the vagina. Urethral injuries in the female are treated with suprapubic drainage and elective repair. Some authors recommend primary operative repair of the urethral rupture with closure of associated vaginal tears. Long-term complications of this injury include urethrovaginal fistula, vaginal stenosis, incontinence, and urethral stricture.

SCROTUM

Scrotal trauma may occur as a result of straddle injuries or bicycle accidents or during sporting events. The patient may present with scrotal tenderness, edema, and ecchymosis. Potential injuries include skin or dartos ecchymoses and lacerations, intrascrotal hematomas, testicular hematomas, testicular dislocation, and testicular rupture. In addition, a testicle may torse after trauma.

When inspection of the scrotum and its contents is obscured by local swelling and pain, ultrasonography is helpful to define the extent of the injury. An intratesticular hematoma may show as an echogenic or hypoechoic testicular mass. A hematocele produces a complex extratesticular fluid collection. Sonographic findings of rupture include presence of hematocele, mixed parenchymal echogenicity, intraparenchymal hemorrhage, and disruption of the tunica albuginea or parenchyma. If the ultrasound exam is inconclusive, radionuclide scanning may provide additional information. Both ultrasonography and nuclear scintigraphy help in the diagnosis of testicular torsion (see Chapter 122).

Patients without evidence of injury to the testes who sustain intrascrotal hematomas, skin ecchymosis, or skin and dartos injury only can be managed conservatively. Treatment consists of ice packs and scrotal support. Minor testicular injuries such as contusions or hematomas can also be treated conservatively. Large testicular hematomas may require surgical management. Delay in surgery may lead to ischemic necrosis, secondary infections, and disruption of testicular function.

Testicular dislocation occurs as a result of an upward blow to the scrotum. In most cases, the dislocated testis lies under the abdominal wall. Associated injuries, such as pelvic fracture, are common. Operative repair is required if closed reduction fails.

Testicular rupture with tear of the tunica albuginea and extravasation of testicular contents into the scrotal sac requires surgical exploration and repair. Testicular salvage is more likely when exploration is performed within 24 hours of the injury. Other injuries requiring surgical management include tense hematoceles and torsion after trauma.

Superficial lacerations of the scrotum can be repaired using absorbable sutures. Local infiltration with lidocaine with epinephrine provides adequate anesthesia. Urologic consultation should be obtained if the laceration extends through the dartos. Physical examination of the scrotal contents determines the need for debridement and primary closure. All penetrating testicular injuries require surgical exploration.

Degloving injuries of the scrotum can be seen after motor vehicle (particularly motorcycle), industrial, or farm machinery accidents. Scrotal injuries are associated with varying degrees of penile skin loss. The underlying penile and scrotal structures are usually spared. Management involves debridement and coverage of the defect by skin flaps or grafting.

PENIS

The most common cause of penile trauma in infants is iatrogenic, especially at the time of circumcision. Complications include transection of the glans, urethrocutaneous fistula, deskinning of the penile shaft, and coagulation necrosis of the entire penis from electrocautery. These injuries usually require extensive surgical repair.

Blunt penile trauma from toilet seats falling on the glans or distal shaft has been described in toddlers. Significant injury to the corporal bodies or the urethra is rare and patients can be managed expectantly with warm soaks. Although the child does not commonly experience urinary retention, he may be more comfortable voiding in a tub of warm water.

Tourniquet injuries may result from bands, rings, or human hair. In the infant, strangulation with a fine hair may be difficult to recognize because of local edema. The initial diagnosis may be balanitis or paraphimosis. Local or general anesthesia may be required to expose and remove the hair. Complications include urethrocutaneous fistula or loss of the penis.

Zipper entrapment of the penis or foreskin can be managed in the ED by cutting the median bar of the zipper with wire cutters and disassembling the zipper mechanism (Fig. 109.9). Conscious sedation may facilitate the procedure. Edema can subsequently be treated with warm soaks.

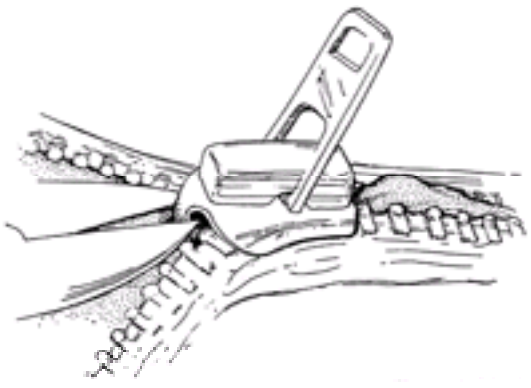

FIGURE 109.9. Penile zipper injury. A wire cutter may be used to cut the median bar of the zipper, releasing the two sides of the zipper and freeing the penis.

Fracture of the penis is produced by traumatic rupture of the corpus cavernosum. This injury usually occurs when the erect penis is forced against a hard surface. The patient may hear a cracking sound and develop pain, edema, and deformity of the shaft of the penis. The urethra is rarely involved. Fracture of the penis can be managed conservatively with bed rest, ice packs, and a pressure dressing. Most injuries require surgical treatment with evacuation of the penile hematoma, repair of the torn tunica albuginea, and a pressure dressing.

Superficial lacerations of the penile shaft can be repaired with absorbable sutures under local anesthesia or penile block. Lacerations extending to the corporal bodies or the urethra, as well as lacerations of the ventral surface of the penis,

require urologic consultation. Diagnostic evaluation includes a retrograde urethrogram to define the extent of the injury. Injuries to the corporal bodies should be repaired primarily to prevent fibrosis and impotence. Injuries to the urethra may require urinary diversion.

PERINEUM

The mechanism most commonly associated with trauma to the female perineum is a straddle-type injury. These injuries may cause vulvar hematomas, which usually respond to treatment with ice packs and bed rest. Patients experiencing mild urinary retention may be more comfortable voiding in a tub of warm water. Massive or expanding hematomas may require evacuation.

Superficial lacerations of the perineum can be treated conservatively at home with sitz baths. Deep lacerations may extend into the rectum or urethra. Rectal penetration requires a diverting colostomy. Suprapubic cystostomy or primary repair should be performed if the urethra is disrupted.

Vaginal lacerations must always be suspected in patients with severe trauma to the external genitalia or penetration by foreign object. If a significant vaginal laceration is noted, endoscopy with sedation or general anesthesia is necessary for a full evaluation. The possibility of extension into the urethra, bladder, or rectum must be investigated. The vaginal laceration is debrided and repaired with fine absorbable sutures.

SEXUAL ABUSE

When common accidental situations fail to explain certain genitourinary injuries, the possibility of sexual abuse should be considered. Injuries resulting from sexual abuse include abrasions and hematomas in the penile shaft, vaginal lacerations, and perineal hematomas (see also Chapter 76 and Chapter 128).

Suggested Readings

Abou-Jaoude WA, Sugarman JM, Fallat ME, et al. Indicators of genitourinary tract injury or anomaly in cases of pediatric blunt trauma. J Pediatr Surg 1996;31:86–90.

Ahmed S, Neel KF. Urethral injury in girls with fractured pelvis following blunt abdominal trauma. Br J Urol 1996;78:450–453.

Avanoglu A, Ulman I, Herek O, et al. Posterior urethral injuries in children. Br J Urol 1996;77:597–600.

Baldwin DD, Landa HM. Common problems in pediatric gynecology. Urol Clin North Am 1995;22:170.

Barach E, Martin G, Tomlanovich M, et al. Blunt pelvic trauma with urethral injury in the female: a case report and review of the literature. J Emerg Med 1984;2:101–105.

Baskin LS, McAninch JW. Childhood urethral injuries: perspectives on outcome and treatment. Br J Urol 1993;72:241–246.

Bass DH, Semple PL, Cywes S. Investigation and management of blunt renal injuries in children: a review of 11 years' experience. J Pediatr Surg 1991;26:196–200.

Bertini JE Jr, Corriere JN Jr. The etiology and management of genital injuries. J Trauma 1988;28:1278–1281.

Bond SJ, Gotschall CS, Eichelberger MR. Predictors of abdominal injury in children with pelvic fracture. J Trauma 1991;31:1169–1173.

Boone TB, Gilling PJ, Husmann DA. Ureteropelvic junction disruption following blunt abdominal trauma. J Urol 1993;150:33–36.

Campbell EW Jr, Filderman PS, Jacobs SC. Ureteral injury due to blunt and penetrating trauma. Urol 1992;40:216–220.

Carroll PR, Klosterman PW, McAninch JW. Surgical management of renal trauma: analysis of risk factors, technique, and outcome. J Trauma 1988;28:1071–1077.

Cass AS. Bladder trauma in the multiple injured patient. J Urol 1976;115:667–669.

Cass AS. Blunt renal trauma in children. J Trauma 1983;23:123–127.

Cass AS. Urethral injury in the multiple-injured patient. J Trauma 1984; 24:901–906.

Cass AS, Luxenberg M. Conservative and immediate surgical management of blunt injuries. J Urol 1983;130:11–16.

Cass AS, Luxenberg M. Accuracy of computed tomography in diagnosing renal artery injuries. Urology 1989;34:249–251.

Cass AS, Luxenberg M. Simultaneous upper and lower urinary tract injury from external trauma. Urology 1990;36:226–227.

Cheng DL, Lazan D, Stone N. Conservative treatment of type III renal trauma. J Trauma 1994;36:491–494.

Deane A. ABC of major trauma. Trauma of the lower urinary tract. Br Med J 1990;301:545–547.

Eastham JA, Wilson TG, Ahlering TE. Radiographic assessment of blunt renal trauma. J Trauma 1991;31:1527–1528.

Feins NR. Multiple trauma. Pediatr Clin North Am 1979;26:759–771.

Fleisher GR. Prospective evaluation of selective criteria for imaging among children with suspected blunt renal trauma. Pediatr Emerg Care 1989;5:8–11.

Geist R. Sexually related trauma. Emerg Med Clin North Am 1988; 6:439–466.

Godec CJ. Genitourinary trauma. Urol Radiol 1985;7:185–191.

Goldman HB, Dmochowski RR, Cox CE. Penetrating trauma to the penis: functional results. J Urol 1996;155:551–553.

Hardeman SW, Husmann DA, Chinn HKW, et al. Blunt urinary tract trauma: identifying those patients who require radiological diagnostic studies. J Urol 1987;138:99–101.

Hendren WH, Peters CA. Lower urinary tract and perineal injuries. In: Touloukian RJ, ed. Pediatric Trauma. 2nd ed. St Louis: Mosby, 1990:371–398.

Hensle TW, Dillon P. Renal injuries. In: Touloukian RJ, ed. Pediatric Trauma. 2nd ed. St Louis: Mosby, 1990:358–370.

Herschorn S, Radomski SB, Shoskes DA, et al. Evaluation and treatment of blunt renal trauma. J Urol 1991;146:274–277.

Hilton SW, Kaplan GW. Imaging of common problems in pediatric urology. Urol Clin North Am 1995;22:13–18.

Hochberg E, Stone NN. Bladder rupture associated with pelvic fracture due to blunt trauma. Urology 1993;41:531–533.

Kawashima A, Sandler CM, Corriere JN Jr, et al. Ureteropelvic junction injuries secondary to blunt abdominal trauma. Radiology 1997;205: 487–492.

Klein S, Johs S, Fujitani R, et al. Hematuria following blunt abdominal trauma. Arch Surg 1988;123:1173–1177.

Knudson MM, McAninch JW, Gomez R, et al. Hematuria as a predictor of abdominal injury after blunt trauma. Am J Surg 1992;164:482–486.

Koraitim MM, Marzouk ME, Atta MA, et al. Risk factors and mechanism of urethral injury in pelvic fractures. Br J Urol 1996;77:876–880.

Kotkin L, Brock JW. Isolated ureteral injury caused by blunt trauma. Urology 1996;47:111–113.

Kristjansson A, Pedersen J. Management of blunt renal trauma. Br J Urol 1993;72:692–696.

Kuzmarov IW, Morehouse DD, Gibson S. Blunt renal trauma in the pediatric population: a retrospective study. J Urol 1988;126:648–649.

Levy JB, Baskin LS, Ewalt DH, et al. Nonoperative management of blunt pediatric major renal trauma. Urology 1993;42:418–424.

Lieu T, Fleisher GR, Mahboubi S, et al. Hematuria and clinical findings as indications for intravenous pyelography in pediatric blunt renal trauma. Pediatrics 1988;82:216–222.

Lim PHC, Chang HC. Initial management of acute urethral injuries. Br J Urol 1989;64:165–168.

Livne PM, Gonzales ET Jr. Genitourinary trauma in children. Urol Clin North Am 1985;12:53–65.

Mansi MK, Alkhudair WK. Conservative management with percutaneous intervention of major blunt renal injuries. Am J Emerg Med 1997;15:633–637.

McAleer IM, Kaplan GW. Pediatric genitourinary trauma. Urol Clin North Am 1995;22:177–188.

McAlister WH, Sisler CL. Scrotal sonography in infants and children. Curr Probl Diagn Radiol 1990;19:201–242.

McAndrew JD, Corriere JN Jr. Radiographic evaluation of renal trauma: evaluation of 1103 consecutive patients. Br J Urol 1994;73:352–354.

McDougal WS. Traumatic injuries of the lower genitourinary tract. In: Libertino JA, ed. Pediatric and Adult Reconstructive Urologic Surgery. Baltimore: Williams & Wilkins, 1987:490–499.

Mirvis SE. Diagnostic imaging of the urinary system following blunt trauma. Clin Imaging 1989;13:269–280.

Monstrey SJM, Vander Werken CHR, Debruyne FMJ, et al. Rational guidelines in renal trauma assessment. Urology 1988;31:469–473.

Moore EE, Shackford SR, Pachter HL, et al. Organ injury scaling: spleen, liver, and kidney. J Trauma 1989;29:1664–1666.

Morey AF, Bruce JE, McAninch JW. Efficacy of radiographic imaging in pediatric blunt renal trauma. J Urol 1996;156:2014–2018.

Neurwith H, Frasier B, Cochran ST. Genitourinary imaging and procedures by the emergency physician. Emerg Med Clin North Am 1989; 7:1–28.

Okur H, Kucukaydin M, Kazez A, et al. Genitourinary tract injuries in girls. Br J Urol 1996;78:446–449.

Peterson NE. Emergency management of urologic trauma. Emerg Med Clin North Am 1988;6:579–598.

Pierce JM Jr. Disruptions of the anterior urethra. Urol Clin North Am 1989;16:329–334.

Pokorny SF, Pokorny WJ, Kramer W. Acute genital injury in the prepubertal girl. Am J Obstet Gynecol 1992;166:1461–1466.

Quinlan DM, Gearhart JP. Blunt renal trauma in childhood. Features indicating severe injury. Br J Urol 1990;66:526–531.

Reda EF, Lebowitz RL. Traumatic ureteropelvic disruption in the child. Pediatr Radiol 1986;16:164–166.

Reinberg O, Yazbeck S. Major perineal trauma in childhood. J Pediatr Surg 1989;24:982–984.

Schneider RE. Genitourinary trauma. Emerg Med Clin North Am 1993; 11:137–145.

Shalaby-Rana E, Lowe LH, Nussbaum Blask A, et al. Imaging in pediatric urology. Pediatr Clin North Am 1997;44:1065–1089.

Smith SD, Gardner MJ, Rowe MI. Renal artery occlusion in pediatric blunt abdominal trauma-decreasing the delay from injury to treatment. J Trauma 1993;35:861–864.

Snyder HM. Urologic emergencies. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 2nd ed. Baltimore: Williams & Wilkins, 1983:1032–1040.

Stein JP, Kaji DM, Eastham J, et al. Blunt renal trauma in the pediatric population: indications for radiographic evaluation. Urology 1994; 44:406–410.

Stevenson J, Battistella FD. The ëone-shot' intravenous pyelogram: is it indicated in unstable trauma patients before celiotomy? J Trauma 1994;36:828–834.

Stine RJ, Avila JA, Lemons MF, et al. Diagnostic and therapeutic urologic procedures. Emerg Med Clin North Am 1988;6:547–578.

Taylor GA, Eichelberger MR, Potter BM. Hematuria. A marker of abdominal injury in children after blunt trauma. Ann Surg 1988;208:688–693.

Terry T. ABC of major trauma. Trauma of the upper urinary tract. Br Med J 1990;301:485–488.

Van Ahlen H, Bruhl P, Porst H. Pediatric blunt renal trauma-surgical or conservative treatment? Eur Urol 1988;14:407–411.

Wessells H, McAninch JW, Meyer A, et al. Criteria for nonoperative treatment of significant penetrating renal lacerations. J Urol 1997;157:24–27.

CHAPTER 110

Facial Trauma and Plastic Surgical Emergencies

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The moment a child sustains a facial injury, a process, which may range from a trip to the local emergency department (ED) to an altered quality or length of life, begins. An appreciation on the part of the emergency physician for the probable overall events that will occur with each particular patient will provide better patient care and more efficient triage. This appreciation comes from an understanding of what happens to the patient not only during the emergency encounter but also days and months later.

Difficulties in decision making in the ED revolve around what constitutes a plastic surgery emergency and which problems can be reasonably treated without specialist consultation. Few plastic surgery problems require an emergent trip to the operating room in the pediatric setting. Traumatic amputations of digits or extremities, severe burns that require emergent escharotomies, or electrical injuries to extremities requiring urgent amputation are some examples of pediatric plastic surgery emergencies. Most injuries in this chapter either require the plastic surgeon, as specialist, to render input regarding the best way to begin treatment, or require no particular specialist input. This chapter covers the evaluation and management of common injuries to the face, with an appreciation for long-term treatment goals.

Patients who have sustained enough force to sustain significant facial injury may have occult injuries elsewhere, and a complete trauma evaluation is often indicated. Patients with obvious facial trauma are sometimes rapidly assigned to plastic surgery services without adequate multisystem evaluation. Diagnostic failures occur in 12% of motor vehicle accident victims and 23% of motorcycle victims. The most common errors include 1) clinical judgment, 2) failure to follow routine systems evaluation, 3) false interpretation of tests, and 4) inadequate radiographs.

In particular, the head and neck should be treated delicately until an injury to the cervical spine is ruled out. In patients with obvious maxillofacial trauma, 10% have a spine fracture. This incidence can be even higher in series of patients with mandibular fractures. If any doubt exists, the patient should be placed in a semirigid collar or the head should be immobilized with sandbags in preparation for radiologic examination. An undiagnosed injury to the cervical spine in this case could lead to serious consequences, especially for patients who receive operative treatment of their facial injuries and undergo neck manipulation under general anesthesia.

Although no trauma book could be written without reference to the ABCs (airway, breathing, and circulation), the airway is obviously of particular relevance in the management of facial injuries. Patients can have airway obstruction resulting from a variety of factors, including blood, loose teeth, the tongue, and pharyngeal edema; therefore, the airway should be cleared and examined for patency as a priority. Loss of support of subglottic musculature can result secondary to severe mandibular fractures, and the tongue can fall posteriorly and occlude the airway in a comatose patient. If necessary, the tongue can be grasped with an instrument or even stitched with a heavy silk suture as a means to pull it outward and relieve the obturator effect.

Children who are hypoxemic are usually agitated and may attempt to sit up from a supine position. If clinically acceptable, they should be allowed to do so because this position may help clear the airway and reduce venous pressure and resultant edema. An oxygen saturation monitor should be used in all cases in which the stability of airway patency is in question, and oral or nasal airways can be used.

Because of the abundant vascular supply to the face, the clinician should never underestimate the amount of hemorrhage that has occurred in cases of lacerations to the face and scalp. In acute hemorrhage without fluid resuscitation, the hematocrit does not reflect the amount of blood lost. Children have an excellent ability to compensate in hemorrhagic shock by vasoconstriction and increased heart rate to a point of rapid decompensation and circulatory arrest. Obtaining serial hematocrits, providing rapid fluid resuscitation, and obtaining a careful history for amount of blood lost at the scene help avoid this potential problem.

Tracheostomy is rarely indicated in the emergency setting in the treatment of pediatric facial injuries. Cricothyrotomy rarely may be necessary but is not to be undertaken except as a last resort because of 1) the small size of the cricothyroid membrane, 2) technical difficulties encountered, and 3) potential damage of important laryngeal structures in the process of airway insertion, all of which may lead to a difficult or unsuccessful laryngeal reconstruction. In some instances, a “needle” cricothyrotomy can be used as a temporizing measure to stabilize a patient before operative intervention. Formal tracheostomy is performed optimally in an operating room; however, in extreme circumstances, a

“slash” tracheostomy is the preferred procedure and can be performed in the ED.

A final point relates to tetanus immune status. Proper documentation in the medical record that a tetanus immunization history was obtained is essential and is a standard of care. If indicated by such history, the administration of any and all antitetanus treatment should also be documented carefully. This procedure not only ensures proper antitetanus treatment but also avoids confusion later and obviates redundancy in treatment. Guidelines for reimmunization are given in Chapter 84 and Appendix D.

FACIAL INJURIES

Background

The cause of facial injuries is varied and includes motor vehicle accidents, accidents at home, and assaults. A distinction should be made and an understanding should be developed regarding facial trauma, analogous to the trauma of gunshot wounds; that is, trauma is either “low velocity” (e.g., fall from a chair, hit with a fist) or “high velocity” (e.g., motor vehicle accident, fall from a height). Distinction between the two types of trauma can aid in diagnosis of injuries and predict efficacy of treatment in many cases. The incidence of fractures in children is low, with those in the range of 0 to 5 years of age accounting for 1% of the facial fractures observed and those in the range of 5 to 12 years of age accounting for 3 to 5% of the fractures observed. The face of a child is small compared with the head, and the face undergoes a twelvefold size increase compared with only a fourfold increase in cranial size over the period of growth. In addition, the face is much stronger because the sinus cavities are not developed and the proportion of cancellous to cortical bone is greater, providing more elasticity. Falls against various objects account for a large number of injuries in children. As children grow, they engage in more adult activities (e.g., tricycles, bicycles, cars) and increase their potential for applying greater and greater forces to their faces. Therefore, as children grow, the severity of facial injuries tends to increase.

Clinical Manifestations

Bony Injuries

Fractures of the mandible are the most common facial fracture, with nasal fractures being more common in some cases. In contrast to adults, the most common site for mandibular fractures in children less than 10 years of age is the condyle or condylar neck (Fig. 110.1). When a fall onto a child's chin results in a parasymphyseal fracture, an associated fracture in the opposite subcondylar region often occurs and should be looked for and documented if present. Pain and difficulty with mouth opening are important signs. The inferior alveolar nerve exits at the mental foramen, an anterior opening in the mandible on a common vertical line with the second premolars. The mental nerve, exiting this foramen, supplies sensation to the lower lips and lower incisor teeth. The sensation in these regions should be evaluated and documented. During the acute period after injury, patients may not recall decreased sensation, and subsequent surgical manipulation may be incorrectly indicted as the cause for such sequelae. Anesthesia in these areas should raise suspicion of injury to the mandible.

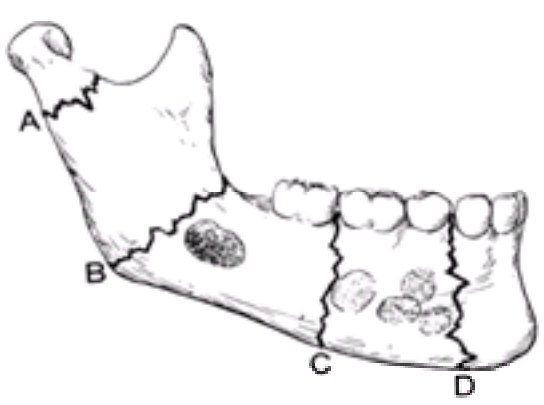

FIGURE 110.1. Examples of various types of possible mandibular fractures. A child's mandible with unerupted dentition is shown. *A,* subcondylar fracture; *B,* fracture at the angle; *C,* body fracture; and *D,* symphyseal fracture.

Powerful muscles of mastication are normally set at resting tensions and rely heavily on proper bony length for strength. When bony anatomy is disrupted, distracting forces are applied to the fractured mandibular segments, often causing acute occlusal disharmony. In a simple subcondylar fracture, the lateral pterygoid on the nonfractured side is unopposed and pulls the mandible medially, or toward the fractured side. In bilateral subcondylar fractures, muscle imbalance causes premature closure of the molar teeth, resulting in an anterior open bite. The growth center for the mandible is located in the area of the condyle, and damage to this area, similar to that from a fracture, can cause significant growth disturbances, especially if sustained before 3 years of age. Therefore, the clinical evaluation of any chin laceration should include examination of the mandible, particularly the condyles.

Fractures of the floor of the orbit, otherwise known as “orbital blowout fractures,” probably result from an acute increase in pressure within the orbital contents, thus pushing down on the thinner bone of the medial orbital floor (Fig. 110.2). Alternate theories suggest direct force applied to the inferior orbital rim, with subsequent buckling of the rim and floor, as a mechanism. The globe often reveals subconjunctival hemorrhage, and occasionally a traumatic meiosis is present in the affected eye. Rarely, retrobulbar hemorrhage with an afferent pupillary defect occurs, and emergency optic nerve decompression would be indicated.

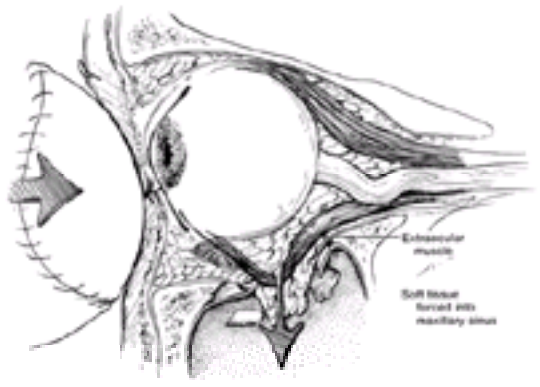

FIGURE 110.2. Mechanism of blowout fracture. In a sagittal view, a ball is shown striking the eye, deforming it, and causing increased pressure of the intraorbital contents. The periorbital fat is forced through the floor of the orbit. Retropositioning of the eye (enophthalmos), lowering of the eye, and extraocular muscle entrapment can result.

Because the volume of the globe and surrounding structures (fat, extraocular muscles) is fixed and because an acute increase in orbital space (an opening in the floor of the orbit) occurs, the globe may sink down in the orbit, producing enophthalmos, a sunken appearance to the eye. Blood and orbital fat may sink into the maxillary sinus, clouding the sinus on the radiograph (Fig. 110.3). Asymmetry in the horizontal level of the eyes (orbital dystopia) may also be present. Both enophthalmos and orbital dystopia may not be present initially, however, because of periorbital edema, which tends to compensate for the deformity. These findings usually become apparent after 5 to 7 days when swelling subsides. The infraorbital nerve, the terminal branch of the maxillary division of the trigeminal nerve, exits the maxilla just below the infraorbital rim and is often injured. Manifestations of injury to this nerve include decreased sensation to the cheek, upper lip, and upper gums on the affected side. If extraocular muscles are entrapped in the fracture gap in the floor of the orbit, voluntary extraocular movements may be limited. The inferior rectus is the muscle most commonly entrapped, which results in an inability to fully gaze upward on the affected side and often in diplopia. The presence of entrapment is one indication to operate on a blowout fracture on an emergent basis. Studies suggest that early repair of orbital fracture and release of the entrapped muscles (within 24 to 48 hours) avoids muscle ischemia and fibrosis and results in better functional recovery.

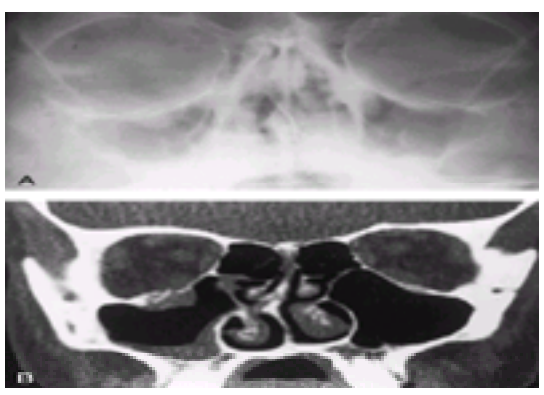

FIGURE 110.3. A. Blowout fracture. The sinus view shows teardrop configuration of the blowout fractures in the right orbit. Note associated fracture through the orbital floor and air–fluid level in the maxillary sinus. **B.** In the same patient as **A,** computed tomography section more clearly demonstrates the multiple fragment fracture through the orbital floor. Teardrop and air–fluid level are evident in the right maxillary sinus. (Courtesy of Soroosh Mahboubi, MD.)

Because of their prominent position on the face, the nasal bones are among the most commonly fractured bones of the facial skeleton. Nasal fractures may be difficult to detect clinically because of the significant swelling. The role of radiographs in nasal fractures is one of documentation only because they contribute little, if at all, to management. Most nasal injuries can be followed by a specialist on an outpatient basis, and evaluation after the swelling subsides dictates the need for further intervention. Two particular nasal injuries that deserve specific comment are the intractable nosebleed and septal hematomas. Because of the rich vascular network in the nose, supplied by branches of both the internal (anterior ethmoidal) and external (superior labial, palatine) carotid arteries, nasal hemorrhage can be difficult to stop, despite usual conservative measures (e.g., elevation, gauze compression). Septal hematomas (see Chapter 112 and Procedures, Section VII) arise because of hemorrhage from an artery beneath the mucoperichondrium, separating it from the septal cartilage. Because the septal cartilage is avascular and relies on the overlying mucoperichondrium for its blood supply, a hematoma results in cartilage necrosis and eventual septal perforation.

The zygoma is the second most commonly fractured bone in the face when all age groups are considered. The zygoma attaches to the craniofacial skeleton at three distinct areas: the 1) zygomaticofrontal, 2) zygomaticotemporal, and 3) zygomaticomaxillary regions (Fig. 110.4). A complete fracture of the zygoma results in fractures at each of these sites, and extends through the floor of the orbit. The masseter muscle is the most powerful muscle that contributes to jaw closure, averaging 200 pounds of bite force in the average male adult. The masseter muscle origin is the malar eminence of the zygoma, and this muscle accounts for the inferior displacement characteristic with these fractures. The resultant increase in apparent orbital volume leads to enophthalmos and orbital dystopia, as seen in blowout fractures. A “bird's eye” view often reveals a loss of anterior projection of the affected malar eminence. Decreased sensation along the distribution of the infraorbital nerve is also common as zygomaticomaxillary fractures usually include the infraorbital foramen. In isolated zygomatic arch fractures, a decrease in temporal width can be appreciated when viewing the face

from the front as a result of buckling of the zygomatic arch. If this buckling is severe, the mandibular condyle may be impinged, with resultant difficulty in mouth opening.

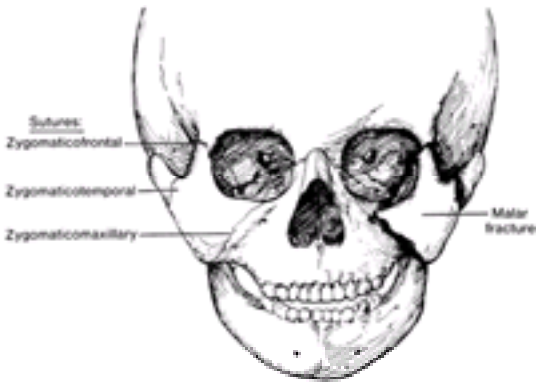

FIGURE 110.4. Zygomatic sutures and a malar fracture are shown. Fractures do not necessarily occur at these points. The zygomaticofrontal suture separation occurs most commonly.

Higher-energy injuries to the craniofacial skeleton can result in a series of fracture patterns, as originally described by Le Fort (Fig. 110.5). The Le Fort I fracture pattern extends through the zygomaticomaxillary region to the base of the pyriform aperture. The Le Fort II pattern, also called a pyramidal fracture, is similar but extends more superiorly to the infraorbital rims and across the nasofrontal sutures. The Le Fort III pattern, also called craniofacial dissociation, extends across the zygomatic arch, zygomaticofrontal region, floor of the orbit, and nasofrontal sutures, effectively separating the face from the skull base. In prepubescent children, facial fractures tend not to follow the classic Le Fort patterns. The fractures in this age group have a more oblique pattern because of the lack of full paranasal sinus development and the presence of undescended secondary dentition reinforcing the midface. In either age group, the fracture patterns are rarely symmetric because impact is sustained on one side more than the other. Because of massive swelling and atypical as well as asymmetric fracture patterns, computed tomography (CT) scans are often essential in the diagnosis of facial fractures in children. Even with good quality CT scans of the face, the definitive diagnosis is sometimes made only in the operating room under direct visualization.

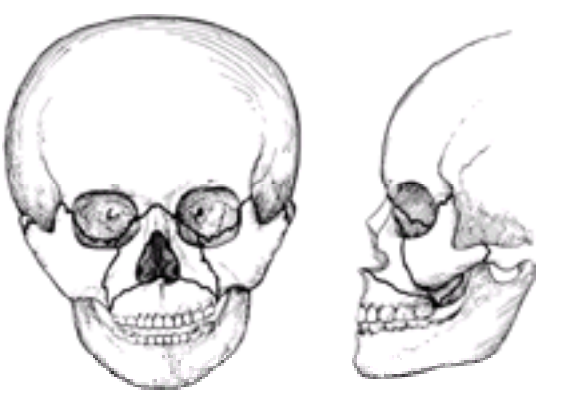

FIGURE 110.5. Le Fort classification of fractures. With type I, the maxilla is separated from its attachments. Type II (pyramidal) produces a mobile maxilla and nose. With type III (craniofacial dysjunction), all attachments of the midface to the skull have been separated. Traction on the anterior maxilla produces motion up to the inferior orbital rims and zygoma. These fractures are not mutually exclusive. For example, a Le Fort I can be present with a type II or type III. Le Fort II may exist on one side with type III on the other side.

Naso-orbito-ethmoid (NOE) fractures involve complete separation of the nasal bones and medial walls of the orbits from the stable frontal bone above and infraorbital rim laterally. These injuries are usually the result of high-velocity trauma to the central midface. The bones are often fragmented and telescoped posteriorly into the ethmoid region. These patients display a characteristic pugnacious nose, with loss of anterior projection on lateral view. Because the medial canthal tendons attach firmly to the medial walls of the orbits, lateral drift of the fracture segments results in traumatic telecanthus (Fig. 110.6). Normal mean intercanthal distance is 16 mm at birth, which increases to 25 mm in a female and 27 mm in a male at full facial growth. A significant increase in intercanthal distance or gross asymmetry in the medial canthal to facial midline distance should raise suspicion for this fracture.

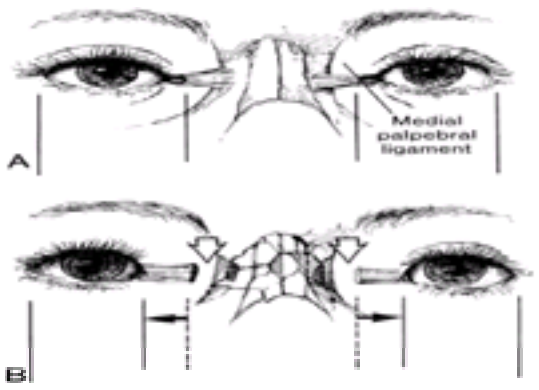

FIGURE 110.6. Mechanism of telecanthus. **A.** Normal attachment of medial palpebral ligament to medial aspect of orbital wall. **B.** Trauma producing comminution of nasal and orbital bones allows the ligaments to drift laterally, resulting in rounding of the medial canthus.

In the forehead, injury to the frontal sinus may reveal a palpable or visible depression if the anterior wall of the sinus has been compressed. Frontal sinus injuries are seen only in children older than 8 years of age because the sinus cells begin to aerate the frontal bone at this age. A forehead depression in a younger child is a depressed skull fracture until proven otherwise. In more severe frontal sinus fractures associated with forehead lacerations, a fracture of the posterior wall of the sinus and dural tear may allow cerebrospinal fluid (CSF) to leak from the wound. Wound fluid can be sent for glucose and chloride levels to help determine this diagnosis, or a “halo test” can be performed at the bedside. A drop of wound fluid is placed on filter paper, and the fluid is allowed to diffuse. CSF travels further from the center of the drop than does serum, and a “halo” will be present. Recently, the ability to test for b_2-transferrin provides an easy means of identifying CSF. b_2-Transferrin is a protein produced by neuraminidase activity in the brain and is uniquely found in CSF and perilymph. The presence of b_2-transferrin in wound fluid, detected by immunofixation electrophoresis, confirms a CSF leak.

Soft-Tissue Injuries

Facial lacerations are usually more obvious in their clinical presentation. However, an appreciation for deeper anatomic structures in the soft tissues aids in the diagnosis of injuries to these areas. Lateral periorbital lacerations should raise suspicion to injury to the frontal branch of the facial nerve, which travels superficially along a line from just above the tragus to a point 1.5 cm above the lateral eyebrow. Lacerations in the medial periorbital region near the medial canthus should raise suspicion for lacrimal duct injury. Because 85% of tears are drained via the lower canaliculus, failure to repair a laceration to it results in excessive tearing (epiphora). If deep lacerations are present in the cheek region, the clinician must determine whether injury to the buccal branch of the facial nerve and to the parotid duct has occurred (Fig. 110.7).

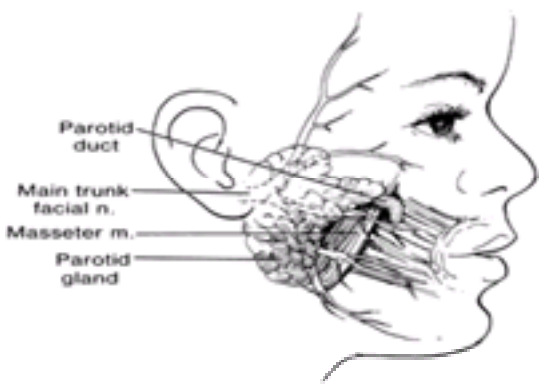

FIGURE 110.7. Deep lacerations to the cheek can injure the facial nerve, parotid gland, or parotid duct. The facial nerve becomes more superficial as it branches and proceeds distally. Distal nerve injuries can thus occur with more superficial wounds.

Once flaps of skin have been raised traumatically, their ultimate fate is directly proportional to their existing blood supply. A random skin flap can survive if the ratio of length to base width is 3:1; in the face, which is privileged with an excellent blood supply, this ratio can be even higher. If the injury included a significant crushing component, flaps of tissue can necrose on the basis of capillary thrombosis. In general, most facial lacerations initially look much worse than they are, and a conservative approach to removal of tissue is advocated. When in doubt, leaving tissue of questionable viability is always preferable. The tissue can always be removed or the resultant scar revised at a later time, if necessary.

Assessment and Management

The keys to successful management are predicated on making a correct diagnosis. Although seemingly mundane, this factor is most pertinent in the ED. The trauma patient makes a “first pass” through the health care system during the initial encounter. Errors in diagnosis and errors of omission at this point can set the stage for a multitude of complications. A methodical, standardized assessment is therefore mandated. Once a diagnosis is made, the decision must be made regarding the necessity for a specialist, based on an appreciation for the complexity of the problem and its long-term course.

History

The importance of a detailed history cannot be overemphasized. If the patient is unable to give a history, witnesses, emergency medical technicians (EMTs), and family should be questioned. If the validity of the facts is in doubt, “allegedly” or “reportedly” can precede the history and may save time in the long run. This situation is mentioned because, in certain criminal cases, attorneys have subpoenaed emergency physicians on the basis of their medical record history (a legal document) and have attempted to discredit the history if everything stated (e.g., “patient shot in face by girlfriend”) cannot be proven. Pictures of the injury should be taken when possible and included in the medical

record. If recent preinjury photographs are available (school pictures), they are of special benefit in the management of major facial injuries and should be sought.

As in all trauma histories, the mechanism and instrument of injury should be determined, as well as the timing and location of the injury and the likelihood of contamination. Specific attention should be given to the possibility of foreign bodies. If lacerations are present, an attempt should be made to determine whether these are caused by a crush mechanism (stellate lacerations from a blunt impact on a windshield) or a sharp object (facial lacerations from the steel trim of a car door).

Physical Examination

Soft-Tissue Injuries. Abrasions of the face should be evaluated for size, likely depth of the wound, presence of foreign bodies, and exposure of underlying structures. Especially in the pediatric population, local anesthesia will be necessary before adequate wound inspection can be performed. Classic “road tattoos” are created from falls out of motor vehicles or off bicycles and demonstrate particulate matter embedded in the dermal elements of the wound.

Probably the most common laceration seen on the face is a simple linear laceration. These injuries usually occur secondary to blunt trauma over bony prominences (e.g., supraorbital ridge, malar eminence, chin), causing the skin to burst. When superficial, this problem is relatively simple. When deep, punctate, or crushing in nature, tissue viability, presence of possible foreign bodies, and possible deeper structure injury must be considered. These wounds tend to be more complex, and flaps of various forms result (Fig. 110.8). For the purposes of efficient communication, lacerations should be described as simple or complex, linear or stellate, superficial or deep, and with an estimation of total length. This method aids in decision making regarding the need for specialist intervention. Wound margins should be inspected carefully for presence of bleeding and for capillary refill. A very pale color implies arterial ischemia. A dark blue color, which refills briskly when compression is released, implies venous engorgement. These flaps must be handled carefully during cleansing and evaluation because any further separation from their blood supply could be the determinant from tissue survival to necrosis.

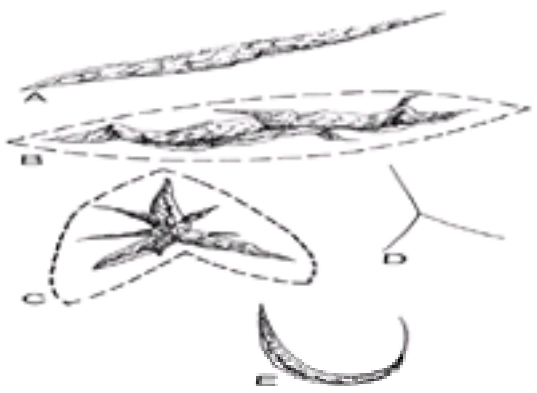

FIGURE 110.8. Variation in laceration injuries and suggestions for management: simple laceration **(A);** elliptical excision of damaged wound margins **(B);** excision **(C)** and closure **(D)** of stellate laceration; and flap-type laceration **(E).**

Local anesthesia is advised before wound exploration. Topical agents are available but do not substitute for local field blocks. Topical agents are most effective in the infant and toddler. With increasing dermal thickness, they become less efficacious. The most commonly used agent for local injection is 1% lidocaine with epinephrine 1:100,000 solution. Epinephrine should be used with caution if tissue viability is questionable or in end-arterial systems such as the earlobe or nasal tip. Lidocaine with epinephrine is supplied at a slightly acidic pH; premixing with bicarbonate has been described and is purported to decrease pain on injection. Practically speaking, the pressure that results from the rapidity of injection is the variable most easily controlled, and slow injections with a small, 30-gauge needle seem to work well. Maximum safe lidocaine doses are 4 mg/kg when no epinephrine is used (maximum dose 300 mg), and 7 mg/kg when epinephrine is used (maximum dose 500 mg). Percutaneous injection along a wound edge, as opposed to within the wound itself, theoretically reduces the potential for bacterial inoculation, although this is probably of significance only in heavily contaminated wounds. Intravenous or intramuscular sedation can be used if necessary.

Regional peripheral nerve blocks of the face can assist in the examination and management of facial lacerations in the pediatric patient. Nerve blocks have several advantages: 1) the total amount of anesthetic injected can be reduced, 2) they can augment local infiltration, 3) distortion of local anatomy can be avoided, and 4) injection sites can often be moved to less painful locations. The peripheral nerves most optimal for blockade by the pediatric emergency physician are the supraorbital and supratrochlear nerves, the infraorbital nerve, the preauricular and postauricular nerves, and the mental nerve. The supraorbital and supratrochlear nerves exit at the supraorbital rim in the medial third of the brow approximately 2 to 3 cm from the facial midline. Local infiltration in this region can effectively block these nerves and provide anesthesia to the ipsilateral hemiforehead.

The infraorbital nerve exits through the infraorbital foramen in the midface approximately 5 mm inferior to the infraorbital rim and in line with the first premolar tooth. Effective anesthetic block of this nerve can deaden sensation to the ipsilateral medial cheek and upper lip. The preauricular and postauricular nerves can be blocked by infiltrating local anesthetic approximately 1 cm inferior to the lobule and posterior to the ramus of the mandible. From this point, the infiltration is continued superiorly, anteriorly, and posteriorly to encircle the ear. With the addition of anesthetic injection in the choncal bowl, the entire ear can be anesthetized. The mental nerve can be blocked as it exits the mandible by injecting local anesthetic through the buccal sulcus to a point approximately 2 to 3 cm superior to the border of the mandible and

between the first and second premolar teeth. This technique achieves anesthesia of the ipsilateral lower lip.

Examination for foreign bodies can be aided by the history and helps determine how extensive a search should be. Radiographs can be a helpful adjunct to exploration, particularly because most glass is radiopaque. Careful probing of the wound digitally for foreign bodies can also be performed. If the wound is too small to accept a gloved finger or if digital probing is contraindicated, probing with a closed small hemostat often transmits the feeling of foreign bodies.

The potential for deep lacerations to involve important underlying structures has been mentioned, and specific maneuvers to rule this condition out are discussed. In general, if only subcutaneous fat is visible in the bed of the wound, it is considered superficial to facial nerves. If cut muscle is seen in the bed of the wound, nerve and glandular duct injury is possible. When facial nerve injury is suspected, patients can be tested by having them move specific muscles of facial expression. This testing should take place before infiltration with local anesthetic. The frontal branch can be tested by asking the patient to frown, looking for symmetry of frontalis action. The buccal branch can be tested by asking the patient to smile because most elevators of the corner of the mouth are innervated by this branch.

The marginal mandibular branch, or ramus mandibularis, may course as much as 1 to 2 cm below the border of the mandible and is responsible for depression of the corners of the mouth. Injury of this branch results in a characteristic upward pull of the corner of the mouth on the affected side, as a result of unopposed levator tone on that side. Examination for potential injury to Stenson's (parotid) duct is accomplished by grasping the commissure between thumb and index finger and gently everting the buccal mucosa to identify Stenson's duct, which lies on a vertical line with the maxillary second premolar. With the opposite hand, gentle massage of the parotid gland is accomplished by pressing in the preauricular region. The appearance of clear fluid from Stenson's duct suggests an uninjured duct. The absence of fluid after several minutes of inspection, or bloody fluid, suggests injury to the gland or duct. In this case, inspection of the depth of the wound may reveal salivary fluid, and cut ends of the duct may be identified. A sialogram can be a useful adjunct in the diagnosis of parotid duct injuries. In wounds with substantial bleeding, clamping all but the most obvious cut ends of blood vessels is to be avoided, for possible injury to a facial nerve branch.

The mouth should be evaluated for lacerations of the mucosa or tongue, injury to the palate, and loose or missing teeth. Chapter 113 details specific oral injuries.

Subperichondrial hematomas should be ruled out when injury to the nose or the external ear is present. These two appendages are based on a specific cartilage framework, and distortion of this framework can lead to functional and aesthetic deformities. Subperichondrial hematomas of the ear commonly occur in boxers and wrestlers and are characterized by a swelling of the conchal portion of the ear. Simple inspection of both sides of the nasal septum with a speculum reveals a septal hematoma, which presents as a boggy swelling, partially or completely obstructing the nasal airway on the affected side.

Bony Injuries. Many patients being examined for facial bone injury have potential head injury, and the Glasgow Coma Scale should be used. This tool aids in communication between specialists and, if operation is necessary, helps weigh the risks and benefits of such a procedure. Patients with a Glasgow Coma Scale score of less than 10 are considered poor risk for general anesthesia, and if possible, surgery for facial injuries should be delayed.

Examination for specific bony injuries begins with observation. Full-face anterior view and a bird's-eye view from the top of the head, with attention to possible asymmetry, aid the trained eye toward potential injury. The malar eminences and zygomatic arches are well visualized for differences in projection from this view. Asymmetry can take the form of swelling or loss of projection or flattening.

Any differences can be further assessed in the second or palpation phase of the examination (Fig. 110.9). Systematic palpation should be performed to avoid errors of omission. It is simple to begin from the top and work down. The forehead should be palpated for depressions or “stepoffs,” seen in frontal sinus fractures. The position of the malar eminences should be palpated simultaneously to aid in detection of asymmetry. Palpation of the zygomatic arches often can detect an outward buckling or depression. The thin skin of the lower eyelid affords easy palpation of the infraorbital rim for possible stepoffs. Intraoral palpation of the lateral maxillary buttress, by placing a gloved finger above the second maxillary premolar, can help diagnose a fracture in this area. Maxillary fractures are further checked for by grasping the anterior maxilla in the area of the incisors and attempting to move it. Alveolar fractures can be looked for in the same fashion. External palpation of the mandicular symphysis, body, angle, and ramus can help diagnose fractures in these areas, and intraoral palpation of the mandible along the lingual surface likewise is useful. The body of the mandible can be grasped on either side, with the right hand of the examiner on the left body and the left hand on the right body of the mandible. The index and long fingers can be placed in the mouth and the thumb in the submental area. An attempt is then made to produce unnatural motion in the mandible by torquing the bodies in opposite directions. In less cooperative or younger patients, gentle compression of the mandibular angles can elicit pain, suggesting the presence of a fracture. Placement of index fingers in the external auditory canals during active jaw opening can elicit differences in condylar position.

FIGURE 110.9. Sequential steps in examination for facial fractures. **A.** The supraorbital ridges are palpated while steadying the patient's head. **B.** The infraorbital ridges are lightly palpated using the index, middle, and ring fingers to determine symmetry or fractures. **C.** The zygomatic arch is palpated on each side to determine continuity and the possible presence of displaced fractures. **D.** The infraorbital rims, zygomatic bodies, and maxilla are palpated and examined from the top of the head to determine depressions and fracture displacement. **E.** The nasal bone and maxilla are examined for stability and possible fracture displacement. **F.** The nose is examined intranasally to determine placement of the nasal septum and possible displacement of nasal bones or disruption of nasal mucosa. **G.** The occlusion is observed to determine any disturbances of normal teeth relations. **H.** The mandible is palpated and then retracted to determine sites of discomfort and possible mandibular fractures.

Examination of the eyes should include observation for pupillary reactivity and size, noting possible asymmetry. Direct trauma to the globe should be excluded. Assessment for orbital dystopia and enophthalmos should be performed. Examination of extraocular motility is obtained if the patient is awake and cooperative. Alternately, passive extraocular motility can be checked by applying topical anesthesia to the conjunctiva and performing forced duction tests, which involve gentle grasping of the inferior fornix of the conjunctiva and passive ranging of the globe in all four directions. Gross visual acuity should be checked and charted. If any concerns are noted, an ophthalmologist should be consulted.

Inspection of the mouth for specific bony injuries is of vital importance because occlusal disharmonies are a tip-off to mandibular or maxillary displacement. Occlusion should be observed and an assessment made about its normalcy. The patient can simply be asked if the bite feels “normal,” if he or she is awake and cooperative. Opposing teeth that do not come together but that exhibit wear facets (smoothing of mamillations along the incisal surfaces of the teeth) suggest a traumatic malocclusion. Preauricular pain and pain or limitation with mouth opening are suggestive of mandibular fracture.

The patient should be checked for anesthesia of the cheek (infraorbital nerve distribution) and lower teeth and lower lip (inferior alveolar nerve distribution). After thorough observation, palpation, and sensory evaluation, the examiner usually can predict a specific area of injury, which is confirmed by radiologic evaluation.

Radiographic Studies

Because of the occult nature of pediatric facial fractures, the emergency physician should have a low threshold for radiographic evaluation. CT scans have virtually replaced radiographs in the assessment of serious acute bony facial injuries. Most medical centers are equipped with CT scanners, and these studies have greater ability to show exact locations of craniofacial fractures. Furthermore, patients with potential facial fractures are also patients with closed head injuries, and a CT scan often is already indicated. CT scans also can help rule out cervical spine fractures, and it is routine in many centers to scan a patient's head, face, neck, and abdomen in one sitting. Axial views provide excellent visualization of the zygomaticofrontal regions, the zygomatic arches (Fig. 110.10), the frontal sinus (Fig. 110.11) and patency of the nasofrontal ducts, the maxillary sinuses and their anterior walls, and the mandible. Coronal cuts with 1.5-mm cuts through the orbits (Fig. 110.12) are useful in viewing the status of the orbital floors and infraorbital rims, nasoethmoid region, malar eminences, and midfacial buttresses, as well as the status of the mandibular condyles and symphyses. Coronal views require considerable neck extension to orient the anterior facial plane as vertical as possible, and neck clearance should precede coronal scanning. Three-dimensional computerized reconstructions are dramatic to view, demonstrate fractures easily, and are useful for teaching, but they are not considered necessary for evaluation of routine fractures.

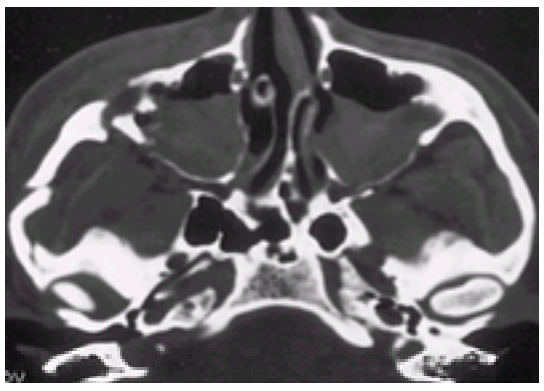

FIGURE 110.10. Axial computed tomography scan of the midface on a patient with a right zygoma fracture. Note the buckling of the zygomatic arch, the fractures of the zygomaticomaxillary and zygomaticotemporal areas, and the opacification of both maxillary sinuses.

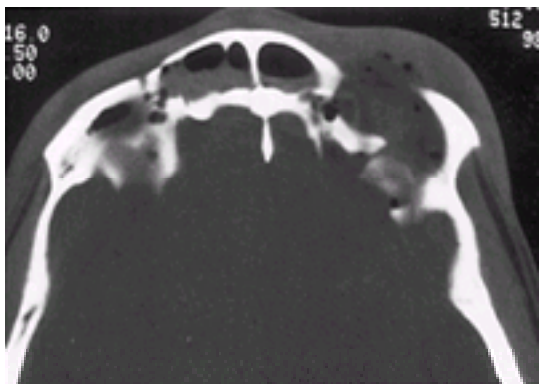

FIGURE 110.11. Axial computed tomography (CT) scan on the upper face in a patient with forehead lacerations. A bony defect in the anterior wall of the frontal sinus and fracture of the posterior wall on the patient's left side (*right side* of photography) suggest a fracture through both walls of the frontal sinus. Note the air–fluid levels in both sides of the sinus. Before CT, such detailed information regarding the status of the posterior wall was unavailable without operative examination.

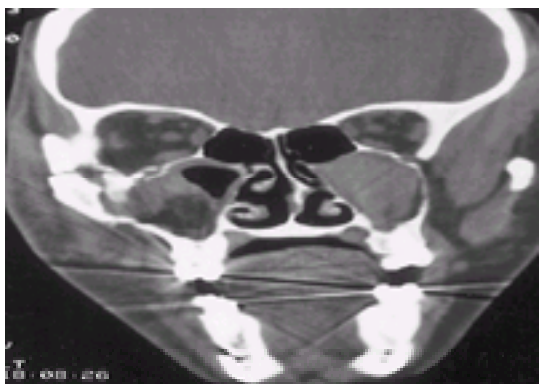

FIGURE 110.12. Coronal computed tomography scan of the same patient as shown in Figure 110.10. Note the excellent visualization of the fractured orbital floor associated with the zygoma fracture and increase in orbital volume. The lower portion of the scan demonstrates artifact created by dental filings (plates can cause similar scatter).

Panorex views show the mandible in its entirety and also clearly demonstrate tooth root anatomy and condylar position. However, Panorex views are limited in acute trauma evaluation because imaging suites are usually located some distance from the ED and require the patient to sit upright and remain still.

The Water's view (occipitomental) is useful for viewing the orbital rims, particularly the infraorbital rims and the maxillary sinuses. Lateral facial views provide reasonable imaging of the mandible on that side as well as a view of the sinuses and nasal bones. The submentovertex, or “jughandle,” view demonstrates the zygomatic arches well. Although, in general, the use of these standard radiographs has been superseded by CT scans.

GUIDELINES FOR CONSULTATION

In many cases, management of a specific injury is best handled in the ED. A telephone call to a consultant can be made to clarify the issue, but the judgment of the primary physician ultimately dictates the eventual outcome of patient care.

Abilities vary greatly, and rather than describe examples of injuries that might be expected to be handled by primary physicians in the ED, general principles are stressed. The decision must be made whether to treat the patient or to consult a specialist to assist in treatment. Vacillation between these two options should be avoided. This decision should be based on a balance of confidence in the diagnosis and the ability to treat it properly, and a realistic appraisal of present conditions in the ED regarding affordability of time. Specialists achieve good results not only because of specialty training but also because they can optimize their working conditions through the use of an operating room.

For the pediatric population, the parent(s) or guardian often requests a specialist consultation for problems that normally would not require specialist intervention. In these cases, instilling confidence is important. The physician should explain to the patient or family that the specialist has no additional techniques to render and that delays in treatment may increase the possibility of complications (e.g., infection, stress of being in the ED). Once patients and families are confident in the primary physician's ability, they usually opt for the fastest way to complete the treatment.

Injuries that should be left to the specialist for repair include 1) lacerations with evidence of injury to deep structures (a major motor nerve or a glandular duct), 2) cases in which a substantial amount of devitalized tissue exists or actual tissue loss has occurred, 3) wounds in which the amount of bleeding cannot be easily controlled, 4) full-thickness defects of the ear and nose, and 5) cases in which it is unclear exactly what tissue to approximate to restore preinjury anatomy and aesthetics (e.g., lips, eyelids, nostrils, ears).

SPECIFIC MANAGEMENT PROBLEMS

Soft-Tissue Injuries

Minor abrasions can usually be handled in the ED. The key principles are adequate anesthesia to perform acceptable cleansing of the wound, removal of devitalized tissue and debris, and proper dressing. Regional peripheral nerve blocks can be essential adjuncts to appropriately anesthetizing large facial abrasions. Normal saline and a mild soap or Betadine are commonly used for irrigation, cleansing, and scrubbing, if necessary, although special isotonic cleansers are available. Injuries on road or gravel can implant particulate matter into dermal layers and create “road tattoo.” These areas require aggressive scrubbing and possible picking out of individual particles with the tip of a No. 11 blade. The best chance of removing road tattoos is at the time of the initial treatment, and attention to detail is key. Minor abrasions represent a loss of epidermal and partial dermal layers. The remaining dermal structures in the bed of the wound should be kept moist and free of infection by using a thin layer of antibiotic ointment, such as Silvadene or Bacitracin. Keeping gauze dressings on the face of a child is difficult unless a head wrap is used, and for small area abrasions, ointment alone is practical. These ointments should be washed off gently with soap and water several times daily and reapplied until reepithelialization occurs. The risk of infection is probably related more to inadequate cleansing and debridement than to the use or choice of antibiotic dressing, underscoring the importance of anesthesia.

Deeper abrasions involve near total or complete loss of all dermal elements. Often, they are seen as part of a large abrasion with more superficial surrounding components. If the resultant full-thickness defect is 1 to 2 cm^2 or less, it is reasonable to allow the defect to contract in for possible scar revision at a later time. Acute primary closure is a less attractive option because increased skin tension on the wound leads to a widened scar, and ischemia could result. An acute skin graft for a defect this small is not indicated because a small graft gives a patchy appearance and it is unnecessary to cover so small a defect.

With larger surface area, full-thickness losses of skin, coverage with autogenous tissue to gain “control of the wound” is a short-term goal. This approach protects deeper structures below, reduces transwound fluid losses, and reduces the likelihood of soft-tissue contamination and infection. Control of the wound does not always guarantee a good aesthetic result, and an understanding of reconstructive planning helps illustrate this problem. For example, a 4 × 4-cm full-thickness abrasion on the cheek of a 10-year-old child might be treated by cleansing, several days of moist dressings, and a split-thickness skin graft to cover the wound. However, this treatment represents only the first phase of the reconstruction. After 4 to 6 months, the skin graft will have contracted slightly, thereby pulling the normal facial skin closer together. Serial excisions of the skin graft could be performed, each time stretching the surrounding normal skin and bringing it closer together. Finally, the remaining graft could be excised completely and the defect covered with facial skin. The complete process may take more than 12 months to complete and may require several operations, but the aesthetic result of replacing “like tissue with like” is superior to the patch of a skin graft. Why not close the original defect primarily? The tension created by such a would closure would be excessive, and tissue necrosis and scar widening would surely result. “Control of the wound” would not be accomplished.

In general, lacerations should be repaired within 8 to 12 hours after the injury was sustained. However, on the face, clean, sharp lacerations can be reapproximated up to 24 hours after the injury, and even late-presenting lacerations (48 to 72 hours after the injury) can be closed after thorough sharp debridement, usually by a specialist. The risks of infection in closing such a wound must be weighed against the benefits of reducing the facial scarring that will result if the wound is allowed to heal secondarily. Factors such as mechanism of injury, immunocompetence, and hygiene must be considered. Similar to abrasions, anesthesia, copious irrigation and cleansing, and exact, tension-free approximation are vital to a successful closure. The purpose of sutures is to reapproximate tissues so wound repair can be facilitated, not to hold the tissues together forever. Excessive tension on sutures simply causes tissue ischemia by exceeding capillary filling pressure. This condition leads to tissue necrosis and a dehisced wound. The capacity of the human body to heal greatly precedes the advent of suturing, and scar revision can always be performed at a later time once the wound has contracted. If in doubt regarding closure of a late-presenting or heavily contaminated wound, the clinician should call a specialist or consider leaving the wound open to heal by secondary intention or delayed primary closure. In this case, cleansing is still important, and moist, saline-soaked sponges can be placed in the wound (to be changed several times a day).

If injury to deeper structures is ruled out, deep lacerations can be closed in layers, using absorbable suture material in the deeper layers. Each layer of closure attempts to reorient the preinjury anatomy and sets up the orientation of the next layer. A poorly approximated subcutaneous layer results in an uneven wound, no matter how well the skin stitches are placed. In fact, a well-closed wound should demonstrate the skin edges to be 1 to 2 mm apart or closer at the layer preceding final skin closure. Suture is a foreign body; more is not necessarily better. Inexperienced stitchers tend to use too many sutures, which leads to tissue ischemia, excessive foreign body in the wound, and unnecessary expense. Deep layers eliminate dead space in the depth of the wound, which, if not eliminated, could lead to formation of a fluid collection and eventual depression in this area. Deep-layer sutures commonly used for facial lacerations are 4-0 or 5-0 chromic, Dexon or Vicryl. Skin suture material is commonly nonabsorbable monofilament suture, such as nylon or prolene, except in the infant or toddler who benefits from the use of absorbable sutures such as 6-0 fast-absorbing gut to avoid the trauma of suture removal.

A vertical simple suture that catches the lower dermis should approximate the wound edges (Fig. 110.13). Forceps should be used delicately to avoid crushing skin edges; therefore, fine-toothed forceps are preferred. An alternative is to use no forceps and to provide traction on the skin by pressing down on the wound with the index finger of the nondominant hand. Skin margins should be everted by placing the needle into the skin at an angle of 90 degrees or greater and by coming out at the same angle. If a portion of the wound is excised because of devitalized tissue, severe contamination, or to join two adjacent wounds into a single wound, care should be taken to orient the incisions parallel to the relaxed skin tension lines (Fig. 110.14). If a laceration extends into the hair-bearing scalp, care must be taken to align the hairline. Shaving of hair before suturing should be avoided in these cases and is not necessary in any case. Lacerations extending across the mucocutaneous junction of the lip should be closed with careful approximation of muscle layers, if evident in the wound. Skin edges should be closed with monofilament suture; mucosa should be closed with absorbable material, which is left in place. Great care should be taken to identify the “white roll,” the segment

between the skin and the pink vermilion (mucosa) because a discrepancy in this area is obvious.

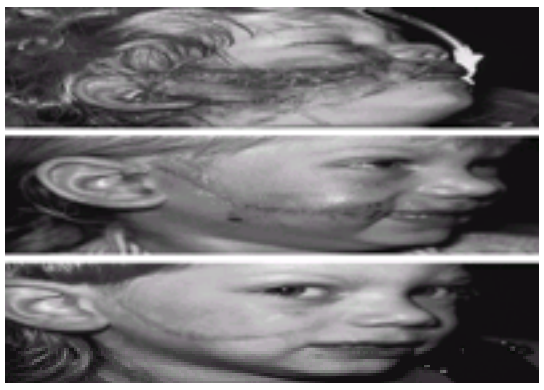

FIGURE 110.13. Photographs of a 3-year-old boy after an attack by a dog. Child was evaluated in the pediatric emergency room, intravenous antibiotics were given, his facial wounds were irrigated, and a plastic surgery consultation was made. The **top** photograph shows the child in the operating room before sharp debridement, facial nerve exploration, and an exacting layered closure of his complex wound. The **middle** panel pictures the child 1 week after his repair and demonstrates the precise reapproximation of the facial soft tissues. The **bottom** photograph was taken 8 months after the attack and demonstrates a nicely healing facial scar that will continue to fade and soften. (Courtesy of David W. Low, MD.) *Please see the color-tip insert (Color Plate 110.13).*

FIGURE 110.14. This illustration shows the natural pattern of skin creases that occurs with aging. If a circular type of wound needs to be converted to an elliptical shape for closure, an attempt should be made to have the wound parallel these lines.

Repair of complex injuries to laminated structures (e.g., ear, eyelid, nose, lip) requires careful attention to reapproximation of each layer of the structure. For example, a full-thickness laceration to the nose at the nostril rim requires closure of three separate layers. The nasal lining is usually closed first with an absorbable suture material. Next, the cartilage must be exactingly repaired. Finally, the overlying skin of the nose can be reapproximated. Similarly, complex injuries of the ear, the eyelid, or the lip require layered closure to achieve the best final results. A specialist's input is often helpful in managing complex injuries to these structures (Fig. 110.14).

Skin sutures can be continuous or interrupted and should be removed in 3 to 5 days. Leaving skin sutures in too long produces stitch marks in the skin and creates suture sinus tracts. If a wound complication such as a hematoma or wound abscess requires drainage, interrupted sutures allow for partial opening of the wound without disruption of the entire suture line. In cases in which these complications cause particular concern, interrupted sutures are advised. In cases in which the difficulties of suture removal outweigh potential aesthetic drawbacks (e.g., scalp sutures in a 4-year-old), absorbable sutures can be used on the skin and simply left in place to dissolve.

Bony Injuries

Facial fractures are more of a diagnostic problem than a treatment problem for the emergency physician. The diagnosis can usually be made by careful history and physical examination, as outlined previously, liberally augmented by CT evaluation. Once the diagnosis is made, a specialist consultation is appropriate for further patient management. Specialists' recommendations can be better understood by a brief overview of standard facial fracture treatment.

In general, fractures of the upper face are managed such that anatomic alignment is restored. For displaced facial fractures, reaching this goal often requires open reduction of the fractures in the operating room. The coronal, lower eyelid, and upper buccal sulcus incisions are the standard incisions used to gain access to the upper facial bones. Because bony healing is especially rapid in children, anatomic reduction becomes increasingly difficult when healing in the displaced position has occurred, and early treatment (within 2 to 4 days) is preferred. After anatomic reduction is achieved, the fractures are rigidly fixed if unstable. In children, facial fractures are often of a “greenstick” type, and reduction alone is adequate. If reduction alone is inadequate, rigid fixation can be accomplished by using small metallic plates and screws or, more recently, plates and screws fabricated from a biodegradable copolymer.

Because of concern regarding mandibular growth retardation and injury to permanent tooth buds, mandibular fractures in children are treated by more conservative measures compared with adults. A substantial proportion of fractures may be

managed by closed reduction and intermaxillary fixation (50%) and a soft diet (25%), and the remaining fractures are treated by either open reduction, internal fixation, or the use of splints.

Suggested Readings

Bartlett S, Delozier JB 3rd. Controversies in the management of pediatric facial fractures. Clin Plast Surg 1992;19:245–258.

Fujino T, et al. Entrapment mechanism and ocular injury in orbital blowout fracture. Plast Reconstr Surg 1980;65:571–574.

Habal M, et al. Facial Fractures. Philadelphia: BC Decker, 1989.

Kruger G, et al. Oral and Maxillofacial Traumatology. Vol. 1. Chicago: Quintessence, 1982.

Lewis V, et al. Facial injuries associated with cervical fractures: recognition, patterns, and management. J Trauma 1985;25:90–93.

Moore M, et al. Oblique craniofacial fractures in children. J Craniofacial Surg 1990;1(1):4–7.

Mulliken J, et al. Management of facial fractures in children. Clin Plast Surg 1977;4:491–502.

Randall P, et al. Fractures of the facial bones in children. J Trauma 1972;12:56–66.

Randle H, et al. Know your anatomy. Local anesthesia for cutaneous lesions of the head and neck: practical application of peripheral nerve blocks. J Dermatologic Surg Oncol 1992;18(3):231–235.

Touloukian R. Pediatric Trauma. St Louis: Mosby, 1990.

CHAPTER 111

Eye Trauma

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When faced with a child who has sustained eye trauma, the pediatric emergency physician must keep in mind four important basic principles:

1. The management of life-threatening systemic illness or central nervous system trauma must always take precedence over the eye injury.
2. The structural integrity of the eyeball must be ensured.
3. Check the vision in both the injured and uninjured eye.
4. When in doubt, seek ophthalmology consultation.

The most serious eye injury may be acutely “neglected” if urgent lifesaving procedures are under way. It is reassuring to know that even if a child lost an eye because of the inability of medical personnel to tend to an eye injury while managing other urgent medical problems, that child will function entirely normally, both at school and in athletic activities, with one remaining eye. If possible, a shield (see Fig. 111.9) can be placed over an obviously injured eye while resuscitative efforts are ongoing to prevent further accidental injury or contamination by the medical staff. This precaution prevents unwanted pressure on the eyelid or eyeball in case of an unrecognized ruptured globe. Such pressure could otherwise lead to extrusion of intraocular contents through an eyeball laceration. This situation would significantly worsen the prognosis for that eye.

FIGURE 111.9. Patient shielded for right ruptured globe, which was caused by a thrown pen.

This chapter is designed to assist the pediatric emergency physician in the diagnosis and management of basic and uncomplicated ocular injuries. However, it is important to recognize when ophthalmology consultation is necessary. Even with the increasing number of emergency departments (EDs) that have slitlamp biomicroscopy available to the nonophthalmologist, the ophthalmologist is more expert at using the slitlamp and has experience with a wide array of diagnostic tools that allow viewing and recognition of intraocular injuries that cannot be seen with a direct ophthalmoscope. In fact, most children who sustain high-risk blunt trauma to the eye, even in the absence of injury that can be seen on external examination, warrant a dilated retinal examination by an ophthalmologist using the indirect ophthalmoscope. This examination allows viewing of the edges of the retina just behind the iris where much of the damage (e.g., retinal tears) is sustained after blunt trauma. Even when ophthalmology consultation is not readily available, it is important to identify an ophthalmologist in the community who is comfortable examining children for outpatient follow-up.

History

Certain questions help identify possible intraocular injury: 1) What was the child's prior visual status? If the child was previously known to have poor vision in the eye, then less concern will be elicited if the same poor vision is noted after the trauma. 2) Has the child ever had a prior eye examination, and if so, what were the results? 3) Has the child ever had a patch over one eye for an extended period? This history would indicate that the child previously had poor vision secondary to amblyopia in the unpatched eye. 4) Has the child ever had eye muscle surgery? Children who have had strabismus are at a greater risk of developing amblyopia and therefore poor vision in one eye is more common. 5) Does the patient wear glasses? If so, they should be worn when visual acuity is tested. 6) Does the patient wear contact lenses? If so, location and removal of the lenses may be necessary.

Additional questions include the following: What was the nature of the injury? How hard was the eye struck? Certain types of trauma have a particularly high risk for causing intraocular damage: significant blunt impact directly to the eyeball (e.g., fist, ball), projectiles, and sharp implements (e.g., pencil, stick). Different antibiotic coverage may be necessary for suspected contamination (e.g., *Bacillus spp.*) by soil or other “outdoor” implements. Hammering is a particularly high-risk behavior for causing intraocular foreign bodies. If an intraocular foreign body is suspected, the clinician must establish whether it is metallic. Magnets are sometimes used by the ophthalmologist during surgical removal of an intraocular foreign body.

Examination

An attempt should always be made to assess the visual acuity in the injured eye before proceeding with the rest of the eye examination. Some patients may be unable to perform this task because of eye pain, noncompliance, an inability to open swollen lids, or obtundation from accompanying head trauma. At the very least, even if the eyelids remain closed, the physician can test for light perception. By shining a bright penlight or direct ophthalmoscope in the direction of the eyeball through the closed eyelid, the physician can ask the patient to indicate whether he or she perceives the additional light on that side. Even without a patient response, a reflex contraction of the lids may be seen, indicating light perception.

If the patient is able to exhibit a greater degree of compliance, the examiner may ask the patient to count fingers that are held before the affected eye at varying distances. The maximum distance at which these fingers can be counted should be noted on the chart (e.g., “counting fingers at 4 feet”). If the patient cannot stand but can identify letters or numbers, a commercially available near card or any other reading material can be used to assess the quality of near vision. Virtually no injury causes very abnormal distance vision but normal near vision. Normal near vision usually indicates that the patient has not sustained a significant ocular injury that is impairing vision at that time.

Should the patient be able to comply, the examiner can try to obtain a standard visual acuity using a distance chart. Letter charts should be used only if the child is known to be able to identify accurately all letters either by parental report or by walking up to the distance chart and identifying them at close proximity. If the child has any trouble with letters, a “tumbling E” chart or a picture chart can be used (see Chapter 120).

The patient's visual acuity in each eye should be tested. Certainly, the presence of bilaterally poor vision in a patient with unilateral eye trauma suggests that the cause of the poor vision is unrelated to the trauma. The eye that is not being tested should be covered well to prevent any conscious or unconscious attempt on the part of the patient to peek around the obstruction. Children will naturally try to do this if their better eye is being covered. To ensure that the child is actually viewing the chart with the eye that is being tested, the examiner should stand by the chart, facing the patient, indicating which letters are to be read while observing the patient's compliance.

If a patient demonstrates poor vision in the traumatized eye, the clinician can readily establish whether this deficit is related to the trauma. When a person looks through a pinhole and experiences improvement in performance on visual acuity testing, he or she can have only a refractive error as the cause of the initially tested poor vision. In other words, if a patient comes in with a traumatized eye that is able to read only 20/400 (needs to stand at 20 feet to see what a normal person can see at 400 feet) but then improves to 20/25 using a pinhole device, the patient has not sustained visual impairment from the ocular injury. Rather, the patient simply needs glasses. The maximum vision obtainable through a pinhole may be only 20/25 to 20/30. A pinhole can be created by poking holes through opaque paper or cardboard with an 18-gauge needle. A cluster of five or six holes may be easier for the patient to use.

After visual acuity has been established, an attempt may then be made to examine the eyeball. Using a stepwise anatomic approach, the examiner should first inspect the periorbital tissues and eyelids for the presence of ecchymosis, lacerations, and ptosis. Eye muscle movements (see Chapter 25) and the anterior surface of the eye should be evaluated next. All attempts should be made to examine these structures without touching or upsetting the child, particularly if by history or examination a ruptured globe is suspected. An upset child creates a Valsalva maneuver while crying, which may lead to extrusion of intraocular contents through a ruptured globe. If a ruptured globe or hyphema has been ruled out, the examiner may proceed with full eye examination, including pharmacologic dilation of the pupil. Regimens are suggested in Table 111.1.

Phenylephrine 2.5%	For brown irides add
Tropicamide 1%	cyclopentolate 1%

[a]May repeat regimen in 30 minutes if needed

Table 111.1. Emergency Department Ocular Dilating Regimen[a]

If pupillary dilation is not to be performed in the ED, a red reflex should be documented. By standing 2 to 3 feet away from the patient, the examiner should shine the largest available circle of illumination from the direct ophthalmoscope onto the patient's face such that both eyes are illuminated simultaneously (Fig. 25.6). The focusing dial is then spun until the patient's face and eyes come into focus. The patient should be instructed to look at the observer. The light should be turned out in the room to encourage maximum physiologic pupillary dilation. The red reflex is usually orange–red or yellow–orange; only rarely is it actually red (Fig. 25.6). The absence of a red reflex may indicate an obstruction within the eyeball to the passage of light, such as a corneal scar, cataract, or hemorrhage within the eyeball. A darkened reflex may also be caused by small pupils or a misaligned eye (Fig. 25.6). Rechecking the reflex after pharmacologic dilation may be helpful in these situations. A white reflex may also indicate the presence of a corneal scar, cataract, or other white intraocular pathology, such as a coloboma or retinoblastoma.

When trauma damages the retina, it often does so at the edges of the retina. This area is out of the field of view of the direct ophthalmoscope and requires indirect ophthalmoscopy to be viewed adequately. Therefore, any patient with severe blunt trauma to the eyeball should be considered for ophthalmology consultation so that the inside of the eye may be examined completely.

The emergency physician often finds the direct ophthalmoscope to be useful for the identification of possible papilledema or retinal hemorrhages in the setting of acute central nervous system injury. Pupillary size should be maximized by the use of dim ambiant illumination and a small circle direct ophthalmoscope beam. Pharmacologic agents (Table 111.1) can also be used. The patient should be told to fixate on a distant target. The examiner must avoid inadvertently lowering his or her head in a way that obstructs the eye that is not being examined, therefore obstructing the fixation of that eye. The child could then begin looking around, making adequate examination of the optic nerve difficult. To avoid this reaction, the examiner should sit or stand such that his or her head is level with and parallel to the patient's head rather than leaning over (Fig. 111.1B). If the examiner is standing above the patient, the patient's head can be tilted away from the examiner such that it becomes parallel to the examiner's head when he or she leans over to look into the eyes (Fig. 111.1C).

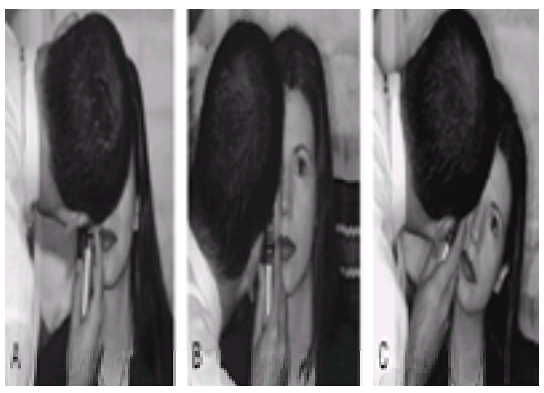

FIGURE 111.1. Use of the direct ophthalmoscope. **A.** Examiner is in incorrect position. His head is blocking patient's ability to fixate with the left eye during examination of right eye. By placing his head at level of patient's **(B)** or by tilting patient's head away **(C),** the left eye can continue to fixate.

The ophthalmoscope may be used with or without the examiner's glasses (or contact lenses) in place. The examiner simply holds the direct ophthalmoscope approximately 1 foot from the patient while standing at his or her side holding the instrument in the same hand that corresponds to the eye to be examined (i.e., right hand for right eye). The focusing dial should be set at zero, and a red reflex should be obtained. Then the examiner approaches the patient slowly until a blood vessel is seen within the red reflex. As the examiner moves closer, he or she should follow the blood vessels while spinning the focusing wheel with the forefinger to keep the blood vessels in focus. The apex of any branching blood vessel points in the direction of the disc (Fig. 111.2). The optic nerve head (disc) is best found in this manner. When the disc is located and in focus, the examiner's forehead should be no more than 1 to 3 inches away from the patient's forehead.

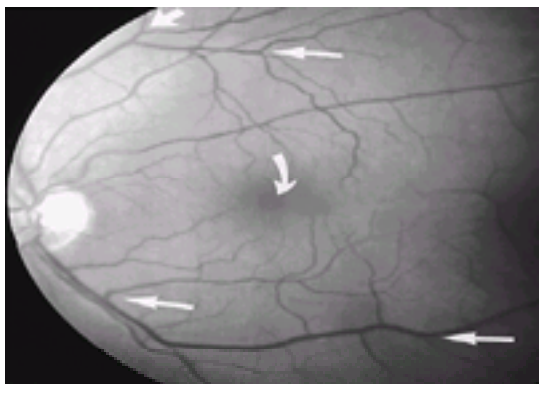

FIGURE 111.2. Normal retina as viewed by indirect ophthalmoscope. Central dark area *(curved arrow)* represents fovea. Note that the apex of the branch point of the blood vessels *(straight arrows)* always points back toward the direction of the optic nerve head.

The optic nerve should be yellow–orange to pink (Fig. 111.2). Centrally, a white depression called the optic cup is apparent. Normally, this depression may be barely visible or may occupy up to 50 to 60% of the optic nerve surface area. Both optic nerves usually have symmetric cup sizes. The retinal blood vessels usually emerge from the center of the cup. In papilledema (Fig. 111.3), the edges of the optic nerve become indistinct and difficult to differentiate from the surrounding retina. The blood vessels may become engorged and tortuous. There may be associated hemorrhages on the optic nerve surface or immediately surrounding the disc. Exudate may also be seen within the retina. The optic nerve cup may not be identifiable because of swelling of the neurons. It may be difficult to focus on the optic nerve surface and the retina at the same time because of the elevation of the optic nerve. However, papilledema may at times be subtle. If any question exists in the mind of the examiner that papilledema may be present, ophthalmology consultation is appropriate. Papilledema is almost always bilateral. Presence of unilateral papilledema suggests an ipsilateral orbital trauma, such as an orbital hemorrhage or direct injury to the optic nerve. The optic nerve can be injured from blunt trauma without a penetrating orbital injury.

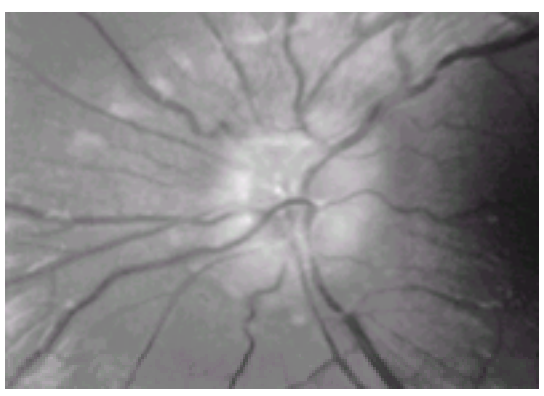

FIGURE 111.3. Papilledema. Note blurred disc margins and loss of view of blood vessels on disc. *Please see the color-tip insert (Color Plate 111.3).*

If the eye has been traumatized such that the patient is unable to voluntarily open the eyelids, attempts should be made to assist the patient in doing so. A warm compress may be gently applied to the eyelashes to loosen any crust or discharge that may be holding the eyelashes together. When opening the eyelids, it is essential to avoid pressure on the eyeball, which might lead to extrusion of intraocular contents via an underlying ruptured globe. By placing his or her thumbs on the supraorbital and infraorbital ridges while exerting pressure against the underlying bone, the examiner's thumbs can then be pulled away from each other such that the eyelids are separated (Fig. 111.4). Various commercially available speculums can also be used to open swollen eyelids (Fig. 111.5). However, in the trauma setting, which is severe enough to cause lid swelling that precludes a readily available view of the eyeball using the techniques previously described, it is probably safer to refer the patient for an ophthalmology consultation. Risking the use of a speculum may upset the patient and cause a struggle that could also contribute to disruption of the intraocular contents in the presence of a ruptured globe.

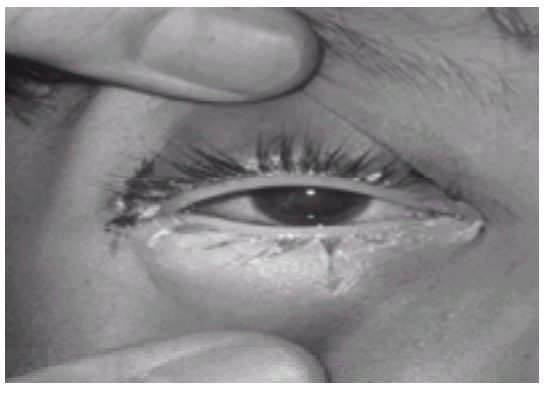

FIGURE 111.4. Opening swollen eyelids from superior and inferior orbital rims.

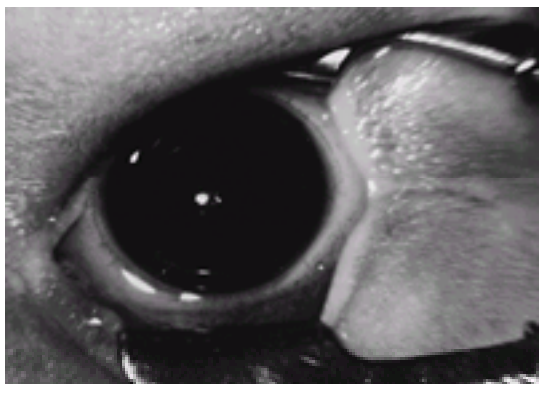

FIGURE 111.5. Commercially available eye speculum in place.

The Desmarres retractor is perhaps one of the least uncomfortable yet effective means for opening swollen lids. After instillation of a topical ophthalmic anesthetic (proparacaine, tetracaine), the cupped end of the retractor (Fig. 111.6A) is slipped under the upper lid and gently retracted with the handle parallel to the forehead (Fig. 111.6B). A second retractor can be simultaneously applied to the lower lid to further improve exposure. Paper clips can also be used (Fig. 120.3).

FIGURE 111.6. A. Desmarres retractor. **B.** Desmarres retractor in use to elevate upper lid.

RUPTURED GLOBE

Clinical Manifestations

Laceration or puncture of the cornea and/or sclera creates a ruptured globe. This condition can occur following trauma by projectile, sharp implement, or blunt trauma. Although severe intraocular disruption may occur, the eyeball has a remarkable ability to maintain its integrity. Immediately upon laceration, the iris or choroid (which is the extension of the iris posteriorly underneath the sclera) plugs the wound. This plug may appear as a blue, brown, or black material on the surface of the sclera (Fig. 111.7) or at the corneal–scleral junction. The iris will come forward and plug a corneal wound (Fig. 111.8). Because of this iris or choroid movement, the pupil may take on a teardrop shape, with the narrowest segment pointing toward the rupture (Fig. 111.7). Hemorrhage within the anterior chamber (hyphema) often accompanies a corneal or anterior scleral laceration (Fig. 111.8). With small lacerations that are plugged by iris or choroid, the eyeball does not deflate but rather takes on a remarkably normal external appearance. Subconjunctival hemorrhage for 360 degrees may obscure an underlying scleral rupture but leave the eye fairly intact. Patients who present following trauma with this finding or with severe 360 degree conjunctival swelling without hemorrhage should be treated as if they had a ruptured globe and referred immediately to an ophthalmologist.

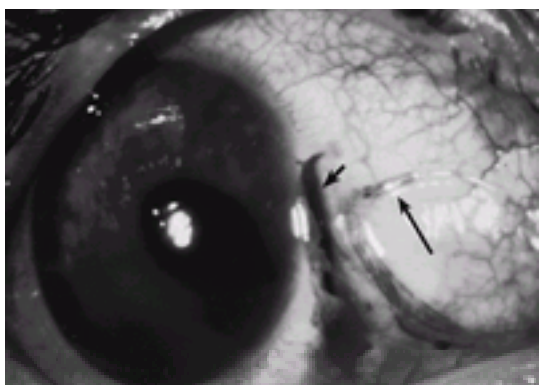

FIGURE 111.7. Ruptured globe. The scleral laceration *(short arrow)* appears as a linear brown line on the white of the eye. The pupil has a teardrop shape, the apex of which points in the direction of the rupture. The *long arrow* points to the upper border of a large conjunctival laceration. Note that the underlying sclera is intact. There is a diffuse hyphema in the anterior chamber, which partially obscures the pupil. *Please see the color-tip insert (Color Plate 111.7).*

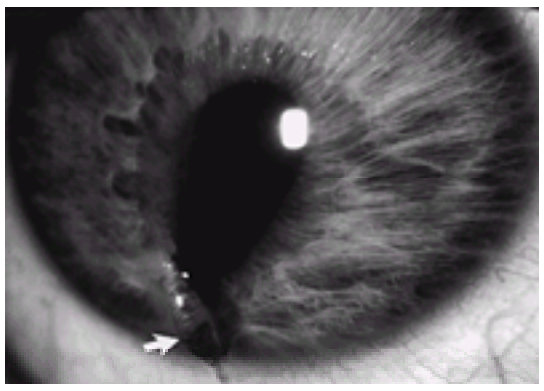

FIGURE 111.8. Corneal laceration (ruptured globe). Note iris protruding through wound *(arrow)* and teardrop-shaped pupil pointing in direction of laceration.

Management

Although the outcome in some ruptured globes, particularly small peripheral corneal lacerations, may be good, eyeball rupture is certainly an ominous sign that warrants emergent referral for ophthalmology consultation. Further ocular examination should be stopped immediately. No eyedrops should be instilled. A patch should never be used in this circumstance. A plastic shield should be placed over the eye such that the edges of the shield make contact with the bony prominences above and below the eyeball (Fig. 111.9). If a commercially marketed shield is not available, the clinician should cut off the bottom of a styrofoam or plastic cup and use it as a shield, resting it against the bony prominences (Fig. 111.10).

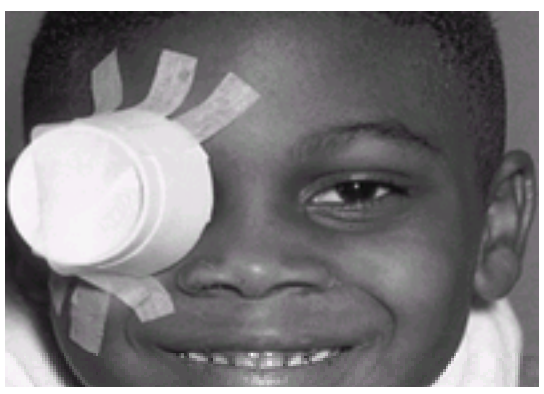

FIGURE 111.10. The bottom of a styrofoam cup is used as an eye shield.

Severe eye trauma may cause sedation or vomiting without head trauma or brain injury. If not, every attempt should be made to keep the child calm, even if sedation must be used. Keep in mind that crying, screaming, and Valsalva maneuvers such as vomiting can result in extrusion of intraocular contents through the rupture. Although broad-spectrum intravenous antibiotic coverage is desirable, particularly if a delay may occur before the patient sees an ophthalmologist, this treatment must be weighed against the potential aggravation of the child, which might accompany the needle puncture for catheter placement.

Even if a ruptured globe is not seen clearly on examination, any patient who has a high-risk history, severe lid swelling, and extreme resistance to examination should be given an eye shield and referred to an ophthalmologist as if a ruptured globe was confirmed.

BLOWOUT FRACTURE

Clinical Manifestations

The pathophysiology and diagnosis of blowout fractures are discussed in Chapter 25. Following blunt compressive eye trauma, the eyeball may be retroplaced in a manner that increases the pressure within the orbit and “blows out” one or more bones of the orbital wall. Direct blunt trauma to the orbital rims may also cause bony fractures that extend back into the orbit. Therefore, orbital fractures may occur after facial trauma with or without eyeball trauma.

The most common orbital fracture is an inferior and/or medial wall fracture. The lateral wall is the least commonly fractured. The intraocular contents often sink back into the fracture, giving an enophthalmic appearance (“sunken eye”). However, proptosis can also occur from orbital hemorrhage. Superior wall fracture (roof fractures) may be associated with pulsating proptosis as a result of communication between the orbit and intracranial cavity. Fractures of the inferior wall may be associated with numbness of the ipsilateral malar region caused by injury to the infraorbital nerve, which travels along the floor of the orbit. Palpation of the bony rim of the orbit is often remarkably normal contrary to the

common teaching about point tenderness and “step off” signs.

The hallmark sign of orbital fracture is a restriction of extraocular movement. Usually, the eye is unable to look away from the fracture site because of a tethering of intraocular muscle or other orbital tissues in the fracture (Fig. 25.4). However, orbital hemorrhage at the fracture site can displace the eyeball away from the fracture and make it difficult for the eye to look in the direction of the fracture.

Management

Some controversy exists among ophthalmologists, otorhinolaryngologists, and craniofacial surgeons regarding the urgency for radiologic evaluation and surgical intervention in the management of orbital wall fractures. Approximately 20% of orbital fractures are associated with eyeball injury. Therefore, ophthalmology consultation for complete dilated retinal examination and slitlamp biomicroscopy is indicated in virtually every case. Axial (proptosis) or coronal displacement of the eyeball is an ominous finding because it may be a sign of orbital hemorrhage, which can cause compression of the optic nerve, requiring emergency surgical intervention. If a decision is made to proceed with radiologic imaging, the optimal test is a computed tomography (CT) scan of the orbit with both axial and coronal views. The brain should be included, particularly when a roof fracture is suspected. Plain skull radiographs have little role in the management of orbital wall fractures and need not be ordered.

EYELID LACERATIONS

Clinical Manifestations

Although eyelid lacerations are usually easy to detect, the clinician must remember that the underlying eyeball also might have been lacerated or injured. Seemingly superficial lacerations of the eyelid may be associated with penetration into the orbit or intracranial cavity, particularly when the injury was caused by a pointed implement such as a tree branch or pencil. If possible, the eyelid should be everted to look for a conjunctival wound indicating that the laceration is actually a complete perforation of the eyelid.

Oblique lacerations that extend into the medial canthal area (juncture of the upper and lower lids medially) may involve the proximal portion of the nasolacrimal duct (Fig. 111.11). Sometimes, the lid margin puncta, which drain tears into the system, is displaced laterally as a result of the laceration (Fig. 111.11). Lacerations in this area should usually be referred for ophthalmology consultation if any question persists regarding whether the tear drainage system is intact.

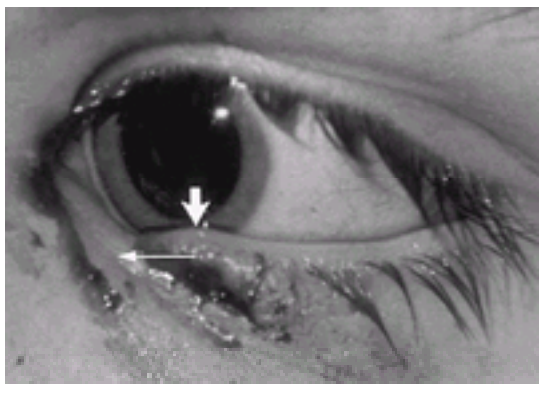

FIGURE 111.11. Lower lid laceration involving tear drainage system. *Large arrow* indicates lower lid punctum, which has been displaced laterally. *Thin arrow* indicates normal course of canaliculus, which drains tears from the puncta to the lacrimal sac located medially.

Management

Lacerations of the periorbital skin and superficial eyelid skin may be managed by standard skin closure techniques discussed elsewhere in this book. However, it is important that sutures not grasp deep tissue within the eyelid because this may result in cicatrical eversion of the eyelid margins. Table 111.2 summarizes those findings that, when associated with eyelid lacerations, should prompt ophthalmology consultation for wound closure.

Consult ophthalmology if laceration associated with:
Full-thickness perforation of lid
Ptosis
Involvement of lid margin
Possible damage to tear drainage system
Tissue avulsion
Eyeball injury

Table 111.2. Eyelid Lacerations

PERIORBITAL ECCHYMOSIS

Clinical Manifestations

Periorbital ecchymosis is usually a benign finding, although it may be associated with bony fracture or eyeball injury. Because of the loose connection of the eyelid skin and underlying tissues, dramatic ecchymosis can occur with mild blunt trauma. It is of forensic importance that bilateral ecchymosis may occur from midline forehead injuries. The dating of injuries should not be made on the basis of the color of periorbital ecchymosis. Because accumulation of blood tends to be more dramatic in the eye than in other body sites, periorbital ecchymosis often looks much darker than what might be expected elsewhere for a bruise of similar age.

Management

No treatment is routinely needed for periorbital ecchymosis. Ice packs applied to the area, with the eyelids closed, can be helpful in reducing swelling. Anticipatory guidance may be given to inform the family that the ecchymosis may persist for more than 2 weeks. An ongoing color change from purple to green and yellow may occur as the blood is resorbed and broken down. A hyperpigmented area may be left for several weeks or months thereafter.

CORNEAL AND CONJUNCTIVAL INJURY

Clinical Manifestations

The conjunctiva can be abraded or lacerated (see Fig. 111.7), although the management of this problem is usually identical to that of corneal abrasion because the tissues heal so rapidly. Corneal or conjunctival abrasions may occur even from mild surface trauma. Self-inflicted abrasions may occur accidentally.

Corneal abrasion can be painful and accompanied by dramatic photophobia and resistance to opening of the eyes. Yet, some children have remarkably few symptoms. They may complain of a foreign body sensation even though no foreign body is present. A drop of topical proparacaine 0.5% or tetracaine 0.5% may have both diagnostic and temporary therapeutic usefulness. Any patient who is made more comfortable by the instillation of these drops must have an ocular surface problem (conjunctiva or cornea) as the cause of pain. The child who is crying and refusing to open the eyes may be compliant and easy to examine just a few minutes after the instillation of a topical anesthetic. Onset of action is approximately 20 seconds, and duration is approximately 20 minutes.

Topical fluorescein is used as a diagnostic agent to stain the affected area. Fluorescein is available as impregnated paper strips and as a solution combined with a topical anesthetic (Fluress, Barnes-Hind, Inc.). Considering the limited use in an ED setting, strips may be a more practical method because bacterial contamination over time may be less likely. When impregnated strips are used, they must be wet before instillation. Otherwise, the strip itself may cause a corneal abrasion, thus preventing the examiner from correctly identifying the patient's problem. Topical anesthetic or saline may be used to wet the strip. The lower eyelid should be pulled down, exposing the pink inner surface (palpebral conjunctiva) against which the strip may be placed. The solution then diffuses off the strip into the area between the lower eyelid and the eyeball (inferior fornix) where it is then displaced across the ocular surface with the next blink. The clinician must avoid placing too much fluorescein because the tear film can be so oversaturated that it may be difficult to find a small abrasion.

Fluorescein, which is orange, fluoresces yellow–green when exposed to blue light. Many modern ophthalmoscopes have a blue light. The examiner can view through the peephole of the direct ophthalmoscope, spinning the focusing dial to allow the ocular surface to be in focus with the examiner 2 to 3 inches away from the patient, inspecting the cornea and conjunctiva for an area that is stained. A Wood or Burton lamp can also be used. Some of these devices have handheld magnifying glasses attached. Although the staining may not be as dramatic, white light from a direct ophthalmoscope or penlight can also be used because some blue wavelengths are incorporated in white light. Green light (red free), available on most direct ophthalmoscopes, may make identifying stained areas more difficult than white light.

If the staining pattern reveals one or more vertical linear abrasions, the examiner should suspect the presence of a retained foreign body under the upper lid. This foreign body may be viewed by upper lid eversion (Fig. 111.12). The patient should be asked to look down repeatedly throughout this procedure. With the eye in downgaze, a cotton swab should be placed against the midbody of the upper eyelid and gently rotated downward toward the eyelashes so that the skin is rolled with the swab by friction. This procedure causes the eyelashes to turn out toward the examiner so that they may be grasped between the examiner's thumb and forefinger. It may be necessary to grab the entire lid margin in some children. If a topical anesthetic is instilled before lid eversion, this procedure is painless. After the lashes (or margin) are grabbed, they should be lifted vertically while the cotton swab is used to apply gentle downward pressure in the opposite direction. The eyelid then flips around the cotton swab. If a foreign body is identified, it can be gently lifted away using a cotton swab or forceps. To revert the eyelid, simply have the patient look upward or massage the lid down.

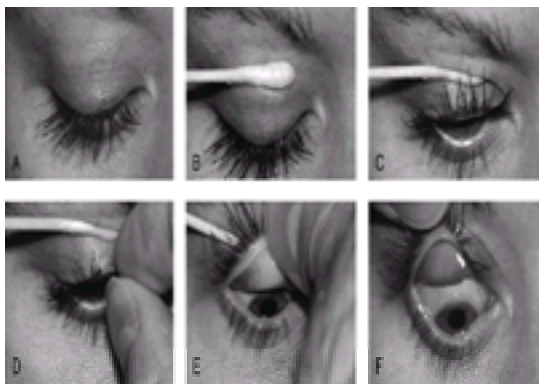

FIGURE 111.12. Upper lid eversion. Note that patient is looking down throughout procedure. In frame **C,** the swab is being rolled clockwise to engage skin and indirectly lift lash line. In frame **E,** the swab is being pushed downward as the examiner lifts the lashes upward in the opposite direction. In **F,** note that patient is wearing a contact lens.

Subconjunctival hemorrhage may result from blunt trauma, conjunctivitis, chemical irritation, and increased intrathoracic pressure (e.g., chest trauma, suffocation). Although usually focal, the lesions may be multiple or diffuse. Hypertension, coagulopathy, or anticoagulant medications may result in subconjunctival hemorrhage out of proportion to the injury. After blunt eyeball trauma, a 360 degree subconjunctival hemorrhage may mask an underlying ruptured globe (Fig. 111.13). No treatment is needed for isolated subconjunctival hemorrhages. They may take up to 2 weeks to resolve, turning yellowish in the process.

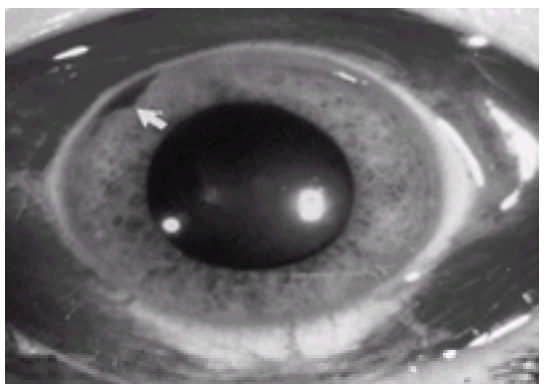

FIGURE 111.13. Subconjunctival hemorrhage extending for 360 degrees. Note the small hyphema *(arrow)*.

Chemical injuries of the cornea and conjunctiva may also occur. These injuries are discussed in Chapter 120.

Management

Several studies have suggested that patching, especially in children with small corneal abrasions, may not accelerate healing or decrease symptoms. Although some controversy exists in this regard, many physicians prefer to apply a lubricating antibiotic ointment (e.g., bacitracin, erythromycin, Polysporin [Burroughs Wellcome]) to the ocular surface, followed by a pressure patch over the closed eyelid. For patients who are in significant pain, a drop of cyclopentolate 1% can be instilled to relieve spasm of the eye's ciliary muscle. Ointments containing steroids or neomycin should not be used. For patients who are relatively asymptomatic, with corneal or conjunctival abrasions that are small and that do not involve the visual axis (i.e., not involving central cornea over the pupil), management with antibiotic or artificial tears alone may be sufficient. If desired, a pressure patch can be created by stacking two or three patches on top of each other over the closed eyelid before applying tape. The patch should be worn overnight. For conjunctival abrasions and small corneal abrasions not involving the central cornea, the patch may be removed by the patient or parents on the following day (approximately 18 to 24 hours later). If the patient is asymptomatic, no follow-up is required. However, if pain or foreign body sensation continues, the patient should be instructed to seek ophthalmologic care. A topical antibiotic ointment may be prescribed for use after patch removal two to three times daily for approximately 3 days. Larger corneal abrasions and those involving the visual axis should be seen the next day by an ophthalmologist.

If concern exists that a superficial abrasion may actually represent penetration into the deeper corneal tissues, a teardrop or irregular pupil may be seen. Urgent ophthalmic consultation is indicated. Any patient with a staining corneal defect who has a history of ocular herpes or who wears contact lenses should be referred urgently for ophthalmology consultation. Patients who wear contact lenses should never be patched under these circumstances. The contact lenses should be removed immediately.

HYPHEMA

Clinical Manifestations

The presence of blood between the cornea and iris is a sign of severe ocular trauma. Although the entire anterior chamber may be filled with blood (“8-ball hyphema”), clots may also be small, requiring careful inspection for detection (Fig. 111.13). Sometimes the blood is more diffuse throughout the anterior chamber (Fig. 111.7) or may even be microscopic, requiring slitlamp examination for detection (microhyphema). The size of the hyphema is directly

proportional to the incidence of secondary glaucoma and is inversely proportional to visual prognosis. Patients with hyphema enter a vulnerable period 3 to 5 days after injury when spontaneous rebleeding may occur. Patients with hemoglobinopathies are also at particular risk for ocular complications of hyphema. Therefore, all patients who are in a high-risk ethnic group should receive a screening test (“shake test”) or formal hemoglobin electrophoresis at presentation unless their status is already known.

Management

All patients who have hyphema must be seen by an ophthalmologist. Although microhyphemas and perhaps some small hyphemas may be managed in select clinical situations as outpatients with careful daily follow-up, hospital admission is often recommended. The eye should be shielded, not patched (see Fig. 111.9), and the patient should be placed at bedrest with the head elevated 45 degrees. This position helps allow blood within the anterior chamber to settle inferiorly, thus allowing a clearance of the visual axis, improvement of vision, and a better view for the ophthalmologist looking into the eyeball. If an ophthalmologist is not readily available, the examining physician may recommend admission and the use of a dilating agent to paralyze the ciliary muscle within the eye. Some ophthalmologists may even recommend sedation of an active or distressed child. Oral antifibrinolytics may be used to prevent spontaneous rebleeding. However, these agents should be used only under the supervision and recommendation of an ophthalmology consultant.

TRAUMATIC IRITIS

Clinical Manifestations

Inflammation within the anterior chamber of the eye often does not present for 24 to 72 hours after blunt trauma to the eyeball. The patient may complain of eye pain, redness, photophobia, and sometimes visual loss. The pupil on the affected side may be constricted (see Chapter 26). The ocular injection may be confined to a ring of redness surrounding the cornea (ciliary flush). Definitive recognition of traumatic iritis requires slitlamp biomicroscopy.

A beam of light projected from a light source can be seen only as it reflects from surfaces. For example, the light from a movie projector in a movie theater would not be visible unless smoke and dust were present in the air to reflect the light as it passes between the projector and the screen. Likewise, the slit beam of light projected from the slitlamp is normally visible only as it reflects off (and through) the cornea and then passes unseen through the optically clear fluid of the anterior chamber, landing on the iris and lens (within the pupil), where it is again visible as it reflects from their surfaces. When white blood cells are floating within the anterior chamber fluid (aqueous humour), along with protein that has leaked from inflamed blood vessels, this beam of light then becomes visible as it passes through the aqueous humour. The white blood cells appear as small specks floating within the beam of light. Red blood cells from microhyphema can also be detected in this manner.

Management

Traumatic iritis may be an indicator that other ocular injuries have occurred. Ophthalmology consultation should be obtained in the diagnosis and management of this condition. The ophthalmologist often recommends dilating drops and topical steroids for treatment. Because of the risks associated with the use of topical steroids, they should not be prescribed except in consultation with an ophthalmologist.

TRAUMATIC VISUAL LOSS

Clinical Manifestations

Some techniques for the recognition of true traumatic visual loss are discussed in pages 1397–1398. Occasionally, the emergency physician is faced with a child who is feigning visual loss. These situations seem to be more common after motor vehicle accidents or other injuries in which legal action may be involved. Functional visual loss can also be idiopathic or associated with other overt or covert stress in the child's life. In the absence of other signs of ocular or head trauma, this diagnosis should be suspected. It then becomes necessary to “trick” the child into demonstrating that he or she can actually see. Patients who are truly acutely blind should demonstrate some degree of anxiety and virtually complete inability to navigate in the new surroundings of the ED. When asked to write their names on a piece of paper, truly blind patients can do so accurately, unlike children who are functionally blind who assume that they are unable to write. When a mirror is held before a truly blind eye and then tilted in the vertical and horizontal planes, the eye will not follow. However, any eye that truly has enough sight to recognize its own image moves involuntarily with the motion of the mirror.

Children who are feigning visual loss but not complete blindness can be more difficult to “trick.” Sometimes, by placing a drop of saline or topical anesthetic in the eye while giving the child the suggestion that these “magic drops” will cause a return of vision, the child then begins to see better. The pinhole test (previously discussed) can also be used in this manner. Ophthalmology consultation is sometimes critical in discovering whether a child has truly sustained visual loss.

A rare cause of visual loss after head trauma is transient cortical visual impairment/blindness. As a result of a direct or centre coup occipital contusion, a child may experience acute blindness despite an otherwise normal eye examination. This centrally mediated phenomena may resolve spontaneously. Ophthalmology consultation can be useful to rule out other causes of the visual loss.

A multitude of intraocular injuries, including traumatic cataract, vitreous hemorrhage, retinal bruising (commotio retinae), retinal detachment, and optic nerve injury, can result in true visual loss or blindness. The pediatric emergency physician is the “gatekeeper” in recognizing that true intraocular injury has occurred, although in most of these circumstances, ophthalmology consultation is then required. Perhaps the best screening test for intraocular injury is the examination of

the red reflex and direct ophthalmoscope exam.

CHILD ABUSE

Clinical Manifestations

Virtually any eye injury can be the result of child abuse. Unusual types of ocular trauma, such as the covert instillation of noxious substances onto the conjunctiva, should also be considered in the differential diagnosis of recurrent red eyes in the absence of an apparent cause. Perhaps the most common ocular manifestation of child abuse is the finding of retinal hemorrhages associated with the shaken baby syndrome (Fig. 111.14). Although these hemorrhages can be seen with the direct ophthalmoscope, ophthalmology consultation is indicated. Children who present to the ED before the age of 4 years with a history of head trauma or sudden unexplained cardiorespiratory arrest should have a full dilated examination to look for retinal hemorrhages that may indicate that a nonaccidental shaking injury has occurred.

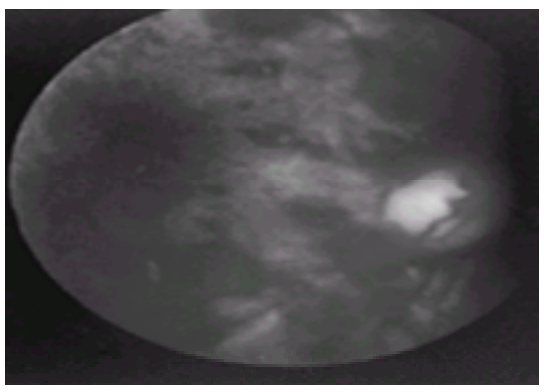

FIGURE 111.14. Retinal hemorrhages in shaken baby syndrome. *Please see the color-tip insert (Color Plate 111.14).*

Suggested Readings

Arbour JD, Brunette I, Boisjoly HM. Should we patch corneal erosions? Arch Ophthalmol 1997;115:313–317.

Dutton JJ, Manson PN, Iliff N, Putterman AM. Management of blow-out fractures of the orbital floor. Surv Ophthalmol 1991;35:279–298.

Frey T. Pediatric eye trauma. Pediatr Ann 1983;12:487–497.

Grin T, Nelson LB, Jeffers JB. Eye injuries in childhood. Pediatrics 1987; 80:13–17.

Kaiser PK. A comparison of pressure patching versus no patching for corneal abrasions due to trauma or foreign body removal. Ophthalmology 1995;102:1936–1942.

Levin AV. Eye emergencies: acute management in the pediatric ambulatory setting. Pediatr Emerg Care 1991;7:367–377.

Levin AV. General pediatric ophthalmic procedures. In: Henretig FM, King CK, eds. Textbook of Pediatric Emergency Procedures. Philadelphia: Williams & Wilkins, 1997;579–592.

Levin AV. Slit lamp examination. In: Henretig FM, King CK, eds. Textbook of Pediatric Emergency Procedures. Philadelphia: Williams & Wilkins, 1997;593–598.

Nelson LB. Pediatric eye trauma. Pediatr Clin North Am 1983;30:1167–1183.

Vaughan D, Asbury T, Tabbara KF. General Ophthalmology. 12th ed. Norwalk, CT: Appleton & Lange, 1989.

CHAPTER 112

Otolaryngologic Trauma

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The ear, nose, and throat are common sites for trauma; therefore, emergency medicine specialists must be familiar with the head and neck region because they will often be called on to evaluate this area. Although the presenting complaints may seem extremely distressing to the patient and cause considerable anxiety for the parents, the conditions prompting the visit are rarely life-threatening. Many instances of trauma may be isolated to the ear, nose, or throat, but associated injuries (e.g., eye, dental, central nervous system, thorax) are common and must be detected when evaluating and treating otolaryngologic trauma.

Evaluation of the patient with otolaryngologic trauma requires a careful examination of the head and neck. The specific methods of examination of each anatomic area are detailed in Chapter 121.

EAR

Foreign Bodies

Foreign bodies in the ear canal are common in children. Solid objects, such as stones, beads, or paper, are the most commonly encountered foreign bodies, but live insects may also enter the ear canal. Foreign bodies should be removed as soon and as safely as possible. Most objects can be gently rolled out of the external meatus with an ear curette or grasped and removed with an otologic forceps (see Procedure 5.3 in Section VII). Round or occluding objects may be removed by irrigation of the canal with body-temperature water. (Irrigation, however, should not be performed if a tympanic membrane [TM] perforation is suspected or if a ventilating tube is in place.) The stream is directed along the side of the foreign body, forcing it to the external meatus. Insects should be killed by filling the ear canal with alcohol or mineral oil before they are removed from the ear canal by the techniques already described. Objects resting against the TM are best removed by irrigation to avoid injury by manipulation.

Care must be taken to remove the foreign material without causing pain; if the removal is unsuccessful, a general anesthetic is usually necessary for removal of the object by the otolaryngologist.

Trauma

External Ear Trauma

External ear trauma is common in children because the pinna is in an exposed position on the side of the head. Reflex turning of the face to the side to avoid a blow or a fall places the ear directly in the line of injury. External blunt trauma often occurs secondary to an athletic injury, a fall, or a direct blow to the ear; the injury may result in simple ecchymosis or it may disrupt perichondrial blood vessels with subsequent hematoma or seroma formation. These collections form a smooth bluish colored mass on the lateral surface of the auricle that obscures its normal contour. Hematomas and seromas must be evacuated immediately to prevent cartilage necrosis (see Procedure 5.4 in Section VII). Lacerations of the pinna should be closed using the same surgical principles applied to repairing lacerations in other end-organ areas of the body. Earrings in pierced ears may be torn from the lobule. These lacerations should be closed like all skin lacerations, reestablishing the normal anatomy.

Thermal injury of the external ear commonly occurs because the ear protrudes from the head and is exposed to burns

and cold. Burns of the ear should be treated in the same manner as burns of other parts of the body (see Chapter 114). Frostbite is suspected when the ear is pale and painful on warming. The frostbitten ear should be rewarmed rapidly by applying warm soaked cotton pledgets at 38° to 40°C (100.4° to 104°F); the ear should be completely thawed and never recooled.

Middle Ear Trauma

A slap to the side of the head (by a hand or a breaking wave) may result in perforation of the TM by sudden compression of the air in the external auditory canal, but traumatic perforation of the drum is usually caused by poking an object into the ear canal. The structures of the middle ear may also be damaged by the penetrating object. The ossicles may be fractured and a perilymph fistula may be created in the footplate of the stapes. This reaction causes immediate vertigo and a sensorineural hearing loss. The facial nerve may be injured and cause facial paralysis. Traumatic perforations of the TM must be examined carefully to be certain that the edges of the perforation do not fold into the middle ear. If they do, skin may grow into the middle ear and a cholesteatoma will develop. Clean perforations with margins that do not fold into the middle ear usually heal spontaneously in 2 to 3 weeks. The perforation should be kept clean and dry. If the ear is draining, topical otic drops (neomycin, polymyxin, and hydrocortisone) should be used for 10 days. Systemic antibiotics are usually unnecessary. Any perforation that does not heal within 3 weeks should be referred to an otolaryngologist for evaluation and repair. Traumatic perforations associated with vertigo, sensorineural hearing loss, or facial nerve paralysis require urgent consultation and possible exploration of the middle ear by an otolaryngologist.

Barotrauma to the ear may occur during an airplane trip or while scuba diving, especially if the child has an acute upper respiratory infection. A direct open communication between the middle ear and the nasopharynx normally permits prompt equalization of changes in ambient pressure. If the eustachian tube is obstructed, however, changes in ambient pressure may not be transmitted to the middle ear, and barotrauma can result. As the child descends in an airplane (or during an underwater dive), the increased ambient pressure is transmitted to the cardiovascular system and, thus, to the vessels of the mucosal lining of the middle ear. The mucosa becomes edematous and the vessels become engorged. If the eustachian tube is obstructed and has not equalized the air pressure, a large differential pressure occurs between the middle ear mucosa and its air-filled cavity. This condition results in a rupture of the blood vessels within the mucosa and bleeding into the middle ear. Serous fluid may also accumulate in the middle ear secondary to eustachian tube obstruction. Rarely, perforation of the TM occurs. These injuries usually resolve spontaneously over several weeks. Antimicrobials may be prescribed to prevent infection of the middle ear fluid/blood. The rare case that does not respond to this regimen should be referred to an otolaryngologist for further evaluation and treatment. Persistent symptomatic fluid may require myringotomy and ventilation tube placement. Barotrauma with acute sensorineural hearing loss and/or vertigo may indicate the presence of a perilymph fistula (previously described). Persistence of symptoms and signs of a fistula require urgent middle ear exploration to close the fistula.

Inner Ear Trauma

Concussive injuries to the head may cause inner ear trauma by disrupting the delicate intracochlear membranes. Sensorineural hearing loss and/or vertigo may occur as a result of such an injury. Occasionally, the losses from these injuries can improve spontaneously, but most are permanent. Temporal bone fractures (especially transverse) have a high incidence of cochlear disruption.

Constant exposure to loud noise or amplified sound may cause a progressive high-frequency sensorineural hearing loss. Loud blasts from explosions may cause sudden permanent sensorineural hearing loss.

Cerebrospinal Fluid Otorrhea

Cerebrospinal fluid (CSF) otorrhea may occur secondary to a temporal bone (usually longitudinal) fracture that results in a fracture through the inner ear and a ruptured TM. Manipulation or instrumentation of the external auditory canal in the presence of CSF otorrhea is discouraged because it could introduce bacteria and contribute to the development of meningitis. If CSF otorrhea is suspected, the child should be placed at bed rest with the head elevated and neurosurgical consultation should be obtained. The use of prophylactic antimicrobials in CSF otorrhea is controversial.

Facial Nerve Paralysis

Facial nerve paralysis may occur as a result of temporal bone trauma (see Chapter 105). Transverse fractures of the temporal bone can cause disruption of the facial nerve in its intratemporal segment. Longitudinal fractures are less likely to cause facial nerve paralysis. Patients with traumatic facial nerve paralysis should be referred to the otolaryngologist for possible exploration and nerve repair.

NOSE AND PARANASAL SINUSES

Nasal Trauma

General Principles/Nasal Fracture

Facial trauma often occurs in children as a result of play activities, contact sports, and automobile accidents. Most of these injuries are minor. Nevertheless, any child with facial trauma should be assessed for associated, possibly more serious, injuries to the cervical spine, central nervous system, and chest (see Chapter 104).

Because of its prominent position on the face, the nose is subject to frequent trauma and accounts for most facial injuries in children. It is important, however, to realize that nasal trauma may also be associated with ocular injury, such as hyphema or retinal detachment. If the initial survey suggests a serious ocular injury, an ophthalmologic consultation must

be obtained.

Trauma to the nose most often causes ecchymosis and edema of the overlying skin. However, a direct blow to the nose can fracture the nasal skeleton with resultant deviation and/or depression of the nasal bones and septum. The deformity should be readily apparent by clinical examination, but the postinjury edema may prevent its recognition for several days until the swelling has subsided (Fig. 112.1). A stepoff or bony irregularity may often be detected in these patients. Radiographs of the nose are notoriously unreliable in the evaluation of nasal injuries and are not recommended in the routine management of simple nasal fractures. Epistaxis commonly accompanies nasal trauma but usually has stopped by the time the child reaches the emergency department (ED). Persistent or severe bleeding may require local pressure, topical vasoconstrictors, or nasal packing (see the section on epistaxis in Chapter 121). The presence of any associated ocular injury, such as hyphema or retinal detachment, must be detected, and ophthalmologic consultation must be obtained.

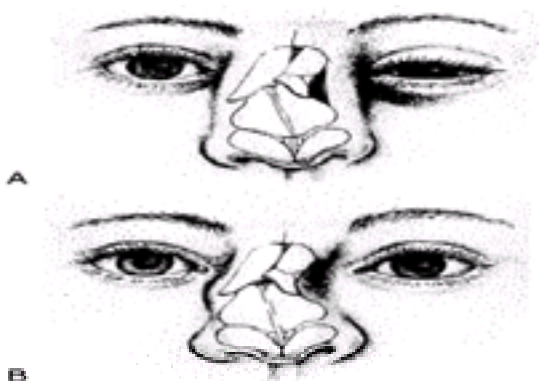

FIGURE 112.1. A. Postinjury edema may mask underlying nasal bone deformity. **B.** Nasal deformity manifests as edema subsides.

In assessing the nasal injury, the emergency physician must determine the nature and extent of trauma to the overlying skin, the nasal skeleton, and the nasal septum. A septal hematoma, if present, requires incision drainage. The amount of nasal deviation and/or depression should be noted. Because this condition can be masked by postinjury edema, it may be best to examine the child again in 3 to 4 days when the swelling has subsided to allow an accurate determination of nasal deviation and/or depression.

If no septal hematoma or associated ocular or intracranial injuries are present, then the deviated nose and septum can be reduced by the otolaryngologist when the swelling has subsided enough to permit an accurate evaluation of the nasal deformity. If more than 7 days elapse between the time of injury and the attempt at reduction, the fracture fragments begin to form a strong fibrous union in their deviated positions, making reduction difficult. Antimicrobials (usually amoxicillin 25 to 50 mg/kg per day for 7 days) are generally administered to these patients to prevent complications from occurring in what is almost always a compound fracture (i.e., open into the nasal cavity).

Septal Hematoma

The presence of a septal hematoma must be recognized as soon as possible after the injury. A septal hematoma appears as a bulging of the nasal septum into one or both sides of the nasal cavity. Accumulation of blood between the septal cartilage and its overlying mucoperichondrium deprives the cartilage of its blood supply. Otolaryngologic consultation should be obtained as quickly as possible if a septal hematoma is suspected. The hematoma is drained as soon as possible and the mucoperichondrium packed back against the septal cartilage to restore its blood supply (see Procedure 7.4 in Section VII). If the hematoma is left for any length of time, septal abscess or cartilage destruction may occur. A saddle nose deformity is the result of an improperly treated septal hematoma.

Cerebrospinal Fluid Rhinorrhea

A clear, watery rhinorrhea occurring after nasal trauma may be CSF rhinorrhea, which would indicate a skull fracture, usually through the cribriform plate. Less commonly, the CSF originates from a temporal bone fracture and enters the nasopharynx through the eustachian tube. If the patient leans forward, allowing the nasal drainage to drip onto a piece of paper, a characteristic target pattern will often appear, with a blood stain in the center of the drop and a clear halo of CSF around it. CSF is high in glucose, which can be detected with the use of a glucose oxidase test paper (used in urinalysis). Care must be taken in interpreting these tests, however, because normal nasal mucus can look like CSF, and the oxidizing substances present in nasal and lacrimal secretions may give a false-positive reaction. If CSF rhinorrhea is suspected by history or clinical examination, the child should be admitted and restricted to bed rest with his or her head elevated in an attempt to decrease the leak, and neurosurgical consultation should be obtained. The use of prophylactic antimicrobials in CSF rhinorrhea is controversial. Further diagnostic studies, such as computed tomography (CT) scans and isotope scans, can be performed to confirm the diagnosis of CSF leak.

Sinus Trauma

Fractures of the paranasal sinuses may occur as isolated injuries or in association with trauma to the nose and orbital structures. Fractures of the ethmoid sinus or anterior wall of the maxillary sinus usually occur as a result of blunt trauma to the nose or cheek, respectively. The otolaryngologist should assist the emergency physician in evaluating these injuries. Subcutaneous crepitance may be felt in the cheek or around the eye. Radiographs may demonstrate air in the cheek or orbit, or air-fluid levels. After determining the absence of associated ocular injury, the patient is usually placed

on oral antimicrobials (usually amoxicillin 25 to 50 mg/kg per day for 7 days) and observed as an outpatient until the crepitance resolves.

Facial Trauma

Blunt facial trauma can result in focal injuries and fractures or more generalized fractures involving much of the midface. Blunt trauma to the orbit may result in the force being transmitted through the globe to break the orbital floor (roof of maxillary sinus) or the medial orbital wall (lamina papyracea of the ethmoid sinus). These blowout fractures are discussed in detail in Chapter 110 and Chapter 111.

Midface fractures include fractures of the malar bone, which affect both the orbital floor and the maxillary sinus. A complete malar (incorrectly called trimalar or tripod) fracture is present when the malar bone fractures at the infraorbital rim, zygomatic arch, and zygomaticofrontal suture line. Isolated malar fractures can also occur at the zygomatic arch or the lateral wall of the maxillary sinus. (For more detail regarding sequential steps and examination for facial fractures, see Chapter 110.) Severe midface injuries often require multidisciplinary cooperation between otolaryngology, ophthalmology, plastic surgery, oral surgery, and neurosurgery.

Sinus Barotrauma

A direct open communication between the paranasal sinuses and the nasal cavities normally permits prompt equalization of changes in ambient pressure. If a sinus ostia is obstructed, however, changes in ambient pressure may not be transmitted to the affected sinus cavity (most commonly the maxillary sinus, although the frontal sinus may be affected in older children and adolescents), and barotrauma can result. As the child descends in an airplane (or during an underwater dive), the increased ambient pressure is transmitted to the cardiovascular system and, thus, to the vessels of the mucosal lining of the sinus. The mucosa becomes edematous and the vessels become engorged. If the sinus is obstructed and has not equalized the air pressure, a large differential pressure occurs between the sinus mucosa and its air-filled cavity. This condition results in a rupture of the blood vessels within the mucosa and bleeding into the sinus. The child usually complains of cheek pain and may have epistaxis. Treatment for this condition involves amoxicillin 25 to 50 mg/kg per day for 7 days (to prevent infection of the blood-filled sinus), antihistamine–decongestant therapy and topical nasal sprays to restore the normal physiologic communication between the sinus and the nasal cavities, and the avoidance of further barotrauma. The rare case that does not respond to this regimen should be referred to an otolaryngologist for further evaluation and treatment.

Foreign Bodies

Nasal foreign bodies are common in children. These children are usually brought to the ED with the history of putting an object into the nose, but the presence of a foreign body may often be unsuspected and may be discovered only during evaluation of a child with persistent, unilateral, foul-smelling, purulent rhinorrhea. This mode of presentation for these problems is so common that any child with a foul-smelling unilateral nasal discharge (even without a history of placing an object in the nose) should be considered to have a nasal foreign body until proven otherwise. The foreign body is usually visible on anterior rhinoscopy. However, purulent secretions may have to be suctioned from the nose before the object is seen. Radiographs are of limited value because most of the foreign bodies are radiolucent (e.g., paper, cloth, sponge, food).

If the object is located in the nasal vestibule, the emergency physician may attempt to remove it (see Procedure 7.3 in Section VII). The child should be adequately restrained, and the necessary equipment, including a nasal speculum, directed light, suction, small hooks, and forceps, should be available. Otologic instruments are often very useful in the removal of nasal foreign bodies. A few drops of 4% lidocaine can be placed in the nostril to provide topical anesthesia before removing the foreign body. An otolaryngologist should be consulted if the foreign body cannot be removed easily. Hygroscopic foreign bodies, such as beans, may swell with nasal secretions and become difficult to remove. The foreign body should never be pushed or irrigated into the nasopharynx, where it could be aspirated by the struggling child. Antimicrobials (usually amoxicillin 25 to 50 mg/kg per day for 7 days) are administered to prevent (or treat) an infection (rhinitis, sinusitis) in this already traumatized area, especially after removal of a long-standing foreign body.

ORAL CAVITY, PHARYNX, AND ESOPHAGUS

Trauma

Oral cavity trauma is usually caused by biting the inside of the cheek or tongue. This condition is painful and can cause a laceration or hematoma formation. Treatment of self-inflicted bites is rarely needed, but a laceration may require suturing if it bleeds excessively or is severe enough to alter intraoral anatomy or physiology (i.e., breathing, deglutition, or speech).

Children may have oropharyngeal lacerations or puncture wounds when they fall with an object, such as a stick, in their mouths. If the injury is restricted to the central portion of the palate, damage to vascular or neural structures of the neck is unlikely. These children are usually safe to send home after confirmation of absence of any retained foreign body (see the following section). However, trauma to the lateral aspects of the palate or the posterior pharyngeal wall may be associated with vascular injuries of the carotid artery or the jugular vein. Expanding hematoma of the neck or pharynx, continued intraoral bleeding, and diminished pulses in the neck are all signs of serious vascular injury. These children need to be admitted and have an urgent angiogram and possible neck exploration. If a lateral pharyngeal or palatal puncture injury is present without signs of vascular injury, the child should be observed closely in the hospital or at home for signs of neurologic deterioration.

In treating puncture wounds of the pharynx, it is imperative to determine whether the foreign body has been recovered intact or if a portion of the foreign body may have been left in the palatal tissues. For example, a portion of pencil lead left

in the palatal tissue causes a chronic foreign body reaction if it is not removed at the time of initial treatment and repair. Plain radiographs may not be useful in determining whether a foreign body has been left in the wound because most of the objects are radiolucent and/or too small to be seen. Inspecting the actual object that caused the wound to make sure that it is intact is more important. If a retained portion of the foreign body is suspected, CT scan may be required, followed by exploration of the wound, usually under general anesthesia.

Clean puncture injuries or simple lacerations do not require surgical repair and usually heal by secondary intention. Large gaping injuries may require formal layered closure to restore normal function to the palate.

Caustic Injuries

Caustic substances (lye or acid) may be ingested, causing burns anywhere from the lips to the stomach. Burns of the oral mucosa appear as patches of erythema, blebs, or ulcerated areas. Although caustic burns are usually visible in the oral cavity and pharynx, large skip areas may exist. Therefore, the absence of oral or pharyngeal burns does not rule out esophageal injury. If a history of significant caustic ingestion exists (see Chapter 88), an esophagoscopy should be performed 6 to 12 hours later to establish the presence of esophageal burns, regardless of the condition of the oral cavity and pharynx. Because burns occur rapidly after ingestion, the child need not be given any oral antidote in the ED. In fact, emesis should not be induced because it only exposes the esophagus to the caustic substance again.

Caustic substances may burn the larynx when ingested and can cause rapidly progressive edema and respiratory distress. Orotracheal intubation for acute airway management should be performed in the ED if necessary. A tracheotomy should be performed as soon as possible after the intubation to minimize the possibility of laryngotracheal stenosis.

Foreign Bodies

Foreign bodies in the oral cavity and pharynx are uncommon because of the child's protective reflexes. The tongue is sensitive and can detect sharp foreign objects that are then spat out. The gag reflex often expels foreign material from the pharynx, but sharp objects, such as fish bones, pins, and pieces of plastic, may get stuck in the oral mucosa, tonsils, or pharynx (Fig. 112.2). If a foreign body is visible and the patient is cooperative, the object may be removed in the ED with a clamp or forceps.

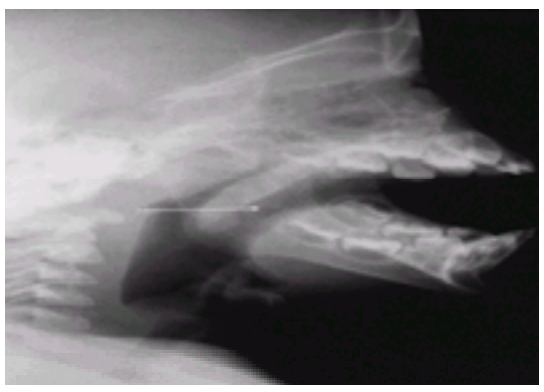

FIGURE 112.2. Lateral neck radiograph of a straight pin lodged in posterior pharyngeal wall.

Objects of all types may lodge in the hypopharynx or esophagus. Esophageal foreign bodies generally lodge at the areas of natural narrowing of the esophagus. The most common sites are the cricopharyngeal area, thoracic inlet, arch of the aorta, and gastroesophageal junction. If the child can breathe and talk, no attempt should be made to remove the object in the ED. The safest method of removal is endoscopically, using a general anesthetic. If the child is gagging and unable to breathe, the Heimlich maneuver should be used (see Chapter 1). If this method is not effective, emergency intubation or tracheotomy may be required to bypass the obstructing object.

Lateral neck and chest radiographs reveal radiopaque hypopharyngeal and esophageal foreign bodies. Plastic and other nonradiopaque objects cause the same foreign body sensation (something stuck in the throat) but are not visible on radiograph). Young children often have dysphagia and drooling because of painful swallowing.

Although ingestion of a foreign body usually causes gagging and choking that last for several seconds and subsequently subside, the initial episode may take place unobserved by an adult. Thus, unexplained dysphagia or drooling should initiate a search for a possible foreign body.

If a child presents to the ED with a history of swallowing an object, such as a toy or a fish bone, and complains of a foreign body sensation, a careful examination of the oral cavity and hypopharynx must be performed. If no foreign body is seen, plain radiographs of the neck should be obtained. Barium esophagrams are rarely helpful in pinpointing sharp foreign bodies in the esophagus but may be useful to confirm esophageal obstruction from an impacted foreign body such as a bolus of food. Foreign bodies easily seen in the oral cavity may be removed by the emergency physician. Consultation with an otolaryngologist (or similarly skilled specialist) should be obtained if a foreign body is detected in the pharynx or esophagus because removal usually requires endoscopic examination under anesthesia.

If the physical examination and radiographs fail to detect a foreign body, management is determined by the child's symptoms. If the child is having significant pain, the otolaryngologist should be consulted to perform esophagoscopy in the operating room. If the pain is mild, the child can swallow his or own saliva, and no evidence of respiratory distress is present, the foreign body sensation may be secondary to a mucosal scratch from a foreign body that has passed into the

stomach. In that instance, it may be appropriate to send the child home to return the next day if the sensation persists, which would indicate the presence of a persistent foreign body and require endoscopic removal.

LARYNX AND TRACHEA

Trauma

Laryngeal trauma can occur in a variety of ways. Blunt or penetrating injuries of the larynx can result in mucosal lacerations, laryngeal hematomas, vocal cord paralysis, or fractures of the thyroid and cricoid cartilages. Proper treatment requires prompt recognition of the presence and nature of a laryngeal injury and protection of the airway. Patients with laryngeal trauma present with varying degrees of neck pain, hoarseness, hemoptysis, and airway obstruction. Physical examination of a child with blunt trauma can reveal anterior neck tenderness, crepitance, and absence of the normal prominence of the thyroid cartilage or “Adam's apple” (Fig. 112.3). The otolaryngologist (or similarly skilled specialist) may be needed to perform an indirect examination of the larynx on the child with a suspected laryngeal injury. A direct laryngoscopy may be required when the child is in respiratory distress. The otolaryngologist should be prepared to intervene with intubation, tracheostomy, and/or surgical exploration of these laryngeal injuries.

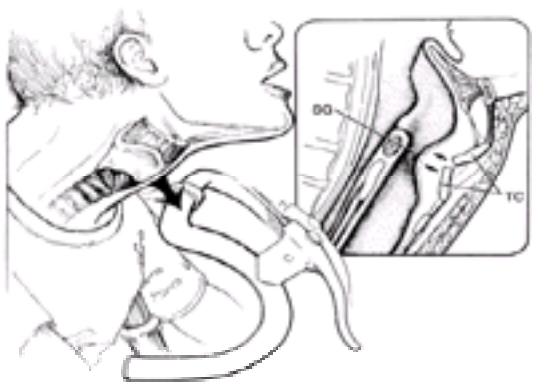

FIGURE 112.3. Loss of thyroid cartilage prominence and associated acute airway obstruction secondary to laryngeal fracture. *SG,* narrowed subglottic space; *TC,* fracture of thyroid cartilage.

Ingestion of caustic substances can cause severe burns of the larynx and pharynx; airway obstruction may occur secondary to the edema related to this injury. Laryngeal burns should be suspected in the child who has hoarseness or stridor after caustic ingestion. The child should be hospitalized, and otolaryngologic consultation should be obtained. If signs of respiratory distress (e.g., tachypnea, stridor) occur, the child should be taken to the operating room, where endoscopy can be performed, and an artificial airway, usually a tracheostomy, can be placed.

Foreign Bodies

Foreign bodies may become trapped in the laryngeal inlet, causing acute upper airway obstruction. The child usually presents with severe coughing, hoarseness, and significant respiratory distress. If the child is able to phonate, air is moving through his or her larynx, indicating only partial obstruction. “Back blows” or the Heimlich maneuver should not be performed in these children because this action may cause the foreign body to lodge more firmly in the larynx and convert a partial obstruction into a complete one. The child should be taken immediately to the operating room, where the otolaryngologist (or similarly skilled specialist) can perform the direct laryngoscopy necessary to remove the foreign body. In contrast, if the child is unable to speak, the foreign body may be causing total obstruction. In this case, back blows or the Heimlich maneuver may be lifesaving. Care must be taken in performing the Heimlich maneuver in young children because of the potential hazard of liver laceration. Emergency laryngoscopy, intubation, or tracheostomy is rarely required, and only if the previously described maneuvers are unsuccessful.

Foreign bodies that pass the larynx to lodge in the trachea or proximal bronchi can present problems in diagnosis and management. A history of coughing or choking on food (e.g., a peanut, raw carrot) or a toy is usually obtained. The child is often in no acute distress but may demonstrate a mild cough and/or wheezing. Inspiratory and expiratory stridor are characteristic of tracheal foreign bodies. Unilateral wheezes and decreased, or even absent, breath sounds are often seen with unilateral bronchial obstruction. Because most of the foreign bodies are radiolucent, they are not identifiable on radiographs. However, a radiographic difference in aeration of the lungs often helps detect the presence and identify the site of bronchial obstruction. Volume decrease, atelectasis, and infiltrate on the involved side may be seen on plain chest films if the bronchus is completely occluded by the foreign body. Hyperaeration (air trapping) secondary to a ball-valve effect of a foreign body that is partially blocking the bronchus is best seen by comparing inspiration and expiration films (Fig. 112.4). If the child will not cooperate to obtain these views, right and left lateral decubitus films can often demonstrate the same phenomena. Although differentiating hyperaeration and contralateral volume loss from atelectasis and compensatory contralateral lung expansion may help predict the location of a possible foreign body, this distinction is not as important as recognizing that any radiographic asymmetry signals a possible foreign body and requires endoscopy. However, a normal chest radiograph does not rule out the possibility of a foreign body. If a foreign body is suspected (by history or clinical examination), the child should be admitted and otolaryngologic consultation should be obtained to perform the endoscopy necessary for the prompt and safe removal of the object, if present.

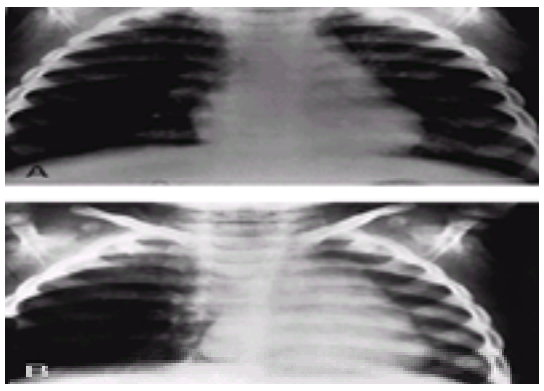

FIGURE 112.4. Chest radiograph of child with bronchial foreign body. **A.** Inspiratory film demonstrates only subtle hyperaeration of right lung. **B.** Expiratory film shows accentuated hyperaeration on the right side secondary to air trapping (“ball-valve” phenomenon) by the foreign body in the right mainstem bronchus. In addition, the mediastinum is displaced to the left.

Suggested Readings

Baker M. Foreign bodies of the ears and nose in childhood. Pediatr Emerg Care 1987;3:67–70.

Bressler K, Shelton C. Ear foreign-body removal: a review of 98 consecutive cases. Laryngoscope 1993;103(4 Pt 1):367–370.

Burstein F, Cohen S, Hudgins R, Boydston W. Frontal basilar trauma: classification and treatment. Plast Reconstr Surg 1997;99(5):1314–1321; discussion 1322–1323.

Canty PA, Berkowitz RG. Hematoma and abscess of the nasal septum in children. Arch Otolaryngol Head Neck Surg 1996;122(12):1373–1376.

Darrow DH, Holinger LD. Aerodigestive tract foreign bodies in the older child and adolescent. Ann Otol Rhinol Laryngol 1996;105(4):267–271.

East CA, O'Donaghue G. Acute nasal trauma in children: causes, diagnosis and treatment. Ear Nose Throat J 1989;68:522–538.

Handler SD. Maxillo-facial injuries. In: Torg J, ed. Athletic Injuries to the Head, Neck and Face. 2nd ed. Philadelphia: Lea & Febiger, 1990.

Handler SD, Wetmore RF. Otolaryngologic injuries. Clin Sports Med 1982;1:431–477.

Hawkins DB. Removal of blunt foreign bodies from the esophagus. Ann Otol Rhinol Laryngol 1990;99:935–940.

Healy GB. Management of tracheobronchial foreign bodies in children: an update. Ann Otol Rhinol Laryngol 1990;99:889–891.

Hester TO, Campbell JP. Diagnosis and management of nasal trauma for primary care physicians. J Kentucky Med Assoc 1997;95(9):386–392.

Kadish HA, Corneli HM. Removal of nasal foreign bodies in the pediatric population. Am J Emerg Med 1997;15(1):54–56.

Kadish HA, Schunk JE. Pediatric basilar skull fracture: do children with normal neurologic findings and no intracranial injury require hospitalization? Ann Emerg Med 1995;26(1):37–41.

Lima JA. Laryngeal foreign bodies in children: a persistent, life-threatening problem. Laryngoscope 1989;99:415–420.

Liu-Shindo M, Hawkins DB. Basilar skull fractures in children. Int J Pediatr Otorhinolaryngol 1989;17:109–117.

Northern JL, Downs MS. Hearing in Children. 4th ed. Baltimore: Williams & Wilkins, 1991.

Potsic WP. Management of trauma of the external ear. In: English G, ed. Otolaryngology. New York: Harper & Row, 1990.

Potsic WP, Handler SD, Wetmore RF, Pasquariello P. Primary Care Pediatric Otolaryngology. 2nd ed. Andover, NJ: J Michael Ryan, 1995.

Reilly JS, Cook SP, Stool D, Rider G. Prevention and management of aerodigestive foreign body injuries in childhood. Pediatr Clin N Amer 1996;43(6):1403–1411.

Rosenfeld RM, Sandhu S. Injury prevention counseling opportunities in pediatric otolaryngology. Arch Otolaryngol Head Neck Surg 1996;122(6):609–611.

Singh B, Kantu M, Har-El G, Lucente FE. Complications associated with 327 foreign bodies of the pharynx, larynx, and esophagus. Ann Otol Rhinol Laryngol 1997;106(4):301–304.

Tong MC, Ying SY, van Hasselt CA. Nasal foreign bodies in children. Int J Pediatr Otorhinolaryngol 1996;35(3):207–211.

CHAPTER 113

Dental Trauma

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ASSESSMENT OF TRAUMATIC DENTAL EMERGENCIES

The care of pediatric patients with oral and maxillofacial and dental trauma should follow the basic tenets of emergency medicine. An initial general assessment includes evaluation of airway patency, breathing adequacy, and cardiovascular status (ABCs). Control of bleeding, assessment of the degree of shock, evaluation of neurologic status, and notation of other injuries must all be done sequentially.

Life Support

The most common cause of airway obstruction in a child with facial injuries is the accumulation of blood in the oral cavity and pharynx. Unconscious children may be unable to clear their airway by coughing or swallowing. A tooth aspirated by a child can block the airway. A fractured mandible may cause the tongue to fall posteriorly and create obstruction. The mouth should be gently suctioned clean. If the tongue of an unconscious child is causing obstruction, the mandible can be pulled forward by pressure at the angles or an oropharyngeal or nasopharyngeal airway can be placed. Chapter 1 details the procedures for establishing a patent airway. If endotracheal intubation fails, a surgical airway must be obtained with either a cricothyrotomy or tracheostomy.

The soft tissues and bones of the lower and midface are well vascularized and bleed profusely when injured. Hemorrhage is best controlled by ligating any vessels that are easily seen. However, vessels of the face often retract when severed, and if a vessel cannot be seen, direct pressure should be applied to control bleeding. With extensive blood loss, the patient should be assessed for signs of shock (see Chapter 3).

History

A thorough history and physical examination are paramount to any treatment considerations. Traumatic orofacial injuries can be dramatic, making a history difficult to obtain. Informants other than the patient may have to be questioned. The practitioner should always be alert to the possibility of “nonaccidental” trauma (i.e., child abuse) if the history is not consistent with the observed injury. One key question of significance is the immunization history. Many traumatic facial injuries occur with concomitant soft-tissue injuries, and the need for tetanus prophylaxis is based on the history of immunization. Antibiotic prophylaxis may be indicated before treatment in the child with congenital heart defects to prevent subacute bacterial endocarditis (see Table 82.25) and in children who have certain hematologic, oncologic, or endocrine disorders (e.g., sickle cell disease, leukemia, diabetes). The history should also give the physician an indication of the preinjury and postinjury neurologic status. A thorough neurologic assessment must be made as early as clinically possible because the risk of neurologic injury is high in all head and neck trauma.

Physical Examination

Children with facial injuries are usually frightened and apprehensive. Hence, the initial contact with the child should be authoritative and reassuring. The examination should be organized to include inspection, palpation, percussion, and auscultation of both extraoral and intraoral aspects.

Extraoral Examination

Inspection

The extraoral examination should start with inspection. The clinician should note the symmetry of the face in the anterior

view as well as in profile. A loss of symmetry is often associated with dental infection or swelling from trauma. The child should be inspected for any nasal or orbital malalignments. The clinician should carefully note the location and nature of any swollen or depressed structures and the color and quality of the skin and should look for lacerations, hematomas, ecchymoses, foreign bodies, or ulcerations. The child should be asked to open and close his or her mouth while facing the clinician to see whether the mandible deviates during function. If the child is unable to open or close his or her mouth because of pain, the action should not be forced because it may increase the extent of injury. The clinician should inspect for lip competency (the ability of the lips to cover the teeth) because loss of competency may indicate displacement of the teeth from trauma.

Palpation

Gentle digital palpation bilaterally, to feel the temporomandibular joints (TMJs), should be the next point of the examination. The clinician should feel the TMJs as the child opens and closes his or her mouth. There should be equal movement on both sides without major deviations. The clinician should palpate to the orbital rim, checking to be sure it is continuous and intact all the way to the inner canthus of the eye. The clinician should then move across the zygoma to the nose and palpate for any crepitus or mobility and move back to the preauricular region and along the posterior border of the mandible, palpating the ramus. The clinician should then move anteriorly from the angles to the symphysis, palpating for any discontinuity, mobility, swellings, or point tenderness. Finally, the clinician should palpate the major lymph nodes of the neck and note any paresthesia or hypoesthesia (numbness) of the lips, nose, and cheek, which may indicate a fracture. Figure 113.1 shows the main nerve supply to facial structures.

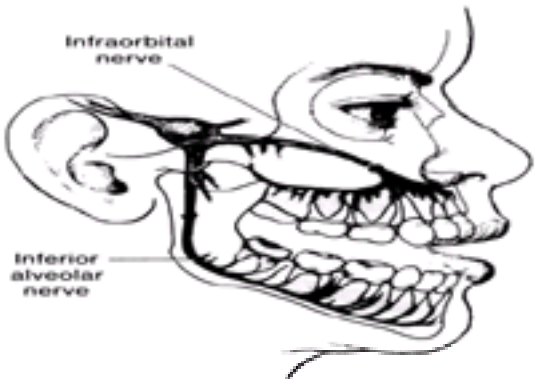

FIGURE 113.1. Infraorbital and inferior alveolar main nerve supplies the teeth.

Auscultation

The clinician should listen for crepitus as the child opens and closes his/her mouth, at the same time palpating the TMJ. Crepitus may indicate TMJ pathology.

Intraoral Examination

Inspection

A good light is essential for the intraoral examination. The clinician should inspect the color and quality of the lips, gingiva (gums), mucosa, floor of the mouth, tongue, and palate. The gingiva should be pink, firm, and stippled (like a grapefruit skin). The mucosa should be pink, moist, and glassy in appearance. The floor of the mouth should be flat and well vascularized. The clinician should inspect the mouth for any swellings of soft tissue that may be bluish as a result of vascularization and/or hematoma. Hematomas or mucosal ecchymoses in the floor of the mouth or vestibular area are highly suggestive of mandibular fractures. The clinician should note any inflamed, ulcerated, or hemorrhagic areas, as well as any foreign bodies or denuded areas of bone. Traumatically displaced teeth often produce the complaint that the child's teeth do not fit together when he or she bites on the back teeth. However, a similar complaint may be expressed by a child when a primary tooth is mobile and about to exfoliate. Figure 113.2 shows key eruption times for primary and secondary teeth. Exfoliation of primary teeth is often confused with traumatic injuries. The child should be inspected for any chipped or missing teeth. If a tooth is chipped or missing, the clinician should check for any fragments of teeth or foreign bodies in adjacent soft tissues. If the child's teeth are missing yet no bloody socket is present, the eruption/exfoliation timetables (Table 113.1A and Table 113.1b) can be helpful in determining whether the loss is normal.

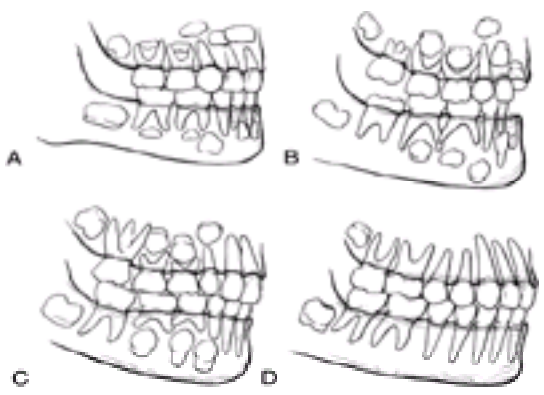

FIGURE 113.2. A. At age 3, 20 primary teeth should be erupted. The permanent teeth are in various stages of calcification underneath and behind the primary teeth. **B.** At age 6, 16 primary teeth should be present. The permanent

6-year molars should be erupting distal to the last primary molars. The permanent anterior central incisors should be erupting. **C.** At age 9, 8 to 12 primary teeth should still be present. The permanent 6-year molars should be totally erupted and in occlusion. The permanent anterior central and lateral incisors are totally erupted and are most prone to fracture at this time. **D.** At age 12, 28 permanent teeth should be present. Only teeth not present should be third molars (wisdom teeth).

	Maxillary	Mandible
Primary*	Mean Age (mo)	Mean Age (mo)
Central incisor	10 (8–12)	8 (6–10)
Lateral incisor	11 (9–13)	13 (10–16)
Canine	19 (16–22)	20 (17–23)
First molar	16 (13–19 boys) (14–19 girls)	16 (14–18)
Second molar	29 (25–33)	27 (23–31)
Permanent†	**Mean Age (yr)**	**Mean Age (yr)**
Central incisor	7–7.5	6–6.5
Lateral incisor	8–8.5	7.2–7.7
Canine	11–11.6	9.7–10.2
First premolar	10–10.3	10–10.7
Second premolar	10.2–11.2	10.7–11.4
First molar	6–6.3	6–6.2
Second molar	12.2–12.7	11.7–12.0
Third molar	20.1	20–20.5

*Mean age in months ±1 standard deviation. Reprinted with permission from Lunt RC, Law DB. J Am Dent Assoc 1974;89:872.
†Reprinted with permission from Gaudi AR. The development and eruption of the human dentition. In Pomeroy DJ, Wagoner ML, Fleming JI, eds. Pediatric Dental Medicine. Philadelphia: Lea & Febiger, 1961.

Table 113.1A. Chronology of Eruption of the Primary and Permanent Dentition

			Mean Age* (yr, mo)	
Rank	Mandibular Arch	Maxillary Arch	Boys	Girls
First	Central incisors		6.0	5.7
Second		Central incisors	6.10	6.7
Third	Lateral incisors		7.2	6.10
Fourth		Lateral incisors	7.10	7.5
Fifth	Canines		10.5	9.7
Sixth	First molars		10.8	10.2
Seventh		First molars	10.11	10.6
Eighth		Canines	11.3	10.7
Ninth	Second molars	Second molars	11.9	11.5

Reprinted with permission from Ripa LW, Leske GS, Sposanto AL, et al. Chronology and sequence of exfoliation of primary teeth. J Am Dent Assoc 1982; 105:641.
*Ages are for right side of mouth; however, exfoliation is generally bilaterally symmetric.

Table 113.1B. Sequence of Primary Tooth Exfoliation

Palpation

Using the thumb and index finger, the clinician should palpate the alveolar ridges, evaluating for any swellings, discontinuity, or mobility of the soft tissues. This procedure should be done circumferentially in all four quadrants. The palate should be palpated for any swellings or point tenderness. The masseter muscle should be palpated with fingers placed intraorally and extraorally and rolling the muscle between the two. Using a gauze pad, the clinician should hold the tongue and lift it gently to better view and examine its dorsal, ventral, and lateral surfaces. The lips should be palpated for any swellings or nodules. The quality of the swellings should be noted (i.e., fluctuance versus induration). The teeth should be palpated for mobility, tenderness, and fracture. The gums should be palpated for any erupting teeth. Finally, the clinician should look for any purulent exudate during palpation.

Percussion

The teeth should be percussed individually with the end of a mouth mirror handle or tongue depressor. Mobile, abscessed, vertically fractured, or traumatized teeth may be hypersensitive and often sound dull on percussion.

Radiographs are a valuable supplement to the clinical examination. However, obtaining diagnostically perfect radiographic surveys in a child with acute orofacial/dental injuries may be difficult. Table 113.2 indicates the radiographic view that would be the preferred diagnostic aid for these injuries.

Radiographic View	Diagnostic Aid for
Right and left lateral oblique	Fractured body and ramus of mandible
Anteroposterior view of mandible	Fracture of mandibular condyles and symphysis
Towne's	Fractured condyles
Waters'	Maxillary fractures
Intraoral radiographs	
Panoramic	Maxillary and mandibular fractures and related pathology
Occlusal	
Periapical/bite wing	Tooth fractures and pathology; alveolar fractures and pathology

Table 113.2. Radiographic Diagnostic Aids

OROFACIAL/DENTAL TRAUMA

Dental trauma occurs in a variety of forms that can be confusing to the primary care provider. The emergency physician needs to know which injuries can be managed without dental consultation, which need follow-up care, and which need emergency dental care. Table 113.3 shows the types of chief complaints associated with pediatric dental emergencies at Children's Hospital of Boston from 1989 to 1990. Trauma represented 21.7% of the chief complaints and 29.2% of the diagnoses. Several factors, including age, occlusion, and agility in sports, predispose pediatric patients to orofacial trauma. Traumatic dental injuries occur at the following peak ages: 1) as the toddler becomes ambulatory (ages 1 to 3), 2) as the child enters school (ages 7 to 10), and 3) in the older adolescent (ages 16 to 18) engaged in athletic activities. In addition, proclined or prominent maxillary anterior teeth are usually more susceptible to being displaced or fractured in orofacial injuries. Use of a preformed or custom-made mouthguard in children with prominent maxillary incisors is helpful in reducing the chance of these accident-prone teeth sustaining traumatic injuries. This section details the management of dental injuries that are most commonly seen in the pediatric emergency patient.

Type of Emergency	Chief Complaint (% of Total)	Diagnosis (% of Total)	Referral Status
Oral pain/infection	46.1	39.0	Stat
Trauma	21.7	29.2	Stat
Caries	3.4	9.4	Next day
Soft-tissue trauma	0.9	2.8	Stat
Erupting/exfoliating teeth	8.2	5.5	Next day
Other	19.7	14.1	N/A[b]

[a]Data courtesy of Corine Barone Cognata D.M.D. Thesis, September 1991.
[b]Not applicable.

Table 113.3. Dental Emergencies at Children's Hospital Boston, 1989–1990 [a]

Soft-Tissue Lacerations

Management of injuries to the soft tissues of the oral cavity follow the same emergency care principles used for other soft tissue injuries. Lip injuries swell alarmingly even after minor trauma. Lacerations of the tongue and frenum bleed profusely because of the richness of their vascularity. However, as with lip injuries, ligating specific vessels is usually unnecessary because the bleeding normally stops with direct pressure and careful suturing. Because they heal well spontaneously, frenum lacerations usually do not require suturing, especially in young children. The injured area should be thoroughly examined for foreign bodies. To confirm the presence of a foreign body in a deep laceration of the tongue or lip, a radiograph should be obtained before suturing. If chipped teeth are present, radiograph examination becomes even more important. The decision to primarily repair a laceration of the oral cavity that is more than a few hours old depends on the relative risk of secondary infection (see Chapter 124).

Suturing of the lip must be done carefully to achieve a precise approximation of the edges of the vermilion border to avoid a disfiguring scar even with superficial lip lacerations. If necessary, the lip must be sparingly debrided and the skin closed with 5-0 or 6-0 nylon sutures. Deep lip lacerations are sutured with 4-0 chromic and then with 5-0 or 6-0 nylon for the skin and vermilion. If the lip laceration is through and through, debridement may be necessary. In children less than 5 years of age, 4-0 chromic on the deeper mucosal aspects of the lip and 6-0 chromic on the superficial aspects are preferred. In children older than 5 years of age, 4-0 chromic is used on the deeper mucosal aspects of the lip and 5-0 or 6-0 nylon on the superficial edges. Most superficial tongue lacerations heal without suturing; however, deep lacerations or those that create a flap need to be sutured. When necessary, tongue lacerations are usually sutured with 4-0 chromic if superficial and 3-0 chromic in deeper wounds. With tongue lacerations, it is important to consider the excessive muscular movements that pull at the sutures; therefore, tongue sutures should be made deep into the musculature. Most superficial mucosal injuries are sutured with 4-0 chromic. If the mucosal laceration is deep, 3-0 chromic can be used.

Injuries to the Teeth

Traumatic dental injuries can be categorized into two groups: 1) injuries to the teeth—hard dental tissues and pulp; and 2) injuries to the periodontal structures—periodontal ligaments and alveolar bones. Figure 113.3A indicates the relative positions of these structures. Andreason further categorized traumatic fractures of the teeth into complicated and uncomplicated fractures.

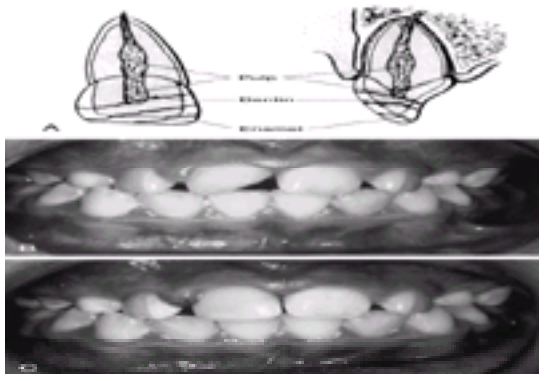

FIGURE 113.3. A. The anatomy of a tooth should be considered during a traumatic injury: enamel fracture, no emergency treatment; dentin fracture, emergency treatment as soon as convenient; and pulpal fracture, emergency treatment as soon as possible. **B.** Enamel and dentin fracture of a permanent incisor. **C.** The same child after bonded restoration.

Injuries to Hard Dental Tissues and Pulp

Uncomplicated tooth fractures are confined to the hard dental tissue (enamel, dentin, cementum). Clinically, there may be a jagged edge of tooth. The fracture line may appear deep, but no sign of bleeding from the central core (pulp) of the tooth is apparent. The child may complain of sensitivity, especially to cold air and fluids. Emergency treatment is aimed at protecting the pulp, even if no frank pulp exposure is noted. The dentist should be called as soon as possible to place a dressing of calcium hydroxide or glass ionomer over the exposed dentin for thermal and chemical insulation and for prevention of (pulpal) necrosis. A temporary restoration is then placed to prevent the insulation material from dissolving. At some later date, an aesthetic resin acid-etched restoration can be placed. The prognosis for uncomplicated tooth fractures is usually good.

A complicated tooth fracture involves the pulp of the tooth. Often, bleeding is noted from the central core of the tooth. To best preserve the viability of that tooth, dental pulpal treatment must be initiated immediately. Prognosis depends on the size of the exposure (less than 1 mm carry the best prognosis) and the time interval between the trauma and therapy (less than 24 hours carries the best prognosis). Thus, calling the dental consultant as soon as possible to institute pulpal therapy is important. Root fractures generally are seen after the tooth has reached full root formation, which is approximately 2 to 3 years after eruption begins (Table 113.1). These fractures usually involve maxillary anterior teeth. The child may clinically have a displaced crown in which the tooth seems to be mobile and extruding from the socket. Definitive diagnosis depends on intraoral dental radiographs; therefore, immediate dental consultation is necessary. Treatment often involves reduction with splinting and pulpal therapy of the involved teeth.

In any injury resulting in fragmentation of teeth, the emergency physician should attempt to account for all of the fragments. Soft-tissue lacerations, especially of the lower lip and tongue, should be evaluated clinically and, if necessary, radiographically to rule out embedded tooth fragments. Infection and poor wound healing are the sequelae of such an oversight.

Displaced Teeth

The tooth is held in the socket by slender elastic and collagen fibers collectively known as the periodontal ligament. These fine, slender fibers are easily injured or broken with trauma to the teeth. Clinically, the physician may note either an increase or decrease in mobility depending on the extent of the cortical plate fracture and/or displacement of the affected teeth. If asked, the child will be able to point to an injured tooth because of the tooth's heightened sensitivity. Periodontal injuries may be further subdivided into five clinical types: 1) concussion, 2) subluxation, 3) intrusion, 4) extrusion/lateral luxation, and 5) avulsion, as noted in Figure 113.4.

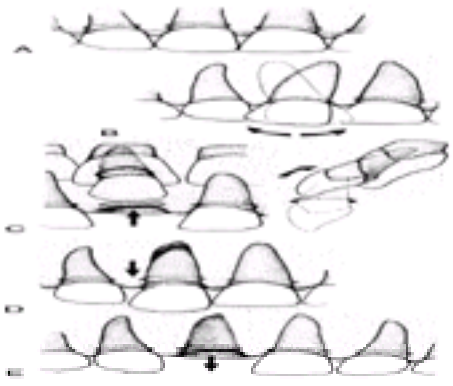

FIGURE 113.4. The various types of trauma to the periodontal structures. Concussion **(A);** subluxation **(B);** intrusion (if primary tooth is intruded note location of developing permanent tooth bud) **(C);** extrusion **(D);** and avulsion **(E).** Refer emergencies **B** through **E** to the dental staff as soon as possible.

Concussion is usually caused by minor damage to the periodontal ligaments, resulting in slight edema. Teeth sustaining lateral displacement injuries exhibit no displacement or excessive mobility. They are often percussion-sensitive when tapped with the blunt end of a metal instrument such as an intraoral mirror. No emergency treatment is indicated for such

injuries, although baseline radiographs are taken to rule out more serious dental injuries. Therefore, a dental consultation should be arranged. The prognosis for concussion injuries is good, although pulpal necrosis is possible over time.

Subluxation is usually more damaging to the periodontal ligaments because of increased edema. There is excessive mobility in the horizontal and/or vertical direction but no displacement within the dental arch. The tooth is often sensitive to percussion. The child will complain that his or her teeth feel like they do not meet when biting down. Because subluxated teeth, especially in the permanent dentition, may require immobilization with a bonded resin splint, this type of injury should be referred to the dental service as soon as possible.

Intrusion, although more commonly seen in the primary dentition, can be seen in the permanent dentition with high velocity or high force injuries. Intruded teeth are teeth displaced directly into the socket. Intruded teeth may not be visible and, thus, give the false appearance of being avulsed. To confirm intrusion and to rule out avulsion, an intraoral dental radiograph is indicated. An intruded primary tooth must be evaluated for its proximity to the developing permanent tooth. Again, immediate dental consultation and treatment are necessary. In addition, compression fractures of the alveolar socket and anterior nasal spine may be seen radiographically. The prognosis for intruded teeth is poor because of pulpal compression and severance, which occurs on impact. Treatment of intruded primary teeth can include extraction, orthodontic repositioning, or allowing for spontaneous reeruption. Intrusive injuries in the permanent dentition usually require repositioning and splinting. Pulpal treatment (endodontics) is almost always needed because the pulp is rendered nonvital as a result of trauma, which can cause root resorption and periapical infection.

Extrusion/lateral luxation is manifested clinically as displacement of the tooth from the alveolar socket in various directions. Usually, the anterior maxillary tooth is extruded and displaced lingually, causing a fracture of the labial cortical plate of the alveolar socket. Luxated permanent teeth must be realigned and immobilized with a splint as soon as possible. Endodontic treatment is often needed in the long term. Extrusive/lateral luxations of the primary dentition usually necessitate extraction to avoid potential injury to the permanent tooth bud during realignment or as a result of eventual pulpal necrosis.

Avulsion is the term used to describe a tooth that has been completely displaced from its socket. Radiographs may show the tooth to be actually intruded, ingested, or aspirated. The best prognosis exists if therapy is instituted within 30 to 60 minutes of the avulsion. The emergency physician or the parent should (as seen in Fig. 113.5): 1) find the tooth; 2) determine whether it is a primary tooth by checking the child's age and the table of tooth eruption (if it is a primary tooth, do not reimplant); 3) if it is a permanent tooth, gently rinse the tooth under running water or saline, taking care to hold the crown of the tooth and not the root (do not scrub the crown or root); 4) insert the tooth into the socket in its normal position (do not be concerned if it extrudes slightly).

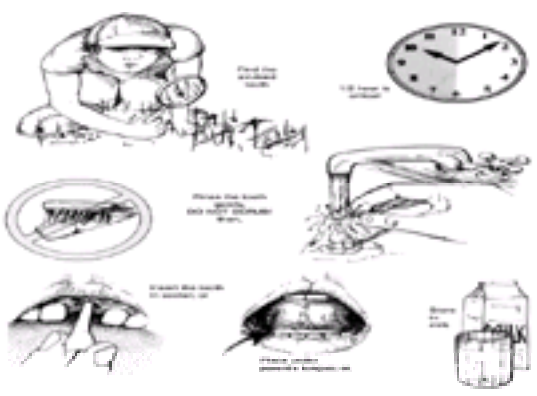

FIGURE 113.5. If a child loses or avulses a tooth. 1. Find the tooth. 2. Determine whether it is a primary or permanent tooth by checking Table 113.1. If it is a primary tooth, DO NOT REIMPLANT. 3. Gently wash but do not scrub the tooth. 4. Insert the tooth back into the socket or place in milk and take immediately to the dentist. Remember, you have only 30 minutes to preserve the vitality of the tooth.

If on-site reimplantation is impossible, the optimal storage to preserve the vitality of the periodontal ligament of the root surface is a cell culture medium such as Viaspan or Hank's balanced salt solution. A commercial product such as the 3M Save-a-Tooth Emergency Tooth Preserving System (Smart Practice, Phoenix, AZ) containing Hank's is available to place the tooth into during transportation to the dental office. If none of these products are available, milk is an excellent alternative transport medium. Although saliva or saline are not ideal, they are alternative mediums that are preferred over allowing the root surface to air dry. The patient should go directly to the dentist for immobilization (splint). Dental follow-up is mandatory to prevent resorption of the root. Prophylactic pulpal therapy (endodontics) helps improve the prognosis by limiting pulpal necrosis and thus root resorption. Avulsed primary teeth are generally not reimplanted because of the close proximity of the permanent tooth and possible negative effects on development of this tooth.

Orthodontic Trauma

Orthodontic trauma often results from loose wires or ligatures that are attached to orthodontic brackets or bands. These emergencies should be seen by the dental service as soon as possible to alleviate any discomfort and soft-tissue trauma. If dental service is unavailable, the physician can bend or cut the wire away from the soft tissues with a hemostat. Softened wax can be molded over the loose wire as a temporary method or to allow the traumatized soft tissues to heal. If no discomfort is noted and no loose foreign bodies are present, the definitive treatment often can be delayed until a more convenient time.

Mandibular Fractures/Dislocations

The incidence of facial fractures in children is low. However, the most common facial bones fractured in children are the nasal bones, followed by the mandible. The emergency physician must therefore be knowledgeable in the diagnosis of mandibular fractures. Physical examination of the head and neck and obtaining radiographs should confirm the diagnosis of mandibular fracture.

The mandible can be compared to an archery bow, which is strongest at its center and weakest at its ends. Thus, most fractures occur at the neck of the condyles. Other areas of the jaw that are predisposed to fracture include the angle of the mandible where deep impacted teeth or unerupted 6-year molars make the mandible more vulnerable. The clinician should examine the teeth for any changes in occlusion and any raised or depressed fragments. Areas of bleeding, gingival/mucosal tears, or sublingual ecchymosis are also clues. Pain when opening the mouth, especially if the child is unable to open it fully, often indicates mandibular fracture. A unilateral condylar fracture should be suspected if the mandible deviates toward the affected side on opening.

Radiographic views detailed in Table 113.2 should be obtained. In the pediatric patient, a mandibular fracture generally necessitates hospital admission. The appropriate consulting service should be called to stabilize the fracture, using either open or closed reduction.

Mandibular dislocation occurs when the capsule and TMJ ligaments are sufficiently stretched to allow the condyle to move to a point anterior to the articular eminence during opening. Dislocation can be unilateral or bilateral and often accompanies a history of extreme opening of the mouth (e.g., deep yawn) or after a long dental treatment session. The muscles of mastication then enter a tonic contraction state, and the patient is unable to move the condyle back into the glenoid fossa and is unable to close the mouth. Treatment consists of reassuring the patient and gently massaging the muscles of mastication. If this approach fails, intravenous diazepam (0.2 mg/kg, maximum 10 mg) can be administered as an adjunct. When relaxation is achieved in the muscles of mastication, gentle downward and backward pressure should be applied by the physician's thumb (wrapped in gauze) on the occlusal surfaces of the posterior teeth (Fig. 113.6). The downward pressure moves the dislocated condyle below the articular eminence; subsequent backward pressure on the molars shifts the condyle posteriorly into the mandibular fossa. Figure 113.7 shows the anatomic landmarks and repositioning of the TMJ.

FIGURE 113.6. Position for reduction of a dislocated mandible.

FIGURE 113.7. Dislocation of the temporomandibular joint occurs when the mandibular condyle moves to a point anterior to the articular eminence during opening. Reduction is accomplished by pushing downward and backward on the occlusal surfaces of the posterior teeth.

Maxillary Fractures

Premaxillary or anterior maxillary alveolar bone fractures are a common finding associated with displacement or avulsion of maxillary anterior teeth. By gentle digital manipulation, the labial plate of bone can often be guided back into position under local anesthesia. (Infiltration with 2% lidocaine with epinephrine is commonly used.) Splinting the loose teeth and suturing the gingival tissue holds the bone fragments in place. After infiltration of local anesthesia and digital manipulation, the bone fragment can be held in place temporarily by aluminum foil (three thicknesses) molded over the teeth and alveolar ridge. This emergency splint should be held in place by having the child bite down. A dental consultant

should be contacted as soon as possible for fabrication of a more permanent dental splint. Figure 113.8 shows an acid-etch composite splint in place. Mandibular and other facial fractures are covered in greater detail in Chapter 110.

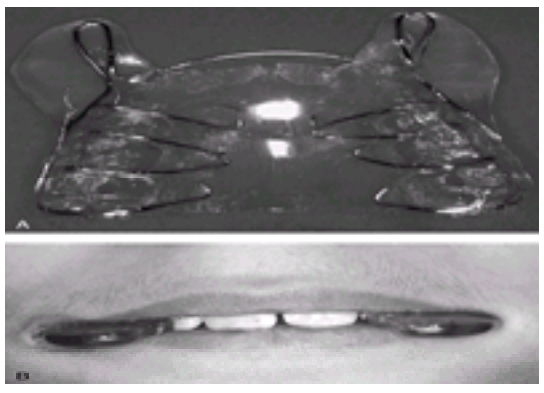

FIGURE 113.8. A. Removable maxillary intraoral acrylic appliance with extraoral commissure extensions. **B.** Appliance in place, separating the upper and lower lip with electrical burns at the commissure.

Electrical Burns

Electrical burns of the mouth occur when children bite on electrical cords. The saliva in the child's mouth acts as a conductor to complete the circuit. In the emergency department, the first consideration is the patient's respiratory status. Next, the patient should be assessed for the presence of shock or other injuries. Although the commissure of the mouth is most likely affected, the tongue, alveolar ridge, and floor of the mouth are occasionally involved. Most children with these injuries can be managed as outpatients. A bland, soft, cold diet is initially recommended. If refusal of food and dehydration are problems, the child requires admission to the hospital for intravenous fluids. Meticulous oral hygiene using a toothbrush with or without toothpaste must be performed three to four times a day, as well as hydrogen peroxide and water (1:1) rinses. With severe burns of the lips and mouth, severe arterial bleeding may continue 5 to 8 days after the burn occurs. The clinician should instruct the parent on the method for digitally compressing the labial artery or consider elective admission to the hospital for wound management. To prevent scarring down of the commissure, electrical burns of this area require the fabrication of an intraoral or extraoral device to separate the upper and lower segments during healing (see Fig. 113.8A and Fig. 113.8B).

Suggested Readings

Andraeson JO, Andraeson FM. Textbook and Color Atlas of Traumatic Injuries to the Teeth. 3rd ed. St Louis: Mosby, 1994.

Croll T, Brooks EB, Schut L, Laurent JP. Rapid neurologic assessment and initial management for the patient with traumatic dental injuries. J Am Dent Assoc 1980;100:530–534.

Kaban L. Pediatric Oral and Maxillofacial Surgery. Philadelphia: Saunders, 1990.

Kelly J, Seldin E. In: Donoff R, ed. MGH Manual of Emergency Medicine. Baltimore: Williams & Wilkins, 1992.

Krasner P. Modern treatment of avulsed teeth by emergency physicians. Am J Emerg Med 1994;12:241–246.

Schultz RC. Facial Injuries. 3rd ed. Chicago: Year Book, 1988.

CHAPTER 114

Burns

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BACKGROUND

Modern technology has increased the exposure of children to potentially injurious thermal energy in their environment. Physicians who care for children must be prepared to treat the many different types of thermal injury. Burns and related injuries are the third leading cause of death in childhood, killing approximately 2,500 children per year. Serious morbidity is three times more common than mortality, generating medical costs exceeding $1 billion per year. Competent management of minor burns can optimize cosmetic results and minimize functional morbidity for children, a benefit that is difficult to estimate in dollars.

Major advances in burn treatment have occurred during the past 40 years despite underfunding of burn-related medical research relative to diseases with far less impact on public health. The LB_{50} (percentage of body surface area [BSA] burn resulting in 50% mortality) has increased from 30% in the 1930s to approximately 70% today. State-of-the-art care of burned children, which has improved survival and reduced morbidity, starts with competent management in the emergency department (ED).

Only 3 to 5% of all burns in children are life-threatening. Most are minor scalds, accounting for about 80% of all thermal injuries. Flames produce 13% of burns and, with associated smoke inhalation, result in the majority of deaths. Electrical or chemical burns account for 2 to 3% of injuries and pose special challenges in management.

Males and children less than 5 years old are at highest risk of thermal injury. Bathing-related scalds are a particular risk during infancy. Hot liquid spills are common in toddlers. School-age children are often injured as a result of playing with matches, with high morbidity and mortality to the child and his or her family. Burns related to volatile agents and high-voltage electrical lines are seen primarily in teenagers.

Several preventive strategies can reduce the risk of thermal injury to children. Lowering the temperature of water heaters from over 130° (54.4°C) to 120°F (48.9°C) increases the time for full-thickness scalding from under 30 seconds to 10 minutes. Burn centers have noted a decrease in full-thickness flame burns since the introduction of flame-resistant children's sleepwear. Cigarette misuse is responsible for more than 30% of house fires. “Fire safe” cigarettes, which are less likely to ignite household materials, are technically feasible but are not yet being manufactured. Smoke detectors and sprinkler systems can greatly reduce deaths, but only if installed and maintained properly. Advances in burn prevention can have a far greater impact on public health than refinements in burn management.

PATHOLOGY AND PATHOPHYSIOLOGY

The vital functions of skin, which accounts for 15% of total body weight, are often unappreciated until it is severely injured. Skin preserves body fluids, efficiently regulates heat loss to the environment, and acts as a barrier to infectious pathogens. Skin is composed of an outer, mostly nonviable epidermis and dermis. The stratum corneum, a layer within the epidermis, prevents passive water loss and is lethal to most viruses and Gram-negative bacteria. It has a fatty acid film that is fungistatic and bacteriostatic. The dermal–epidermal junction prevents loss of macromolecules through the skin. Dermis contains eccrine sweat glands that actively secrete fluid to increase evaporative heat loss. The vasculature in the dermis can regulate radiant heat loss through vasodilation and vasoconstriction, with a 100-fold variation in skin perfusion. Therefore, children with extensive burns have difficulty retaining body fluids and regulating temperature (Fig. 114.1).

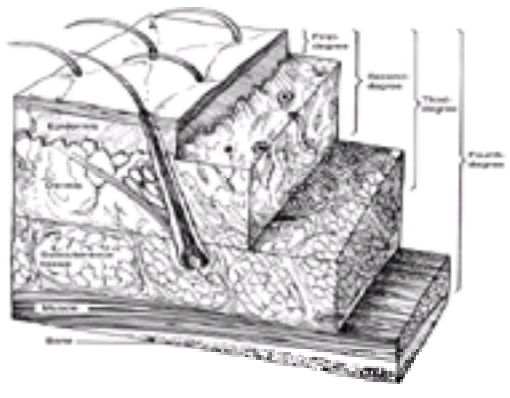

FIGURE 114.1. Degree of burn wound depth. First-degree involves only epidermis, second-degree extends into the dermis, third-degree into subcutaneous tissue, and fourth-degree to muscle, tendons, or bone.

Larger burns can have systemic effects. Capillary permeability in burned tissues is greatly increased. Burning releases osmotically active molecules to the interstitial space, further driving the extravasation of fluid. In patients with large burns, vasoactive mediators released from burned tissue result in systemic capillary leakage. Edema develops in both noninjured and burned tissue. Cardiac output is decreased by circulating factors that depress myocardial function. Acute hemolysis of up to 15% of red blood cells occurs from direct heat damage and a microangiopathic hemolytic process. The profound circulatory effects of severe burns can result in life-threatening shock early after injury.

Hair follicles, sweat glands, and sebaceous glands in the dermis play a crucial role in the healing of partial-thickness burns. After an inflammatory phase characterized by leukocyte infiltration, cytokine release, and complement activation, epithelial cells in these structures undergo metaplasia to produce the stratified squamous epithelial cells required for healing of skin. Neovascularization and fibroblast migration occur 1 to 3 weeks after partial-thickness burns. Overproduction of collagen can result in hypertrophic scarring. In full-thickness burns, the absence of intact dermal appendages precludes reepithelialization by this mechanism and necessitates skin grafting.

Thermal energy damages the skin structures in proportion to the intensity and duration of exposure. Hot grease or thick soup cause deeper injuries because they usually cling to the skin longer than scalding water. Ignition of synthetic fabrics often causes melting and adherence to the skin, resulting in more serious burns than ignition of cotton garments. Skin thickness is also a variable in the severity of injury with a given thermal exposure. Submersion of the hand may result in deep burns of the dorsum, with relative sparing of the thick skin of the palm. The thinner skin of young children accounts for deeper burns when compared with adults with similar heat exposures.

A first-degree burn is characterized by redness and a mild inflammatory response confined to the epidermis, without significant edema or vesiculation. First-degree burns are not included in the calculation of burn surface area used for therapeutic decisions. These minor burns are somewhat painful, healing in 3 to 5 days without scarring.

Most burns treated in EDs are partial-thickness or second-degree burns. Superficial second-degree burns involve destruction of the epidermis and less than half of the dermis. Blistering is often present. Increased capillary permeability, resulting from direct thermal injury and local mediator release, results in edema. These injuries are usually painful because intact sensory nerve receptors are exposed. The capillary network in the superficial dermis gives these burns a pink–red color and moist appearance. Healing occurs in about 2 weeks and scarring is usually minimal.

Deep partial-thickness burns involve destruction of epidermis and greater than 50% of the dermis. Edema can lessen the exposure of sensory nerve receptors, making some partial-thickness burns less painful and tender. Deep partial-thickness burns have a paler, drier appearance than superficial injuries. They are sometimes difficult to distinguish from areas of full-thickness injury. Thrombosed vessels often give the deep partial-thickness burn a speckled appearance. Burns evaluated immediately may appear to be partial-thickness and subsequently become full-thickness injuries, especially if secondary damage from infection, trauma, or hypoperfusion ensues. Deep partial-thickness burns can take many weeks to heal completely. Unacceptable scarring is not uncommon. Skin grafting is often necessary to optimize cosmetic results.

Full-thickness or third-degree burns involve destruction of the epidermis and all of the dermis. They usually have a pale or charred color and a leathery appearance. Destruction of the cutaneous nerves in the dermis make them nontender, although surrounding areas of partial-thickness burns may cause pain. Full-thickness burns cannot reepithelialize and can only heal from the periphery. Most require skin grafting. Fourth-degree burns are those full-thickness injuries that involve underlying fascia, muscle, or bone.

All burns become colonized by potentially pathogenic organisms. Most organisms are acquired from the skin and intestinal flora of the burned patient and not from exogenous sources. Heat causes coagulation necrosis of tissue, producing a protein-rich medium that nourishes bacterial growth. Removal of this material by cleansing and debridement reduces substrate for bacterial proliferation. Aggressive topical antimicrobial therapy, with or without systemic antibiotics, can reduce the number of microorganisms but cannot sterilize a burn.

FIRST AID AND PREHOSPITAL CARE

Emergency physicians may be consulted about the immediate care of minor burns. The first step is to stop the burning and help dissipate the heat. Running cool water over the injured area accomplishes both of these goals. Applying ice directly to the wound is painful, and the extreme cold can worsen the injury. Parents should be reminded not to put grease, butter, or any ointment on the burn because they do not dissipate heat well and may contribute to the

contamination. Blisters should not be broken. The burn should be covered with a clean cloth or bandage.

Small burns from mechanisms unlikely to cause full-thickness injury can be managed at home with a topical antibiotic and a bandage. Burns of larger size and burns involving the face, hands, feet, or perineum should be evaluated promptly by a physician. Telephone advice, without the benefit of physical examination, should always err on the side of caution with recommendation for a medical evaluation.

The concerns for children with major burns are different. Prehospital care providers should initially forget about the burn and focus on airway, breathing, and circulation as they would for any other trauma victim. Rapid transport to a hospital setting is still crucial and should be emphasized. Oxygen should be administered. The trachea should be intubated if there are signs of upper airway obstruction apnea or severe hypoventilation. If transport time is likely to be prolonged, intravenous fluids should be given.

MAJOR BURNS

Evaluation and Management

During the first few seconds after arrival, the physician must determine if a burned patient requires aggressive therapy for major burns (greater than 15% BSA) (Fig. 114.2). In children with severe injuries, the evaluation and initial management take place simultaneously. Smoldering clothing or other sources of continued burning must be removed. Information about the circumstances of the burn and the potential for associated injuries should be sought from prehospital care providers, police, or family members but should not delay the initial treatment.

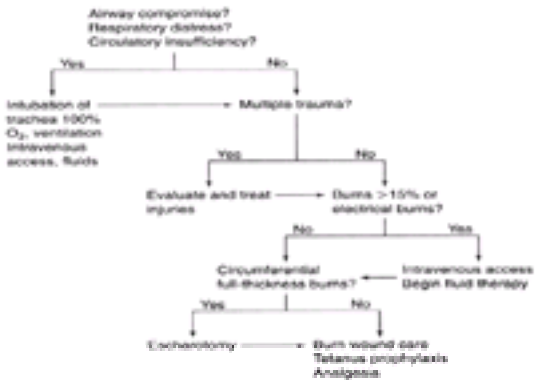

FIGURE 114.2. Diagnostic approach to the burn patient.

Airway

There are several causes of airway obstruction in the severely burned patient. Most life-threatening burns are the result of house fires. The inhalation of hot gases can burn the upper airway, leading to progressive edema and airway obstruction. Any child with burns of the face, singed facial hairs, or hoarseness is at high risk, but airway burns can occur in the absence of these signs. Edema of the burned airway will worsen over the first 24 to 48 hours. Knowledge of the time course of airway swelling warrants intubation of the trachea for subtle signs of airway compromise that occur shortly after the injury. Early intubation may prevent the later difficult intubation of a child with severe pharyngeal and airway swelling. Endotracheal tubes of smaller diameter than expected for age should be available in anticipation of a narrowed airway.

Children who have jumped or fallen in house fires, been burned in motor vehicle accidents, or been burned by explosions are at risk for associated cervical spine injuries. A history of trauma may not be available at the time of airway management. Physicians should manage the airway with the neck in a neutral position, avoiding any flexion or extension. Radiographs can be obtained later to help exclude cervical spine injury (see Chapter 106). Children with severe burns may have depressed levels of consciousness for many reasons. Airway obstruction by the mandibular block from loss of pharyngeal tone is not uncommon. A chin-lift is preferable to a jaw-thrust maneuver in patients with possible cervical spine injury.

Breathing

A rapid assessment of ventilation includes respiratory effort, chest expansion, breath sounds, and color. Pulse oximetry is useful, but patients with significant levels of carboxyhemoglobin will look pink and have normal oxygen saturation as measured by a pulse oximeter. Every severely burned child should receive 100% oxygen. Arterial blood gases with cooximetry should be obtained promptly. Patients whose ventilatory status is questionable should receive careful assisted ventilation. Avoidance of high inflating pressures and application of cricoid pressure can minimize gaseous distension of the stomach and reduce the risk of regurgitation with pulmonary aspiration.

Children burned in house fires or in any closed space are at high risk of inhalation injury. Facial burns, singed facial hairs, and carbonaceous sputum are not always present in children with significant inhalation injury. Chest radiographs may be normal initially, even if pulmonary injury has occurred. Smoke is responsible for most of the lower airway abnormalities in burned patients and management of smoke inhalation is covered in Chapter 89. The efficient heat-exchanging function of the upper airway can dramatically reduce the temperature of inspired dry gases, protecting the lower airway from thermal injury. Inhalation of steam, with its higher heat capacity, is more likely to result in burns of

the lower airway.

Circulation—Burn Shock

The physiology of circulatory impairment in severely burned patients is complex. Burn shock occurs in adults with burns over 30% of BSA but occurs in children with burns over 20% of BSA.

The rapid assessment of circulation includes skin color, capillary refill time, temperature of the peripheral extremities, heart rate, and mental status. Blood pressure is often maintained until decompensation occurs, making it an unreliable measure of early circulatory impairment. Hypertension from increased systemic vascular resistance has been reported immediately after severe burns, particularly in pediatric patients.

Vascular access should be obtained soon after arrival of the severely burned child. Intravenous lines in the upper extremity through intact skin are preferred because they are easier to secure, but access through burned areas may be necessary. Attention to aseptic technique when starting intravenous lines in the ED can prevent infectious complications during subsequent care. Circumferential taping is dangerous because the swelling that occurs during the first 24 hours can cause circulatory insufficiency distal to the constriction. Sites for central line placement should be saved, if possible, for future use for hyperalimentation. In severe burns with associated inhalation injury, however, central venous pressure monitoring is useful.

An initial bolus of 20 mL/kg Ringer's lactate solution is recommended while assessment of the extent of the burns takes place. A urinary catheter should be placed, and urine output monitored to assess the adequacy of fluid therapy. Major burns cause decreased splanchnic blood flow and ileus. After ensuring that the airway is protected with an endotracheal tube or an adequate gag reflex, the clinician should place a nasogastric tube. Hypothermia can occur rapidly in small children, especially in those whose skin injury impairs normal thermoregulation. Core temperature should be monitored and the child kept covered, except as necessary for examination and burn assessment. Tetanus toxoid is indicated if the child has not been immunized in the preceding 5 years; unimmunized patients require tetanus immune globulin as well.

Assessment

Major burns are three-dimensional injuries. To estimate the size of a burn one must assess the surface area and depth of burned skin. Decisions about fluid therapy, referral, disposition, and prognosis are based on the size of the burn. After stabilization of vital functions in the primary survey, a systematic evaluation of the surface area and depth of burns follows. The rule of nines used to estimate burn surface area in adults cannot be applied to children with their different body proportions. Young children have relatively larger heads and smaller extremities. Areas of partial- and full-thickness injury should be recorded on an anatomic chart (Fig. 114.3) and then total percentage burn surface area computed using age-appropriate proportions. First-degree burns are not included. A child's palm (not including the fingers) is approximately 1% of BSA and can be used to estimate the extent of smaller burns (Fig. 114.3).

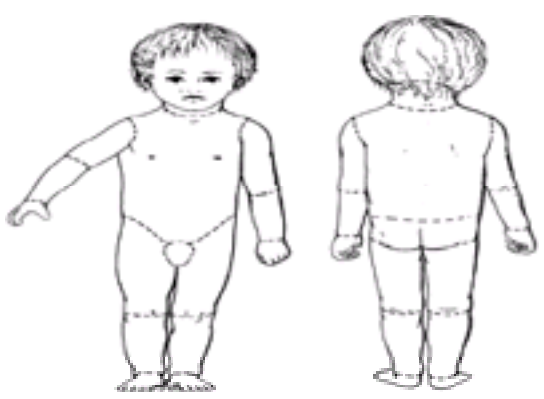

FIGURE 114.3. Estimation of surface area burned based on age. This modification by O'Neill of the Brooke Army Burn Center diagram shows the change in surface of the head from 19% in an infant to 7% in an adult. Proper use of this chart *(above)* provides an accurate basis for subsequent management of the burned child.

Fluid Therapy

Prompt treatment of the hypovolemia that occurs early in children with severe thermal injuries is of prime importance. The fluid status of burned children is a dynamic process that requires careful reevaluation and therapeutic adjustments. Extravasation of water and sodium through abnormally permeable capillaries continues for about 24 hours after injury. Capillary integrity then improves and intravascular volume stabilizes. Most burn centers recommend crystalloid during the first 24 hours because colloid may extravasate through the leaky capillaries and worsen interstitial edema. Once capillary integrity is restored, colloid is used for volume expansion and for preservation of serum oncotic pressure. The sodium ion is critical for maintaining adequate intravascular volume, so isotonic crystalloid solutions are recommended in the resuscitation phase. Hyperosmolar therapy with 3% saline appears to have little benefit and increased risk. Potassium is released from damaged cells and may elevate measured serum levels shortly after injury. Potassium replacement is not recommended during the early phase of fluid therapy.

Several formulas for calculation of fluid therapy exist (Table 114.1). The Parkland formula recommends 4 mL/kg per %BSA of crystalloid over the first 24 hours, half during the first 8 hours and half over the next 16 hours. This formula underestimates the needs of young children. Adding maintenance requirements to this volume more accurately estimates the requirements of burn victims younger than 5 years of age. The Carvajal formula uses BSA rather than weight to

calculate fluid therapy. Carvajal recommends 5000 mL/m^2 per %BSA, half in the first 8 hours and half over the next 16 hours, plus 2000 mL/m^2 per day as maintenance. Formulas for fluid therapy in burn patients must be used carefully in small children. The calculated volume requirements are useful to the clinician in providing an initial rate of fluid infusion. Adjustments of infusion rates are the rule, not the exception. Many pediatric burn centers prefer to follow urine output rather than central venous pressure to assess the adequacy of fluid therapy. Children should produce at least 1 mL/kg per hour of urine. Oliguria, as determined by this measure, is almost always the result of inadequate fluid administration. Intrinsic renal disease is sometimes noted after electrical injuries because of myoglobinuria. Hyperglycemia may cause an osmotic diuresis and complicate care of the burned patient. Before infusions are decreased in response to excessive urine output, a measurement of blood sugar should be made.

Parkland: 4 mL/kg/%BSA second- and third-degree burns, half in the first 8 hr, half in the next 16 hr. Add maintenance in children <5 yr old.
Carvajal: 5000 mL/m^2/%BSA second- and third-degree burns, half in the first 8 hr, half in the next 16 hr. Add 2000 mL/m^2/day maintenance.

Table 114.1. Fluid Resuscitation Formulas

Trauma associated with burns may increase fluid requirements. Additional fluids are often necessary when burns are associated with an inhalation injury. Fractures or other traumatic lesions causing blood loss and edema also increase the need for fluids. Neurogenic shock from unrecognized cervical spine or head injury may cause hypotension, usually with a relative bradycardia. Toxins that were ingested before the burn or inhaled during the fire can depress myocardial function or vascular tone. Any patient with shock that appears out of proportion to the extent of the burn injury or who is poorly responsive to fluid therapy should have an aggressive diagnostic workup for concurrent problems.

Antibiotics

Burn sepsis continues to be the major cause of mortality after a burned patient survives the period of resuscitation. Meticulous antiseptic techniques can lessen colonization of burns with potential pathogens. Topical antibiotics further reduce bacterial number. Streptococcal cellulitis does not occur as frequently in the first days of burn therapy as in previous years, before the development of topical antibiotics for burns. Most burn centers do not routinely treat patients with prophylactic penicillin and have not seen a rise in early streptococcal infections. Pediatric patients may have greater risk than adults, however, and some pediatric burn centers continue to use intravenous penicillin for the first 3 to 5 days after injury. Broad-spectrum antibiotics should not be used prophylactically because they do not significantly reduce the incidence of infections and they increase the likelihood of acquiring resistant organisms. Frequent examination of healing burns for signs of infection and cultures to monitor colonization can direct specific antibiotic therapy for documented infections.

Care of the Burn Wound

Early surgical management of some partial-thickness and most full-thickness burns with excision and grafting has been an important advance in burn treatment. In the ED a few basic measures initiate the wound care. Burns should be covered loosely with clean sheets during the early phase of resuscitation in severe injuries. Once the cardiorespiratory status is stabilized, the wounds are uncovered and assessed for size and depth. The goals of burn wound care are to promote rapid healing and prevent infection. Cleansing with large volumes of lukewarm sterile saline reduces contamination. Loose tissue can often be wiped away with sterile gauze, simplifying and expediting burn debridement. In general, bullae should be left intact to preserve the barrier to bacterial invasion. Certain large bullae in locations that are likely to rupture may benefit from debridement.

Full-thickness burns cause a loss of skin elasticity. The burned skin cannot expand as tissue edema develops during the first 24 to 48 hours of fluid therapy. Circumferential injury can cause vascular insufficiency of the distal extremities. Assessment of blood flow with a Doppler device is useful for monitoring peripheral circulation because the usual methods of assessment, including capillary refill and temperature, may be difficult in the severely burned child. Absent flow is an indication for escharotomy. Extensive full-thickness burns of the trunk may restrict expansion of the chest and impair ventilation. Respiratory embarrassment in this setting is also an indication for escharotomy. Escharotomy involves incision through the depth of the eschar on the medial and lateral aspects of the extremities, including the hands and fingers. It is especially important to extend across the joints because at these locations the skin is tightly adherent to the underlying fascia where vascular obstruction is likely to occur. The procedure does not require anesthesia because the wounds are full thickness and insensate. If reperfusion of the extremities is not immediate, hypovolemia should be suspected. Reperfusion of the extremities after escharotomy may reduce intravascular volume and require adjustment of fluid therapy. With extensive, full-thickness thoracic burns, incision along the anterior axillary lines allows adequate chest expansion.

Pain Management

Reducing pain is an important consideration in the management of children with burns. Pain is a subjective experience

influenced by the preceding events. Children rescued from house fires, separated from their parents, transported in ambulances, and brought to EDs are usually extremely anxious. Calm, developmentally appropriate verbal reassurance, even to preverbal children, can reduce anxiety and dramatically reduce the perception of pain.

The exposure of sensory nerve receptors in partial-thickness burns makes them sensitive to environmental stimuli. Movement of cool air across burned tissue increases pain significantly. The simple measure of covering burns with a clean sheet, only exposing them when necessary for burn assessment, is extremely effective and safe analgesia.

Many children will still have significant pain after reassurance and covering of the burns. Narcotic analgesics are useful when administered appropriately. Morphine may reduce the blood pressure, especially in patients who are hypovolemic. Narcotics should not be given until adequate circulation is established.

Analgesic medications given intravenously are preferred because they are more effective and predictable. Intramuscular injections or oral doses should not be given to patients with significant burns. Circulation to muscle and gut is reduced and absorption of medication given via these routes is delayed and unpredictable. Morphine (0.1 to 0.15 mg/kg) is the drug of choice for most burn patients who require analgesia. In children who do not respond well to the initial dose of morphine, a careful assessment for other causes of pain or agitation should be sought. Hypoxemia, early shock, and occult injuries should be excluded before repeating doses of analgesics. Analgesic administration just before debridement of the burn wound is recommended.

Disposition

Guidelines for admission must be individualized when treating burned children. Hospitals, physicians, and parents have varying capabilities for managing pediatric burn patients. In general, children with burns of smaller percentages of BSA than adults require admission, especially patients less than 2 years old.

Children with partial-thickness burns greater than 10% of BSA should be admitted to a hospital. Partial-thickness injury over 20% of BSA warrants admission to a children's hospital or burn center. Full-thickness burns over 2% of BSA require inpatient treatment. Burns in certain locations are higher risk for disability or poor cosmetic outcome and should be considered for treatment in the hospital. These include over 1% burns of the face, perineum, hands, feet, circumferential burns, or burns overlying joints. Children with inhalation injury or associated trauma require admission with burns involving lesser percentages of BSA. Any time the physician suspects that the burns cannot be adequately cared for in the home, admission to the hospital is warranted.

OUTPATIENT MANAGEMENT OF BURNS

A small minority of all burns in children require therapy in the hospital. Once a careful assessment has led to a decision to manage a burn as an outpatient, preparations for treatment at home should begin. Parents become the physician's partner in this context and need to be instructed carefully. If no one in the household seems able to adequately manage the burn wound, inpatient care must be considered.

A first-degree burn usually requires no therapy. Moisturizers and acetaminophen or ibuprofen can be given as needed. Partial-thickness burns are first cleansed with mild soap and water, one-quarter strength povidone–iodine solution, or saline alone. Devitalized tissue can usually be removed by wiping with gauze. Large bullae that are likely to rupture because of their location can be debrided. Clean partial-thickness burns less than 2% BSA can be dressed with petrolatum gauze. Topical antibiotics are recommended for larger or more contaminated burns. Silver sulfadiazine cream (Silvadene) or bacitracin are the topical antibacterial agents of choice at most burn centers. A $^{1}/_{16}$ to $^{1}/_{8}$ inch layer of silver sulfadiazine is applied to the burn with a sterile tongue blade or gloved hand. Silver sulfadiazine is soothing to the burn and has few side effects. Mild bleaching of the skin may occur. Bacitracin is often chosen for burns of the face. About 5% of children are allergic to sulfa and can be treated with bacitracin or povidone–iodine ointment. Leukopenia has also been reported in patients treated with silver sulfadiazine. Mafenide acetate (Sulfamylon) is a topical antimicrobial agent that is more penetrating than silver sulfadiazine. It causes pain when applied, cannot be used in sulfa-sensitive patients, and inhibits carbonic anhydrase, which can cause a metabolic acidosis. Some experts recommend mafenide acetate for burns overlying cartilaginous structures such as ear and nose. Small, superficial partial-thickness burns may be treated with sterile dressings and no antibiotics.

A loose gauze dressing should be placed over the burn and secured with tape. Burns of the face can be treated with an open technique. Dressings should be changed twice each day. The parent should rinse off residual antibacterial cream with warm water and inspect the wound. Signs of infection, such as redness and tenderness around the margin of the burn, warrant immediate evaluation by a physician. A greenish material formed by serous drainage from the burn mixing with the silver sulfadiazine cream is often mistaken for purulence. If the burn is healing well, the parent should reapply the antibiotic cream and dress the wound as demonstrated by the physician or nurse in the ED. Burns should be examined by a physician every 2 or 3 days until healing is well under way. Large burns or burns of the hands, feet, perineum, or overlying joints that are managed as an outpatient should be referred for follow-up to a burn specialist and evaluated more frequently. Prophylactic antibiotics are not recommended.

INFLICTED BURNS—CHILD ABUSE

Physicians who treat children with burns must consider child abuse in patients with specific patterns of injury. Between 10 and 20% of burns in children are inflicted, accounting for 10% of child abuse cases. Most inflicted burns are scalds. Forced submersion of the hands or feet often causes burns that are deep, have a clear line of immersion, and are symmetric. Scald burns of the buttocks and thighs in toddlers are usually the result of child abuse. Forcible submersion in a tub of hot water is often punishment for toilet-training mishaps. Inflicted contact burns also have characteristic patterns. Small, round, deep burns result from cigarettes intentionally applied to the skin. Deep injuries with distinctive patterns

may be noted in children held against portable heaters or burned with irons.

In many children with inflicted burns the pattern of injury is nonspecific. A clear history of abuse is usually not offered. A deep wound with a geometric pattern and sharply demarcated borders suggests a contact burn. Scald burns usually have scattered splash lesions. In burns from spilled hot beverages there is often a pattern of injury spreading downward from the falling liquid. Physicians need to consider if the characteristics of the burn correspond with the reported mechanism in a plausible way. Identifying suspicious injuries and notifying the appropriate authorities can prevent subsequent injuries to abused children.

ELECTRICAL BURNS

Burns that result when electrical current passes through the body have unique characteristics. Each year there are more than 4000 ED visits caused by electrical injuries, most in children (see Chapter 89). Electrical burns account for 3% of burn center admissions and are increasing in number. Most injuries occur in young children from contact with low voltage (less than 120 V) alternating household current, often from mouthing plugs or extension cords. Severe high voltage (greater than 500 V) injuries are seen most often in adolescent males as a consequence of risk-taking behaviors.

Thermal energy is released in proportion to the amount of electrical current that passes through tissue. Current flows preferentially through tissues of low electrical resistance, such as blood vessels, nerves, and muscles. Moisture on the skin decreases resistance, accounting for the greater severity of injury in the antecubital and axillary areas in victims of electrical burns. Current arcing through the skin can ignite clothing and cause severe thermal burns in addition to the electrical injury. In some electrical burns, a depressed entrance wound and a blown out exit wound can be identified. The concentration of current at these points results in higher temperatures and more severe tissue damage. If the flow of current traverses the heart at certain points of the cardiac cycle, ventricular fibrillation or asystole can occur. Electrical injury, especially direct current, can cause prolonged tetany of the musculature, including the respiratory muscles, leading to suffocation.

The initial approach to victims of electrical burns is similar to that in other severely burned children. The potential for arrhythmias requires close cardiac monitoring. Electrical burns are usually more severe than they appear. Significant deep and internal injuries may occur in patients with relatively small entrance and exit wounds. Fluid requirements are higher than predicted by formulas based on estimates of percentage of BSA because a larger portion of the injury is internal. Destruction of muscle often causes myoglobinuria. Renal failure usually can be prevented with forced diuresis and alkalinization. Electrical injury and edema within fascial compartments can cause a compartment syndrome with vascular insufficiency. Severe electrical injuries require extensive evaluation for internal injuries, which should be done at a children's hospital or regional burn center.

A common electrical injury occurs to the lips and mouth of toddlers who suck on plugs or extension cords. Deep burns at the corner of the mouth require specialized attention to prevent severe scarring and contracture. Bleeding from the labial artery 1 to 2 weeks after injury, when the eschar separates, can result in significant blood loss. In previous years these children were hospitalized for 2 weeks, but some burn specialists now manage these children as outpatients. See also Chapter 89 for additional discussion of electrical burns, including lightning strikes.

CHEMICAL BURNS

More than 25,000 different caustic products can cause burns. Most are either acidic or alkaline. Acids cause coagulation of tissue proteins, which limit the depth of penetration. Alkali results in liquefaction and deeper injury. Caustic chemicals on the skin cause a prolonged period of burning compared to most thermal burns. Edema of the underlying tissue can make full-thickness injuries appear deceptively superficial. Treatment of caustic burns, whether acid or base, involves copious irrigation to dilute the chemical and stop the burning. Attempts at neutralization are usually ineffective and should be avoided. The pH of the effluent can be monitored to help determine whether irrigation has been adequate. A thorough examination is necessary to identify other areas of skin exposed from splashes or contact that also require irrigation. Consultation with a burn specialist and admission are recommended at smaller percentages of BSA with chemical burns than with thermal injuries. Chemical burns to the eye can threaten vision and, after starting irrigation, require prompt consultation with an ophthalmologist.

Suggested Readings

Boss W, Brand D, Acampora D, et al. Effectiveness of prophylactic antibiotics in the outpatient treatment of burns. J Trauma 1985;25:224–227.

Carvajal HF. Fluid resuscitation of pediatric burn victims: a critical appraisal. Pediatr Nephrol 1994;8(3):357–366.

Coren C. Burn injuries in children. Pediatr Ann 1987;16:328–339.

Harmel RP, Vane DW, King DR. Burn care in children: special considerations. Clin Plast Surg 1986;13:95–105.

Herndon DN, Thompson PB, Desai MH, et al. Treatment of burns in children. Pediatr Clin North Am 1985;32:1311–1332.

McLoughlin E, Crawford JD. Burns. Pediatr Clin North Am 1985;32:61–75.

Merrell S, Saffle J, Sullivan J, et al. Fluid resuscitation in thermally injured children. Am J Surg 1986;152:664–669.

Mozingo DW, Smith AA, McManus WF, et al. Chemical burns. J Trauma 1988;28:642–647.

O'Neill JA. Fluid resuscitation in the burned child--a reappraisal. J Pediatr Surg 1982;17:604–607.

Osgood PF, Szyfelbein SK. Management of burn pain in children. Pediatr Clin North Am 1989;36:1001–1013.

Raine P, Azmy A. A review of thermal injuries in young children. J Pediatr Surg 1983;18:21–26.

Walker AR: Emergency department management of house fire burns and carbon monoxide poisoning in children. Curr Opin Pediatr 1996;8(3):239–242.

CHAPTER 115

Orthopedic Trauma

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Orthopedic trauma currently accounts for 10 to 15% of emergency department (ED) visits in urban pediatric hospitals. The number and spectrum of musculoskeletal injuries sustained by children and adolescents appear to be on the rise in recent years, in part because of the rapid growth of organized sports. As the result of a number of anatomic and physiologic differences, the array of orthopedic injuries seen in pediatrics differs greatly from that seen in adult practice. An understanding of these differences allows the emergency physician to make accurate diagnoses and to avoid complications. The purpose of this chapter is to provide a set of principles and guidelines to be used in the initial evaluation, diagnosis, and treatment of common orthopedic injuries in children.

GENERAL PRINCIPLES OF PEDIATRIC ORTHOPEDICS

Structural and Physiologic Differences between the Musculoskeletal Systems of Children and Adults

The bony architecture in children includes a thick and active periosteum, a growth plate (physis), an epiphysis (secondary ossification center), and perichondrial rings (Fig. 115.1). The bones of a child are much more porous and thus more pliable than those of an adult. On the other hand, because of this increased porosity, stiffness and overall bony strength are less and the incidence of fractures is greater in children. During growth, the skeleton undergoes changes that cause different anatomic regions to be more susceptible to fracture at certain stages of development. In general, the ligaments attaching one bone to another have greater strength than the epiphyseal plates and perichondrial rings. As a result, although the number of fractures is greater, the incidence of sprains, ligamentous injuries, and dislocations is much reduced in children.

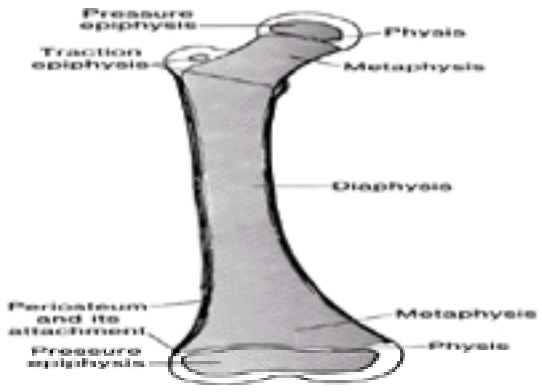

FIGURE 115.1. Diagrammatic representation of the femur in late childhood.

The periosteum plays an important role in the reparative process of fracture healing. In children, the periosteum is thick and physiologically active and is easily stripped from the bony cortex during injury. When injuries occur, the periosteum is often torn on the convex side of the fracture while remaining intact on the concave side. The intact periosteum on the concave aspect often aids the orthopedist in the reduction of the fracture fragments. Callus formation is exuberant in the young and declines with age as the physiologic activity of the periosteum decreases. Nonunions almost never occur in children.

Remodeling, although rare in adults, is expected to a degree in children. Significant remodeling can be anticipated in younger children and when the fracture occurs in the metaphysis of growing bones. Deformities occurring in the plane of motion of the adjacent joint remodel to the greatest degree. Fractures that occur in the diaphysis of long bones in adolescents, away from the plane of motion of the joint, cannot be expected to correct spontaneously with growth. In general, it is important to obtain as near an anatomic reduction of fracture fragments as possible in all age groups and not to rely on remodeling to align angulated fractures.

Fractures Unique to Children

The anatomic and physiologic differences between adults and children are reflected in a number of fractures and injuries unique to the pediatric age group, including physeal fractures, torus fractures, greenstick fractures, bowing deformities, and avulsion fractures.

Physeal Fractures

Fractures often occur at the physis (growth plate) in children. Most such fractures occur through the zone of provisional calcification, a relatively weak area of the germinal growth plate. Overall, up to 18 to 30% of pediatric fractures involve the physis. Physeal injuries are more common in adolescents than in younger children, with a peak incidence at 11 to 12 years of age. Most growth plate injuries occur in the upper limb, particularly in the radius and ulna.

Several classification systems have been described for physeal fractures. The most widely used is that of Salter and Harris, who described five types of growth plate fractures, each having specific prognostic and treatment implications (Fig. 115.2).

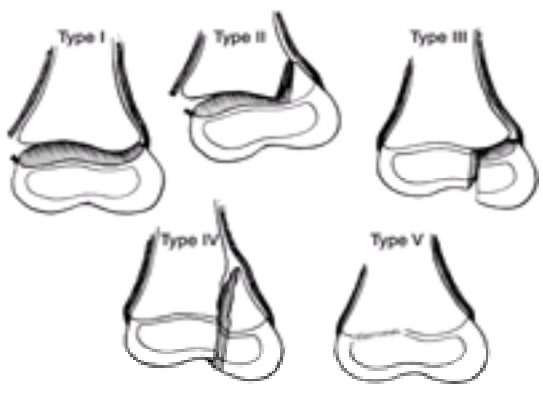

FIGURE 115.2. The Salter-Harris classification for physeal fractures. The prognosis for growth disturbance worsens from type I through type V.

Salter-Harris Type I Fracture

Salter-Harris type I fracture involves separation of the metaphysis from the epiphysis through the zone of provisional calcification. Diagnosis is often difficult if displacement is minimal. Type I fractures are generally benign, with little chance of growth disturbance if near anatomic reduction is achieved. Exceptions include type I injuries of the proximal radius, the proximal and distal femur, and the proximal tibia, all of which are subject to premature physeal closure and posttraumatic growth arrest. In general, when radiographic studies are negative but physical findings are suggestive of a Salter-Harris type I injury (e.g., point tenderness over a growth plate), immobilization and a follow-up examination are essential.

Salter-Harris Type II Fracture

Salter-Harris type II fracture is the most common type of pediatric physeal fracture. It is similar to a type I fracture except that a portion of metaphyseal bone is displaced with the epiphyseal fragment. The fracture line crosses the germinal growth plate as it courses toward the metaphysis. Like the type I injuries, these fractures generally carry a good prognosis.

Salter-Harris Type III and IV Fractures

Salter-Harris type III and IV fractures are intra-articular injuries that also involve the growth plate. Anatomic position must be reestablished to restore normal joint mechanics and prevent growth arrest. Because of the increased incidence of growth disturbance, altered joint mechanics, and functional disability following Salter-Harris type III and IV fractures, an orthopedic consultation is usually obtained for all but the most minor type III and IV injuries while the patient is in the ED.

Salter-Harris Type V Fracture

Salter-Harris type V fracture results from axial compression of the germinal growth plate. It is often difficult to diagnose; the radiograph may be normal or may demonstrate any of the above Salter-Harris growth plate fractures. The diagnosis is often made on hindsight after a growth arrest becomes evident.

Torus Fractures

Torus (buckle) fractures are common fractures in young patients. They occur in the metaphyseal region of bone from a compressive load. The cortex of the bone buckles in a small area, resulting in a stable fracture pattern (Fig. 115.3). As the child matures, the stiffness of the metaphyseal region increases, and the incidence of this fracture pattern decreases.

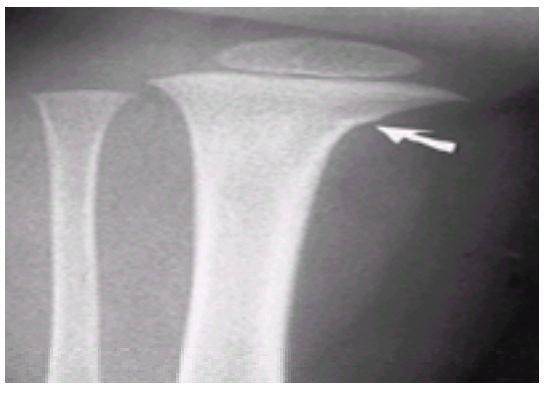

FIGURE 115.3. Torus fracture of the proximal right tibia in a 1-year-old child *(arrow).*

Greenstick Fractures

Greenstick injuries are the most common fracture pattern in children, accounting for up to 50% of fractures before 12 years of age. They are incomplete fractures that occur at the diaphyseal–metaphyseal junction and in which the cortex remains intact on one side. Angulation and rotation are common. To obtain an anatomic reduction, the fracture must often first be completed (Fig. 115.4).

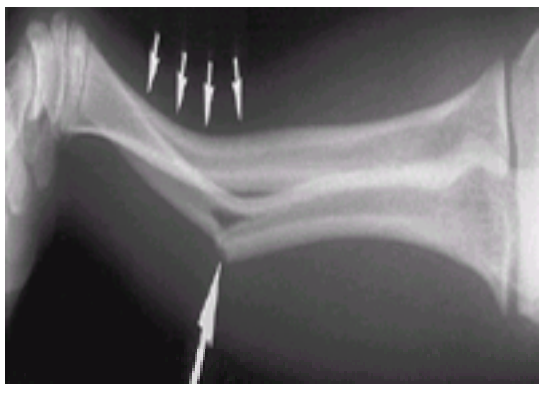

FIGURE 115.4. Greenstick fracture of the ulna *(large arrow)* and a bowing fracture *(small arrows)* of the radius. The extent of bowing can often be fully appreciated only with comparison views of the opposite extremity.

Bowing Fractures

Bowing fractures occur uniquely in children. Recent evidence suggests that the force causing the deformation is longitudinal. The force stops short of creating a fracture but does cause persistent plastic deformation (bowing) of the bony structure (Fig. 115.4). Little remodeling can be expected from the injury, and both cosmetic and functional deficits

are common. Anatomic reduction produces the most satisfactory result. All bowing deformities should be referred to an orthopedic surgeon.

Avulsion Fractures

Avulsion fractures are common in children. Strong muscular attachments adhere to secondary centers of ossification known as apophyses in the developing skeleton. During intense muscular contraction, fractures occur through the apophyseal plate. Most avulsion fractures occur in the pelvis and heal uneventfully. Other common sites include the tibial tubercle and the phalanges. Rarely do avulsion fractures require open or closed reduction. Conservative care is the mainstay of treatment.

Physical and Radiographic Examination

Approach to the Physical Examination

A systematic approach to the child with a suspected fracture is necessary to avoid overlooked injuries and undue complications. The basic principles of a history and physical examination should be followed. In all cases, it is necessary to consider the possibility of associated head and truncal injuries. Often the history is obtained from a parent or bystander who witnessed the accident. At times, no history is available. Attention to the mechanism of injury and the force causing the injury gives clues to the severity of the fracture and soft-tissue injury. The activity of the patient following the injury also helps define the likelihood and nature of orthopedic injuries. Whether the child is able to provide any details of the accident obviously depends on both age and the extent of associated head and internal injuries.

The physical examination should begin with careful observation of the patient's behavior. Is he or she guarding or not moving an extremity? Is there pain? How is the patient's color? Is the child interacting normally with his or her parents and the environment? After observing the child and determining the area or areas of injury, the physician can look for swelling and deformity. It is best to always begin with the extremities that do not appear to be injured. An effort should be made to gain the patient's trust by gently moving all joints and extremities that appear uninjured while trying to distract the child from the examination itself. This also allows detection of unsuspected areas of injury. Attention should then be turned to the injured extremity. Swelling, ecchymosis, deformity, and the presence of lacerations and puncture wounds should be noted. When open wounds are present, the exact location, degree of contamination, presence of fat globules, and rate of active bleeding should be documented. It is not always obvious whether there is an open fracture or simply a laceration that does not communicate with the fracture; operative exploration may ultimately be required.

While continuing to distract the child, the physician should then carefully palpate the soft tissues and bones above and below the area of injury. The point of maximal bony tenderness should next be gently defined. Evidence of increased compartment pressures should be sought both by palpation and by careful assessment of the pulses and capillary blood flow distal to the injury. However, the compartment syndrome can occur in the presence of palpable pulses (see Compartment Syndrome, following). If no significant deformity is discovered, joint motion both proximal and distal to the fracture should be assessed. As detailed a sensory and motor examination as the age and overall condition of the child will permit should be performed.

Radiographic Examination

All unstable and significantly deformed fractures *must* be immobilized before the initiation of radiographic studies. By so doing, further deformity and soft-tissue injury is avoided, and patient discomfort during positioning for the radiographs is decreased. A plaster or fiberglass splint can be applied quickly and does not prevent adequate radiographic assessment of the fracture (Fig. 115.5). Unless a specific contraindication exists, pain medication, often parenteral, should be administered as well. Recent studies continue to demonstrate that many children with skeletal injuries receive no or inadequate pain medication, both in the ED and upon discharge. No patient should be allowed to take any food or drink by mouth until all examinations and referrals are completed.

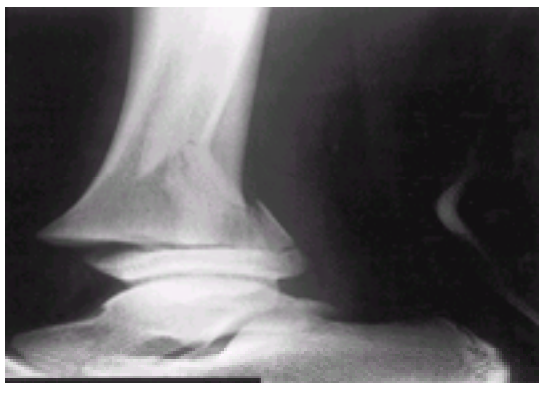

FIGURE 115.5. Lateral radiograph through a fiberglass splint showing angulated distal tibial and fibular fractures in a 13-year-old boy. Splinting before radiologic studies provides fracture stability and comfort but does not prevent adequate visualization of bony injury.

After a complete history and physical examination, the physician should be able to order specific radiologic views to identify the injury. In some instances (e.g., a toddler who is refusing to bear weight but who has no localizing signs), the history and a knowledge of the most common injuries for a given age have to guide the choice of radiographic studies. A complete examination should include the joints above and below the fracture and at least two views taken at 90 degrees

to one another (generally anteroposterior and lateral views). Oblique and other additional views are necessary at times for certain body parts (e.g., the hand, ankle, foot, phalanges) and when routine views are normal but suspicions of a fracture are high. Given the degree of normal variability in bony anatomy, particularly in growing bones, comparison views are indicated on occasion (Fig. 115.6).

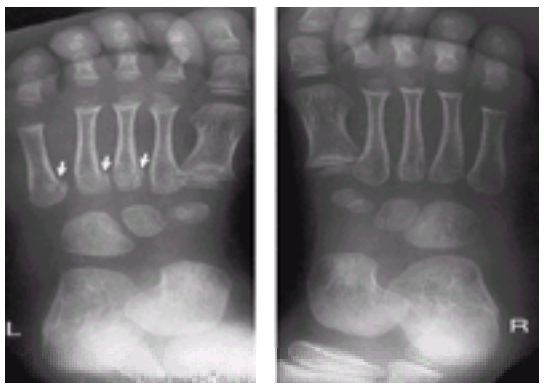

FIGURE 115.6. Radiographs of the feet of a 3-year-old child who sustained a crush injury of the left foot. The soft-tissue swelling of the left foot *(L)* is readily apparent. That the irregularities of the proximal third, fourth, and fifth left metatarsals *(arrows)* are indisputably fractures is immediately apparent when the injured foot is compared with the uninjured right foot *(R).* A proximal second metatarsal fracture was suspected as well and was better visualized on other views.

Although plain film radiography will no doubt remain the primary imaging technique used in fracture evaluation, other modalities, notably bone scintigraphy, computed tomography (CT), and magnetic resonance imaging (MRI) have come to play an important adjunctive role. Scintigraphy is more sensitive than plain films in certain settings, for example, when a stress fracture is suspected (Fig. 115.7). CT plays an important role in the definition of complex fractures, particularly intra-articular ones, as well as in the evaluation of spine injuries. The roles of MRI in the same settings continue to be defined and expanded.

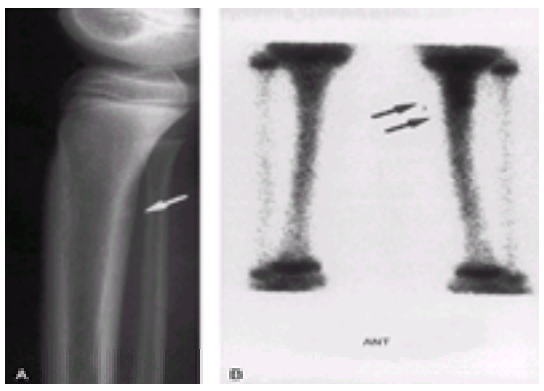

FIGURE 115.7. A. Routine radiographs of this 10-year-old girl revealed only a minor cortical irregularity of the posterior aspect of the proximal left tibia *(arrow).* **B.** Bone scintigraphy was performed. The increased isotope uptake seen along the proximal left tibia *(arrows)* confirmed the clinical suspicion of a stress fracture.

Fracture Description

When obtaining an orthopedic consultation, the emergency physician must relay accurate and descriptive information to allow the orthopedist to make appropriate treatment recommendations. A clinical description should include the patient's age and sex, the mechanism of injury, the anatomic location, the status of the neurovascular structures, and the extent of associated soft-tissue injury. A careful and precise radiographic description should include the anatomic location of the fracture; the type of fracture (e.g., transverse, spiral, oblique); the amount of displacement; the degree of angulation, shortening, or malrotation; the degree of comminution; and the extent of involvement of the joint and growth plate. Accurate descriptions using appropriate terminology are helpful in assisting the orthopedist in his or her recommendations (Fig. 115.8).

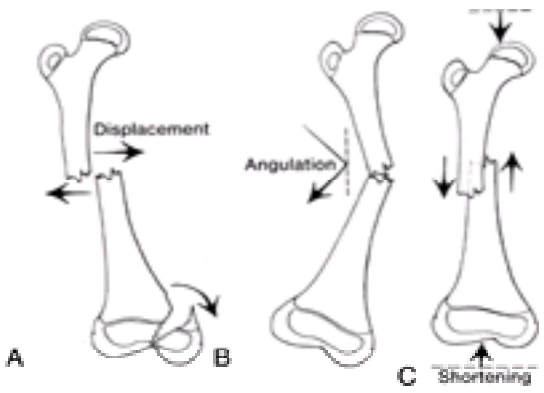

FIGURE 115.8. Diagrammatic representation of fracture deformities: displacement **(A);** angulation **(B);** overriding with

shortening **(C).**

Orthopedic Referral and General Principles of Acute Fracture Care

Indications for Orthopedic Referral

The indications for an orthopedic consultation vary somewhat with the ability and experience of the emergency physician and the availability and preferences of the orthopedist. Certainly, if any question exists regarding the diagnosis, treatment, complications, or follow-up of an orthopedic injury, a consultation should be obtained. An orthopedic surgeon should be called to evaluate all fractures that are open, unacceptably displaced, or causing neurovascular compromise. Other indications for immediate orthopedic referral include significant growth plate or joint involvement, many fractures of the long bones of the lower extremity, pelvic fractures (other than avulsions), spinal injuries, and dislocations of major joints other than the shoulder. On the other hand, the emergency physician should be expected to provide the initial, if not the definitive, care for many pediatric fractures. Most nondisplaced Salter I fractures; clavicular injuries; nondisplaced upper extremity, foot, and phalangeal fractures; incomplete, nondisplaced fractures of the long bones of the lower extremity; and routine dislocations of minor joints and the shoulder can all be managed initially by physicians other than orthopedists (Table 115.1).

Injuries That Require Immediate Orthopedic Referral
Open fractures
Unacceptably displaced fractures
Fractures with associated neurovascular compromise
Significant growth plate or joint injuries
Complete or displaced fractures of the long bones of the lower extremities
Pelvic fractures (other than minor avulsions)
Spinal fractures
Dislocations of major joints than the shoulder
Injuries That Can Be Managed Initially by the Emergency Physician
Nondisplaced Salter-Harris type I fractures (exceptions are femur, proximal tibia)
Clavicular fractures
Nondisplaced upper extremity fractures
Incomplete, nondisplaced fractures of the long bones of the lower extremities
Nondisplaced fractures of the hand and foot
Routine dislocations of the shoulder and minor joints (no fractures)

Table 115.1. Indications for Orthopedic Referral

Acute Fracture Care

Immobilization is the mainstay of the initial treatment of any fracture. Plaster and fiberglass materials are both satisfactory, although the greater strength of fiberglass has advantages in splinting of lower extremities. Immobilization of the joints above and below the fracture provides both the greatest degree of comfort and the best guarantee against additional injury or deformity. (An exception is a minor torus fracture of the distal forearm, for which a short arm splint or cast is often adequate.) Several layers of padding material should always be applied before the actual splint or cast because the padding provides greater comfort, and the risk of neurovascular compromise, if swelling occurs, is diminished. (See the Procedures section for detailed descriptions of splinting techniques.) The degree of actual or anticipated swelling, the propensity of the fracture to lead to a compartment syndrome, and the training of the emergency physician dictate whether a splint or a cast is applied at the time of initial evaluation.

Most fractures that are initially casted should be reevaluated within 24 hours for signs of neurovascular compromise, either in the ED or by the orthopedist. Otherwise, when orthopedic follow-up is necessary, an appointment within the week following injury is generally reasonable (assuming the immobilization is adequate). Certain injuries (e.g., any fracture that could possibly displace) should be seen more promptly. Other fractures need no orthopedic referral at all. Discharge instructions should include a review of the need for elevation and ice application and a discussion of the signs of neurovascular compromise. The need for pain medication in children should not be ignored. All radiographs should be reviewed by a radiologist, and parents should be routinely informed that further evaluation and radiographs will be necessary despite initially negative studies should symptoms persist. The importance of immobilization of all but the most minor injuries and of careful documentation of follow-up instructions cannot be overemphasized.

Postfracture Care

Two factors influence the function regained following fractures in children: the establishment of a bony union and the restoration of normal alignment and growth. Unlike postfracture care for the adult, physical therapy usually is not necessary to regain normal range of motion. Stiffness rarely becomes a long-standing problem for children. The child acts as his or her own physical therapist through normal activities. On occasion, it may become necessary for the parents or physical therapist to “supervise” active range of motion exercises until normal range of motion is obtained. If the orthopedic injury is associated with tissue loss, head injury, nerve damage, or vascular compromise, physical and occupational therapy may play a major role in reestablishing normal or near-normal function in the child.

SPECIAL CONSIDERATIONS

Open Fractures

Several considerations dictate that the emergency physician approach open fractures with special concern. Such fractures generally result from high-energy accidents, namely falls, motor vehicle collisions, and auto–pedestrian accidents. Multiple injuries are common in such settings. The physician should not allow an open fracture to distract from the detection and orderly management of other less apparent but potentially life-threatening injuries. A complete examination is imperative.

The incidence of complications is higher with open fractures, and a complete evaluation for neurovascular compromise and for signs of compartment syndrome should be performed. In addition, the incidence of infection is increased with open fractures. Management should include cleansing the wound, applying a sterile Betadine dressing, administering prophylactic intravenous antibiotics (e.g., broad-spectrum cephalosporins), and immobilizing the fracture. Tetanus prophylaxis should be administered according to the usual guidelines. Clearly, open fractures must be regarded as true orthopedic emergencies. Surgical debridement, irrigation, and definitive care of the wound and fracture are uniformly necessary. The patient should be given nothing by mouth and an urgent orthopedic consultation obtained. The laceration over a fracture should never be closed, even if the fracture is in good alignment.

Compartment Syndrome

The compartment syndrome is a devastating fracture complication that, if left untreated, may progress to muscle necrosis and nerve palsies. It occurs when a buildup of intracompartmental pressure results in ischemia of the muscle and neurovascular tissue. The pressure initially blocks venous outflow, resulting in increased pressure in the nonelastic compartment. Eventually, the small arterioles and capillaries are occluded and irreversible muscle and nerve damage results.

The compartment syndrome can occur in the forearm, hand, thigh, leg, or foot; the most common site is the anterior compartment of the leg. The fracture need not be a severe one; indeed, the compartments are often torn with significantly displaced fractures and thus are less subject to pressure buildup. Pain, particularly pain with passive extension, is the earliest sign of the compartment syndrome. With any fracture or blunt tissue injury presenting with pain out of proportion to the injury, the compartment syndrome must be suspected. On palpation, the muscular compartment may feel hard, swollen, and tense. Other physical signs, including pulselessness, paresthesia, pallor, and paralysis, may or may not be present. Direct measurement of compartmental pressures confirm the diagnosis. When clinical and objective signs of compartment syndrome are present, a fasciotomy should be performed as soon as possible.

In the patient with multiple injuries, it is imperative to palpate every muscular compartment to rule out impending compartment syndrome. An orthopedic consultation should be obtained in every case of suspected compartment syndrome.

Multiple Trauma

Although fractures are common in the child with multiple injuries, only rarely are they life-threatening. There is no question that orthopedic injuries are often the most obvious. And there is no question that more children require operative orthopedic surgical procedures than general surgical procedures after major trauma. On the other hand, it is definitely a mistake to disregard the usual tenets of trauma management and forsake an orderly and thorough evaluation of a child's respiratory, cardiovascular, and neurologic status in a rush to provide fracture care. The B of the ABCs is not for bone.

Only in a few instances is the blood loss associated with a fracture significant enough to cause signs of shock. Exceptions include pelvic fractures and multiple long bone fractures (even an isolated femur fracture rarely causes hemodynamic compromise). Clearly, then, signs of significant volume loss in a child thought only to have sustained fractures should prompt an immediate search for other injuries.

In most instances, initial fracture management in the ED should consist simply of immobilization. Traction splints are extremely useful for lower extremity fractures. The role of pneumatic antishock garments continues to be debated; their use should be considered for unstable pelvic injuries. On occasion, application of an external fixator device may help tamponade bleeding from such fractures.

Many fractures, primarily nondisplaced ones, go undetected during initial ED management. Little harm occurs as a result. On the other hand, the consequences of missing a thoracic or lumbar spine fracture can obviously be much greater. Physical signs of such fractures are generally lacking, and the status of the child often precludes an assessment of pain and neurologic function. When the mechanism of injury is unknown or suggests the possibility of a spinal injury, radiographs should be ordered and careful immobilization maintained.

Child Abuse

Although the diagnostic significance of skeletal injuries in child abuse has long been recognized, only 5 to 18% of abused children sustain fractures. On the other hand, a large percentage of fractures in infants less than 1 year of age are not results of accidents. Careful consideration of the details of the injury, particularly from the viewpoint of the child's developmental stage, provides the first clue regarding the likelihood of abuse. Only a limited number of fracture types and patterns can be considered almost uniformly specific for child abuse (Table 115.2). In most cases of abuse, the radiographic findings will not by themselves confirm suspicions of an intentional injury. Although spiral fractures in nonambulating children and metaphyseal–epiphyseal injuries are essentially diagnostic, transverse fractures are common and diaphyseal fractures predominate in abused as in nonabused children (Fig. 115.9, Fig. 115.10 and Fig. 115.11).

1. Fractures inconsistent with history
2. Fractures inconsistent with developmental stage of child
3. Fractures with associated injuries suggestive of abuse
4. Multiple fractures, particularly in various stages of healing
5. Multiple, complex, or depressed skull fractures
6. Epiphyseal-metaphyseal rib fractures
7. Spiral fractures of the femur or tibia in preambulating children
8. Spiral fractures of the humerus
9. Metaphyseal chip (corner) fractures
10. Avulsion fractures of clavicle and acromion process

Table 115.2. Fractures Strongly Suggestive of Child Abuse

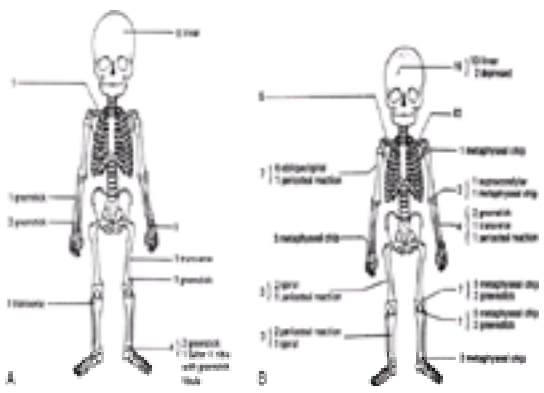

FIGURE 115.9. Comparison of the locations and types of accidental **(A)** and nonaccidental **(B)** fractures in infants less than 18 months of age. (Reprinted with permission from Worlock P, Stower M, Barbor P. Patterns of fractures in accidental and nonaccidental injury in children. Br Med J 1986;293:100–102.)

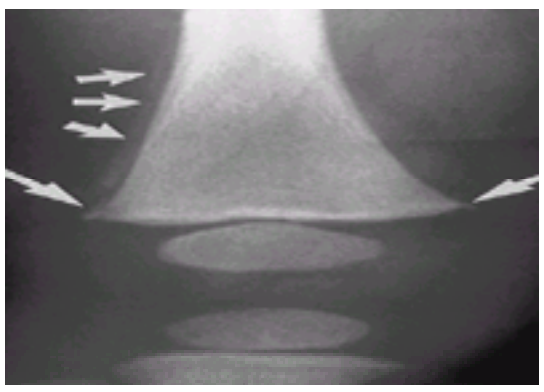

FIGURE 115.10. Radiograph of the right knee of a 3½-month-old victim of child abuse. The metaphyseal corner fractures *(large arrows)* are considered diagnostic of abuse. Also evident is periosteal new bone formation *(small arrows),* proof of a significant delay between injury and medical evaluation.

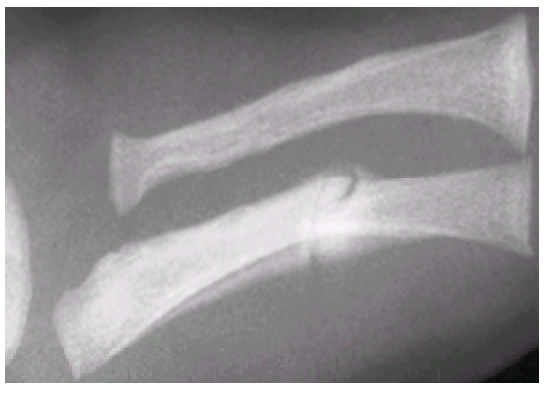

FIGURE 115.11. Radiograph of the left forearm of a 3-month-old victim of child abuse. Although the ulnar fracture is transverse and thus by itself is not diagnostic of intentional injury, both the child's age and the extent of periosteal bone formation (again establishing a significant delay between diagnosis and medical evaluation) should strongly suggest the possibility of child abuse to the examining physician.

As for diagnostic studies, any clinically suspected fracture should be evaluated using the radiographic views customary for the site in question. In addition, a skeletal survey must be performed routinely as part of the evaluation of all cases of strongly suspected abuse in children less than 2 years of age. For children 2 to 5 years of age, individual considerations determine whether a skeletal survey is needed. Skeletal surveys have little role in the evaluation of children older than 5

years of age. Routine radionuclide bone scans are not indicated, although this technique may be an adjunct to the detection of subtle rib and long bone shaft fractures, as well as of some spine injuries.

Pathologic Fractures

A pathologic fracture is one that occurs through abnormal bone. Many conditions, including tumors, hereditary diseases, metabolic disorders, neuromuscular diseases, and infections, can cause either generalized or localized bone weakness (Table 115.3, Fig. 115.12 and Fig. 115.13). On occasion, the predisposing condition does not become obvious until a fracture occurs. All pathologic fractures require orthopedic consultation. The nature of the underlying disease that is identified or suspected determines the need for consultation of other specialists. In most instances, the initial treatment parallels that of a nonpathologic fracture in the same site.

Tumors and Cysts—Benign	**Metabolic Disorders**
Aneurysmal bone cyst	Copper deficiency
Endochondroma	Cushing's syndrome
Eosinophilic granuloma	Hyperparathyroidism
Fibrous dysplasia	Renal osteodystrophy
Giant cell tumor	Rickets
Nonossifying fibroma	Scurvy
Osteochondroma	**Neuromuscular Diseases (osteoporosis from disuse)**
Unicameral bone cyst	Cerebral palsy
Tumors—Malignant	Muscular dystrophy
Chondrosarcoma	Poliomyelitis
Ewing's sarcoma	Severe head injury
Neuroblastoma	Spina bifida with paraplegia
Osteogenic sarcoma	Traumatic paraplegia or quadriplegia
Hereditary Diseases	**Infections**
Gaucher's disease	Osteomyelitis
Neurofibromatosis	
Osteogenesis imperfecta	
Osteopetrosis	
Sickle cell disease	

Table 115.3. Differential Diagnosis of Pathologic Fractures

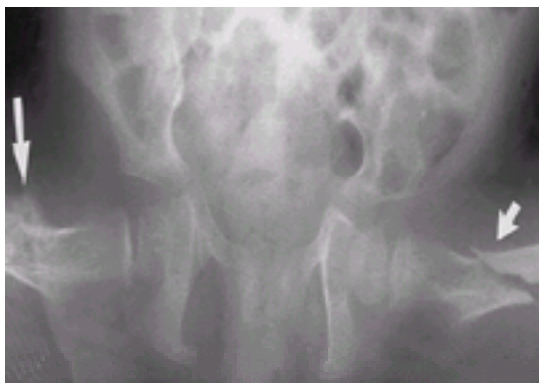

FIGURE 115.12. Radiograph of the pelvis and femur of an 18-month-old girl with osteogenesis imperfecta. There is a healing fracture of the right femur *(large arrow),* as well as an acute fracture of the left femur *(small arrow).*

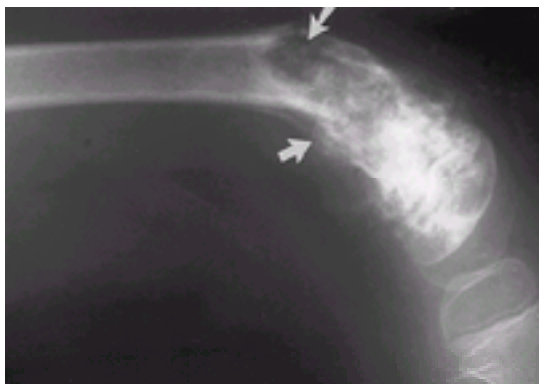

FIGURE 115.13. Radiograph of a 5-year-old girl with an osteosarcoma of the left femur showing an acute pathologic fracture *(arrows).* Amputation was ultimately necessary.

INJURIES OF THE UPPER EXTREMITIES

Injuries of the Shoulder Region

For the purposes of this discussion, injuries of the shoulder region are grouped as follows: 1) clavicular fractures, 2) scapular fractures, and 3) shoulder dislocations.

Clavicular Fractures

The clavicle ranks as the most commonly fractured bone in children. More than half of all clavicular fractures occur in children less than 10 years of age. In children younger than 2 years (excluding the newborn period), such fractures are

uncommon and should provoke consideration of intentional trauma. For the sake of discussion, clavicular injuries can be divided into fractures of the shaft, the medial end, and the lateral end.

Fractures of the clavicular shaft result from direct trauma and from indirect forces transmitted by falls onto an outstretched hand. Most are greenstick injuries of the midshaft; the thick periosteum enveloping the clavicle prevents significant displacement or angulation. The diagnosis is usually self-evident. Typically, a child complains of shoulder pain and is cradling the arm on the injured side with the opposite one. Occasionally, the initial injury is unnoticed and comes to attention only when a lump appears as callus forms. Radiographs are confirmatory, although visualization of nondisplaced fractures may require several views. (It can be debated whether radiographs are strictly necessary when the diagnosis is self-evident on physical examination.) Despite the proximity of the brachial plexus and subclavian vessels, neurovascular injury is rare other than when a violent direct blow results in significant displacement of the fracture fragments.

Medially, strong ligaments anchor the clavicle to the sternum. Eighty percent of the growth of the clavicle occurs at the medial physis. The last epiphysis in the body to close, the medial clavicular epiphysis, is rarely visible radiographically before 18 years of age. Apparent dislocations of the sternoclavicular joint are invariably epiphyseal separations in children and young adults. With such fractures, either anterior or posterior displacement can occur. The direction of displacement can often be determined by direct palpation. Radiographic visualization may be difficult; special views and/or CT scans are often required to define the degree and direction of displacement. Posterior displacement is of particular concern as compression of the mediastinal vessels and the trachea can result. Should there be evidence of neurovascular or respiratory compromise, prompt orthopedic consultation and closed reduction are indicated.

Laterally, the clavicle is anchored by the coracoclavicular and acromioclavicular ligaments. Once again, fracture through the physis rather than dislocation is the rule. The usual mechanism of injury is a direct blow to the point of the shoulder. Typically, the proximal fracture fragment is displaced superiorly; the radiographic appearance suggests acromioclavicular separation. However, the periosteum remains whole inferiorly, its ligamentous connections intact. As a result, most distal clavicular fractures heal uneventfully with no loss of joint stability (Fig. 115.14).

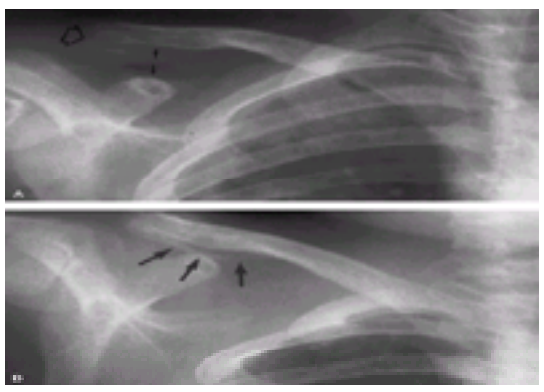

FIGURE 115.14. Radiograph of the right clavicle of a 5-year-old child. **A.** A lateral clavicular fracture *(open arrow)* and widening of the space between the clavicle and the coracoid process *(small arrows)* are evident on the initial film. **B.** The pattern of new bone formation *(arrows)* seen on the follow-up radiograph demonstrates that the periosteum and the ligaments have remained intact inferiorly.

Only rarely is orthopedic consultation or, for that matter, any follow-up necessary for a clavicular injury. Exceptions include significantly displaced midshaft fractures, for which closed reduction is occasionally desirable; posteriorly and significantly anteriorly displaced medial fractures; grossly unstable distal injuries; and all open fractures. Immobilization in a figure-of-eight dressing or a sling and swathe for 3 weeks followed by 3 weeks of restriction from sporting activities is adequate treatment for most shaft fractures. (See Section VII, Procedures, Application of Figure-of-Eight Harness .) Repeat radiographs are usually unnecessary given that nonunion is extremely unusual. It is best to inform parents that a lump will appear as callus forms and may persist for as long as a year. With medial and distal fractures, a sling is recommended along with progressive motion as the pain subsides.

Scapular Fractures

Fractures of the scapula are unusual in adolescents and rare in children. In the isolated instances in which they do occur, the usual mechanism is a severe direct blow, such as can be sustained in a fall from a height or a motor vehicle accident (Fig. 115.15). The same force that produces the scapular fracture may result in more concerning and potentially life-threatening injuries to the chest, neck, or head. Fractures of the body and neck of the scapula are usually well visualized on plain radiographs; adequate definition of glenoid injuries may require a CT scan. Although a sling and swathe is usually the only treatment necessary, orthopedic consultation is suggested given the rarity of these injuries.

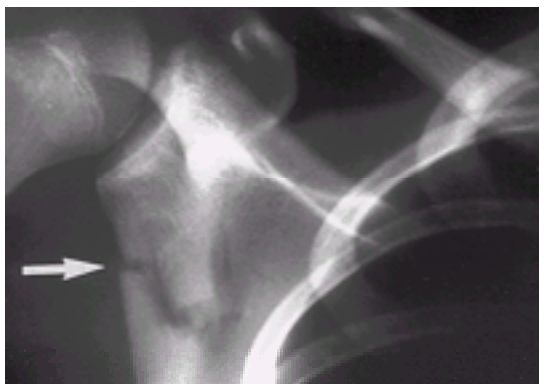

FIGURE 115.15. Radiograph of a 13-year-old boy who sustained an isolated right scapular fracture as the result of a skateboarding accident *(arrow).*

Shoulder Dislocations

Dislocations of the shoulder are unusual before physeal closure. Other than medially, the proximal humeral physis runs external to the shoulder capsule, and injuries that in an adult would cause dislocation result in fractures in children and skeletally immature adolescents. Most dislocations that do occur are anterior, as is the case with adults. Findings on physical examination include swelling and deformity with loss of the usual rounded contour of the shoulder. Palpation generally reveals the displacement of the humeral head anterior to the glenoid fossa. Signs of axillary nerve injury may be present. Radiographic studies should include an axillary (Y) view in addition to the customary views of the shoulder to best define the direction of displacement. As for treatment, closed reduction of anterior dislocations can be accomplished by any of a number of techniques, one of which is reviewed in detail in Section VII, Procedures. Postreduction radiographs should be performed routinely in part to ensure that no fracture has occurred in conjunction with the dislocation. Given their rarity, posterior dislocations merit orthopedic consultation before reduction. The rate of chronic shoulder instability and recurrent dislocation is high; even seemingly routine anterior dislocations should be immobilized in a sling and swathe for several weeks and referred to an orthopedist for subsequent care.

Fractures of the Humerus

In this section, humeral fractures are grouped as follows: 1) proximal humeral fractures and 2) humeral shaft fractures. Supracondylar fractures are discussed under Injuries of the Elbow.

Proximal Humeral Fractures

About 80% of the growth of the humerus occurs at the proximal humeral physis. As a result, the potential for fracture healing and remodeling with fractures that involve the proximal humeral shaft and physis is remarkable. Nonunion is unheard of; malunion is rare, other than with significantly displaced or angulated injuries in older adolescents. Before adolescence, most proximal humeral fractures are metaphyseal (Fig. 115.16), although Salter-Harris type I injuries are seen occasionally. With the onset of adolescence, rapid growth makes the physeal region relatively weak and thus vulnerable to injury. The incidence of proximal humeral fractures is highest in this age group; most are Salter-Harris type II injuries. Type III, IV, and V injuries are most unusual. Common mechanisms of injury include falls on an extended, adducted arm and direct blows to the shoulder.

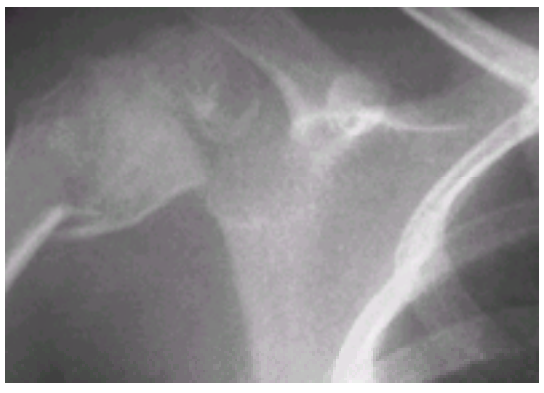

FIGURE 115.16. Impacted proximal right humeral fracture with approximately 25 degrees of angulation in a 3-year-old child. Full remodeling can be anticipated.

Physical findings with proximal humeral fractures range from mild swelling to obvious deformity and shortening of the arm. Routine radiographs are generally sufficient. Care must be taken not to confuse the normal variations in the epiphyseal line with a fracture; comparison views can be useful. Conservative management is the rule. Before adolescence, as much as 50 degrees or even 70 degrees of angulation is satisfactory. In younger children, even totally displaced fractures can remodel completely. Recommendations regarding the degree of deformity acceptable in adolescents vary somewhat; 20 to 50 degrees of angulation and 50% apposition are generally tolerable. The indications for open reduction are limited. A sling and swathe for several weeks is usually the only treatment necessary. Orthopedic follow-up is recommended.

Humeral Shaft Fractures

Fractures of the humeral shaft are much less common than those involving either the proximal or distal segments. The pattern of fracture reflects the mechanism of injury; transverse fractures result from direct blows, whereas spiral fractures are caused by indirect twisting, as with a fall. When a child less than 3 years of age sustains a spiral fracture of the humerus, the strong possibility of child abuse must be considered seriously (Fig. 115.17).

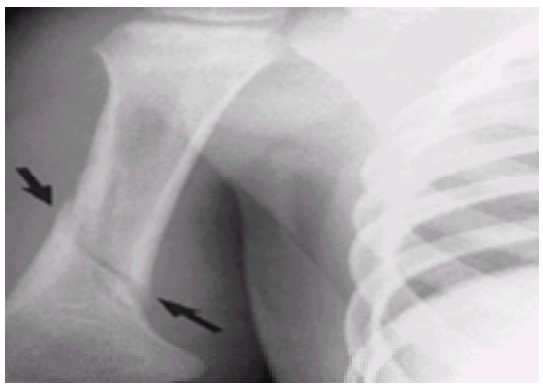

FIGURE 115.17. Spiral fracture of the right humerus in an 18-month-old girl. Although in this case the injury was accidental, spiral humeral fractures in children less than 3 years old must always evoke concerns about child abuse.

Many humeral fractures are obvious on physical examination, although only minimal swelling and tenderness may be present with buckle and greenstick injuries. Vascular injury is relatively uncommon. On the other hand, evidence of radial nerve injury must always be sought, particularly with a fracture that involves the distal two-thirds of the humeral shaft. Physical findings suggestive of damage to the radial nerve include loss of motor strength in the extensors of the wrist and fingers and loss of sensation on the dorsum of the hand in the web space between the thumb and index finger. Of note is that, with proper fracture management, nearly all cases of radial nerve palsy resolve. As for radiographs, anteroposterior and lateral views usually suffice. A prominent vascular groove in the distal humerus is a normal finding that should not be confused with a fracture.

The thick periosteal sleeve of the humeral shaft limits fracture displacement and promotes rapid healing. A sling and swathe is all that is needed for incomplete fractures. For complete or minimally displaced fractures, application of a sugar-tong splint of the upper arm, followed by a sling to support the forearm is recommended. In older children and adolescents, a hanging long arm cast is an alternative. Given the potential for overgrowth with healing, overriding of the fracture fragments by up to 2 cm is acceptable. Remodeling of as much as 40 degrees of angulation can be expected in younger children. Immediate orthopedic consultation is suggested for any completely displaced fracture, any fracture angulated more than 20 degrees in children and 10 degrees in adolescents, and any fracture with evidence of radial nerve injury. All humeral fractures should be referred for orthopedic follow-up within 5 days.

Injuries of the Elbow

Normal Anatomy and Radiographic Diagnosis

Of all the fractures encountered in the pediatric age group, those of the elbow rank as the most problematic in terms of diagnosis, treatment, and complications. For the emergency physician, an understanding of normal anatomy and normal radiographic findings ensures that misdiagnoses and untoward outcomes are uncommon. The elbow is a complex hinge joint composed of three separate articulations, namely those between the trochlea of the humerus and the ulnar notch, the capitellum and the radial head, and the proximal radius and ulna (Fig. 115.18). To further complicate matters, there are four growth centers within the distal humerus alone, and ossification of these growth centers begins at different but predictable times (Table 115.4). The ages shown in Table 115.4 are averages; ossification begins at an earlier age in girls than in boys, and much variation exists overall. When there is confusion about what is a normal growth center and what is a fracture fragment, comparison views of the uninjured elbow can be extremely helpful.

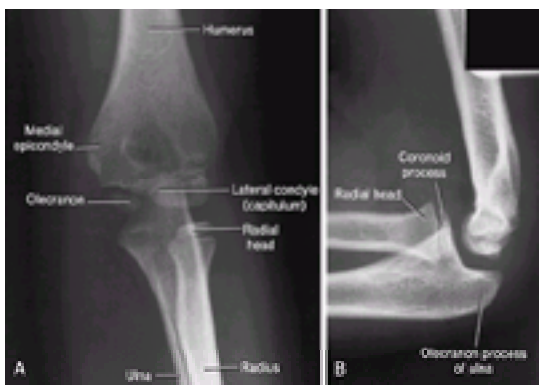

FIGURE 115.18. A. Anteroposterior radiograph of a normal elbow of a child. **B.** Normal lateral radiograph.

Capitellum	11 mo
Medial epicondyle	4-6 yr
Trochlea	9-10 yr
Lateral epicondyle	10-12 yr
Radial head	5-6 yr
Olecranon	6-8 yr

Table 115.4. Growth Centers of the Elbow: Average Age for Onset of Ossification

When a fracture is grossly displaced, the radiographic diagnosis is straightforward. Radiographic detection of subtle torus and nondisplaced fractures of the elbow joint is difficult. Close inspection of the radiographs for the presence of abnormalities of the fat pads and the anterior humeral line will prevent missed diagnoses in most cases. These radiographic signs are reliable only if the elbow has been properly positioned for the radiographs; the importance of a true lateral view in particular cannot be overemphasized. Of the two fat pads overlying the joint capsule along the distal humerus, only the anterior fat pad is normally visible on a lateral radiograph (Fig. 115.19). When fluid is in the joint space, as with a hemarthrosis from a fracture, these fat pads are displaced upward and outward (Fig. 115.20). If soft-tissue edema is extensive, one or both of the fat pads, although elevated, may be obscured. In the setting of known or suspected trauma, the presence of an abnormal fat pad sign should be considered a marker of an occult fracture and an indication for careful immobilization and close follow-up. Of note is that fractures of the distal humerus and of the proximal radius and ulna can all produce a hemarthrosis and thus positive fat pad signs. On occasion, oblique views will reveal the fracture line.

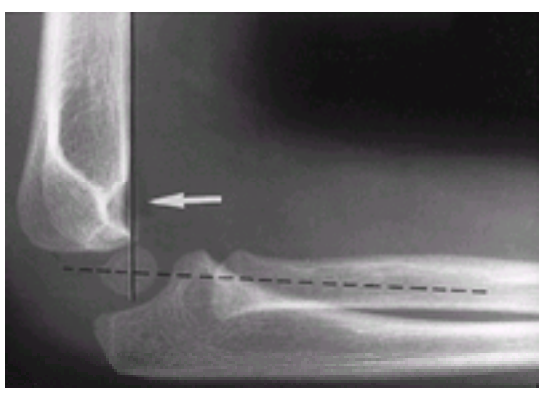

FIGURE 115.19. Normal lateral radiograph of the elbow of a 2-year-old child. The anterior fat pad is readily seen *(arrow);* the posterior fat pad is not visible. A line drawn along the anterior cortex of the humerus intersects the capitellum in its middle third *(solid line).* A line drawn along the axis of the radius also passes through the center of the capitellum *(dashed line).*

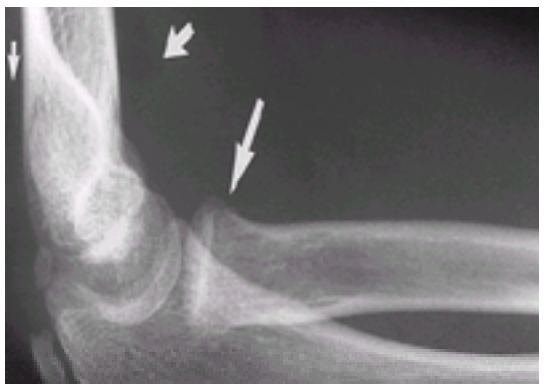

FIGURE 115.20. Lateral radiograph of the elbow of a 12-year-old girl, demonstrating marked elevations of both the anterior and posterior fat pads *(small arrows).* A subtle radial neck fracture is also visible *(large arrow).*

Close inspection of a true lateral view of the elbow for abnormalities of the anterior humeral line is also essential. In the normal elbow, a line drawn through the anterior cortex of the humerus intersects the capitellum in its middle third (Fig. 115.19). Because the most common mechanism of injury to the elbow is hyperextension, posterior displacement of the distal humerus is to be expected when a fracture occurs. As a result, the anterior humeral line, rather than intersecting the middle third of the capitellum, passes through its anterior third or even fails to intersect it all together (Fig. 115.21). Detection of abnormalities of the anterior humeral line in children less than 2½ years of age is complicated by the variable rates of ossification of the capitellum; once again, comparison views can be helpful in uncertain cases.

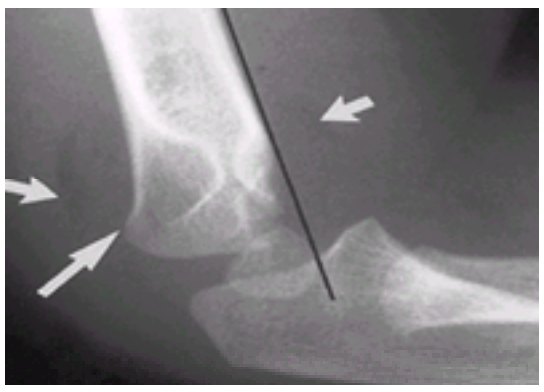

FIGURE 115.21. Lateral radiograph of the elbow of a 2-year-old girl. Once again, both the anterior and posterior fat pads are elevated *(small arrows).* In addition, the anterior humeral line passes along the anterior edge of the capitellum rather than through its center. Mild buckling of the posterior cortex of the distal humerus can be seen *(large arrow).*

In summary, errors in the management of pediatric elbow injuries can be minimized by an understanding of normal anatomy and development, careful interpretation of properly obtained radiographs, and immobilization with careful follow-up when there is even the mildest suspicion of a fracture. For the sake of discussion, elbow injuries are divided as follows: 1) supracondylar fractures, 2) lateral condylar fractures, 3) medial epicondylar fractures, 4) distal humeral physeal fractures, 5) olecranon fractures, 6) radial head and neck fractures, 7) elbow dislocations, and 8) radial head subluxation.

Some of these injuries are presented diagrammatically in Figure 115.22. Fractures of the medial humeral condyle and the lateral humeral epicondyle are rare and are not discussed.

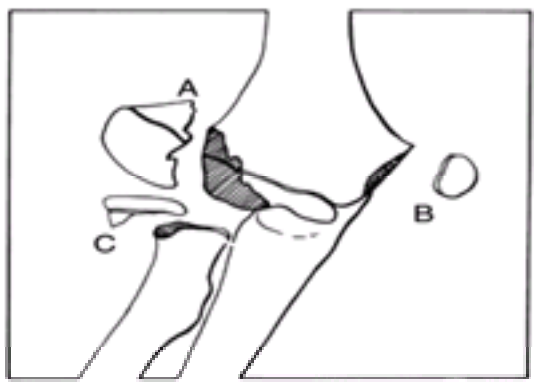

FIGURE 115.22. Common elbow fractures in children: lateral condylar fracture *(A);* medial epicondylar fracture *(B);* radial neck fracture *(C).*

Supracondylar Fractures

Supracondylar fractures account for more than 50% of the fractures of the elbow in the pediatric age group. Most are sustained by children 3 to 10 years of age. A fall on the outstretched arm with hyperextension of the elbow is the most common mechanism. Accordingly, posterior angulation or displacement of the distal fracture fragment nearly always occurs (Fig. 115.23). A direct blow to the posterior aspect of the elbow can lead to anterior angulation or displacement of the distal fragment, but such injuries are rare by comparison. With minimally displaced or nondisplaced fractures, recognition can be difficult. There may be only mild soft-tissue swelling. A suggestive history coupled with localized tenderness should prompt a radiologic examination. The radiographic findings also may be subtle. Close attention to the fat pads and the anterior humeral line, as detailed previously, facilitate diagnosis. At times, the actual fracture may be visualized only with an oblique view.

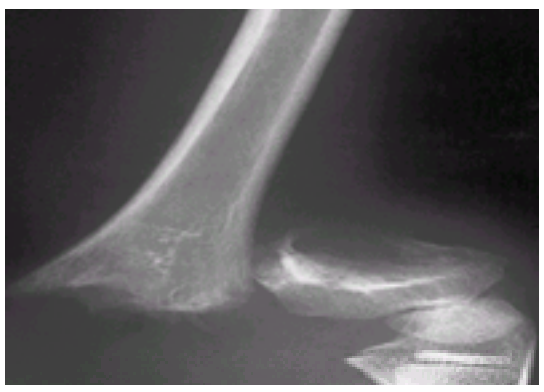

FIGURE 115.23. Displaced and rotated supracondylar fracture in an 8-year-old girl. The distal pulses were absent in this case but returned with reduction of the injury.

With more severe supracondylar injuries, the problem is not that of diagnosis (although a dislocated elbow may have a similar clinical presentation) but that of the recognition and prevention of complications. The complications associated with supracondylar fractures are multiple, ranging from immediate neurovascular compromise to long-term deformities and range-of-motion abnormalities. For the emergency physician, the first priorities are those of neurovascular assessment and fracture stabilization. The vascular examination should begin with palpation of the distal pulses and assessment of capillary refill. Use of a Doppler may allow detection of distal arterial flow when no pulse can be palpated. Absence of a pulse by itself is not extremely worrisome. Direct vascular injury is uncommon. In most instances, vasospasm or arterial compression has occurred instead, and arterial flow will resume with fracture reduction. On the other hand, significant muscle ischemia can be present even when pulses and capillary refill are judged to be normal. Forearm pain, pain with passive extension of the fingers, paralysis of finger extension, and paresthesias are each worrisome and should be considered evidence of an impending compartment syndrome.

Neurologic deficits, usually transient, are also common with supracondylar fractures. Radial, medial, and ulnar nerve palsies all occur, as do isolated injuries of the anterior interosseous nerve (a motor branch of the median nerve). Table 115.5 outlines the innervation of these nerves and should be used as a guide to the neurologic examination. Once the neurovascular examination is completed and *before* radiographic studies, all displaced supracondylar fractures must be immobilized. It is usually best to simply splint the limb in the deformed position in which it lies. More than 20 to 30 degrees of elbow flexion will place undue tension on the neurovascular structures and should be avoided. All patients must have nothing by mouth because reduction, whether open or closed, requires general anesthesia. Frequent repeat neurovascular examinations should be performed and documented.

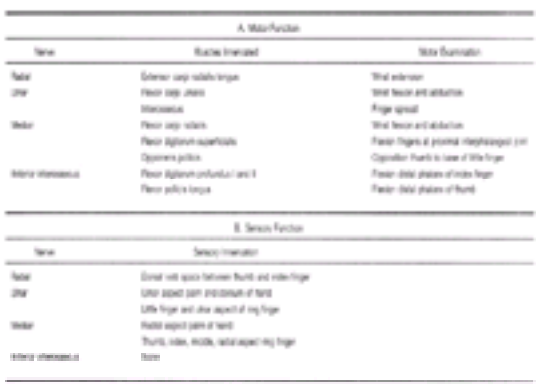

Table 115.5. Guide to the Neurologic Examination of the Distal Upper Extremity

Minimally displaced or nondisplaced supracondylar fractures may be immobilized in a well-padded long-arm posterior splint with the elbow at 90 degrees and the forearm in pronation or neutral rotation. Orthopedic referral for casting is suggested when the swelling subsides. Immobilization for a total of 3 weeks is adequate in most cases. All nonminimally displaced supracondylar fractures require immediate orthopedic referral.

Lateral Condylar Fractures

Fractures of the lateral condyle, like those of the supracondylar region, are prone to poor functional outcome if misdiagnosed or mismanaged. Unlike supracondylar fractures, lateral condyle injuries involve the articular surface; they are true Salter-Harris type IV injuries. The most commonly proposed mechanism of injury is a varus stress on the elbow, as can occur with a fall on an extended and abducted arm. The lateral ligament and the common extensor tendon remain attached to the fracture fragment, which can be partially or totally avulsed from the distal humerus (Fig. 115.24). Clinically, swelling, ecchymosis, and tenderness localized over the lateral aspect of the elbow should suggest a lateral condylar fracture. With severely displaced fractures, routine anteroposterior and lateral views usually provide adequate fracture definition. With less severe injuries, the fracture line and the degree of displacement may be evident only on oblique views. On occasion, stress views, an arthrogram, or CT scan may be needed to adequately visualize the extent of injury.

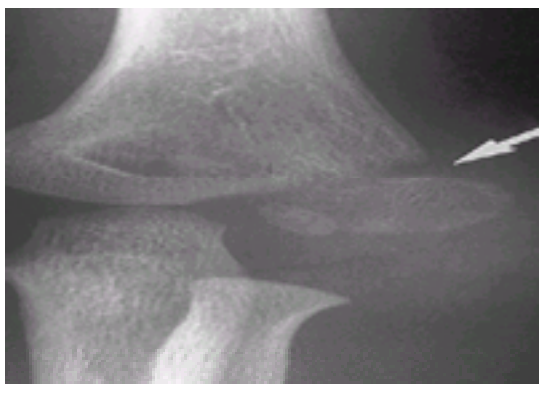

FIGURE 115.24. Lateral condylar fracture in a 2-year-old girl *(arrow).*

For minimally displaced and nondisplaced injuries, immobilization in a posterior splint with the elbowflexed to 90 degrees

and the forearm in pronation (some authorities suggest supination instead) is satisfactory emergency management. Lateral condylar fractures as a group are inherently unstable and prone to displace despite immobilization; orthopedic follow-up within 3 to 4 days is essential. All fractures displaced more than 2 mm require reduction and often pinning.

Medial Epicondylar Fractures

Fractures of the medial epicondyle occur both as the result of falls directly onto the elbow and falls onto the outstretched arm in which the elbow is subjected to a valgus stress. With the latter mechanism, the flexor muscles of the forearm avulse the medial epicondyle from the humerus (Fig. 115.25). Medial epicondyle injuries are particularly common with elbow dislocations. The physical findings are those that would be expected, namely swelling and tenderness localized to the medial aspect of the elbow. Valgus instability may be evident. Given its proximity, paresis of the ulnar nerve can occur. Oblique views and comparison views may be needed on occasion. The diagnosis is particularly problematic before the onset of ossification of the medial epicondyle at 4 to 6 years of age; fortunately, it is an uncommon injury in younger children. Open reduction is almost invariably necessary for displaced fractures. Nondisplaced fractures can be placed in a posterior splint with the forearm in pronation. Orthopedic follow-up is encouraged strongly, as it is with most elbow injuries.

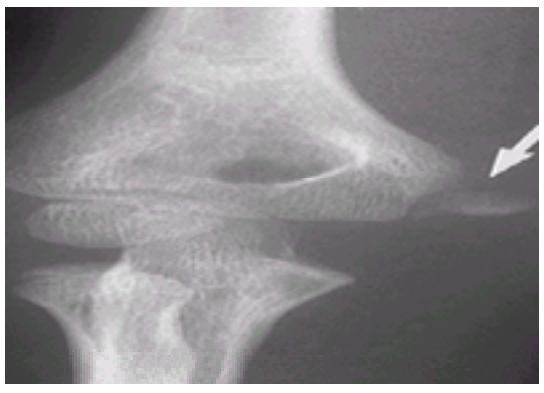

FIGURE 115.25. Displaced fracture of the medial epicondyle in an 8-year-old girl *(arrow).* Note the extensive soft-tissue swelling.

Distal Humerus Physeal Fractures

Fractures of the entire distal humerus physis are relatively uncommon. Most such injuries take place in children less than 2½ years of age; nearly all the remainder are sustained by children less than 7 years of age. Recognition is both difficult and important, especially in infants, in whom this particular injury is often the result of child abuse. The proposed mechanism in abused children is forceful twisting of the arm that shears off the distal epiphysis. In children 5 to 7 years of age, a fall on an extended arm with hyperextension of the elbow usually results in a supracondylar injury but occasionally can lead to a fracture of the distal humerus physis instead.

Elbow swelling without significant deformity is the usual clinical finding. When displacement is significant, the appearance may mimic that of an elbow dislocation. The latter, however, is an injury of early adolescence. Radiographic diagnosis can be difficult, particularly in infants in whom the capitellum has not yet begun to ossify. Posteromedial displacement of the ulna and radius in relation to the humerus is the most important finding. Recognition of this displacement may necessitate comparison views. Given the difficulty in recognition and the frequent need for reduction and pinning, all suspected epiphyseal separations of the distal humerus merit immediate orthopedic referral. In addition, the strong possibility of abuse needs to be considered seriously with this injury in children under 3 years of age.

Olecranon Fractures

Isolated fractures of the olecranon are seen only rarely. More often than not, they occur in conjunction with another injury of the elbow, in particular a fracture or dislocation of the radial head. Various mechanisms have been described, including sudden flexion of the elbow when the triceps is strongly contracted (essentially an avulsion injury) and direct trauma. Physical findings range from swelling localized to the olecranon to a marked hemarthrosis. Elbow extension may be weak or lacking altogether. Nondisplaced fractures may be somewhat difficult to discern radiographically; fat pad abnormalities are commonplace, however, and should be viewed as presumptive evidence of a bony injury (Fig. 115.26). A nondisplaced olecranon fracture can be splinted in partial extension and referred for orthopedic follow-up. Displaced fractures often require open reduction and internal fixation. Isolated olecranon fractures almost invariably heal quickly and without significant complications.

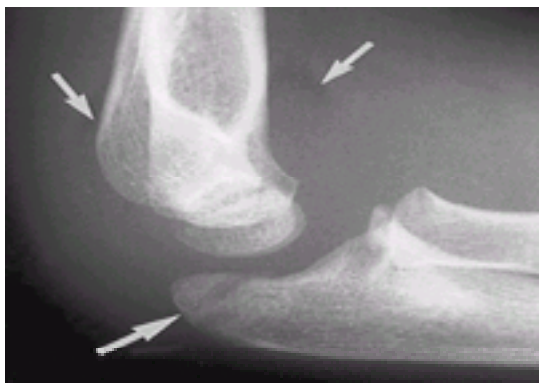

FIGURE 115.26. Nondisplaced fracture of the olecranon in an 8-year-old boy *(bottom arrow).* Note the elevated fat pads *(top arrows).*

Radial Head and Neck Fractures

Falls on an outstretched, supinated arm account for most fractures of the radial head and neck. Salter-Harris types I and II and pure metaphyseal (i.e., radial neck alone) injuries are the most common. Involvement of the epiphysis (i.e., radial head), which is largely cartilage in childhood, is rare. The physical examination typically reveals localized swelling and ecchymosis. Tenderness overlying the proximal radius strongly suggests the diagnosis. Of note is that pain may be referred to the wrist and thus distract from the true injury (Fig. 115.27). As for radiographic diagnosis, oblique and comparison views can clarify the diagnosis in uncertain cases. When the metaphysis alone is injured, a hemarthrosis may be absent and the fat pads normal. Associated fractures are common. The incidence of complications, especially loss of motion and overgrowth of the radial head, is significant. For this reason, orthopedic referral is recommended for all radial head and neck fractures. Immobilization with the elbow in 90 degrees of flexion and the forearm in neutral rotation is acceptable emergency management for minimally displaced or nondisplaced fractures. Angulation of greater than 15 degrees is an indication for immediate orthopedic consultation.

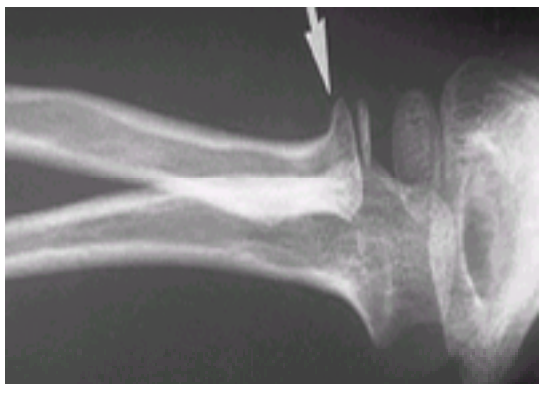

FIGURE 115.27. Buckle fracture of the radial neck in a 9-year-old girl. Wrist pain was the chief complaint. The treating physician failed to identify the proximal radial fracture, which was, however, noticed by the radiologist.

Elbow Dislocations

The elbow is dislocated more often than any other major joint in children and adolescents. Nonetheless, it is an unusual injury. As discussed previously, the ligaments and tendons are relatively stronger than the neighboring bones (particularly the physeal plates) in children; injuries that would lead to dislocations in adults almost invariably result in fractures in the younger age group. It is not surprising, then, that dislocations of the elbow are accompanied by significant soft-tissue and bony damage. A fall on an extended or partially flexed arm with the forearm in supination is the usual mechanism of injury. Accordingly, the radius and ulna are displaced posteriorly and, in most cases, laterally (Fig. 115.28). The anterior capsule is torn and the medial collateral ligament typically ruptured. Fractures of the medial epicondyle, coronoid process, olecranon, and proximal radius are the most commonly associated bony injuries.

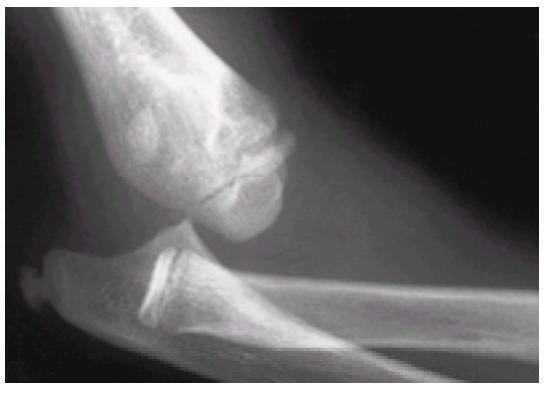

FIGURE 115.28. Elbow dislocation in an 8-year-old girl. A displaced fracture of the medial epicondyle was evident on the postreduction radiographs.

Major neurovascular compromise often accompanies elbow dislocations. True arterial rupture is seen almost exclusively with open dislocations but has been described on occasion with closed injuries. When reduction of the dislocation fails to relieve arterial compromise, further investigation regarding the extent of vascular injury is warranted. Overall, the risk of compartment syndrome with dislocations of the elbow is such that some authors recommend hospitalization for close observation even after successful closed reduction. Nerve injury, particularly of the ulnar nerve, is even more common than vascular injury. Ulnar nerve lesions typically occur when the medial epicondyle is avulsed and then entrapped in the joint. Early recognition and appropriate treatment of such entrapment nearly always lead to complete recovery of ulnar

nerve function. Median nerve entrapment is much rarer, but when it occurs, the degree of nerve damage is such that full recovery cannot be guaranteed. Moreover, recognition of median nerve injury is made difficult by the relative lack of pain and the subtlety of the initial motor and sensory deficits.

Clinical findings with dislocation of the elbow include obvious deformity and significant swelling. The forearm appears shortened. Often, the ulnar notch can be palpated posteriorly, and the humeral head can be detected as a fullness in the antecubital fossa. The importance of a thorough and well-documented neurovascular examination should be obvious from the preceding discussion. Immobilization before radiographic studies is recommended to minimize the risk of further neurovascular injury. Standard radiographic views are satisfactory. They should be closely inspected for the direction of the dislocation and for the presence of associated fractures. Although most elbow dislocations can be reduced uneventfully, the risks of entrapping a fracture fragment or a nerve in the joint space during the procedure are such that immediate orthopedic consultation is recommended. A number of techniques for reduction have been described. Either conscious sedation or general anesthesia should be considered. If orthopedic consultation is unavailable, the child should be placed prone and the forearm allowed to hang over the side of the stretcher. The physician should then encircle the upper arm with his or her hands and use the thumbs to push the olecranon forward and downward. Whatever technique is used, hyperextension should be avoided at all times. Postreduction films are mandatory because only then will many of the associated fractures be evident. Finally, the arm should be immobilized in a posterior splint with the elbow at 90 degrees and the forearm in midpronation.

Radial Head Subluxation

Of all the injuries discussed in this section, by far the most common is radial head subluxation, otherwise known as “nursemaid's elbow” or “pulled elbow” (see Procedures). Pathologically, radial head subluxation occurs when the annular ligament becomes partially detached from the head of the radius and slips into the radiohumeral joint where it is entrapped. The usual mechanism is that of axial traction on an extended and pronated arm. Radial head subluxation is an injury of children a few months to 5 years of age. After 5 years of age, the strength of the annular ligament is such that the injury is uncommon.

The classic history is that of a child who cries with pain and refuses to use an arm after being pulled or lifted by that same arm. With some regularity, however, the history is one of a fall. In infants, radial head subluxation can occur when an extended arm is trapped beneath the trunk as the child is rolled over. The chief complaint is typically that the child is not using the arm; concerns about a wrist or a shoulder injury are common. Children with radial head subluxation uniformly hold the arm in pronation with the elbow slightly flexed. Much more often, the degree of distress is minimal, although supination, pronation, and elbow flexion usually elicit pain. Mild tenderness may be noted with palpation of the radial head. Significant point tenderness or swelling should suggest an alternative diagnosis (e.g., a supracondylar fracture). The radiographic findings with radial head subluxation are minimal at best. Radiographs are not routinely recommended when the history and clinical presentation are classic. Indeed, the subluxation is often reduced when the radiology technician places the forearm in supination for the anteroposterior view.

As for treatment, various reduction techniques have been described. The most widely used is that described in Section VII, Procedures, namely, supination and flexion. Should that approach prove unsuccessful, either supination or pronation with elbow extension should be attempted. When reduction succeeds, the child typically uses the arm normally within 5 to 10 minutes. The delay until normal use is longer in younger children and when there has been greater than a 4- to 6-hour period between injury and treatment. When there is no evidence of recovery, the diagnosis must be reconsidered; fractures of the elbow and clavicle in particular should be excluded because the clinical presentations can be similar. With recurrent subluxations, immobilization for a few weeks in a posterior splint with the elbow at 90 degrees and the forearm supinated is suggested. Note that even when efforts at closed reduction fail, spontaneous reduction almost invariably occurs. The need for open reduction is exceedingly rare.

Fractures of the Forearm and Wrist

Children fracture the radius and ulna more often than all bones other than the clavicle. Fortunately, the incidence of neurovascular complications is low and the potential for healing with proper management high. In many instances, the emergency physician can provide the satisfactory initial, if not definitive, management for forearm injuries. However, certain types of fractures require immediate orthopedic referral, and as such they receive particular emphasis. In this section, forearm fractures are divided as follows: 1) fractures of the radial and ulnar shafts, 2) Monteggia and Galeazzi fracture dislocations, 3) fractures of the distal radius and ulna, and 4) fractures of the bones of the wrist.

Fractures of the Radial and Ulnar Shafts

The usual mechanism of injury with forearm fractures, including those of the radial and ulnar shafts, is a fall on an outstretched hand. Direct blows account for some injuries, displaced and open shaft fractures in particular. Approximately three-quarters of all shaft fractures involve the distal third of the shaft; most of the remainder involve the midshaft. The clinical findings generally make the diagnosis self-evident. A number of fracture patterns are seen; greenstick injuries are especially common. Standard radiographic views are sufficient other than with suspected bowing fractures when comparison views may be necessary. The emergency physician should insist that radiographs of the forearm always include both a true lateral and a true anteroposterior view and both the elbow and the wrist. In general, isolated ulnar fractures do not occur, as is discussed further in the next section.

The periosteum and remaining intact cortex limit the degree of angulation with greenstick injuries. Keep in mind that many greenstick fractures have a significant rotational deformity and that the degree of angulation alone should not determine the need for closed reduction. It must also be remembered that the potential for remodeling decreases with the distance from the epiphysis and with the age of the child. Less angulation is therefore accepted in midshaft fractures than in more distal injuries and in adolescents than in younger children. Although it is hard to make any absolute rules, any shaft fracture angulated more than 10 degrees merits immediate orthopedic consultation, at least by telephone. This is

not to say that all such fractures will require reduction. (Another simple rule is that any forearm that looks crooked should be straightened.) Dorsal angulation is usual; immobilization with the arm in supination minimizes the tendency of the forearm muscles to cause further deformity.

Complete fractures can be particularly problematic, again because significant angulation can occur. Should the ends of the bones be well opposed and angulation and rotation be minimal, a well-applied sugar-tong splint is adequate initial treatment. Otherwise, immediate orthopedic referral is necessary. Closed reduction, although not always as simple as it may appear, is preferable (Fig. 115.29). In children older than 10 to 12 years, adequate alignment is often obtained only with open reduction and internal fixation.

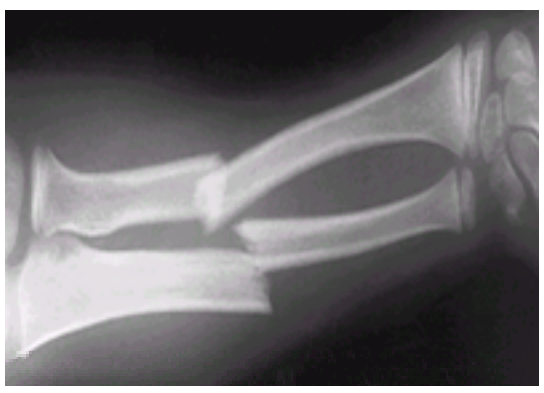

FIGURE 115.29. Complete fractures of the midshafts of the radius and ulna in a 9-year old boy. Efforts at closed reduction failed; internal fixation was necessary.

Recognition of when a bowing fracture has occurred is crucial simply because the potential for remodeling with such injuries is minimal (see Fig. 115.4). Failure to correct bowing can result in permanent loss of supination and pronation. As already mentioned, in the absence of obvious deformity, comparison views may be necessary before the true extent of bowing can be appreciated. Again, no hard and fast rules regarding indications for closed reduction are offered; however, any bowing fracture that causes obvious forearm deformity or restrictions of pronation or supination certainly merits immediate orthopedic referral.

Monteggia and Galeazzi Fracture Dislocations

In general, isolated fractures of the ulna do not occur. Instead, the same force that causes the ulnar fracture leads to a radial injury, in some instances a dislocation of the radial head. It is this combination of an ulnar fracture and a radial head dislocation that is known as a Monteggia fracture. Recognition is most important because failure to reduce the radial head dislocation results in permanent disability. Clues to the diagnosis on physical examination include elbow pain and swelling, which accompany signs of any ulnar fracture. Palpation may confirm the dislocation of the radial head, which may be displaced anteriorly, posteriorly, or laterally, depending on the mechanism of injury. A palsy of the posterior interosseous nerve, a motor branch of the radial nerve, may also be present.

If the radial head dislocation is to be recognized radiographically, the rule that a line drawn through the axis of the radius should pass through the center of the capitellum on all projections must be remembered (Fig. 115.30). Once again, the need for a true lateral view that includes the elbow with all forearm studies must be emphasized. Even bowing fractures of the ulna, which may require comparison views for recognition, are associated with radial head dislocation. Any suspected Monteggia injury requires immediate orthopedic referral.

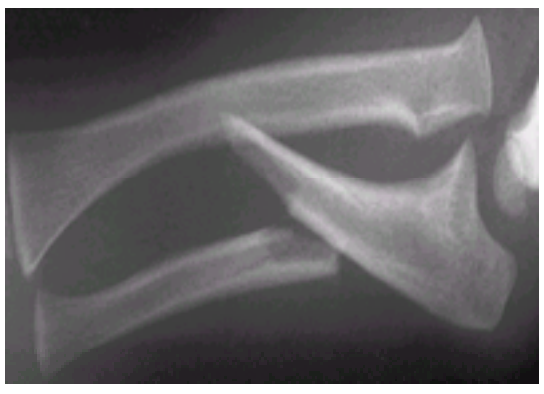

FIGURE 115.30. A Monteggia fracture in a 3-year-old boy. Note that a line drawn along the axis of the radius would fail to intersect the capitellum (compare with Fig. 115.29.)

The Galeazzi fracture is a radial shaft fracture, generally at the junction of the middle and distal thirds, that is accompanied by disruption of the distal radioulnar joint. It is relatively rare. Physical examination reveals prominence of the distal ulna and joint instability. Radiographs are confirmatory. Once again, orthopedic consultation is necessary. The complications are few with proper management.

Fractures of the Distal Radius and Ulna

Distal radial and ulnar fractures bear special mention, not because of any undue rate of complications, but rather because of their overall frequency. Of all the fractures that occur in childhood and adolescence, those of distal forearm are by far the most common. Except for the occasional instance of nerve entrapment at the time of reduction of a complete fracture, significant neurovascular complications are rare. Overall, the capacity for remodeling is significant. The difficulties facing the emergency physician are those of diagnosis with subtle fractures and of recognition regarding when reduction is necessary with displaced fractures.

More often than not, localized swelling and tenderness accompany distal radial fractures and can guide interpretation of the radiographic studies. However, wrist pain can be the chief complaint with more proximal injuries, for example, radial head fractures. Once again, the need for studies that include the whole forearm must be reinforced. Torus fractures are most often overlooked. Often, the location of the soft-tissue swelling on the radiographs helps highlight the position of the fracture, which may be evident on only one projection and then only as a minor irregularity in the contour of the cortex. A fracture of the ulnar styloid should also prompt a diligent search for a radial injury. Ulnar styloid fractures only rarely occur in isolation; as a rule, they are accompanied by either a torus or physeal fracture of the radius. When a torus fracture is identified, a volar splint or, if the swelling is minimal, a short arm cast for 3 to 4 weeks is recommended. Orthopedic referral is optional.

Greenstick and complete fractures are readily recognized. What must be remembered is that such fractures have a tendency to displace if not properly immobilized. The distal fragment is angulated posteriorly in most greenstick and complete fractures of the distal forearm. Angulation of greater than 10 to 15 degrees is an indication for immediate orthopedic referral (Fig. 115.31). Otherwise, immobilization with orthopedic follow-up within 3 to 5 days is adequate emergency management. Although there is some disagreement, immobilization with the forearm in supination is thought to decrease the likelihood of further displacement. Accordingly, either a long arm posterior splint or a well-applied sugar-tong splint are recommended with all greenstick and complete radial and ulnar fractures. Short arm volar splints should be reserved for torus injuries.

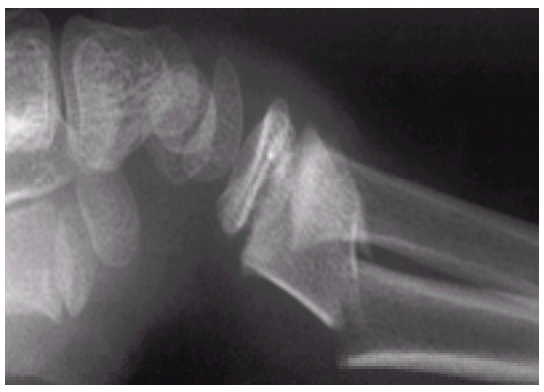

FIGURE 115.31. Complete fracture of the distal radius of a 9-year-old child demonstrating 35 degrees of posterior displacement of the distal fragment. Closed reduction was uneventful.

Salter-Harris type I and II injuries of the distal radial physis rarely lead to growth disturbance, which is fortunate because they are common injuries, particularly from 6 to 12 years of age. The issue again is one of recognition with these fractures. When point tenderness on the physical examination is accompanied by swelling localized to the distal radius on the radiograph, the presumptive diagnosis should be a Salter-Harris type I injury even when there is no obvious displacement of the epiphysis. Immobilization and orthopedic referral are recommended. Closed reduction is needed for all displaced physeal fractures. Of note is that the risk of growth disturbance increases with repeated and delayed manipulations.

Fractures of the Bones of the Wrist

The carpal bones are rarely fractured during childhood and adolescence. Adolescents in the later stages of skeletal maturity sustain scaphoid (navicular) fractures. Most injuries of the scaphoid in adolescence are nondisplaced fractures through the distal third of the bone (Fig. 115.32). The rate of nonunion is much lower than in adults, in whom scaphoid fractures generally involve the middle third of the bone and are more often displaced. The usual mechanism is a fall on an outstretched arm with extreme hyperextension of the wrist. Physical findings that should suggest the possibility of a scaphoid fracture include snuffbox tenderness, pain with supination against resistance, and pain with longitudinal compression of the thumb. As with adults, radiographic visualization of a nondisplaced scaphoid fracture may be difficult even with special views. Should the physical signs suggest a scaphoid fracture, immobilization in a thumb spica splint or cast for 2 weeks is recommended, regardless of the radiographic findings. At that time, radiographs should be repeated; fractures not detectable on the initial films should now be evident. Should radiographs remain normal but clinical suspicions high, a bone scan or an MRI study should be considered. Although the likelihood of complication is low, orthopedic referral is recommended once a scaphoid fracture is identified.

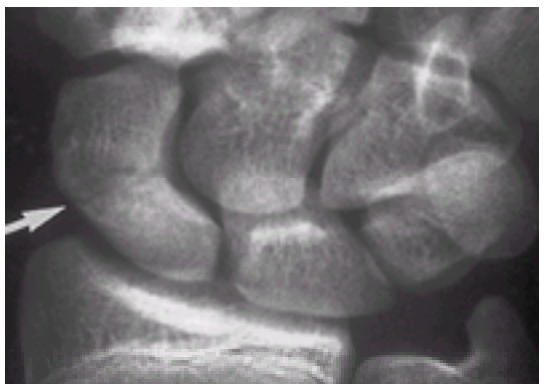

FIGURE 115.32. A scaphoid fracture in a 16-year-old boy *(arrow).* In this case, the fracture is through the middle third of the scaphoid; fractures through the distal third are actually more common during adolescence.

Injuries of the Hand and Fingers

By comparison with adults, children sustain relatively few bony injuries of the hand. The most commonly encountered hand injuries in the pediatric ED are crush injuries of the distal phalanx, in which lacerations and fractures often coexist. It has been stated that such injuries are often undertreated; definitive management often entails removal of the nail, repair of any nailbed injury identified, careful immobilization, and close follow-up. (See Chapter 116 for additional discussion of the management of such crush injuries.)

A wide variety of other injuries, including a whole array of avulsion and physeal fractures, also occur. The types of injuries seen at a given joint reflect the underlying complex anatomy of the tendons and ligaments of the hand, a full discussion of which is beyond the scope of this chapter. However, by adhering to a few basic principles of physical and radiographic diagnosis, the emergency physician should have little difficulty in recognizing which injuries merit referral to a hand specialist.

Given the risks of permanent deformity and stiffness, all displaced fractures, particularly those extending intra-articularly, should be referred. Such fractures are generally self-evident. Before concluding that a fracture is a simple nondisplaced one and thus amenable to routine splinting, the practitioner must first assure himself that there is no accompanying malrotation or joint instability. Malrotation is not always that apparent when the fingers are extended; it becomes much more obvious with finger flexion (Fig. 115.33). Joint stability must also be assessed in flexion and extension, as well as in both the lateral and anteroposterior planes. Adequate examination may be possible only after performance of a digital block. Should either malrotation or joint instability be detected, consultation is again in order.

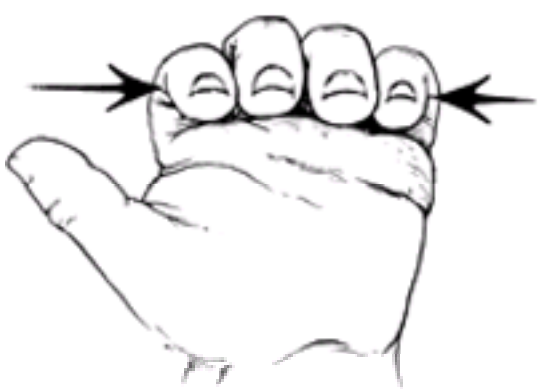

FIGURE 115.33. It is important to check for malrotation with all fractures of the metacarpals and phalanges. When flexed as shown, the fingers should all point in the same direction. If malrotation is present, overlapping will occur.

When the hand is radiographed, oblique views should be included in addition to the usual anteroposterior and lateral projections. Interpretation of the radiographs is complicated by the presence of multiple epiphyses and secondary ossification centers. It is essential to remember that the epiphyses of the phalanges and of the thumb metacarpal are located at the proximal ends of the bones. The growth centers of the remaining metacarpals are distal. Once again, the number of fractures missed will be minimized if close attention is paid to the physical findings and the presence of soft-tissue swelling on the radiographs. For the purposes of further discussion, hand injuries are divided as follows: 1) metacarpal fractures and 2) phalangeal fractures and dislocations.

Metacarpal Fractures

Perhaps the most commonly encountered metacarpal fracture in pediatrics is one of the distal fifth metacarpal in an adolescent who has struck someone or something with a closed fist. The equivalent of a boxer's fracture in an adult, these fractures are metaphyseal rather than physeal injuries and are typically angulated. Closed reduction is usually performed if the angulation is more than 30 to 40 degrees. Salter-Harris type I and II injuries occur on occasion, primarily in the second, third, and fourth metacarpals. Nondisplaced injuries may be immobilized in a gutter splint with the wrist neutral and the metacarpal phalangeal joints at 70 degrees and then referred (Fig. 115.34 and Fig. 115.35). If they are not displaced or rotated, metacarpal shaft fractures can be managed similarly. Proximal metacarpal fractures of the second through fifth metacarpals are rarely displaced; recognition is more of an issue than management (Fig. 115.36).

However, angulation is common with proximal fractures of the thumb metacarpal. Metaphyseal and Salter-Harris type II and III injuries occur and when displaced require closed reduction.

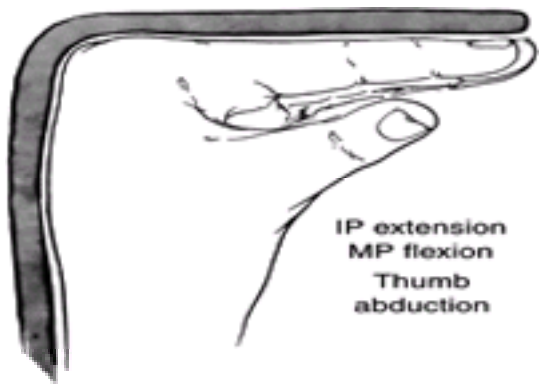

FIGURE 115.34. The hand should be splinted in this position to prevent extension contractures of the metacarpophalangeal *(MP)* joints and flexion contractures of the interphalangeal *(IP)* joints.

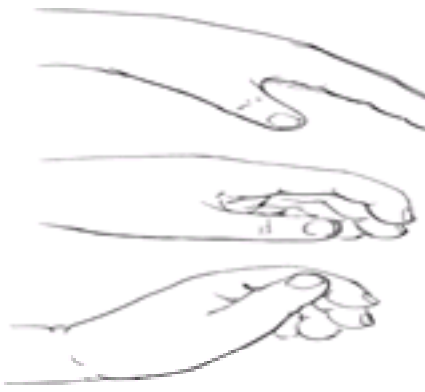

FIGURE 115.35. Splinting the hand in any of the positions shown here tends to promote joint contractures and recurrence of deformity.

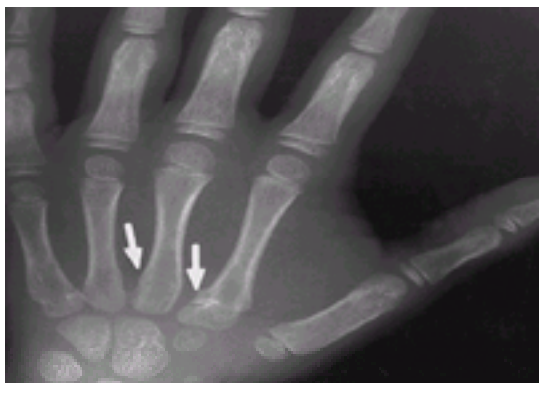

FIGURE 115.36. Radiograph of the left hand of an 8-year-old girl showing fractures of the proximal second and third metacarpals *(arrows).* Significant displacement is unusual with proximal metacarpal fractures.

Phalangeal Fractures and Dislocations

As already mentioned, distal phalanx fractures typically accompany crush injuries of the fingertip. If only the distal tuft is fractured, anatomic closure of the laceration usually results in adequate realignment of the fracture. Displaced physeal fractures merit immediate referral (Fig. 115.37). Hyperflexion injuries of the distal phalanx, leading to so-called mallet finger deformities, are also common. In the child, Salter-Harris type I or II injuries result, whereas type III injuries are the rule in adolescents. The latter often require open reduction and internal fixation. In either case, examination reveals an extension lag at the distal interphalangeal joint. Recognition of the tendonous disruption is important, given that proper treatment entails 6 to 8 weeks of continuous splinting in hyperextension.

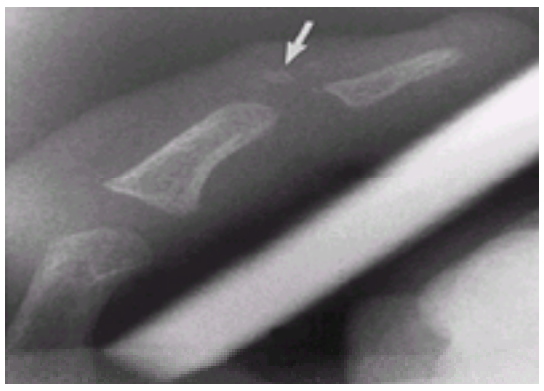

FIGURE 115.37. Displaced Salter-Harris type I fracture of the distal phalanx in an 11-month-old girl that resulted from a crush injury. The *arrow* points to the epiphysis, which is only beginning to calcify. Careful reduction is necessary if the risk of growth disturbance is to be minimized.

A whole range of proximal and middle phalangeal fractures occur, many of which the emergency physician can manage successfully (Fig. 115.38). Gutter splints that incorporate both the injured and an adjacent uninjured finger are used commonly with the positioning discussed previously. Complete fractures almost invariably angulate as the result of the actions of the intrinsic muscles, the direction of angulation determined by the position of the fracture relative to the flexor and extensor tendons. Phalangeal neck fractures are of particular concern in that complete fractures can rotate by as much as 90 degrees and unicondylar fractures are prone to displacement. The radiographic findings may be subtle; the consequences of improperly evaluating such injuries are certainly substantial. In the proximal phalanx, particularly that of the fifth finger, laterally angulated Salter-Harris type II fractures are common. If the displacement is minimal, splinting with follow-up in 3 to 5 days is acceptable.

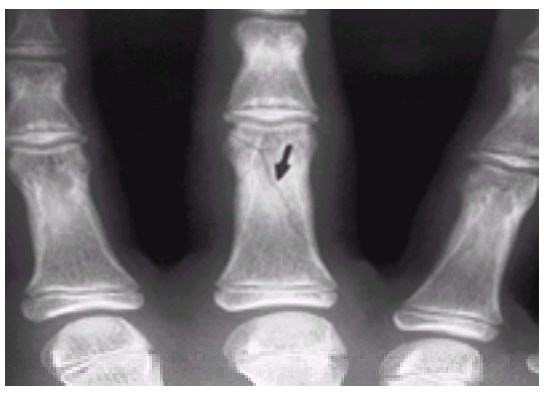

FIGURE 115.38. Oblique fracture of the proximal phalanx of the right third finger in a 12-year-old boy *(arrow).* Before splinting such an injury, the emergency physician must first make certain that no malrotation is present.

Special mention should be made of the so-called gamekeeper's or skier's thumb, an avulsion of the ulnar collateral ligament of the proximal phalanx of the thumb. Localized tenderness should raise concerns about this injury and prompt an assessment of the joint for adduction stability with the metacarpal joint extended and in 30 degrees of flexion. In the pediatric age range, this is a Salter-Harris type III injury. When there is evidence of only minor instability (firm endpoint, increased laxity of less than 30 degrees), thumb spica splinting for 3 to 6 weeks is generally sufficient. More severe injuries require operative intervention. Consultation is suggested when such injuries are suspected.

Despite the strength of the ligaments and tendons, hyperextension can lead to dislocations of the metacarpophalangeal and proximal interphalangeal joints in children. Dislocations of the proximal interphalangeal joints can usually be readily reduced (Fig. 115.39). After a digital block, the joint should be gently hyperextended and the distal bone then pushed back into place. Radiographs both prereduction and postreduction should be scrutinized for fractures and the stability of the collateral ligaments carefully assessed. Buddy taping for 3 weeks is adequate for routine dislocations (Fig. 115.40).

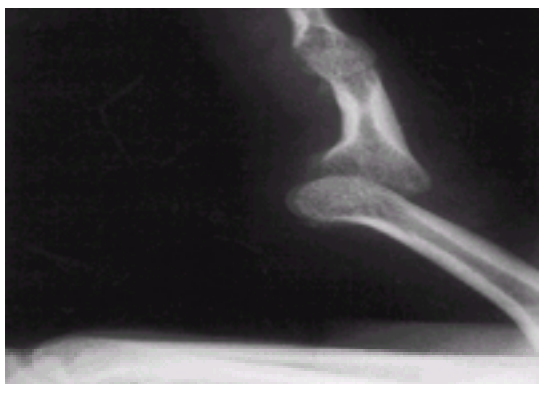

FIGURE 115.39. Dislocation of the right fourth proximal interphalangeal joint in a 15-year-old boy. Most such injuries can be readily reduced, which is not the case with metacarpophalangeal joint dislocations.

FIGURE 115.40. Buddy taping is a convenient way of splinting uncomplicated dislocations postreduction, as well as minor finger fractures.

Metacarpophalangeal dislocations are particularly problematic. Although closed reduction may be successful, often the volar plate is entrapped in the joint and open reduction is therefore necessary. Such volar entrapment can be suspected when physical examination reveals puckering of the palmar skin adjacent to the affected joint. Visualization of a sesamoid bone within the joint space is pathognomonic of volar plate entrapment. (See Section VII, Procedures, for a more in-depth review of techniques for reduction of finger dislocations.) Should an initial attempt at reduction of a metacarpophalangeal dislocation fail, the finger should be immobilized and a hand specialist consulted.

FRACTURES OF THE PELVIS

When evaluating a child with a suspected pelvic fracture, attention to surrounding viscera and to signs of blood loss are the most important immediate considerations. Pelvic fractures are caused by high-energy accidents and are often associated with head, abdominal, and vascular injuries. Overall, pelvic fractures in children have a favorable outcome and rarely require any more treatment than bed rest for 4 to 6 weeks. Exceptions to this rule include severely displaced sacral or sacroiliac joint dislocations and displaced acetabular fractures. An immediate orthopedic consultation is required for all pelvic fractures other than minor avulsions.

Pelvic fractures in children can be divided into three groups: 1) avulsion fractures, 2) pelvic ring fractures, and 3) acetabular fractures.

Avulsion Fractures

Avulsion fractures occur most commonly from sporting activities. The muscular attachments to the secondary centers of ossification (i.e., the anterior superior iliac spine, the anterior inferior iliac spine, and the ischial tuberosity) can be pulled off during strong, active contractions against resistance (Fig. 115.41). Localized tenderness is usually present. The diagnosis is usually readily apparent on plain film radiographs, although bone scintigraphy may be necessary to confirm the diagnosis on occasion (Fig. 115.42). Treatment is based on symptoms. Often, crutches with partial or no weight bearing for 4 to 6 weeks with slow resumption of activities are all that is required even with significantly displaced fractures. With widely separated (more than 2 cm) avulsion fractures of the ischial tuberosity, some authors recommend open reduction and fixation; others continue to advocate conservative treatment.

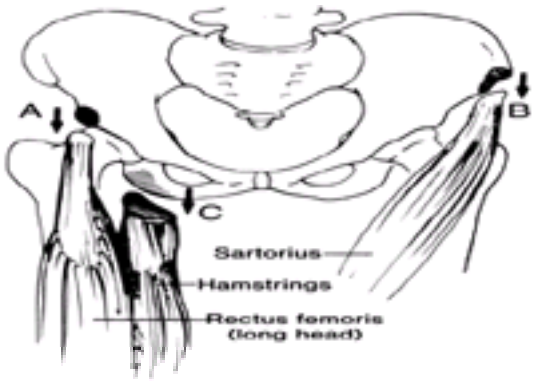

FIGURE 115.41. Common avulsion injuries of the pelvis: anterior inferior iliac spine *(A);* anterior superior iliac spine *(B);* ischial tuberosity *(C).*

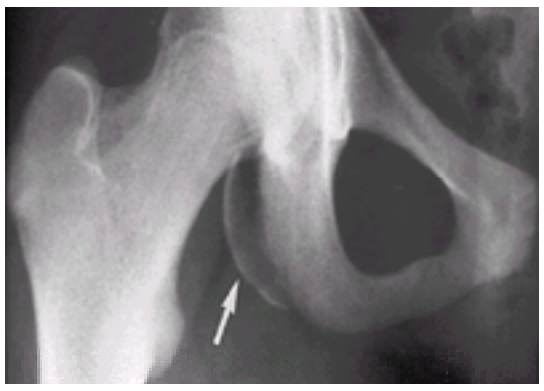

FIGURE 115.42. Avulsion fracture of the right ischial tuberosity in a 13-year-old girl *(arrow).*

Pelvic Ring Fractures

Single Breaks in the Pelvic Ring

Symphysis pubis diastasis, superior and inferior pubic rami fractures, and straddle fractures are classified as single breaks in the pelvic ring. These common childhood fractures often seem worse than they are. Although they are caused by high-energy accidents, they are generally stable fractures. A careful search for accompanying genitourinary and neurovascular injuries must be made. Fractures of the superior and inferior pubic rami rarely require any treatment in the child or adolescent as long as the sacroiliac joints and sacrum remain intact. One exception to this rule is a diastasis of the pubic symphysis, which often is associated with anterior disruption of the sacroiliac joint. This fracture configuration with pubic diastasis and anterior sacroiliac joint disruption is called the open book deformity. If significant displacement occurs through the symphysis pubis, closed reduction with an external fixator or a pubic plate must be considered.

Double Breaks in the Pelvic Ring

Fractures of the pubic rami or symphysis pubis associated with displaced sacroiliac joint dislocations or sacral fractures are classified as Malgaigne's fractures (Fig. 115.43). The hemipelvis is unstable and displaced cephalad. This group of fractures is associated with a high incidence of complications, including genitourinary, abdominal, and vascular injuries. Life-threatening hemorrhage can occur from pelvic vein disruption. In severe cases of bleeding, emergent application of an external fixator or a pneumatic antishock garment in the ED with compression of the pelvis may slow bleeding by a tamponade effect. Angiographic embolization should also be considered in the face of persistent bleeding.

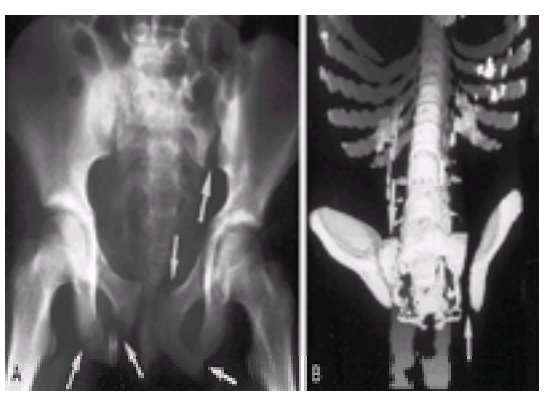

FIGURE 115.43. An unstable pelvic injury. **A.** On the plain film, multiple fractures of the pubic rami *(small arrows)* and widening of the right sacroiliac joint *(large arrow)* are apparent. **B.** On the three-dimensional reconstruction of the CT scan, the left sacroiliac fracture is even more obvious *(small arrow),* and a right-sided sacral fracture is seen as well *(large arrow).*

Initial treatment of the unstable pelvic fracture is bed rest. Special radiographic views consisting of an inlet and outlet view or CT scan assist the orthopedist in deciding whether to place the child or adolescent in traction or to undertake an open reduction and internal fixation of the posterior fracture–dislocation.

Acetabular Fractures

Fractures involving the acetabulum are rare in children. They often are associated with a dislocation of the hip joint. Attention should be directed toward obtaining an early congruent reduction and evaluating the stability of the hip. Acetabular fractures associated with major pelvic disruption should be treated like those involving double breaks in the pelvic ring. An orthopedic consultation should be obtained early and treatment of life-threatening complications initiated.

INJURIES OF THE LOWER EXTREMITIES

Injuries of the Hip and Proximal Femur

Hip dislocations and femoral neck fractures in children and adolescents are the result of high-energy accidents. The care

and resuscitation of the child is paramount before addressing the orthopedic injury. In many instances, the child can be managed in a traction splint until definitive care is given.

Injuries of the hip and proximal femur in children can be divided into five groups: 1) hip dislocation, 2) proximal femoral physeal fractures, 3) slipped capital femoral epiphysis, 4) femoral neck fractures, and 5) intertrochanteric fractures.

Hip Dislocation

Dislocation of the hip in children and adolescents is uncommon. It probably occurs more often than it is diagnosed, however, because of spontaneous reduction at the time of injury. A dislocation or fracture–dislocation is rather obvious if the injured limb is shortened, externally rotated, and painful. Radiographic examinations make the diagnosis (Fig. 115.44). In evaluating suspected dislocations of the hips with spontaneous reduction, attention must be directed to the radiographic medial clear space of the hip. If a suspected dislocation with spontaneous reduction has occurred, the medial clear space is often wider than the normal contralateral side. The posterior labrum and capsule of the joint may be detached when a dislocation occurs. At the time of reduction (either spontaneous or after closed reduction), tissue may get trapped in the joint space, resulting in asymmetry of the joint space and an incongruent reduction. Further evaluation should consist of a CT scan or MRI (Fig. 115.45).

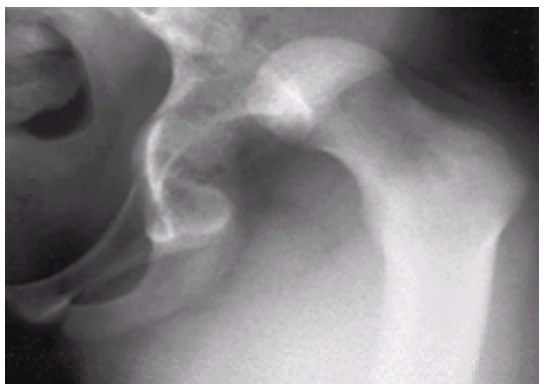

FIGURE 115.44. Dislocation of the left hip in a 9-year old boy. If dislocation is delayed beyond 6 hours, the risk of osseous necrosis rises.

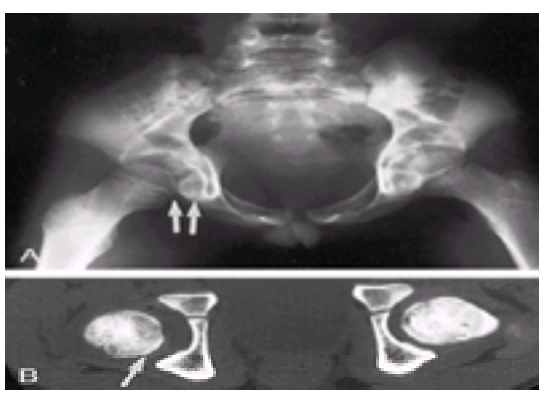

FIGURE 115.45. Radiographs taken following closed reduction of a right hip dislocation. **A.** The plain film demonstrates residual widening of the joint space *(arrows).* **B.** Widening is also apparent with magnetic resonance imaging, as is entrapment of a portion of the posterior joint capsule. Under general anesthesia, further efforts at closed reduction were successful.

A patient presenting with a dislocated hip should undergo a closed reduction in the ED or under general anesthesia. Reduction within 6 hours of the accident is essential to decrease the incidence of osseous necrosis. The technique of closed reduction consists of hip and knee flexion to 90 degrees and axial distraction of the thigh. When closed reduction is unsuccessful or when it is suspected that tissue is trapped in the joint space, open reduction is necessary. Congruency of both hips is imperative to a good result. Complications of traumatic hip dislocation in children include osseous necrosis of the femoral head, posttraumatic arthritis, and persistent instability of the hip joint.

Proximal Femoral Physeal Fractures

Proximal femoral physeal fractures occur through the zone of provisional calcification of the proximal femoral growth plate. The degree of displacement can be mild to complete (Fig. 115.46). Anatomic reduction, either by open or closed means, is essential. Unfortunately, the incidence of osseous necrosis approaches 100% in totally displaced fractures and can lead to long-term disability. In minimally displaced fractures, it may be far better to accept mild displacement than to further compromise the vascularity of the femoral head by performing a reduction.

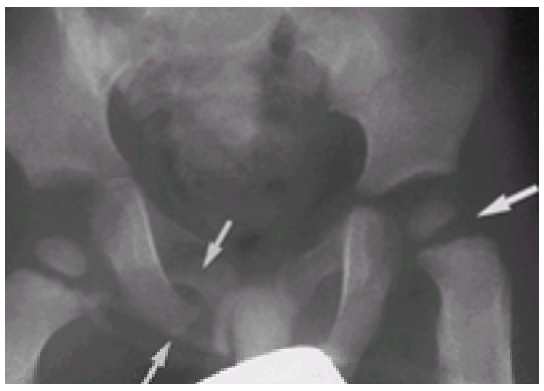

FIGURE 115.46. Displaced Salter-Harris type I fracture of the left proximal femur in a 2-year-old boy *(large arrow).* Also seen are fractures of the right pubic rami *(small arrows).* The pelvis is disrupted posteriorly as well.

Slipped Capital Femoral Epiphysis

Although most cases of slipped capital femoral epiphysis (SCFE) present with chronic pain, a significant percentage present acutely. Several studies have suggested that structural weakness is present in the capital femoral physis during the onset of puberty. Others have identified a genetic or hormonal influence predisposing to SCFE. This malady occurs predominantly in children 8 to 15 years of age, with a male:female predominance of 2:1 to 4:1. Obese children and African-Americans are particularly susceptible.

The diagnosis of SCFE should be considered in any preadolescent or adolescent complaining of hip or knee pain. The history is often one of minimal trauma, causing pain in the hip, thigh, or knee region. Vague hip or knee pain and a limp in the preceding weeks is common. The diagnosis is made by the physical and radiographic examination. Range of motion abnormalities of the hip, in particular limitation of internal rotation, abduction, and flexion, are almost universal. When flexing the hip from the extended position, the examiner will often note external rotation. Range of motion in all directions may be painful. The radiographic examination should include anteroposterior and frog-leg views of the pelvis. Changes on the anteroposterior film may be obscure. The slip is often seen more easily on the frog-leg view. Comparison with the normal side may assist in the diagnosis. However, 10 to 25% of slips may be bilateral (Fig. 115.47).

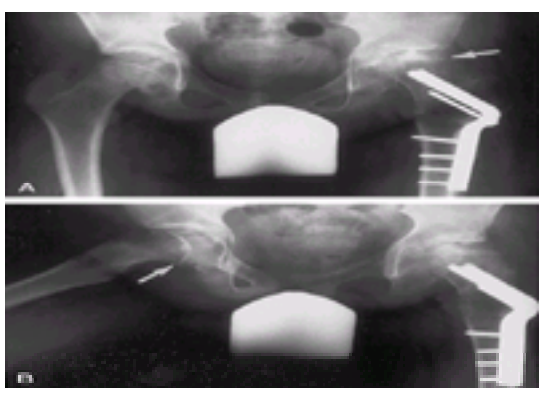

FIGURE 115.47. Radiographs of the hips of a teenager with bilateral slipped capital femoral epiphysis. **A.** Despite surgical intervention, significant avascular necrosis of the femoral head has occurred *(arrow).* The right hip appears normal. **B.** On a simultaneous frog-leg view, however, it can be seen that the right capital femoral epiphysis has slipped as well *(arrow).*

Once diagnosis has been made, treatment should consist of strict non–weight bearing and an urgent orthopedic consultation. Prompt pinning is required to prevent further slippage. This may be performed the night of assessment or shortly thereafter, depending on the availability of anesthesia.

Femoral Neck Fractures

Fractures of the femoral neck are relatively common. Initial treatment is traction and splinting followed by either closed or open reduction, depending on the position of the fracture. If the blood supply to the femoral head is damaged at the time of injury, osseous necrosis can occur. As would be expected, this complication is more likely with displaced than nondisplaced fractures. Overall, the incidence of osseous necrosis in this setting is 40% (Fig. 115.48). Stress fractures of the femoral neck are also being increasingly reported, generally in adolescents involved in repetitive activities such as long distance running. Exercise-induced hip pain should prompt consideration of the diagnosis, which may require bone scanning or MRI study for confirmation. Early recognition is important because restriction of activity may allow healing and thus prevent progression to more complete fractures with displacement.

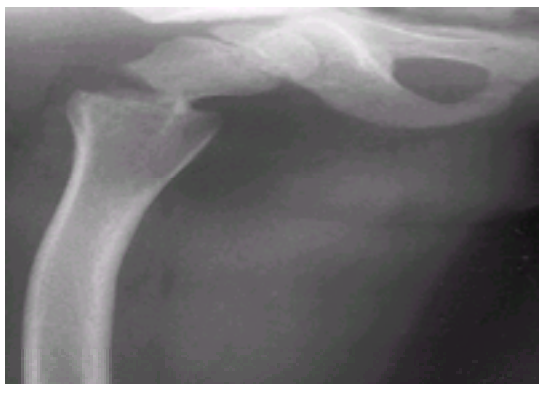

FIGURE 115.48. Fracture of the right femoral neck in a 3-year-old girl.

Intertrochanteric Fractures

Although common in adults, intertrochanteric fractures are uncommon in children and adolescents. Nondisplaced or minimally displaced fractures can be treated easily in a spica cast for 6 to 8 weeks. If significant displacement occurs, internal fixation to restore the normal anatomy may be the best approach to treatment. The incidence of complications is low in this group of patients. The rate of osseous necrosis is approximately 5%.

Fractures of the Shaft of the Femur

Femoral shaft fractures occur in all age groups, from newborn to adolescents. Each group has its specific mechanisms of injury, complications, and treatments. The following age groups are considered: 1) birth to 2 years of age, 2) 2 to 10 years of age, and 3) adolescents.

Birth to 2 Years of Age

Most femoral fractures in the first 2 years of life result from either a slow twisting motion or a direct blow (Fig. 115.49). A large percentage of femoral fractures in this age group are the result of intentional trauma. Overhead skin traction, once the cornerstone of the treatment, has fallen out of favor because of reports of neurovascular compromise and skin problems. Treatment options include immediate spica casting or a short period of Buck's traction followed by spica casting. Shortening and angulation are rarely problems in this age group, although rotational deformity can occur if careful alignment is not maintained during casting.

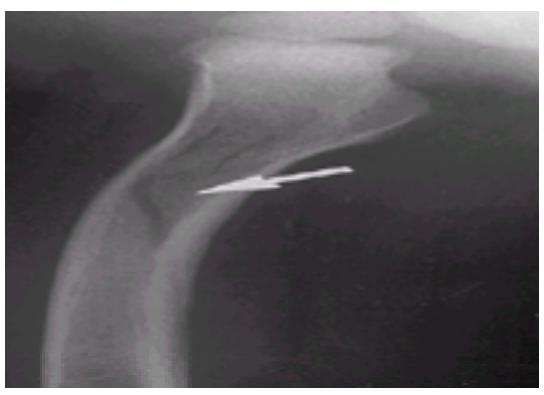

FIGURE 115.49. Spiral fracture of the right femur in a 20-month-old boy *(arrow).* In this instance, the injury occurred as the result of a motor vehicle accident. In general, spiral femur fractures in young children should prompt consideration of child abuse.

2 to 10 Years of Age

Femoral fractures in children 2 to 10 years of age are most often the result of high-energy motor vehicle or vehicle–pedestrian accidents. Concomitant injuries are common. Only rarely does an isolated femur fracture cause hemodynamically significant blood loss. Neurovascular evaluation should be performed and documented at regular intervals. Initial treatment consists of traction or splinting and care of other injuries (Fig. 115.50).

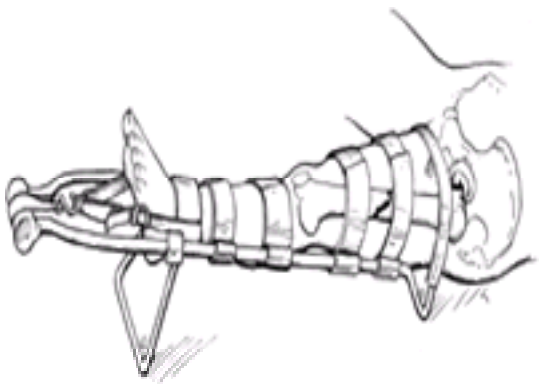

FIGURE 115.50. Use of a traction splint to stabilize femoral fractures is strongly recommended. Both adult and pediatric sizes are available.

Distal femoral skeletal traction for several weeks followed by spica cast application has been the cornerstone of treatment in the past. Recently, early or immediate spica casting under general anesthesia has replaced traditional methods. This has reduced the hospital stay and alleviated the need for the invasive intervention of the traction pin placement. Contraindications to immediate or early spica casting are shortening greater than 2.5 cm, open fractures, and major concomitant injuries. The long-term complications of femur fractures in this age group include excessive shortening or overgrowth, malrotation, and malunions of the healing femur. It is usually desirable to leave the bone fragments overlapping by 1 cm to allow for some “overgrowth” of the healing femur. Stiffness of the knee and hip has been reported following prolonged spica casting treatment but is usually not a long-term complication.

Adolescents

Femur fractures in adolescents are also caused by high-energy accidents. Once again, attention to other injuries should precede treatment of the femoral fracture. Stabilization with traction splints is adequate until an orthopedic consultation can be obtained. The management of these fractures has changed over the last several years. Closed reduction and intramedullary rodding are currently recommended to improve alignment and promote an early return to activity.

Injuries of the Knee

Although relatively uncommon, fractures about the knee arguably rank as the most serious long bone injuries in children and adolescents (Fig. 115.51). The growth centers of the distal femur and proximal tibia together account for two-thirds of the length of the lower extremity. Growth arrest and deformity can occur after physeal injuries about the knee; the resultant limb length discrepancies are hardly trivial problems. On the other hand, ligamentous injuries are uncommon. For the purposes of discussion, pediatric knee injuries can be divided into the following groups: 1) ligamentous injuries and avulsion fractures, 2) distal femoral physeal fractures, 3) proximal tibial physeal fractures, 4) knee dislocations, and 5) patellar fractures and dislocations.

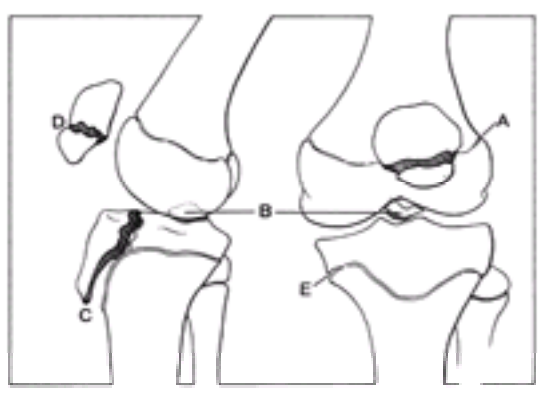

FIGURE 115.51. Common fractures of the knee in children: distal femoral physis *(A);* tibial spine *(B);* tibial tubercle *(C);* patella *(D);* proximal tibial physis *(E).*

Ligamentous Injuries and Avulsion Fractures

Compared with fractures of the epiphyses and physes about the knee, ligamentous injuries are relatively uncommon before growth plate closure. Such injuries do occur, however, both in isolation and in conjunction with fractures. Most ligamentous injuries result from direct trauma to the knee, typically when a child is struck by a motor vehicle while walking or riding a bicycle. Others occur during vigorous sporting activities when the knee is subjected to significant valgus or varus stress. The medial collateral and anterior cruciate ligaments are the ones injured most often, and injury to the latter almost invariably occurs in conjunction with an avulsion of the tibial spine.

Given the propensity for knee injuries in children younger than 14 years to result in fractures rather than ligamentous injuries, radiographs should be ordered routinely for all but the most minor injuries. Stress views after adequate sedation are necessary when routine views are normal, but the history is that of a significant valgus or varus stress. Evidence of distal femoral or proximal tibial epiphyseal separation or of collateral ligament instability may thus be uncovered. Of the growth plates about the knee, the distal femoral physis is particularly vulnerable to injury. Both the medial and lateral

collateral ligaments attach proximally to the distal femoral epiphysis. Their attachment to the tibia and fibula is distal to the epiphysis (Fig. 115.52). Given the relative strengths of the ligaments and the physeal plate, forceful valgus or varus stress results in distal femoral epiphysis separation rather than proximal tibial epiphysis injury or ligament rupture. Such injuries are discussed later in this chapter.

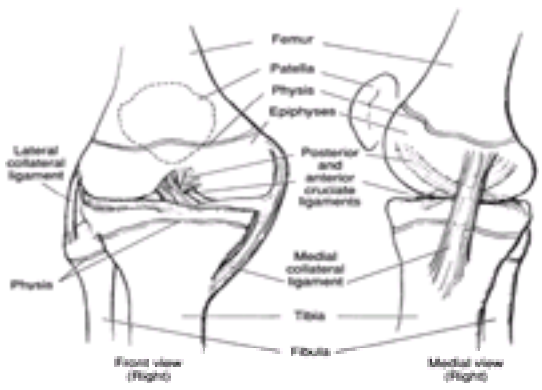

FIGURE 115.52. The ligaments of the knee. Proximally, both collateral ligaments attach to the epiphysis, whereas distally they attach to the tibia and fibula below the tibial epiphysis.

Avulsion of the tibial spine is the pediatric equivalent of an anterior cruciate ligament injury in an adult. The most commonly described mode of injury is hyperflexion of the knee during a fall from a bicycle. Significant pain and a refusal to bear weight are typical; a hemarthrosis is invariably present. Radiographic findings vary from minimal elevation of the anterior portion of the tibial spine (best seen on lateral views) to complete separation (Fig. 115.53). Incomplete separations generally can be managed by closed reduction with the knee held in extension. Open repair is necessary for complete avulsions and when closed manipulation does not lead to a satisfactory reduction. Arthroscopic evaluation of all tibial spine avulsions with instability on Lachman testing (see Chapter 39) has been advocated. Such an approach allows anatomic reduction of the injury with internal fixation and is thought to lead to a better long-term outcome. Immediate management in the ED should include splinting in extensions and arthrocentesis under sterile conditions when the hemarthrosis is causing severe pain.

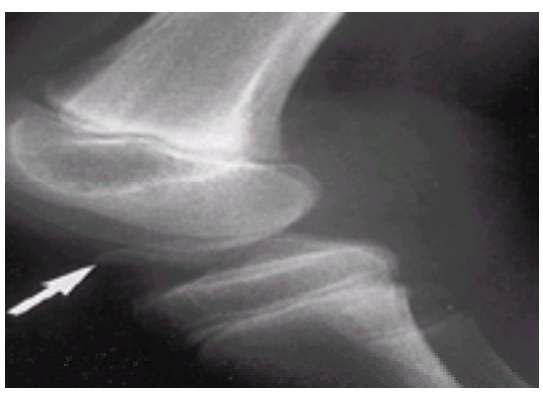

FIGURE 115.53. Avulsion fracture of the tibial spine in a 9-year-old girl. A significant hemarthrosis was present and was aspirated. In an adult, the same mechanism of injury would have resulted in a tear of the anterior cruciate ligament.

Another uncommon but severe knee injury observed in adolescents is an avulsion fracture of the tibial tuberosity. This fracture occurs essentially exclusively in boys 12 to 17 years of age who are involved in vigorous sporting activities. Most such injuries occur during jumping when the quadriceps is strongly contracted. If extension is impeded, as when a basketball player jumps to shoot but is blocked, or if the contraction of the quadriceps is particularly violent, as in high-jumping, the tibial tubercle can be torn either in part or in its entirety from the proximal tibial epiphysis. The result is a Salter-Harris type III fracture. Of note is that the patient often has an antecedent history of Osgood-Schlatter's disease. Once again, the severity of the injury dictates whether closed or open management is chosen.

Distal Femoral Physeal Fractures

Historically, fractures of the distal femoral epiphysis occurred when the leg of a child was caught between the wagon and the spokes of the wheel and thus were known as “wagon wheel injuries” during the 19th century. Today these injuries are caused by high-energy sports injuries, motor vehicle accidents, and falls from a height. Overall, this injury is rare because of the undulating course of the physis and the strong perichondrial ring that surrounds it. Of all the fractures involving the growth plate, however, injuries of the distal femoral physis have the highest incidence of posttraumatic growth arrest.

These injuries are described according to the direction of displacement of the epiphysis and the corresponding Salter-Harris classification. Most common is medial or lateral displacement with a fracture of the adjacent metaphysis (a Salter-Harris type II injury) (Fig. 115.54). As already mentioned, such injuries reflect a marked valgus or varus stress. The risk of neurovascular compromise is low, but peroneal nerve damage can accompany severe medial displacement. Even with adequate reduction, the incidence of premature growth arrest is significant. Somewhat less common is an anterior displacement of the distal epiphysis caused by hyperextension (Fig. 115.55). The risk of neurovascular compromise is high with this injury, which is the counterpart of a knee dislocation in an adult. Both compartment

syndrome and direct compression of the neurovascular structures are well-recognized complications. Posterior displacement of the femoral epiphysis is uncommon but can occur as the result of a direct blow to the flexed knee. The preferred treatment of these injuries in the ED includes a thorough evaluation with splinting in place followed by prompt orthopedic consultation. Gentle closed reduction, often using general anesthesia, is usually successful. Postreduction remodeling cannot be assured because of the high rate of posttraumatic growth arrest associated with these injuries.

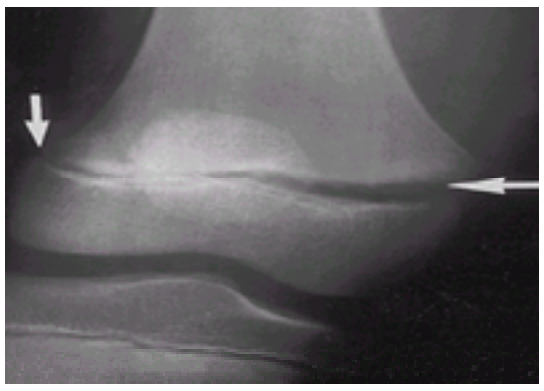

FIGURE 115.54. A Salter-Harris type II fracture of the right distal femoral physis in a 9-year-old boy. Widening of the growth plate is seen medially *(large arrow),* and a small metaphyseal fragment has been displaced laterally *(small arrow).* Closed reduction was successful. In an adult, the same mechanism of injury would have resulted in a medial collateral ligament sprain or tear.

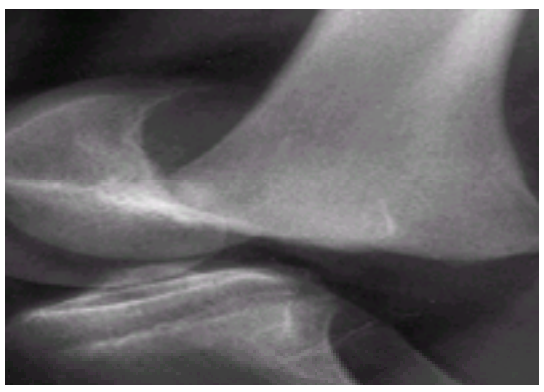

FIGURE 115.55. A Salter-Harris type I fracture of the right distal femoral physis with anterior displacement of the epiphysis in a 14-year-old boy. The injury resulted from a snowboarding accident.

Proximal Tibial Physeal Fractures

Fractures of the proximal tibial physis are also rare. Hyperextension is the usual mechanism of injury. The sheer force tears the posterior periosteum and capsule of the knee, allowing a Salter-Harris fracture to occur through the growth plate. The emergency physician must recognize that the popliteal structures are tethered at this point and are therefore vulnerable to stretch or direct contusion at the time of injury. Careful, sequential neurovascular examinations are mandatory. Compartment syndrome should be considered (p. 1441). Closed or open reduction will be necessary after stabilization. Complications after the injury include recurrent deformity, growth arrest, and limb length inequality (Fig. 115.56).

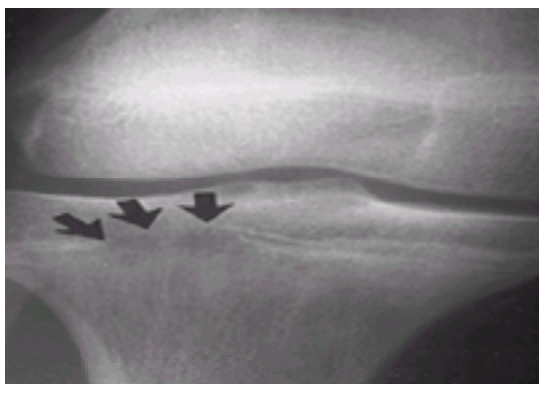

FIGURE 115.56. Although the original injury was only a Salter-Harris type I fracture of the proximal tibial physis, premature closure of the physis occurred *(arrows).* All fractures involving the proximal tibial growth plate require orthopedic referral.

Knee Dislocations

Complete dislocation of the femorotibial joint, another hyperextension injury, is extremely uncommon in children. As a

rule, hyperextension is much more likely to cause a distal femoral epiphyseal separation than a dislocation. Given the high likelihood of neurovascular compromise or compartment syndrome in this setting, femorotibial dislocation is considered a true emergency. A reduction maneuver may be attempted in the ED under intravenous sedation. Axial traction of the tibia with slow flexion of the knee from an extended position may lead to a reduction. Following a closed reduction, an arteriogram must be obtained to rule out an intimal tear of the popliteal artery. Definitive care of the torn ligaments resulting from this injury will ultimately be necessary.

Patellar Fractures and Dislocations

Unlike in the adult, the patella in the child is rarely fractured because of the thick covering of cartilage overlying the patella during growth and development. Fractures of the patella in adolescents are more common and present as avulsion fractures from dislocations, osteochondritis desiccans caused by overuse, symptomatic bipartite conditions, avulsion or “sleeve” fractures, and the occasional transverse displaced fracture (Fig. 115.57).

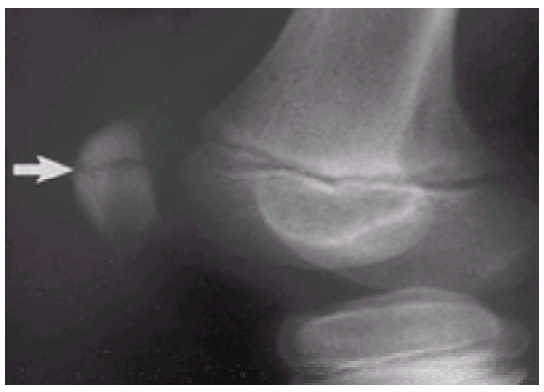

FIGURE 115.57. Transverse fracture of the patella in an 11-year-old victim of a motor vehicle accident *(arrow).*

Diagnosis of a patellar fracture may be difficult. A congenitally bipartite patella can be easily confused with a fracture. In this case, an accessory ossification center is located along the superior lateral margin of the patella. The margins are smooth and rounded. A comparison view of the opposite knee may assist in the diagnosis. Sleeve fractures of the patella, in particular, can easily be misdiagnosed on radiograph. The sleeve fracture occurs when the lower half of the cartilage cap is pulled free by the patellar ligament. The visible bony portion of the patella is displaced cephalad by the quadriceps mechanism. Often, a small fleck of bone is identified at the superior margin of the patellar ligament (Fig. 115.58). Pain usually prevents active extension of the knee.

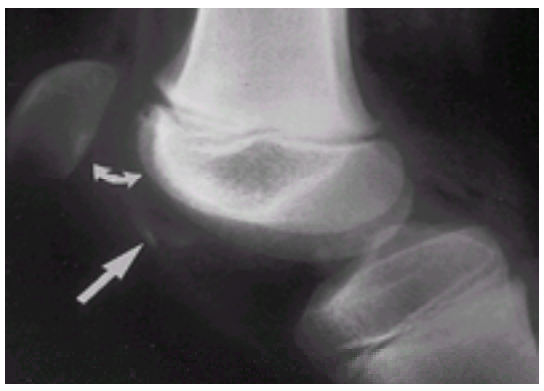

FIGURE 115.58. Radiograph demonstrating a sleeve fracture of the patella in a 10-year-old male. The inferior pole of the patella is displaced anteriorly *(curved arrow).* The bone fragment seen *(large arrow)* was avulsed by and remains attached to the patellar tendon.

The preferred treatment of patellar fractures parallels that of adults. Conservative care is the cornerstone of treatment in nondisplaced fractures. Cylindrical cast treatment from 4 to 6 weeks will result in union. Fractures that are displaced more than 3 to 4 mm are best treated with open reduction and internal fixation. Complications after patellar fractures include knee stiffness, quadriceps atrophy, extensor lag, and persistent pain.

Dislocation of the patella can be classified as an acute or chronic recurrent subluxation or as a dislocation. An acute traumatic dislocation of the patella results from a force displacing the patella laterally while the foot is planted. The patella may reduce spontaneously or may remain dislocated. Examination of the patient reveals an acutely swollen knee with pain to palpation noted along the medial patellar retinaculum. When reduction has already occurred, displacement of the patella laterally will usually elicit an apprehension sign. The patient may state that he or she feels like the knee cap is going to “pop out.” When the patella remains dislocated, the diagnosis is readily apparent by clinical and radiographic examination. Reduction of a dislocated patella usually is accomplished easily with extension of the knee and a medial upward force on the lateral patella (see Section VII, Procedures). Following reduction of an acutely dislocated patella, the physician must exclude the presence of an osteochondral fracture of the lateral femoral condyle or the medial patellar facet. Such fractures may be difficult to identify from a radiographic examination. Physical findings consistent with intra-articular loose bodies should suggest the diagnosis.

Chronic recurrent patella subluxation or dislocation is much less likely to result in osteochondral fractures. Predisposing factors include lateral femoral condyle hypoplasia, a loose medial patellar retinaculum, genu valgum, external tibial torsion, and quadriceps weakness.

The initial treatment of a dislocated patella should consist of a thorough examination, followed by closed reduction. Immobilization in an above-the-knee posterior splint or a commercially available knee immobilizer for 4 weeks is the appropriate ED management. Orthopedic referral is recommended.

Fractures of the Tibia and Fibula

Fractures of the tibia and fibula in children can be divided into the following groups: 1) proximal tibial metaphyseal fractures, 2) tibial and fibular shaft fractures, and 3) toddler's fractures.

Fractures involving the distal growth plates of the tibia and fibula are discussed with ankle injuries.

Proximal Tibial Metaphyseal Fractures

Although usually easy to manage, proximal tibial fractures can lead to two major complications, namely compartment syndrome and progressive posttraumatic valgus deformity (Fig. 115.59). As in other settings, a careful clinical evaluation followed by direct measurement of compartment pressure are the keys to the diagnosis of compartment syndrome. Progressive valgus deformity can develop after any proximal tibial injury, including greenstick and nondisplaced fractures. Deformity has been known to develop even after anatomic reduction of fracture fragments. It is speculated that stimulation of the physis from hyperemia causes asymmetric growth of the proximal tibial physis. Given the propensity for growth deformity, all fractures of the proximal tibial metaphysis should be managed by an orthopedic surgeon.

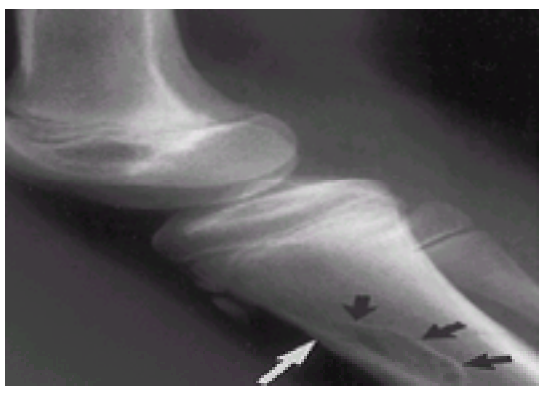

FIGURE 115.59. Nondisplaced proximal tibial metaphyseal fracture in an 11-year-old girl *(large arrow)* through a nonossifying fibroma *(small arrows).* Orthopedic consultation is mandatory both because the fracture is a pathologic one and because it is located in the proximal tibia.

Tibial and Fibular Shaft Fractures

Fractures of the tibial and fibular shafts are the most common fractures of the lower extremity in children. The diagnosis usually is apparent by physical and radiographic examination. Most tibial and fibular fractures are stable and in acceptable alignment (Fig. 115.60). Discussion with an orthopedic consultant helps decide whether any reduction is necessary. In children, these fractures rarely persist to delayed or nonunion. Healing time is quick, averaging 6 to 8 weeks. If the neurovascular status is normal, no signs of compartment syndrome are present (p. 1441), and the fracture configuration is deemed acceptable, a long leg posterior splint may be applied and orthopedic referral within the next few days arranged. Otherwise, more immediate consultation should be sought. (Most cases of compartment syndrome result from minor closed tibial fractures; with more severe injuries, the interosseous membrane typically is torn, allowing decompression of the anterior compartment.)

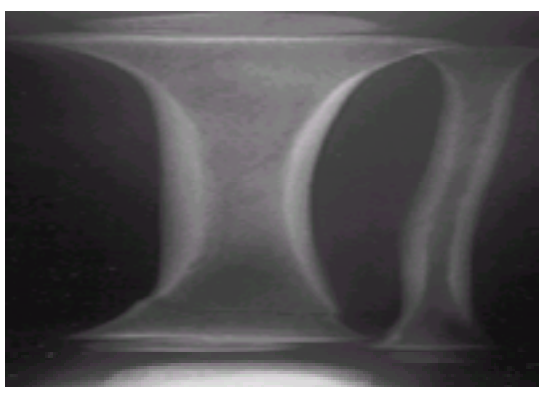

FIGURE 115.60. Nondisplaced transverse fracture of the distal tibia in a 3-year-old girl. Immobilization in a long leg posterior splint with orthopedic follow-up within 3 to 5 days would be adequate emergency treatment. The potential for growth deformity is low.

The indications for open treatment of tibial and fibular shaft fractures in children include open fractures, compartment syndrome, ipsilateral femur fractures, and concomitant severe head injuries. Complications of treatment after tibial and fibular fractures include malunion, limb length inequality, malrotation, and neurovascular deficiency.

Special consideration must be given when evaluating tibial injuries in children with paraplegia. They present with warmth and swelling over the leg or joint, which may suggest infection or inflammatory conditions rather than fractures. When identified, the fracture may be treated in a conservative manner with splints or a short 3- to 4-week period of casting.

Particular note should be made as well of the fact that the tibia and fibula are the most common sites of stress fractures in children. Overall, the proximal third of the tibia is most often affected. The history is usually that of pain and a limp of gradual onset in a child 8 to 15 years of age. Localized swelling and tenderness are present to varying degrees. Radiographs may appear normal, show limited cortical changes, or demonstrate subperiosteal new bone formation. Bone scintigraphy confirms the diagnosis in uncertain cases (Fig. 115.7). Rest is the only treatment needed.

Toddler's Fractures

Occasionally, the ED physician is asked to evaluate a young child with an acute gait disturbance, namely a limp or a refusal to walk. The differential diagnosis is a broad one (see Chapter 43). One possibility that should always be considered is that of a toddler's fracture. Originally, the term *toddler's fracture* referred to an oblique nondisplaced fracture of the distal tibia in children 9 to 36 months of age. The term is now used more loosely.

In most cases, the history is that of a minor accident, such as a fall from a seemingly insignificant height or while walking or running. No history of injury may be recalled in some instances. The physical findings are often subtle and at best difficult to elicit unless a gentle, unhurried examination is performed while the child is calm. The degree of swelling is minimal; warmth and tenderness are more commonly detected but are not uniformly present. Gentle twisting of the lower leg will elicit pain on occasion.

Like the physical findings, the radiographic abnormalities often are subtle. The anteroposterior or lateral views may reveal a spiral or oblique fracture extending downward and medially through the distal third of the tibia. If a toddler's fracture is suspected clinically but the routine radiographic views are normal, an internal oblique projection should be ordered. Consideration should also be given to the possibility of a fracture elsewhere in the limb; fractures of the femur, the foot, and rarely, the pelvis can also present with an acute limp. If no fracture is visualized on routine radiographs, a bone scan may be considered. Alternatively, if symptoms persist, it is certainly reasonable to repeat the plain films after 10 days, at which point subperiosteal new bone formation may be evident or enough sclerosis may have occurred at the fracture edges to render it visible (Fig. 115.61). Immobilization provides symptomatic relief and promotes healing, although no treatment at all may be needed if the history suggests that the fracture occurred several weeks before actual diagnosis.

FIGURE 115.61. Toddler's fracture of the distal tibia *(arrows).* Not until 2 weeks after the onset of symptoms could the fracture line be demonstrated radiographically.

If a toddler's fracture is a spiral one and the caretakers can recall no specific time of injury, suspicions of child abuse may understandably arise. Notably, midshaft fractures are more common in abused children, and most toddler's fractures are distal injuries. When other circumstances or injuries suggestive of abuse are found or when spiral fractures occur in children who are not yet ambulating, the strong possibility of nonaccidental injury should obviously be considered seriously.

Injuries of the Ankle and Foot

Injuries of the ankle and foot in the pediatric age range can be divided into the following groups: 1) ankle sprains, 2) distal tibial and fibular fractures, 3) hindfoot and midfoot fractures, and 4) metatarsal and phalangeal fractures.

Ankle Sprains

Adolescents often present to the ED complaining of ankle injuries (see Chapter 37). The differential diagnosis includes ligamentous injuries; nondisplaced Salter-Harris type I fractures; osteochondral fractures of the tibia, fibula, or talus; and avulsion injuries. Once again, before growth plate fusion, physeal injuries are much more likely than ligamentous injuries. Ligamentous injuries certainly are observed in older adolescents. The most common mechanism is adduction and inversion of the foot while it is held in plantar flexion. Of the three lateral ankle ligaments, the anterior talofibular ligament is the one most commonly injured. Injury to this ligament should be suspected when palpation just anterior to the distal fibula elicits an area of maximal tenderness. Ankle sprains are graded from I to III. In grade I injuries, ligaments are stretched but not torn. Grade II injuries include partial ligament tears without loss of stability. Complete tears of the ligamentous complex with loss of stability are present in grade III injuries. Other than with minor injuries, a three-view radiographic examination should be performed. Should the stability of a ligament be in question, stress views are

recommended.

Controversy exists regarding the appropriate care of ligamentous injuries. One schema is based on the severity of the ligamentous damage. Grade I mild sprains can be treated with an elastic wrap or air splint followed by ice, elevation, and compression for 72 hours. Crutches may be used until the patient is able to walk without a limp. Grade II and grade III injuries should be immobilized either in a cast or a posterior splint. (Because posterior splints break relatively easily, use of fiberglass and/or reinforcement with a stirrup is recommended.) Crutches are used initially. Ambulation in a cast for 3 weeks aids in initial scar formation and healing. This conservative approach with more severe sprains may help prevent recurrent ankle sprains in active athletic adolescents, as may physical therapy once the injury has healed.

Distal Tibial and Fibular Fractures

Although any Salter-Harris type I through V fracture may occur in distal physes of the tibia and fibula, several specific injury patterns are described and discussed here. Fractures involving both the growth plate and the ankle joint often need open reduction and internal fixation to ensure adequate reduction of both the physis and the joint. Only minimal amounts of displacement can be accepted at the articular surface, or altered joint mechanics will develop with possible posttraumatic pain, stiffness, and arthritis (see also Chapter 106).

Of the fractures of the distal fibula, a Salter-Harris type I injury is the most common. Tenderness and swelling are present over the growth plate on physical examination. Often the only radiographic finding is soft-tissue swelling overlying the distal fibula (Fig. 115.62). When suspicions of a Salter I injury are high, a short leg cast may be applied at the time of initial evaluation. When the diagnosis is less certain, immobilization with a repeat examination in a week to 10 days is recommended. If tenderness persists, a presumptive diagnosis of a type I fracture should be made and a walking cast applied. After 10 days, repeat radiographs may reveal periosteal changes confirming the presence of a fracture. In most cases, a total of 3 weeks of immobilization is adequate.

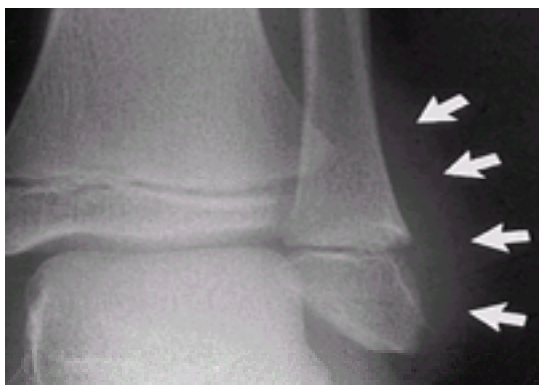

FIGURE 115.62. Radiograph of the left ankle of a 10-year-old boy notable only for soft-tissue swelling localized to the distal fibula *(arrows).* The presumptive diagnosis must be a Salter-Harris type I injury of the fibula.

Although type I injuries of the tibia are uncommon, type II injuries are often observed, usually in combination with a greenstick fracture of the fibula. The mechanism of injury is plantar flexion with eversion. Closed reduction and a long leg cast application usually lead to a satisfactory recovery. Growth disturbance is unusual.

The Tillaux fracture is a Salter-Harris type III injury of the ankle joint that occurs as the medial distal tibial physis begins to close in adolescents who are nearing skeletal maturity (Fig. 115.63). During external rotation of the foot, the anterior tibiofibular ligament avulses the lateral epiphysis from the medial malleolus. When displacement occurs, open reduction with internal fixation is required to ensure restoration of joint anatomy.

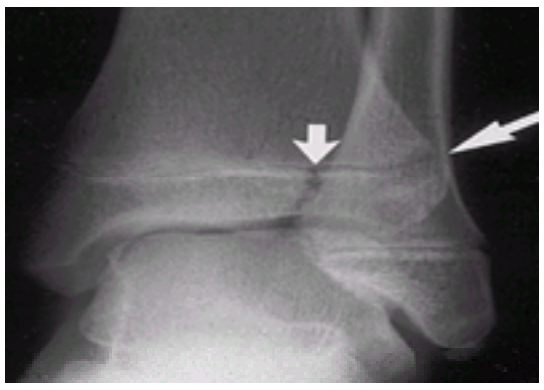

FIGURE 115.63. Classic Tillaux fracture of the distal tibia in a 14-year-old boy. The fracture line runs vertically through the epiphysis *(small arrow)* then laterally along the physis *(large arrow).* The lateral portion of the physis is widened.

The triplane fracture is a complex but uncommon ankle injury that is a combination of a Salter-Harris type II fracture and a Tillaux fracture. The resultant type IV injury may appear innocuous on the anteroposterior and lateral radiographs, but the degree of growth plate damage is generally significant. Suspected triplane fractures should be evaluated by CT scan to delineate the amount of displacement at the physis and the articular surface and to determine the number of fracture

fragments (Fig. 115.64).

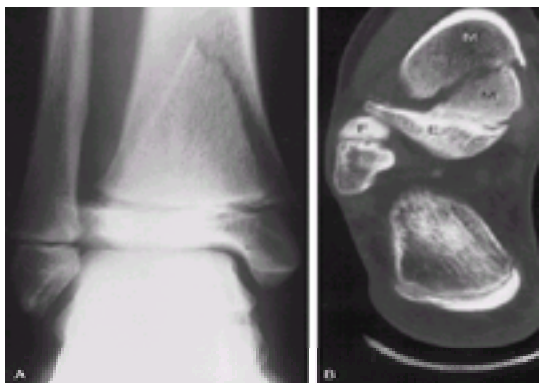

FIGURE 115.64. A. A triplane fracture was suspected from the plain film of the right ankle of this 11-year-old girl, although no clear epiphyseal fracture line could be seen. **B.** The coronal computed tomography scan made it possible to establish that the injury was indeed a Salter-Harris type II injury rather than the triplane fracture suspected. The fracture line runs laterally along the growth plate. *E,* epiphysis; *F,* fibula; *M,* tibial metaphysis.

The treatment of physeal and ankle fractures depends on the type of fracture, the amount of displacement, and patient's the age. Nondisplaced fractures may be treated with a bulky posterior splint, crutches, and a referral to the orthopedist. Immediate orthopedic referral is otherwise necessary.

Hindfoot and Midfoot Fractures

Fractures of the foot in children are uncommon and lead to few complications. Fractures of the hindfoot, which consists of the talus and the calcaneus, are particularly uncommon. When they do occur, they are usually obvious because of swelling, pain, and occasionally, deformity. “Occult” fractures of the calcaneus have been increasingly recognized in children less than 3 years of age. Pain with dorsiflexion may indicate a talar neck fracture. Should suspicions of a fracture be high but routine radiographs normal, additional views and/or bone scintigraphy may be necessary. Because calcaneal fractures generally occur as the result of a fall from a height, associated compression fractures of the spine can occur and must be considered. Treatment of hindfoot fractures are dictated by the amount of displacement. Often, a bulky posterior splint, crutches, and no weight bearing will suffice until an orthopedic consult can be obtained. Complications include osseous necrosis of the talus and chronic pain from calcaneal fractures.

Fractures of the midfoot include the navicular; the cuboid; and the first, second, and third cuneiforms. Fractures of these bones are extremely unusual and usually form part of a more severe injury to the foot (Fig. 115.65). They can be produced by blunt trauma, in which case soft-tissue damage is usually significant and a potential for neurovascular compromise results. Occasionally, an accessory ossification center on the medial side of the navicular may be confused with a fracture.

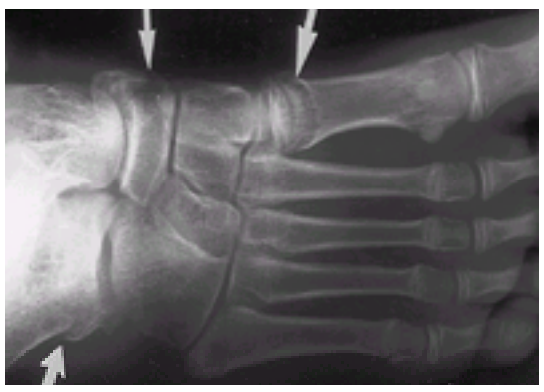

FIGURE 115.65. Radiograph of the right foot of a 13-year-old boy demonstrating fractures of the calcaneus *(small arrow),* scaphoid *(medium arrow),* and the first metatarsal *(large arrow).*

Although well documented in the adult literature, tarsal/metatarsal fracture–dislocations have received little attention in children. These injuries present with swelling and tenderness over the dorsum of the foot. Radiographs, including anteroposterior, lateral, and oblique views, may be necessary to identify subtle abnormalities. Once again, treatment is based on the severity of the injury with reduction and stabilization necessary if more than 2 mm of displacement is identified.

Metatarsal and Phalangeal Fractures

Metatarsal and phalangeal fractures are common in children. The diagnosis is not difficult because pain, swelling, and occasionally, a deformity accompany the fracture. Radiographic evaluations should include anteroposterior, lateral, and oblique views. The possibility of compartment syndrome must be kept in mind with crush injuries or multiple fractures in the midfoot or forefoot.

Two fractures that occur commonly at the base of the fifth metatarsal bear mentioning. The Jones fracture is a fracture at the diaphyseal–metaphyseal junction at the base of the fifth metatarsal. Although more common in adults, reports in adolescents can be found. This fracture has a high incidence of delayed or nonunion and should be splinted and referred to an orthopedist. An avulsion fracture of the base of the fifth metatarsal at the site of attachment of the peroneus brevis is relatively common in children. This fracture occurs more proximally than the Jones fracture and has a better prognosis. The usual treatment is 3 to 6 weeks of immobilization in a weight-bearing cast. When considering the possibility of a proximal fifth metatarsal fracture, care should be taken to distinguish a fracture fragment from the accessory ossification center found in the same location.

The care of most metatarsal and phalangeal injuries is relatively straightforward. If the fracture is nondisplaced or minimally displaced with little angulation, as is the usual case, a bulky splint can be applied and crutches prescribed. Intra-articular fractures of the big toe and significantly displaced fractures of the other toes often require pinning (Fig. 115.66). Buddy taping and hard-soled shoes provide adequate stabilization for most other phalangeal fractures.

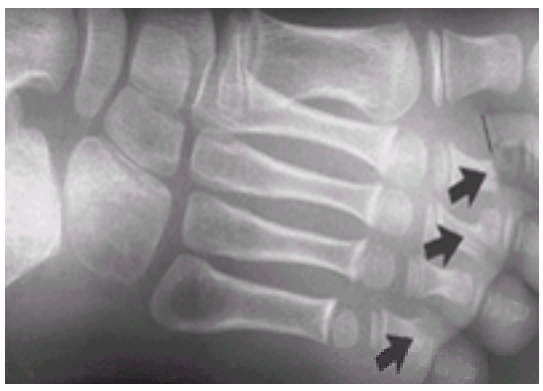

FIGURE 115.66. Displaced fractures of the right second, third, and fifth, proximal phalanges in a 7-year-old girl *(arrows).* Stability and proper healing of the fractures of the second and third toes could be guaranteed only with pinning.

INJURIES OF THE THORACOLUMBAR SPINE

Fortunately, injuries to the spine are rare in children and adolescents. When a child does have a spine injury, distinguishing what is normal from what is abnormal on plain radiographs can be extremely challenging. The advent of CT scans and MRI has made evaluation of spinal injuries much less problematic.

The child's spine differs from the adult's in that growth plates are present, the proportion of cartilage is higher, and the overall flexibility is greater. Because of the overall high elasticity of the pediatric spine, significant spinal cord injury can occur in the absence of radiographic signs of bony injury. By adolescence, the spine has mechanical qualities more like those of the adult, and the fracture patterns are similar. Unlike in adults, the risk of posttraumatic scoliosis after a complete spinal cord injury in children is extremely high.

In general, any child with significant head or multisystem injury should be assumed to have a spinal injury until proven otherwise. Diagnosis of a spinal injury in the child with a severe brain injury can be particularly problematic. However, certain physical findings can suggest the possibility of a coexisting spinal cord injury and should be sought routinely whenever a child with a severe head injury is examined. These findings include asymmetry of movement and reflexes between the arms and legs, absence of sacral reflexes, lax anal tone, priapism, spinal shock, autonomic hyperreflexia, diaphragmatic breathing, and urinary retention, as well as any evidence of a motor or sensory deficit level. When a child is sedated and/or paralyzed, as often is the case when a child has a severe brain injury, many of these findings will be particularly difficult to elicit. Therefore, spinal immobilization must always be maintained until a more detailed neurologic examination becomes possible. Another setting in which spinal cord injuries are overlooked on occasion is that of the lap belt complex, which is discussed in the following.

It is important to classify spine injuries according to the neurologic deficits to make possible a determination of prognosis. A complete lesion involves the entire cord at a given level with no motor or sensory function below that level. An incomplete lesion is associated with sparing of function or sensation below the level of injury; the degree of motor deficit is generally greater than the degree of sensory loss. After a thorough neurologic examination, a complete radiographic evaluation is necessary to determine the stability of the fracture. When faced with equivocal radiographs, flexion–extension views, CT scans, and MRI should be considered. Patients with a neurologic injury should be considered to have an unstable fracture until proven otherwise.

Initial treatment in the ED must focus on maintaining the stability of the spine with a backboard and a cervical collar. Patients should not be moved unless absolutely necessary. Patients can be log-rolled to inspect the spine as long as flexion, extension, or twisting movements do not occur. After stabilization of the patient and the spinal column, orthopedic and neurosurgical consultations should be obtained. Early intervention can decrease the risk of further injury to the spine in unstable, incomplete spinal injuries. Current data suggest that administration of high-dose corticosteroids within 8 hours to teenagers and adults with acute spinal cord injury improves neurologic recovery. Although data for younger children are lacking, it seems reasonable at this time to recommend that steroid therapy be strongly considered for any pediatric patient with evidence of an acute spinal cord injury. According to the protocol, methylprednisolone 30 mg/kg should be administered intravenously over 15 minutes. An infusion of 5.4 mg/kg per hour should then be begun 45 minutes after the completion of the bolus.

Fractures of the spine in children and adolescents can be divided into the following groups based on the mechanism of injury and the radiographic appearance: 1) compression fractures, 2) flexion and distraction fractures, 3) shear fractures, and 4) neurologic injuries without fractures.

Compression Fractures

Compression fractures result from hyperflexion producing an axial load and causing failure of the anterior vertebral body. Multiple fractures are the rule rather than the exception; the first lumbar vertebra is the most commonly injured segment (Fig. 115.67). Compression fractures heal quickly in children and have little tendency to progress. Many children do not require hospitalization for compression fractures and can be treated with bed rest and symptomatic mobilization. Occasionally, a well-molded thoracolumbar sacral orthosis may be necessary for persistent pain or multiple areas of compression.

FIGURE 115.67. Compression fractures of the lumbar vertebrae in a 3-year-old boy *(asterisks).* Multiple fractures are the rule in children.

Flexion and Distraction Fractures

Flexion–distraction injuries are rare in the immature spine. When they do occur, the most common mechanism of injury is hyperflexion over a seat belt during sudden deceleration in a motor vehicle accident. Associated intra-abdominal injuries, particularly tears and transections of the duodenum, jejunum, and mesentery, are common. This combination of spinal and abdominal injuries is often referred to as either the seat belt syndrome or the lap belt complex. The tendency for lap belts to ride higher on children than the recommended position across the iliac crest combined with their relatively higher center of gravity predisposes them to both the abdominal and spinal injuries. A so-called seat belt sign, an abdominal contusion in a band corresponding to the seat belt, is often observed. Instances in which diagnosis of the spinal injury has been delayed because of the presence of the abdominal injury, as well as the reverse, have been described; the discovery of one component of the lap belt complex should obviously prompt a search for the other.

The lumbar spine is most commonly injured as the result of hyperflexion over a seat belt. Among the spinal injuries that can occur are distractions, subluxations, facet dislocations, and ligamentous ruptures, as well as fractures, including compression fractures as previously discussed. One particular fracture, the Chance fracture, merits special mention. A horizontal splitting through both the body and posterior elements of a vertebra, the Chance fracture was formerly thought to occur exclusively in adults. It is now recognized that the same fracture pattern occurs in children, almost always as a seat belt injury (and, once again, often in combination with an abdominal injury). Most Chance fractures are stable; associated neurologic injury is uncommon. Immobilization with a well-molded orthosis for several months is generally adequate treatment.

Shear Fractures

Although the cervical spine is most vulnerable to shear injuries, violent trauma can cause such injuries in the thoracic and the lumbar spine as well (Fig. 115.68). Unfortunately, neurologic deficits are common in this setting. All shear fractures should be considered unstable injuries that will need stabilization procedures to avoid progressive deformity and enhance any possibility of neurologic recovery.

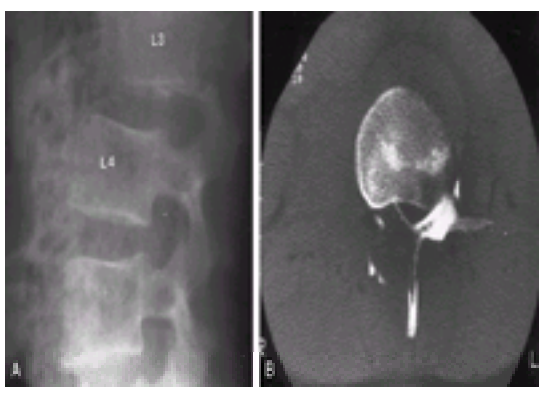

FIGURE 115.68. A. Fracture dislocation of the third *(L3)* and fourth *(L4)* vertebrae in a 12-year-old girl, the result of a shear injury in a motor vehicle accident. **B.** Permanent paralysis of the lower extremities resulted, as suggested by the

degree of collapse of the spinal canal seen on the computed tomography scan.

Neurologic Injuries without Fractures

It is well known that the immature spine is more flexible than the spinal cord. Injuries causing hyperflexion or extension may produce damage to the cord and neurologic injury while leaving the bony, cartilaginous, and ligamentous structures intact. Termed SCIWORAs (Spinal Cord Injury WithOut Radiographic Abnormality), two-thirds of such injuries occur in children less than 8 years of age. Cervical and thoracic spine injuries are the most common. Both incomplete and complete neurologic deficits can occur. Any history of neurologic deficit following spinal trauma should prompt consideration of a SCIWORA. As for radiographic evaluation, MRI can best identify cord damage in the absence of a fracture. To reemphasize, in all cases of severe trauma, spinal immobilization must be maintained until the patient's condition permits a satisfactory neurologic examination.

Suggested Readings

TEXTBOOKS

Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997.

Rang M. Children's Fractures. 2nd ed. Philadelphia: JB Lippincott, 1983.

Rockwood CA Jr, Wilkins KE, King RE. Fractures in Children. 3rd ed. Philadelphia: JB Lippincott, 1991.

Swischuk LE. Emergency Radiology of the Acutely Ill or Injured Child. 3rd ed. Baltimore, Williams & Wilkins, 1994.

Tachdjian MO. Pediatric Orthopedics. 2nd ed. Philadelphia: WB Saunders, 1990:4.

REVIEWS

Carty H. Children's sports injuries. Eur J Radiol 1998;26:163–176.

Kao SC, Smith WL. Skeletal injuries in the pediatric patient. Radiol Clin North Am 1997;35:727–746.

Micheli LJ. Sports injuries in children and adolescents. Clin Sports Med 1995;14:727–745.

GENERAL PRINCIPLES

Light TR, Ogden DA, Ogden JA. The anatomy of metaphyseal torus fractures. Clin Orthop 1984;188:103–111.

Mabrey JD, Fitch RD. Plastic deformation in pediatric fractures: mechanism and treatment. J Pediatr Orthop 1989;9:310–314.

McConnochie KM, Roghmann KJ, Pasternak J, et al. Prediction rules for selective radiographic assessment of extremity injuries in children and adolescents. Pediatrics 1990;86:45–57.

Rang M, Wright J. Pitfalls in fractures. Pediatr Ann 1989;18:53–68.

Rivara FP, Parish RA, Mueller BA. Extremity injuries in children: predictive value of clinical findings. Pediatrics 1986;78:803–807.

Salter RB, Harris WR. Injuries involving the epiphyseal plate. J Bone Joint Surg 1963;45A:587–622.

Shapiro F. Epiphyseal disorders. N Engl J Med 1987;317:1702–1710.

Wilkins KE. Changing patterns in the management of fractures in children. Clin Orthop 1991;264:136–155.

Worlock P, Stower M. Fracture patterns in Nottingham children. J Pediatr Orthop 1986;6:656–660.

CHILD ABUSE

Beals RK, Tufts E. Fractured femur in infancy: the role of child abuse. J Pediatr Orthop 1983;3:583–586.

Carty H. Non-accidental injury: a review of the radiology. Eur Radiol 1997;7:1365–1376.

Haller JO, Kleinman PK, Merten DF, et al. Diagnostic imaging of child abuse. Pediatrics 1991;87:262.

King J, Diefendorf D, Apthorp J. Analysis of 429 fractures in 189 battered children. J Pediatr Orthop 1988;8:585–589.

Merten DF, Carpenter BLM. Radiologic imaging of inflicted injury in the child abuse syndrome. Pediatr Clin North Am 1990;37:815–837.

Rosenberg N, Bottenfield G. Fractures in infants: a sign of child abuse. Ann Emerg Med 1982;11:178–180.

Thomas SA, Rosenfield NS, Leventhal JM, et al. Long-bone fractures in young children: distinguishing accidental injuries from child abuse. Pediatrics 1991;88:471–476.

Worlock P, Stower M, Barbor P. Patterns of fractures in accidental and non-accidental injury in children. Br Med J 1986;293:100–102.

INJURIES OF THE SHOULDER REGION

Eidman DK, Siff SJ, Tullos HS. Acromioclavicular lesions in children. Am J Sports Med 1981;9:150–154.

Riebel GD, McCabe JB. Anterior shoulder dislocation: a review of reduction techniques. Am J Emerg Med 1990;9:180–188.

Zaslav KR, Ray S, Neer CSN II. Conservative management of a displaced medial clavicular physeal injury in an adolescent male. Am J Sports

Med 1989;17:833–836.

INJURIES OF THE ELBOW

Asher MA. Dislocations of the upper extremity in children. Orthop Clin North Am 1976;7:583–591.

Bernstein SM, King JD, Sanderson RA. Fractures of the medial epicondyle of the humerus. Contemp Orthop 1981;3:637–641.

De Jager LT, Hoffman EB. Fracture separation of the distal humeral epiphysis. J Bone Joint Surg 1991;73B:143–146.

Fowles JV, Kassab MT. Displaced fractures of the medial humeral epicondyle in children. J Bone Joint Surg 1980;62A:1159–1163.

Matthews JG. Fractures of the olecranon in children. Injury 1981; 12:207–212.

Micheli LJ, Skolnick MD, Hall JE. Supracondylar fractures of the humerus in children. Am Fam Physician 1979;19:100–114.

Millis JM, Singer IJ, Hall JE. Supracondylar fractures of the humerus in children: further experience with a study in orthopaedic decision making. Clin Orthop 1984;188:90–97.

Murphy WA, Siegel MS. Elbow fat pad signs with new signs and extended differential diagnosis. Radiology 1977;124:659–665.

Ring D, Waters PM. Management of fractures and dislocations of the elbow in children. Acta Ortho Belgica 1996;62(Suppl 1):58–65.

Rogers LF, Malave S, White H, et al. Plastic bowing, torus and greenstick fractures of the humerus: radiographic clues to obscure fractures of the elbow in children. Radiology 1978;128:145–150.

Schunk JE. Radial head subluxation: epidemiology and treatment of 87 episodes. Ann Emerg Med 1990;19:1019–1023.

Webb AJ, Sherman FC. Supracondylar fractures of the humerus in children. J Pediatr Orthop 1989;9:315–325.

FRACTURES OF THE FOREARM AND WRIST

Anderson TE, Breed AL. A proximal radial metaphyseal fracture presenting as wrist pain. Orthopedics 1982;5:425–428.

Crawford AH. Pitfalls and complications of fractures of the distal radius and ulna in childhood. Hand Clin 1988;4:403–413.

Davis DR, Green DP. Forearm fractures in children. Clin Orthop 1976;120:172–184.

Gumucio CA, Fernando B, Young L, et al. Management of scaphoid fractures: a review and update. South Med J 1989;82:1377–1388.

Imatani J, Hashizume H, Nishida K, et al. The Galeazzi-equivalent lesion in children revisited. J Hand Surg 1996;21B:455–457.

Light TR. Injuries to the immature carpus. Hand Clin 1988;4:415–424.

Tredwell SJ, Van Peteghem K, Clough M. Patterns of forearm fractures in children. J Pediatr Orthop 1984;4:604–608.

Vahvanen V, Westerlund M. Fracture of the carpal scaphoid in children: a clinical and roentgenological study of 108 cases. Acta Orthop Scand 1980;51:909–913.

Waeckerle JF. A prospective study identifying the sensitivity of radiographic findings and the efficacy of clinical findings in carpal navicular fractures. Ann Emerg Med 1987;16:733–737.

INJURIES OF THE HAND AND FINGERS

Barton NJ. Fractures of the phalanges of the hand in children. Hand 1979;11:135–143.

Grad JB. Children's skeletal injuries. Orthop Clin North Am 1986; 17:437–449.

Innis PC. Office evaluation and treatment of finger and hand injuries in children. Curr Opinion Pediatr 1995;7:83–87.

Mastey RD, Weiss AP, Akelman E. Primary care of hand and wrist athletic injuries. Clin Sports Med 1997;16:705–724.

Torre BA. Epiphyseal injuries in the small joints in the hand. Hand Clin 1988;4:113–121.

Vicar AJ. Proximal interphalangeal joint dislocations without fractures. Hand Clin 1988;4:5–13.

FRACTURES OF THE PELVIS

Fernbach SK, Wilkinson RH. Avulsion injuries of the pelvis and proximal femur. Am J Roentgenol 1981;137:581–584.

Reichard SA, Helikson MA, Shorter N, et al. Pelvic fractures in children—review of 120 patients with a new look at general management. J Pediatr Surg 1980;15:727–734.

Watts HG. Fractures of the pelvis in children. Orthop Clin North Am 1976;7:615–624.

INJURIES OF THE HIP AND PROXIMAL FEMUR

Boyd KT, Peirce NS, Batt ME. Common hip injuries in sport. Sports Med 1997;24:273–288.

Chung SMK. Diseases of the developing hip joint. Pediatr Clin North Am 1986;33:1457–1473.

Morrissy R. Hip fractures in children. Clin Orthop 1980;152:202–210.

Offierski CM. Traumatic dislocation of the hip in children. J Bone Joint Surg 1981;638:194–197.

Paletta GA, Andrish JT. Injuries about the hip and pelvis in the young athlete. Clin Sports Med 1995;14:591–628.

St. Pierre P, Staheli LT, Smith JB, et al. Femoral neck stress fractures in children and adolescents. J Pediatr Orthop 1995;15:470–473.

FRACTURES OF THE SHAFT OF THE FEMUR

Anderson WA. The significance of femoral fractures in children. Ann Emerg Med 1982;11:174–177.

Gross RH, Strengler M. Causative factors responsible for femoral fractures in infants and young children. J Pediatr Orthop 1983;3:341–343.

INJURIES OF THE KNEE

Balmat P, Vichard P, Pem R. The treatment of avulsion fractures of the tibial tuberosity in adolescent athletes. Sports Med 1990;9:311–316.

Bertin KC, Goble EM. Ligament injuries associated with physeal fractures about the knee. Clin Orthop 1983;177:188–195.

Christie MJ, Dvanch VM. Tibial tuberosity avulsion fracture in adolescents. J Pediatr Orthop 1981;1:391–394.

Clanton TO, DeLee JC, Sanders B, et al. Knee ligament injuries in children. J Bone Joint Surg 1979;62A:1195–2101.

Crawford AH. Fractures about the knee in children. Orthop Clin North Am 1976;73:639–656.

Larson RL. Epiphyseal injuries in the adolescent athlete. Orthop Clin North Am 1973;4:839–851.

FRACTURES OF THE TIBIA AND FIBULA

Blatt SD, Rosenthal BM, Barnhart DC. Diagnostic utility of lower extremity radiographs of young children with gait disturbance. Pediatrics 1991;87:138–140.

Coady CM, Micheli LJ. Stress fractures in the pediatric athlete. Clin Sports Med 1997;16:225–238.

Dunbar JS, Owen HF, Nogrady MB, et al. Obscure tibial fracture of infants—the toddler's fracture. J Can Assoc Radiol 1964;15:136–144.

Hansen BA, Greiff J, Bergmann F. Fractures of the tibia in children. Acta Orthop Scand 1976;47:448–453.

Mellick LB, Reesor K. Spiral tibial fractures of children: a commonly accidental spiral long bone fracture. Am J Emerg Med 1990;8:234–237.

Oudjhane K, Newman B, Oh KS, et al. Occult fractures in preschool children. J Trauma 1988;28:858–860.

Singer J, Towbin R. Occult fractures in the production of gait disturbances in childhood. Pediatrics 1979;64:192–196.

Tenenbein M, Reed MH, Black GB. The toddler's fracture revisited. Am J Emerg Med 1990;8:208–211.

INJURIES OF THE ANKLE AND FOOT

Salter RB. Injuries of the ankle in children. Orthop Clin North Am 1974; 5:147–152.

Schindler A, Mason DE, Allington NJ. Occult fracture of the calcaneus in toddlers. J Pediatr Orthop 1996;16:201–205.

Schmidt TL, Weiner DS. Calcaneal fractures in children. An evaluation of the nature of the injury in 56 children. Clin Orthop 1982;171:150–155.

Starshak RJ, Simons GW, Sty JR. Occult fractures of the calcaneus—another toddler's fracture. Pediatr Radiol 1984;14:37–40.

Trott AW. Fractures of the foot in children. Orthop Clin North Am 1976; 7:677–686.

INJURIES OF THE THORACOLUMBAR SPINE

Bracken MB, Shepard MJ, Collins WF, et al. A randomized, controlled trial of methylprednisolone or naloxone in the treatment of acute spinal-cord injury. N Engl J Med 1990;322:1405–1411.

Dickman CA, Rekate HL, Sonntag VKH, et al. Pediatric spinal column: vertebral column and spinal cord injuries in children. Pediatr Neurosci 1989;15:237–256.

Gallagher DJ, Heinrich SD. Pediatric Chance fracture. J Orthop Trauma 1990;4:183–187.

Knapp JF, Harakas AP, Edwards RK, et al. Unusual mechanism of Chance fracture in an adolescent. Pediatr Emerg Care 1987;3:256–257.

Newman KD, Bowman LM, Eichelberger MR, et al. The lap belt complex: intestinal and lumbar spine injury in children. J Trauma 1990;30: 1133–1140.

Reid AB, Letts RM, Black GB. Pediatric Chance fractures: association with intra-abdominal injuries and seatbelt use. J Trauma 1990;30:384–391.

Sneed RC, Stover SL. Undiagnosed spinal cord injuries in brain-injured children. Am J Dis Child 1988;142:965–967.

Walsh JW, Stevens DB, Young AB. Traumatic paraplegia in children without contiguous spinal fracture or dislocation. Neurosurgery 1983;12:439–445.

CHAPTER 116

Minor Trauma—Lacerations

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LACERATIONS

Each year an estimated 12 million wounds are treated in emergency departments (EDs) in the United States. Lacerations account for 30 to 40% of all injuries for which care is sought in a pediatric ED. Broken glass, wooden furniture, asphalt or concrete, or other sharp objects cause most of these lacerations. Animal bites also account for many. More than 40% of the wounds involve a fall. Boys are the injured victims twice as often as girls. The mechanism of injury varies with patient's age. Household items, fences, and trees most likely injure preschoolers; violent encounters injure older children.

Two-thirds of the injuries occur during warm weather months, although half of the injuries in an urban environment occur indoors. Deaths from minor lacerations are rare; however, complications occur in about 8%. Complications include infection, hypertrophic scarring or keloid formation, and poor cosmetic results.

Pathophysiology

Wound Healing

Normal skin is under constant tension, produced, in part, by underlying joints and muscles. The amount of tension varies by anatomic location and position of a body part. For example, skin overlying a joint will vary in tension, depending on whether the joint is flexed or extended. Lacerations that run parallel to joints and normal skinfolds usually heal more quickly and with better cosmetic results. Wounds under a large amount of tension, crossing joints or perpendicular to wrinkle lines often heal with wide, unattractive scars. When skin is injured, sutures may be placed to provide temporary support until the skin can regenerate and overcome tension to allow wound closure.

A wound such as a laceration regains about 5% of its previous strength 2 weeks after injury and 30% after 1 to 2 months. It reaches full tensile strength 6 to 8 months after the original injury. Many factors, such as infection, tissue edema, and poor nutrition, may delay this progression.

All wounds deeper than the dermis have the potential for scar formation. Scar formation involves the laying down of collagen, which is a complex process essential in restoring tensile strength of the skin. Collagen synthesis begins within 48 hours of the injury and reaches a peak within the first week afterward. Anything that interferes with collagen synthesis, such as infection, may lead to wound dehiscence at this time. Wound contraction is expected with all healing wounds through the action of fibroblasts. Therefore, eversion of suture lines is desired at the time of repair so that the skin will contract to a flat wound during healing. Remodeling may occur for up to 12 months. Thus, the scar may fade and recede over the first 3 months, and the final appearance of the scar may not be apparent until 6 months after injury.

Wound Infection

Wound infection plays a major role in wound healing. Bacteria inhabit normal intact skin. This is the usual source of infection when skin tissue is disrupted. The amount of bacteria on the skin varies by anatomic location. High counts of bacteria are in moist areas such as the axilla and perineum. Low counts of bacteria are in dry areas such as the back, chest, and abdomen. High bacteria counts can also be expected in areas of exposed skin such as the hands, face, and feet. Areas colonized with high bacterial contamination are most prone to infection. Wounds in regions of high vascularity, such as the scalp and face, more easily resist bacterial infection despite the high bacteria count. Certainly, the oral cavity is highly contaminated with bacteria, and this is an important source of infection when a child sustains a

bite wound.

Wounds inflicted by shearing forces with a sharp object such as a knife cause minimal devitalization of adjacent areas and thus are less likely to lead to infection. Wounds caused by a blunt object striking the skin at an angle of less than 90 degrees result in a tension injury such as an avulsion or flap. These injuries involve a larger force applied to the skin than that of a shearing injury, and there is more devitalized tissue. They are more likely to become infected than shearing injuries and are often more difficult to repair. Finally, compression injuries from blunt trauma to the skin at about a 90-degree angle, cause the most tissue disruption and devitalization. They are characterized by ragged edges and lead to the highest infection rates and unacceptable scarring.

Clinical Manifestations

History

In the evaluation of a laceration, it is important to learn the *mechanism* of the injury because this may radically change management plans. For instance, if the wound was caused by an animal bite, the likelihood of devitalized tissue and infection is higher and repair may be omitted (see Chapter 91). Also, a wound caused by a blunt object may be associated with an underlying fracture or crush injury. Certain crush injuries, such as wringer injuries, are inherently more complicated and may require surgical consultation and hospital admission. A wound caused by a sharp object may have injured deeper tissues. The *age* of the wound should be determined, as well as the possibility of a *foreign body* in the wound.

The *location* of the wound must also be considered. If the wound is in the neck area, the physician should consider possible extension through the platysma muscle, with potential for a serious injury to underlying structures. If the wound involves the chest, the physician should look for crepitance in the subcutaneous tissue, suggesting injury to the underlying lung. An injury to the lower extremities is more likely to result in infection because of the relatively poor blood supply. Likewise, a wound overlying a joint space can be complicated if the joint cavity is violated. Injury to distal body parts such as the ear, nose, and fingers may threaten the viability of more distal tissues because of vascular compromise. Conversely, in areas where the vascular supply is good, such as the face, scalp, and tongue, the infection rate is low regardless of the mechanism of injury.

The *environment* in which the injury occurred should also be assessed. If the injury occurred on the street, it is possible that small particulate matter may be embedded in the wound. If this debris is left in place, tattooing of the skin could result, leaving an unfavorable appearance to the healed wound. Injuries that occurred in a field; farm; or a wet, swampy area may have high bacterial loads.

The patient's *health status* should be addressed. If the patient has diabetes, immunosuppression, malnutrition, or other chronic conditions, such as cyanotic heart disease, chronic respiratory problems, or renal insufficiency, higher infection rates may be anticipated. Bleeding disorders and current medications should be determined because some drugs, such as ibuprofen and corticosteroids, may have an impact on the wound. *Allergies* to latex, antibiotics, and local anesthetics, as well as the child's *tetanus status,* should be ascertained.

Physical Examination

A careful physical examination is essential before local anesthesia is given. First, determine whether there is an associated injury distant from the obvious wound. Wound management should not preempt care of more life-threatening injuries. It is important to assess the wound for *vascular damage* and to control bleeding if present. Brisk flow of dark blood may indicate injury to a major superficial vein. These vessels can usually be safely tamponaded and later ligated or sutured. Arterial bleeding is suspected when there is rapid flow of bright red blood. The bleeding site must be identified, although it is often obscured by profuse bleeding. Pressure applied to the site or temporary use of a tourniquet or inflated blood pressure cuff (less than 2 hours) controls hemorrhage and allows identification of the bleeding vessel. Blind clamping of an artery should be avoided except in the scalp. Palpation of pulses and capillary refill distal to the site of injury must be checked.

Next, potential *nerve damage* must be assessed. In an older, cooperative child, the physician should always test the median and ulnar nerve of an injured upper extremity. If a young child does not permit this, sensation may be tested with use of pinprick. Fortunately, when sensation is intact, motor function of the nerve is usually intact as well.

Next, the wound must be evaluated for possible *tendon injury.* The superficial location of extensor tendons of the dorsum of the hand predisposes them to injury. Tendon injuries are sometimes visible if the wound is wide and deep. For example, a torn tendon on the flexor surface of the forearm may be seen when the patient with a laceration to the wrist is asked to flex the hand and wrist. Unless the tendon injury is obvious, wounds over joints and tendons should be put through a full range of motion. A young patient may be too uncooperative to flex and extend the fingers on command. Therefore, it is important to inspect the resting position of the injured hand in a young child to note a flexor tendon injury to the finger. One digit may be found extended at rest, while the other uninjured digits are flexed (Fig. 116.1). Applying a noxious stimulus and noting inability to withdraw the finger that is tested may show injury to the extensor tendons.

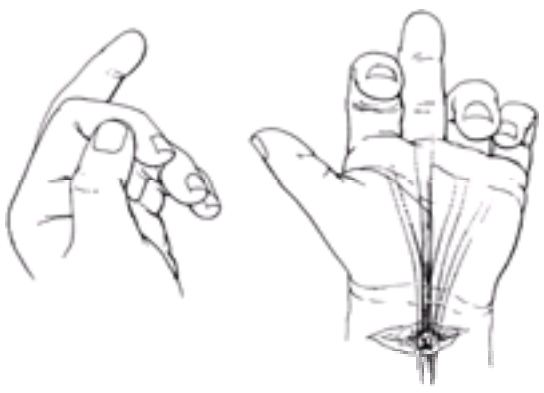

FIGURE 116.1. A seemingly superficial laceration at the wrist might be treated simply by closure of the subcutaneous tissue and skin, unless one appreciates the abnormal posture of the middle finger when the hand is at rest. The loss of normal flexor tone as a result of a divided superficial tendon results in the involved finger lying in a position of relative extension.

It should be determined whether *foreign material* is in the wound. If the history or physical examination suggests a radiopaque foreign body, obtaining a radiograph for confirmation should be considered. This is especially important in assessing a wound caused by glass. A deeply embedded piece of glass may be missed without radiographs. Some recommend obtaining such films in all cases in which glass is involved, except for the most superficial wounds. Ultrasound is also useful in selected cases for detecting and localizing foreign bodies. Further inspection for foreign material should take place after the wound is anesthetized.

Finally, *bones* nearby the wound should be palpated for crepitance, tenderness, or deformity, which may suggest a fracture. Radiographs should be obtained to confirm suspicious findings. Wounds overlying a fracture deserve consultation with a specialist for possible repair in the operating room. Table 116.1 summarizes general principles of wound assessment.

Primary survey—control bleeding	Physical Examination
Secondary survey—other injury?	Location
History	Muscle function
Mechanism	Tendon involvement
Age of wound—time of injury	Vascular injury
Possible foreign body	Nerve injury
Environment	Foreign material
Health status—tetanus	Laboratory
	Consider radiographs or ultrasound if a foreign body or fracture is suspected

Table 116.1. Wound Assessment—General Principles

Patients found to have vascular, nerve, or tendon injury or deep, extensive wounds to the face merit consideration for referral to a surgical specialist for possible repair in the operating room.

Decision to Close the Wound

Children are less likely to get wound infections compared with adults. In children, the infection rate is about 2% for all sutured wounds. Thus, most wounds may be closed primarily, meaning the wound edges are approximated as soon after the injury as possible to speed healing and improve the cosmetic result. If primary closure is long delayed, the risk of subsequent infection increases. However, the length of time before the risk of infection becomes significant is variable. Some authors suggest that the “golden period” for wound closure is 6 hours. However, wounds at low risk for infection (e.g., a clean kitchen-knife injury) can be closed even 12 to 24 hours after the injury. In a study from a developing country where patients presented with wounds after variable delays in care, it was found that wounds of the face and scalp heal well in more than 90% of cases, regardless of the time from injury to repair.

Most wounds of the face are best closed primarily, even up to 24 hours after injury to achieve an optimal cosmetic effect. If the wound is extensive or has a high potential for infection (e.g., a dog bite on the face), thorough irrigation is essential, and in some cases, the operating room may be the best site for this. On the contrary, wounds at high risk for infection such as those in anatomic locations with poor blood supply, contaminated or crush wounds, and those involving immunocompromised hosts should be closed promptly, within 6 hours of injury. Some contaminated wounds (animal or human bites or those occurring in a barnyard) in an immunocompromised host should not be sutured, even if the patient presents immediately for care. Thus, the decision to close a wound must be individualized.

Some wounds should be allowed to heal by *secondary intention* (secondary closure), although scar formation may be more unsatisfactory. Infected wounds, ulcers, and many animal bites are best left to heal by granulation and reepithelialization. Puncture wounds to the foot, with only a small laceration and a low concern for cosmetic results, may also be left open. A small sterile wick of iodoform gauze may be placed inside the wound to keep the edges open. This

gauze can be removed after 2 to 3 days, and the subsequent granulation tissue will aid healing.

If a wound is not closed initially, *delayed primary closure* (tertiary closure) should be considered after the risk of infection decreases, about 3 to 5 days later. This is recommended for selected heavily contaminated wounds and those associated with extensive damage such as high-velocity missile injuries, crush injuries, explosion injuries of the hand, and perhaps bite wounds. The wound should be cleaned and debrided and covered at the time of initial presentation, then reassessed in a few days for infection. It is believed that a contaminated but healing wound gradually gains sufficient resistance to infection to permit uncomplicated closure at a later time. This may still reduce discomfort and lead to a better cosmetic result than no repair at all. Tertiary closure is used rarely in pediatrics because children have few severely contaminated wounds from farm or industrial injuries.

Management of Lacerations

Preparing the Child and Family

It is important to reassure the child and the family that everything will be done to care for the wound appropriately and to relieve the patient's pain and anxiety. In many cases, early removal of blood and foreign material from the surface of the wound is reassuring. Also, carefully chosen words will reduce fear and pain from the procedure. The physician must honestly warn the patient of an impending painful stimulus but may leave open the possibility that it may not hurt as much as the child thinks. Appearing unhurried and confident, giving the child some control of the situation, and explaining the upcoming procedure seems to help reduce anxiety and pain. The parent(s) and child should be informed that steps will be taken to make the procedure as quick and painless as possible, such as with the use of topical anesthetics. The clinician should provide an age-appropriate empathic explanation, rather than give cold, impersonal instructions about a painful procedure to reduce anxiety. Prepare frightening instruments, such as needles and scalpels, away from the child. Allow the child to listen to music or view age-appropriate, entertaining videos during the procedure because this may serve as a distraction (see Chapter 4).

Some debate exists about the value of allowing parents to remain in the room during the procedure. Most parents want to be present during wound repair in the ED, and most can be a stabilizing force if properly oriented. The parent can reassure or distract the child with a story while maintaining physical contact under necessary drapes and restraints. It is usually best if the parent is sitting down and focusing on the child, rather than directly observing the procedure.

Many young children less than 4 years old will need to be placed in a restraining device such as a papoose board for better immobilization. Restraint is needed to ensure the child's safety, protect him or her from self-injury, and allow for more rapid completion of the procedure. Because the child may get excessively warm while in the papoose board, it is important to ensure proper ventilation and assess the child's comfort during the restraint process. A caring, but firm nurse or assistant is often needed to further immobilize the injured body part and complete the procedure successfully. It is better to use such hospital personnel instead of parents to immobilize a child. Sedation may be appropriate in some cases (see Chapter 4). A school-age child can usually cooperate without restraint.

Preparing the Wound

Appropriate use of conscious sedation and *local anesthetics* are essential for successful repair of lacerations in children (see Chapter 4).

Hair near the wound usually creates minimal difficulty during repair and generally does not need to be removed. In any case, nearby hair should not be shaved because this may damage hair follicles and increase infection. Instead, the hair should be clipped with scissors when necessary. Alternatively, petroleum jelly can be used to keep unwanted scalp hair away from the wound while suturing. Hair over the eyebrows should not be removed because this may lead to abnormal or slow regrowth.

It is essential to *clean the wound periphery* at the time of wound evaluation. Povidone–iodine solution (a 10% standard solution) is often used because it is a safe and effective antimicrobial with little tissue toxicity. This solution may be diluted with saline 1:10 to create a 1% solution. Use of chlorhexidine or povidone–iodine surgical scrub preparations, hydrogen peroxide, or alcohol in the wound itself is not recommended. These may be irritating to tissues, and they may increase infection by damaging white cells.

Wound irrigation is extremely important to reduce bacterial contamination and prevent subsequent infection. It is often necessary to anesthetize the wound before thoroughly cleansing. Using universal precautions, the wound should be irrigated with normal saline, about 100 to 200 mL for the average 2-cm laceration. More may be needed if the wound is unusually large or contaminated. Use a large syringe (20 to 50 mL) with a splash-guard attached to the end to reduce splatter during the irrigation. With the splash-guard almost touching the skin surface and the tip of the syringe about 2 cm from the wound, the clinician should apply firm pressure to the plunger. Soaking the injured body part should be avoided because this may lead to maceration of the wound and edema.

Scrubbing the wound should be reserved for particularly “dirty” wounds in which contaminants are not effectively removed with irrigation alone. It may be necessary to extract some foreign material with fine forceps if it remains adherent after copious irrigation. This will avoid tattooing of the skin and reduce the risk of infection.

In rare cases, the wound must be extended with a scalpel to allow proper exploration and cleaning. The physician should trim irregular lacerations and excise necrotic skin but should not make dramatic changes in the wound. Devitalized tissue should be removed only if it looks ischemic or is otherwise clearly indicated. More than a small amount of tissue removal should be attempted only by an experienced physician. Subcutaneous fat can be safely and easily removed if this seems to interfere with wound closure. It is wise to remove this carefully, in small quantities, to avoid disruption of small vessels and cutaneous nerve branches. Debridement is advantageous because it creates well-defined wound edges that can be

more easily opposed. However, excessive removal of tissue can create a defect that is difficult to close or may increase tension at the wound margin such that scarring is more likely. Removal of facial fat should be avoided because this may leave an unsightly depression.

Further examination of the wound should take place after cleansing and debridement. After exploration, it is wise to reevaluate the decision to close the wound primarily. When proceeding further, the clinician should wash hands carefully before donning sterile gloves. Sterile masks are not helpful in reducing wound infections, but a facial splash-shield is useful to protect the clinician. The area surrounding the wound should be appropriately draped before surgical repair. However, if a young child is particularly upset by facial drapes, they can be omitted. Proper cleaning of the wound is more important to uncomplicated healing than meticulous attempts to avoid introduction of small numbers of bacteria by preserving a sterile field.

Wound Closure

Equipment

Suture material must have adequate strength while producing little inflammatory reaction. Nonabsorbable sutures such as monofilament nylon (Ethilon) or polypropylene (Prolene) retain most of their tensile strength for more than 60 days and are relatively nonreactive. Thus, they are appropriate for closing the outermost layer of a laceration. With monofilament nylon, it is important to secure the knot adequately with at least 4 to 5 throws per knot. Polypropylene is useful for lacerations in the scalp or eyebrows because it is more visible and thus easier to remove. It is somewhat more difficult to control while suturing. Silk is rarely used now because of increased tissue reactions and infection.

In many cases, it is appropriate to use fine, absorbable (synthetic) sutures such as Dexon or Vicryl in deeper, subcuticular layers. These materials may elicit an inflammatory response and may extrude from the skin before they are absorbed if they are placed too close to the skin. When subcuticular sutures are used, they should be placed on the deeper surface of the dermis, and epithelial margins may be approximated with tape strips. Synthetic absorbable sutures are less reactive than chromic gut and they retain their tensile strength for long periods, making them useful in areas with high dynamic and static tensions. Absorbable sutures are advantageous for intraoral lacerations. Some recommend using rapidly absorbable sutures (fast-absorbing gut) for skin closure of facial or scalp wounds in children because suture removal is avoided.

A 3-0 suture is recommended for tissues with strong tension, such as fascia, and 4-0 is recommended for deep tissues with light tension, such as subcutaneous tissue. Skin is best closed with 4-0 to 7-0 and oral mucosa with 3-0 to 4-0 sutures. The physician should use the finest sutures (6-0) for wounds of the face; heavier sutures for scalp, trunk, and extremities (4-0 or 5-0); and 3-0 or 4-0 for thick skin such as the sole of the foot or over large joints such as the knee.

Needles are available in a variety of forms, including cuticular, plastics, and “reverse cutting.” The reverse cutting needle is used most for laceration repair. Its outer edge is sharp to allow for atraumatic passage of the needle through the relatively tough dermis and epidermal layers; this minimizes cutting of the skin where suture tension is greatest. A higher grade plastic needle (designated *P* or *PS*) should be used for repairs on the face. A small needle (such as P3) should be used for wounds that require fine cosmesis. Needles come in a variety of sizes such as $^{3}/_{8}$ and ½ circle. Clinicians may develop a preference for a specific needle. In general, a $^{3}/_{8}$ reverse cutting needle satisfies most needs.

General Principles

Perhaps the two most important goals of suturing are to match the layers of the injured tissues and to create eversion of the wound margins so that they will flatten as the wound heals. Layers on one side of a wound should be sutured to the corresponding, matching layers on the other side. First, all layers of skin that have been injured should be identified. Then an attempt to appose each layer (muscles, fascia, subcutaneous tissue, and skin) as nearly as possible back to its original location should be made. This is achieved by carefully matching the depth of the bite taken on each side of the wound when suturing.

Proper *suture placement* should result in slight eversion of the wound so that there is not a depressed scar when remodeling takes place. Eversion may be achieved by slight thumb pressure on the wound edge as the needle is entering the opposite side. Sutures should take equal bites from both wound edges so that one margin does not overlap the opposite margin when the knot is tied. Wound edge eversion is best achieved by taking proper bites while suturing, not by pulling the knot tightly (Fig. 116.2).

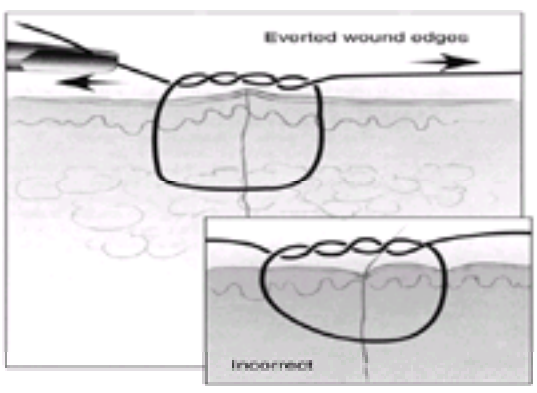

FIGURE 116.2. Suturing technique for wound edge eversion.

Suture placement may be deep or superficial. Deep sutures reapproximate the dermal layers of skin and do not penetrate the epidermis. They help relieve skin tension and improve the cosmetic appearance by reducing the width of the scar. They should be avoided in wounds prone to infection because they will further increase the risk of infection. To place a deep suture, the needle is placed at the depth of the wound and removed at a more superficial level. The needle is then inserted superficially into the opposite side of the wound and exits deeply so that the knot is buried within the wound. The needle end and free end of the suture should be on the same side of the loop before the knot is tied (Fig. 116.3). The simple interrupted technique (described next) with absorbable suture material should be used.

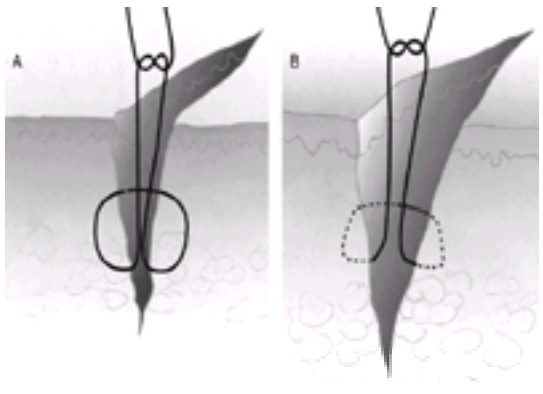

FIGURE 116.3. A. The buried subcutaneous suture. **B.** The horizontal dermal stitch.

Superficial or percutaneous sutures are passed through the dermis and epidermis and leave the knot visible at the skin surface. Skin should be closed with a minimal amount of tension. Sutures should be pulled tightly enough to approximate the wound edges, but not so tightly that they cause tissue necrosis. Sutures that seem well placed initially may begin to cut into the tissue in the next few days because of swelling and inflammation. There is no need to tightly close the skin if other layers have been well sutured. Scalp wounds are an exception. They are under considerable tension, and the knots in this location should be pulled firmly to keep the skin together. The wound will be hidden by hair, so the skin can be pulled more tightly than elsewhere. Firm, but not strangulating, apposition of the wound will help with hemostasis as well.

To ensure proper alignment, the first suture may be placed at the midpoint of the wound, with subsequent sutures then placed in a bisecting fashion lateral to the midpoint. Use of forceps to hold tissue should be encouraged because this allows the operator to precisely pass the needle through the desired points alongside the wound edge. However, forceps use should be kept to a minimum during the repair to avoid tissue damage.

Suture Technique

Skin wounds can generally be repaired using interrupted suturing. To place a *simple interrupted suture,* the needle is held upside down and the wrist is pronated as the needle enters the skin at a 90-degree angle. The needle tip will then move farther away from the wound margin and penetrate deeply. Thus, more tissue is at the depth of the wound and this causes the wound to evert. Sutures should be placed about 2 mm apart and 2 mm from the wound edge on delicate areas such as the face. More sutures placed closer together decrease wound tension and leave a less noticeable scar. Larger bites should be used for body parts where cosmesis is less important.

The physician should use an instrument tie to secure the suture (Fig. 116.4). The knots should ideally be placed on one side of the wound. Knots placed directly over the wound increase inflammation and scar formation. On the first throw, the physician should wrap the needle holder twice to create a surgeon's knot and then wrap subsequent throws a single time. The first and second throws should be snug enough to approximate the wound edges, but not so tight that tissue is strangulated. All subsequent knots are squared to maintain the closure. Four or five throws are usually required to keep the knot from unraveling. A “loop knot” is effective in apposing the wound edge with minimal tension. This involves placing a surgeon's knot, using the instrument tie, followed by a loop. The surgeon's knot will “give” slightly should edema develop subsequently. The loop knot allows easier, painless removal of sutures because it creates a free space between the suture and the skin (Fig. 116.5).

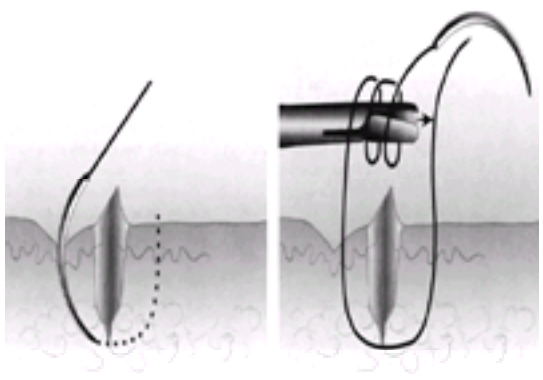

FIGURE 116.4. Simple interrupted skin suture secured with instrument tie.

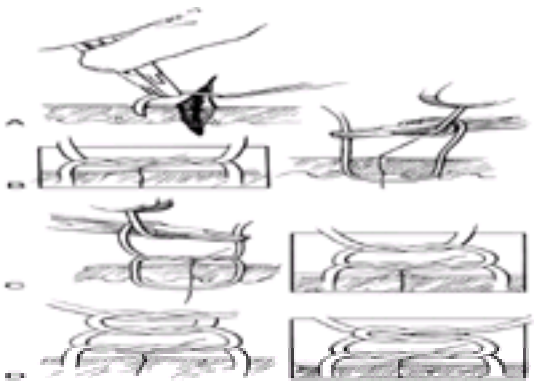

FIGURE 116.5. Placement of a “loop knot” in conjunction with simple sutures of the skin using an eversion technique. **A.** The needle enters the skin at a right angle in a way that allows somewhat less skin and more subcutaneous tissue to be caught in the passage of the needle. The needle should incorporate the same amount of skin and subcutaneous tissue on each side. The ideal suture material for placing a “loop knot” is 4-0 nylon. One can also use 5-0 nylon. **B.** The first knot should be a surgeon's knot drawn down gently so as to barely coapt the skin edges. **C.** The second tie should be placed so as to produce a square knot but should be drawn so as to produce an approximate 2- to 3-mm loop. **D.** The third tie should be placed so as to produce a square knot. This third tie can be secured tightly against the second tie, preserving the loop and allowing for some loosening of the surgeon's knot spontaneously as later edema develops.

Running or continuous sutures can be applied rapidly to close large, straight wounds or multiple wounds. With this technique, the suture is not cut and tied with each stitch. The first suture is placed at one end of the wound and a knot tied, cutting only the end of thread not attached to the needle. The next loop is placed a few millimeters away and continuous loops of equal bites are made to close the wound. On the final loop, the suture is not completely pulled through so that a small loop remains on the opposite side of the wound. Now, the knot can be tied using the preceding loop of suture (Fig. 116.6). This type of stitch is more likely to leave suture marks if not removed in 5 days. Apposition of the edges and eversion are more difficult to achieve with this stitch, and the entire suture line can unravel if the suture breaks anywhere along the repair. However, the technique gives the advantage of having equal tension on the wound edges.

FIGURE 116.6. Continuous skin sutures. **A.** The simple continuous running stitch. **B.** The continuous interlocking skin stitch. **C.** The running lateral mattress stitch or continuous half-buried horizontal mattress stitch.

The *vertical mattress stitch* is useful for deep wounds in which it may be difficult to tie a simple, deep, interrupted suture. It reduces tension on the wound and may close dead space within the wound. It essentially combines a deep and superficial stitch in one suture. The needle is placed deep within the wound (about 3 mm from the wound edge) and brought out to the opposite skin surface. It is then brought across the epidermis to approximate the epidermal edges (Fig. 116.7). This stitch takes more time to accomplish and produces more crossmarks, but it provides excellent wound eversion and apposition of the wound edge. Too tight of a knot will pucker the wound.

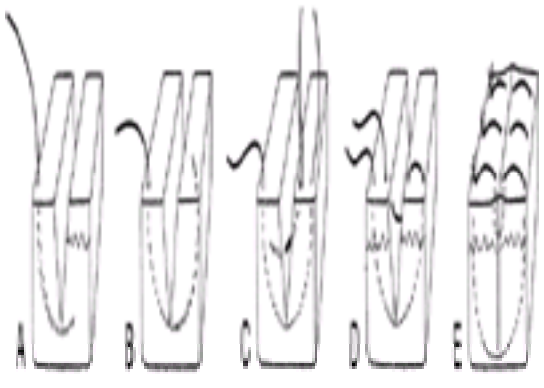

FIGURE 116.7. A–E. The vertical mattress suture. After initially placing a simple interrupted stitch with a somewhat larger bite, make a backhand pass across the wound, taking small, superficial bites. When the knot is tied, the edges of the laceration should evert slightly. (Reprinted with permission from Grisham J. Wound care. In: Dieckmann RA, Fiser DH, Selbst SM, eds. Illustrated Textbook of Pediatric Emergency & Critical Care Procedures. St. Louis: Mosby,

1997:665–679.)

The *horizontal mattress stitch* reinforces the subcutaneous tissue and effectively relieves tension from the wound edges. It does not provide wound-edge approximation as well as the vertical mattress stitch. The needle is passed ½ to 1 cm away from the wound edge deeply into the wound. It is then passed through the opposite side and reenters the wound parallel to the initial suture. To avoid “buckling” and to provide some eversion of the wound edges, the skin must be entered perpendicularly, and the wound must be entered and exited at the same depth (Fig. 116.8).

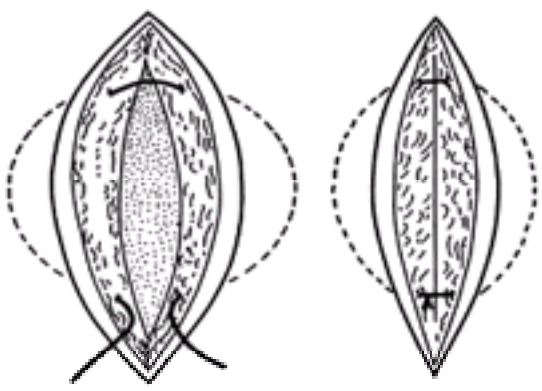

FIGURE 116.8. The horizontal mattress stitch is useful for closing the deep layer in shallow lacerations and in body areas with little subcutaneous tissue. Certain dyed suture materials may cause a tattooing of the skin if placed in such a shallow position. (Reprinted with permission from Grisham J. Wound care. In: Dieckmann RA, Fiser DH, Selbst SM, eds. Illustrated Textbook of Pediatric Emergency & Critical Care Procedures. St. Louis: Mosby, 1997:665–679.)

The *modified horizontal mattress stitch* (half-buried) is often used to close a flap. It is also called the corner stitch. It relieves intrinsic tension and vascular compromise when approximating the tip of the flap. Using 5-0 or 6-0 nylon, the physician should enter intact skin across from the apex of the flap and exit the wound just below the subcuticular plane. The needle should be brought to the tip of the flap, entering and exiting at the subcuticular plane. Then, the needle is brought across the edge of the flap in the subcuticular plane and the skin is exited. A knot should be tied in the usual manner and the tip of the flap brought to the apex of the wound (Fig. 116.9).

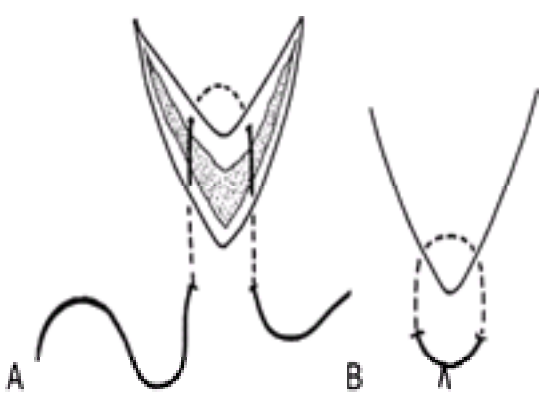

FIGURE 116.9. A and **B.** The corner stitch. Also called the half-buried horizontal mattress stitch, this technique allows repair of flap-type lacerations without further compromising blood flow. Place additional simple interrupted sutures along the sides of the flap if necessary. (Reprinted with permission from Grisham J. Wound care. In: Dieckmann RA, Fiser DH, Selbst SM, eds. Illustrated Textbook of Pediatric Emergency & Critical Care Procedures. St. Louis: Mosby, 1997:665–679.)

Placing the needle in the flap edge first can also repair wounds with flaps. The edge of the flap can then be moved back and forth until proper alignment with the opposite fixed side is obtained. After the tip of the flap is sutured, the sides of the flap are brought together. For wounds with several stellate flaps, subcuticular or subcutaneous sutures should be used to hold the tips of the flap together. Then, a single suture at the tip will provide good apposition without further damaging the tip of the flap. Other interrupted sutures can be placed on the lateral margins of the wound to provide further support. If the wound has many narrow-based stellate flaps or necrotic flap tips, the wound may be better managed with excision and simpler repair (Fig. 116.10).

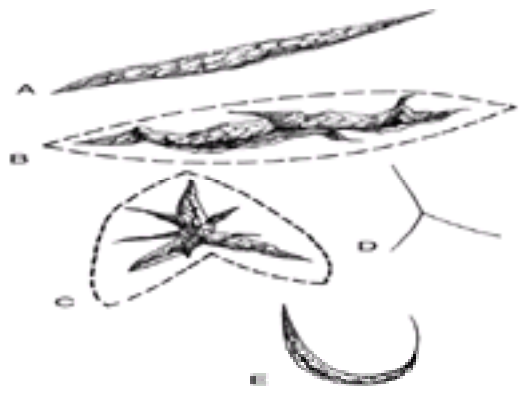

FIGURE 116.10. Variation in laceration injuries and suggestions for management: simple laceration **(A),** elliptical excision of damaged wound margins **(B),** excision and closure of stellate laceration (**C** and **D**), and flap-type laceration **(E).**

Alternative Wound Closure Techniques

Tape causes no suture marks, minimal tissue reaction, and fewer wound infections than sutures. Tape strips, cut to size, can be used to take up tension at the wound margins and can be placed between sutures. These strips are also useful as the only means to close simple lacerations that barely extend through the dermis. Multiple tangential, triangular skin flaps (e.g., those created when an unrestrained passenger hits the windshield of a car) are closed well with tape strips. Likewise, old or contaminated wounds, such as dog bites on the extremities, can be loosely approximated with skin tape.

When tape is used, the wound should be cleansed as any other wound. Care must be taken to properly realign the dermis and epithelium. If the tape is pulled too tightly, the margins of the wound may overlap, causing the wound to heal with a raised ridgelike area where the overlap occurred. The tape is applied perpendicularly across the wound with some space between to allow the wound to drain. In some cases, an adhesive such as benzoin is applied to the adjacent skin (not the wound) to keep the tape strips more securely in place. Some recommend leaving the taped wound uncovered because a bandage may increase moisture and cause the tape to fall off prematurely.

Tape strips should not be used on wounds subject to tension, such as those over flexor surfaces of joints. They should not be applied in areas of the body that are moist, such as the palms or axillae because they will not adhere. They may be impractical for small children, who may inadvertently remove them from the face.

Staples can be applied more rapidly than sutures and have a lower rate of infection, with less of a foreign body reaction. They are best for wounds of the scalp, trunk, and extremities when saving time is important. Therefore, they are particularly helpful when treating mass casualties. Staples are left in place for the same length of time as sutures. They are somewhat more painful to remove and should be removed with a specially designed instrument to avoid tissue damage. Staples do not allow for meticulous cosmetic repair as with sutures. Thus, they should not be used for lacerations of the face, neck, hands, or feet. They should also not be used if the patient requires magnetic resonance imaging (MRI) or computed tomography (CT).

Tissue adhesives, or skin glues, such as cyanoacrylates (Histoacryl Blue) have recently been approved for use in the United States. Skin glues have been used to close wounds for many years in Europe and Canada. They allow rapid and painless closure of wounds. Anesthesia is unnecessary, unless painful irrigation or exploration of the wound is anticipated. No removal is needed because the adhesives slough off after 7 to 10 days. They provide an excellent cosmetic result, comparable with sutures. One study using plastic surgeons blinded to the method of repair graded the wounds repaired with Histoacryl Blue to be cosmetically equal to sutured wounds at 2-month and 1-year follow-up visits.

Tissue adhesives act to decrease wound infections because they have antimicrobial effects against Gram-positive organisms. Dehiscence rates (1 to 3%) are similar to that of sutured wounds. They are less expensive than sutures because little equipment is needed and personnel time is reduced. Studies have noted that patients and families of small children prefer them to sutures. Routine follow-up is not needed for uncomplicated wounds, and no long-term complications have been reported.

Before application of the tissue adhesive, the wound is cleaned and hemostasis achieved with dry gauze and pressure. The wound's edges are held together manually or with forceps while the tissue adhesive is applied drop-wise along the surface of the wound. The tissue adhesive should not be applied to the inside of the wound because it will act as a foreign body and inhibit healing. The wound is then held in place for about 20 to 30 additional seconds to obtain adequate bonding. One study reported that if malalignment is noted, the adhesive can be removed with forceps and reapplied without further complication. The wound is then covered carefully so that bandage removal will not pull off the tissue adhesive.

Tissue adhesives should be used only to close skin of superficial wounds. For many lacerations, deep absorbable sutures will also be needed because the glue has less strength than most sutures. Skin glues should not be used for wounds subject to great tension, such as on the hands or joints.

Table 116.2 summarizes advantages and disadvantages of several techniques available for wound closure.

Technique	Advantages	Disadvantages
Sutures	Greatest tensile strength Meticulous closure Low dehiscence rate	Painful Removal needed Slow application Increased tissue reaction Risk of needle stick (clinician)
Staples	Rapid application Low cost Low tissue reaction	Not for use on face (less meticulous closure)
Tissue adhesive	Rapid application Painless No removal needed Low cost No risk of needle stick (clinician)	Lower tensile strength Not for use on joints
Tape strips	Rapid application Painless Low cost Low infection risk Least tissue reaction	High risk of dehiscence Not for use in moist areas, young children

Table 116.2. Common Techniques of Wound Closure

Dressings

Dressings protect the wound from further injury and contamination. They also help absorb secretions (not likely with small wounds) and immobilize the injured part. Some use a nonadherent sterile dressing (e.g., Telfa, Xeroform) to cover the wound. This prevents the wound edges from sticking to the dressing. Then, a second layer of absorbent gauze is applied, and a third layer of gauze wrap or tape is used to stabilize the other two. This protects and immobilizes the area while absorbing exudate from the wound surface.

For most simple wounds, it is adequate to cover the wound with dry sterile gauze after applying topical antibacterial ointment. Some studies indicate that topical antibiotic ointments may reduce infection and prevent scab formation by lubricating the wound edges. This allows for more rapid epithelialization of the wound.

For the face and trunk, a large bulky dressing is not practical. Thus, for small wounds in those areas, a clear plastic adhesive such as Tegaderm should be used to secure the bandage. Rolls of cotton or stretchable tube gauze can be used for larger wounds to keep the sterile dressing in place. This keeps the young child from touching the wound. Scalp wounds are usually not dressed. Patients can generally wash their hair gently after 24 hours.

For children who are active, it may be best to keep the wound covered until sutures are removed. The dressing should remain in place for 24 to 48 hours after which epithelialization is usually sufficient to keep the wound from gross contamination. Then, the bandage should be changed daily and the wound inspected. Any dressing should be changed if it becomes soiled, wet, or saturated with drainage because the wet dressing may be a source of infection.

It may be advisable to splint the wound if it overlies a joint. This is most important for active children who will likely resume full activity soon after the injury. Some even recommend splinting nearby joints for any large laceration of an extremity to reduce stress across the wound even if it does not involve a joint itself. This should be done for no more than 72 hours to facilitate function. The injured extremity should be elevated to provide comfort and reduce edema.

Systemic Antibiotics

Use of prophylactic systemic antibiotics for wound management is controversial. Studies demonstrating proven benefit to use of antibiotics are lacking. They may lead to allergic reactions, growth of resistant organisms, and unnecessary expense. Thus, they are not recommended for routine use. Decontamination with proper irrigation is more efficacious than use of antibiotics to prevent wound infection. Antibiotics should be considered for heavily contaminated wounds that are at greater risk for infection. They are often used for human and cat bites (see Chapter 91), crush injuries, stellate lacerations, and very long wounds (exceeding 5 cm). Other high-risk wounds include intraoral lacerations and wounds of the hands, feet, and perineum. Similarly, open fractures, exposed joints and tendons, and any tetanus-prone wound may benefit from antibiotics. Likewise, wounds that result in exposed cartilage of the nose or ears or extensive facial wounds that may involve contamination from adjacent nasal passages are often treated with antibiotics. It may also be reasonable to use antibiotics for wounds (other than scalp lesions) when repair takes place more than 12 hours after injury. They may be justified for wounds that occurred in a contaminated environment, such as a farm or roadside. Injured immunocompromised patients may warrant antibiotics.

Usually a first-generation cephalosporin or a penicillinase-resistant penicillin is used to cover staphylococci and streptococci. Amoxicillin–clavulanic acid is recommended for wounds created by mammalian bites (see Chapter 91). Additional coverage for Gram-negative organisms with an aminoglycoside may be worthwhile for heavily contaminated open fractures.

Tetanus

Immunization status of all injured patients should be documented in the medical record. If the wound is clean and minor and the patient has received three previous doses of tetanus toxoid, a booster of tetanus toxoid is given only if 10 or more years have passed since the last dose. If a patient has received three or more previous tetanus immunizations but the wound is not a clean, minor laceration, tetanus toxoid is indicated if the last dose was more than 5 years prior.

In many cases, the tetanus immunization record is unknown. If tetanus status is unknown and the wound is not clean or minor, tetanus toxoid and tetanus immunoglobulin (TIG) are indicated. Wounds involving massive tissue destruction and contamination may also require TIG. Patients with such wounds should be admitted to the hospital (see Table 116.3).

Prior Tetanus Toxoid Immunization (Doses)	Clean Minor Wound	All Other Wounds
Uncertain (or less than 3)	DTP or Td	DTP or Td and TIG or TAT
Three or more (most recent more than 10 years ago)	Td	Td
Three or more (most recent within past 5 years)	None	None
Three or more (most recent between 5 and 10 years)	None	Td

DTP, diphtheria, tetanus, pertussis toxoid; *Td*, adult formulation of diphtheria, tetanus toxoid; *TIG*, tetanus immunoglobulin (dose: 250–500 units IM); *TAT*, tetanus antitoxin; should be used only if TIG is not available and after testing (dose: 3000–5000 units intramuscularly).

Table 116.3. Tetanus Prophylaxis

Discharge Instructions and Suture Removal

Careful discharge instructions, regarding wound care, covering the wound, when to get the wound wet, how to dry it, and so on, are extremely important. The family should be warned about signs of infection. Specifically, they should be told to return for medical care if the wound develops increasing pain, redness, edema, and/or wound discharge, or if the child develops a fever. Analgesics may be given for minor pain, but worsening pain should always prompt a wound check. The family should also be informed that the wound was inspected for a foreign body but that there is still a possibility of a retained foreign body or an undetected injury that may require further treatment. Parents should be told that no matter how skillful the operator, every laceration leaves some scar. The appearance of the scar will change during the next several months and the scar's appearance will not be complete for about 6 to 12 months. Patients and parents should be advised to keep the injured part elevated when possible. A sling can be provided to accomplish elevation of the upper extremities. Some recommend that healing skin not be exposed to sunlight for 6 months after injury because this could lead to permanent hyperpigmentation.

Follow-up care should be arranged for 24 to 48 hours in all but very simple wounds. The wound can then be reinspected for signs of infection, and healing can be assessed.

Wounds closed with tape strips do not require removal of the tape because these will fall off spontaneously. Skin glue also sloughs spontaneously. However, nonabsorbable sutures should be removed at the appropriate time, depending on the location of the injury. The importance of timely removal should be stressed to the patient and family. Sutures should be removed when fibroblastic proliferation at the wound interface is strong enough to take the place of sutures. Removing sutures too early may lead to dehiscence and widening of the scar. Sutures left in too long may create an unnecessary tissue reaction and result in visible cross-hatching (“railroad ties”).

Wounds on the scalp or face are nourished by a better blood supply and generally exhibit more rapid healing. Sutures in these areas are removed more quickly than other locations to avoid unsightly tracts. When sutures are subject to considerable tension (over joints and on the hands), they should be left in longer (Table 116.4). After removal of sutures, it is often necessary to reinforce the healing wound with tape strips to prevent dehiscence.

Wound Location	Time of Removal (days)
Neck	3–4
Face, scalp	5
Upper extremities, trunk	7–10
Lower extremities	8–10
Joint surface	10–14

Table 116.4. Timely Suture Removal

In the first 24 to 48 hours, wound dressings should be changed only if wet or soiled. After that, bathing can be permitted as long as the wound is then patted dry and covered again. There is no proven harm to exposing the sutures to soap and water for short periods.

Table 116.5 summarizes an approach to reduce risk in managing wounds in the ED.

1. Take thorough history.
2. Perform a careful examination.
3. Obtain a consult for complex wounds.
4. Obtain radiographs if foreign body or fracture is suspected.
5. Document carefully (inspection, irrigation, and function).
6. Communicate with parents (likely scar).
7. Arrange follow-up, recheck.

Table 116.5. Reducing Risk in Wound Management

CARE FOR COMMON WOUNDS

The principles of wound care discussed earlier should be applied in repairing any of the wounds discussed in this section. These principles include evaluation of the wound by history, physical examination, and when indicated, radiographic examination. After the wound is evaluated, the feasibility of closure and the possible need for consultation with a surgeon should be addressed. The following section discusses some of the commonly encountered wounds in children.

Facial and Oral Wounds

Forehead Lacerations

Forehead lacerations are common in early childhood. Most of these injuries occur secondary to falls on objects or furniture such as coffee tables. Most of these lacerations are simple and not associated with any other significant injuries. However, complete evaluation of the head and neck should be carried out. Superficial transverse lacerations of the forehead are easy to manage, and the outcome is usually favorable. Closure with simple or continuous cuticular sutures using 6-0 nonabsorbable material is recommended. Deeper transverse lacerations involving the deep fascia, the frontalis muscle, or the periosteum should be repaired in layers. Absorbable 5-0 material such as coated vicryl or catgut can be used. If the deeper tissue plains are not closed, the function of the frontalis muscle, eyebrow elevation, may be hampered. Other facial expressions can also be affected because the skin may tether to the scar tissue bridging the unrepaired gaping tissues.

Vertical forehead lacerations tend to have a wider scar because they traverse the tension lines. Complex forehead wounds, such as stellate lacerations from windshield impact and those with tissue loss, particularly secondary to animal bites, may require consultation with a plastic surgeon. Forehead lacerations are rarely associated with skull fractures, but facial or intracranial injuries should be ruled out.

Lacerations of the Eyebrow

Eyebrow lacerations are common. Repairing an eyebrow laceration is complicated by the presence of hair. It is advisable not to shave the eyebrow for wound preparation because it serves as a landmark during repair. Also, eyebrow regrowth is unpredictable; it may be either slow or incomplete, potentially leading to poor cosmetic outcome. Debridement, if required, should be minimal and along the same axis of the hair shafts to avoid damage to hair follicles; otherwise, alopecia of the brow will result. Closure with simple interrupted stitches using nonabsorbable material is usually sufficient. Attention must be paid to avoid inverting the hair-bearing edges into the wound. It is also important to pay attention to proper alignment of both ends along an eyebrow wound.

Lacerations of the Eyelid

Most eyelid lacerations are simple transverse wounds of the upper eyelid just inferior to the eyebrow. Repairing these wounds does not require any special skills. On the other hand, recognizing complicated eyelid lacerations is crucial for proper repair and good outcome. Vertical lacerations involving the lid margin require precision in approximation to avoid deformity and malfunction of the eyelid. Injuries potentially involving the levator palpebrae muscle, the medial canthal ligament, or the lacrimal duct should be considered for ophthalmologic referral. Evaluation for an associated injury of the globe is a must, particularly if periorbital fat is exposed or tarsal plate penetration is present.

Lacerations and Blunt Trauma of the External Ear

Although the ears are subject to trauma because of their exposed position, lacerations involving the ears are rather rare. To obtain the best results in caring for injuries involving the external ear (auricle or ear lobe), attention must be paid to certain anatomic and physiologic facts. The auricle contains a cartilaginous structure that provides the framework for the complex shape of the ear. The pericondrium covering the cartilage provides it with nutrients and oxygen. Separation of the cartilage from the pericondrium because of trauma may lead to necrosis of the cartilage, leaving the auricle deformed. The overlying skin, although thin and with no or little subcutaneous tissue, is well-vascularized. Skin flaps with small pedicles often survive and should not be hastily debrided.

Simple auricular lacerations can be repaired without difficulty. To avoid chondritis, approximation of the skin is important so that no cartilage is exposed. Occasionally, debridement of the cartilage is needed to obtain complete coaptation of the

wound; however, cartilage debridement should be kept to a minimum. It is imperative to avoid catching the auricular cartilage with the needle tip because the skin and the pericondrium are in close proximity to each other.

Complex auricular lacerations with significant skin damage and involvement of the auricular cartilage can be difficult to repair and may require consultation with a plastic surgeon. In general, when repairing auricular cartilage, a few 5-0 absorbable sutures should be used to approximate the edges. Landmarks of the auricle should be used for proper alignment. The pericondrium should be included in the sutures so that the suture material does not tear through the friable cartilage and also to ensure restoration of nutrient and oxygen supply. For the same reason, excessive tension should be avoided. Closure of the skin should follow as described previously. If the laceration involves the anterior and posterior aspects of the ear, closure of the posterior aspect first is recommended.

To avoid a deep scar line (notching) in repairing the ear lobe or the auricular rim, the skin edges should be everted at the time of closure because fibrotic tissue will eventually pull the scar line down, leading to notching.

Partial avulsion or *total amputation of the ear* is possible. Every effort should be made to reattach the amputated part because tissue survival and cosmetic outcome are usually favorable. Furthermore, blunt ear trauma can lead to a simple contusion or a significant *subperichondrial hematoma* that can comprise the auricular cartilage. Classically, a significant perichondrial hematoma is tense and appears as smooth ecchymotic swelling that disrupts the normal contour of the auricle. This injury is particularly common among wrestlers. Auricular hematoma should be promptly drained to avoid necrosis of the cartilage and deformed auricle or cauliflower ear.

Blunt trauma can drive or *bury an earring* or its backing into the ear lobe. This can also result from contact dermatitis reaction to the earring material with swelling. After local intradermal lidocaine, the earring part is usually easily navigated out of its port of entry. Occasionally, a small incision is needed to facilitate the extraction of the earring. If an incision is needed, it should be done on the posterior aspect of the ear lobe and should be left to heal by secondary intention. If the patient desires to maintain the ear lobe pierced, a sterile nonabsorbable suture should be ringed in the existing hole for a few days until the hole is epithelilized.

After repair of ear lacerations or evacuation of an auricular hematoma, a pressure dressing should be applied. Follow-up in 24 hours to evaluate vascular integrity to the area is recommended.

Lacerations of the Nose

Unlike blunt injuries, lacerations to the nose are unusual. When a laceration results from blunt trauma, careful evaluation of underlying nasal bones and examination for a nasal septal hematoma are essential. Other associated injuries, such as facial bone fractures or injuries to the orbit, should also be ruled out.

The skin overlying the nose is taut and stiff. Approximating the edges of simple, nongaping nasal wounds, mostly along the upper half of the nose, is usually straightforward. Wounds with any gaping, commonly in the lower part of the nose, can be difficult to coapt because of the nature of the skin in this location. The suture material can tear through the skin easily. Absorbable subcutaneous stitches are recommended before skin closure to relieve tension and prevent tearing through the wound edges. Skin closure should be with simple interrupted 6-0 nonabsorbable material. Early removal of the sutures is advised for the same reason.

Full-thickness nasal lacerations involving the alae nasi or entering the vestibule require layered closure. The procedure should be begun with the nasal mucosa, using absorbable material and finished with the skin, preferably using continuous subcuticular intradermal suture technique.

The nasal cartilage, when involved, rarely requires sutures. When alignment is difficult, a few fine sutures (vicryl or plain catgut) will help hold it in place. When the free rim of the nares is involved, precise alignment is imperative for good cosmetic outcome. For complex nasal lacerations, lacerations associated with fractures, or when there is tissue loss, consultation with an otolaryngologist or a plastic surgeon is recommended.

Lacerations of Lip

Lip lacerations are a particular concern because of the importance of the lip as a facial landmark. The lip is a vascular structure with multiple layers. The vermilion border, the junction of the dry oral mucosa and the facial skin, serves as an important landmark for proper repair when involved. The relative pallor of the vermilion border to the lip and to the skin easily identifies it. Therefore, the use of epinephrine with local anesthesia should be avoided, so the landmark is not abolished. When parted, the vermilion border should be precisely reapposed using a 6-0 suture. The buccal mucosal surface is then closed with 5-0 absorbable material, followed by the skin, using 6-0 nonabsorbable sutures. The parents should be warned that while the lip is still anesthetized, there is a chance that the child will bite the sutures off and that they should distract the child from doing so. Typically after the local anesthesia has worn off, the site is sore enough that the child will not attempt to manipulate the area.

In general, lip lacerations should be closed in layers, depending on the depth of the wound. In full-thickness lip lacerations, a three-layer repair is required. The physician should begin with the oral mucosa, using 5-0 absorbable material, followed by the orbicularis oris muscle layer to include the inner and outer fibrofatty layers, and finish with the skin, using 6-0 nonabsorbable interrupted sutures. Small wounds, less than 2 cm, on the inner aspect of the lip without communication to the skin surface need not be repaired. External lip wounds not communicating with the mucosal surface can be closed by either single- or double-layer closure, depending on the depth and degree of gaping of the wound. Absorbable sutures (5-0) for the subcutaneous layer and nonabsorbable (6-0) for closure of the skin can be used.

Extensive lip injuries with tissue loss or those caused by electric burns, especially those that involve the angle of the mouth, should be referred to a plastic surgeon. Associated injuries such as dental trauma, mandibular fractures, and

closed head injuries should be ruled out.

Lacerations of the Cheeks

When managing lacerations involving the cheeks, the physician must evaluate the integrity of the underlying structures. The parotid gland and duct, the facial nerve, and the labial artery are in close proximity of the surface of the skin and can be injured often as a result of an animal bite. If parotid gland or duct injury is identified, consultation with a surgeon is advised. Puncture wounds resulting from animal bites should be debrided and irrigated thoroughly. Some of these puncture wounds are better off left without closure to reduce infection rate, especially if the cosmetic outcome is not compromised. Otherwise, simple interrupted 6-0 nonabsorbable sutures can be used to close uncomplicated lacerations of the cheeks.

Lacerations of the Tongue

The tongue is a vascular and muscular organ. Tongue lacerations often hemorrhage excessively in the beginning, but the bleeding usually ceases quickly as the lingual muscle contracts. Lacerations of the tongue can pose a challenge to repair, not only because of their inaccessibility but also because of the controversy surrounding the indications for closure.

Most tongue lacerations can be left alone with good results. However, large lacerations involving the free edge may heal with a notch causing dysfunction of the tongue. Generally, this type of laceration should be repaired. Large flaps and lacerations that continue to bleed should also be repaired. Patients with tongue lacerations requiring repair should be assessed for potential airway problems, as well as for need of conscious sedation or even general anesthesia. Often, local or regional anesthesia is sufficient. The mouth should be retained open using a padded tongue depressor placed on the side between the upper and lower teeth or by using a Denhardt-Dingman side mouth gag. The tongue can be maintained in the protruded position by a gentle pull using a towel clip or by placing a suture through the tip. Interrupted 4-0 absorbable suture, with full-thickness bites to include the two mucosal surfaces and the lingual muscle between, will close the tongue wound and will provide hemostasis. Multiple knots and inverted sutures are recommended to prevent the untying of the sutures. Some authors suggest that only deep muscle closure is required because the mucosal surface heals rapidly. As in lip lacerations, children may chew off the stitches. Parents must be warned of this possibility and should attempt to distract the child at least until the local anesthesia is worn off.

Lacerations of the Buccal Mucosa

Small, isolated lacerations of the buccal mucosa, mostly from impaction of teeth following falls, require no suturing. Lacerations 2 to 3 cm in length or with flaps are best closed with simple interrupted absorbable material. Coated Vicryl (4-0) on a round needle is preferred because it is less irritating to the child and is easier to work with than chromic gut. Closure of the mucosal surface in through-and-through lip lacerations should be carried out before closure of the muscle and skin layers. After repair, a soft diet and avoidance of irritating foods should be advised, as well as vigilant mouth hygiene. Evaluation for associated injuries of the teeth or the alveolar margin is imperative.

Fingertip Injuries

Fingertip Avulsions

Fingertip injuries are rather common in children. In the young child, most of these injuries are blunt and are secondary to entrapment of the finger in closing doors. Most of these injuries are contused lacerations or partial avulsions. Complete amputation of the fingertips is not as common. Sharp injuries are more common in the older child and less likely to be associated with fractures. Fingertip injuries should be evaluated clinically for an associated nailbed injury and radiographically for possible fractures of the phalanges. In general, this type of injury is manageable by the emergency physician, especially in the preadolescent child because tissue regeneration is remarkable and management is mostly conservative.

The management of amputations of fingertips (distal to the distal interphalangeal joint) can be approached based on the absence or the presence of bone exposure. If no or minimal bone is exposed, conservative management is advised. The wound should be cleansed, dressed in nonadherent gauze, and splinted for protection. Frequent dressing changes and appropriate follow-up should be planned. Antibiotic coverage is recommended when a significant amount of bone is exposed. Consultation with a surgeon should be considered. Shortening of the distal phalanx and covering the tip with volar skin flap is usually the treatment of choice. However, some hand surgeons increasingly advocate for a variety of skin-grafting procedures to avoid permanent shortening and deformity. Amputations proximal to the distal interphalangeal joint should be considered for microscopic reimplantation by a surgeon.

Nailbed Injuries

Trauma to the distal fingers is often associated with nail and nailbed (matrix) injuries. Nail avulsion can be partial or complete and may or may not be associated with nailbed laceration. An underlying fracture of the distal phalanx can occur. Injury to the fingertip is often associated with subungual hematoma. In evaluating these injuries, the emergency physician should determine the need to explore the nailbed for a laceration. Unrepaired nailbed lacerations may permanently disfigure the growth of the new nail from the cicatrix nailbed. If the nail is partially avulsed but is firmly attached to its bed, exploring the nailbed is difficult and is probably not warranted. Good outcome is expected because the nail holds the underlying lacerated nailbed tissues in place.

If a subungual hematoma exists, it should be drained (see the following). When the nail is completely avulsed or is attached loosely, the nail should be removed and the nailbed assessed for laceration. If the nailbed is lacerated, it should be repaired using 6-0 absorbable material. After cleansing and trimming its soft proximal portion, the nail should be

replaced between the nailbed and the nail fold (eponychium), then anchored in place with a few stitches. This will splint the nail fold away from the nailbed, which will prevent the obliteration of the space between the nailbed and the nail fold. By preserving this space, the new nail is allowed to grow undisturbed. The preferred method of local anesthesia for nailbed repair is digital block, and the use of a finger tourniquet during the repair allows a bloodless field. Application of a finger splint after repair, especially if there is an associated fracture, is recommended.

Subungual Hematoma

Subungual hematoma is a collection of blood in the interface of the nail and the nailbed. It is commonly seen with blunt fingertip injuries. The usual presentation is throbbing pain and discoloration of the nail. Subungual hematoma may be associated with nailbed injury or fracture of the distal phalanx. The presence of a fracture may complicate the decision to drain a subungual hematoma because the closed fracture then may be transformed into an open fracture.

Usually, drainage of the hematoma provides relief of the symptoms. Generally, no local anesthesia is required for a simple trephination by cauterization of the nail. After drainage, care for simple subungual hematoma includes elevation of the hand and warm soaks for a few days. The family should always be told about the possibility of nail deformity in the future. When the injury is more involved, digital block is advised. If the hematoma is large and extends to the tip of the nail, separation of the nail from the nailbed using either a sharp or blunt method will allow drainage. In the presence of a distal phalangeal fracture, the physician has to be concerned about transforming a closed fracture to an open one by communicating the subungual, and hence the fracture hematoma, to the exterior surface of the nail. If there is a possibility of an underlying fracture, antibiotic coverage and close follow-up should be sought.

Suggested Readings

Baker MD, Selbst SM, Lanuti M. Lacerations in urban children. Am J Dis Child 1990;144:87–92.

Berk WA, Welch RD, Bock BF. Controversial issues in clinical management of the simple wound. Ann Emerg Med 1992;21:72–80.

Bruns TB, Simon HK, McLario DJ, et al. Laceration repair using tissue adhesive in a children's emergency department. Pediatrics 1996;98: 673–675.

Callahan JM, Baker MD. General wound management. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:1125–1139.

Edlich RD. Soft tissue injuries to the face. In: Tintinali JE, Krome RL, Ruiz E, eds. Emergency Medicine: A Comprehensive Study Guide. 4th ed. New York: McGraw-Hill, 1992:302–311.

Edlich RF, Rodeheaver GT, Morgan RF, et al. Principles of emergency wound management. Ann Emerg Med 1988;17:1284–1302.

Fleisher G, Ludwig S. Textbook of Pediatric Emergency Medicine. 3rd ed. Baltimore: Williams & Wilkins, 1993.

Grisham J, Perro M. Laceration repair. In: Dieckmann RA, Fiser DH, Selbst SM, eds. Illustrated Textbook of Pediatric Emergency & Critical Care Procedures, St Louis: Mosby, 1997:669–679.

Grisham J. Principles of wound healing. In: Dieckmann RA, Fiser DH, Selbst SM, eds. Illustrated Textbook of Pediatric Emergency & Critical Care Procedures. St Louis: Mosby, 1997:665–668.

Hanke BK. Advances in wound management in the emergency department. Res Staff Phys 1993;39:23–27.

Lammers RL. Principles of wound management. In: Roberts JR, Hedges JR, eds. Clinical Procedures in Emergency Medicine. 2nd ed. Philadelphia: WB Saunders, 1991:515–565.

Leibelt EL. Reducing pain during procedures. Curr Opin Pediatr 1996; 8:436–441.

McGrath PJ. Annotation: aspects of pain in children and adolescents. J Child Psychol Psychiat 1995;36:717–730.

McGregor IA. Fundamental Techniques of Plastic Surgery. 8th ed. New York: Churchill Livingstone, 1989.

McNamara R, Loiselle J. Laceration repair. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:1141–1168.

Platt SL, Foltin GL. Ingrown toenail repair. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997;1217–1222.

Roberts J. Evaluation and treatment of subungual hematoma. Emerg Med News 1993;15:2.

Roberts J, Hedges J. Clinical Procedures in Emergency Medicine. 2nd ed. Philadelphia: WB Saunders, 1991.

Rothman DL. Pediatric orofacial injuries. J Calif Dent Assoc 1996;Mar(3):37–42.

Selbst SM, Henretig FM. The treatment of pain in the pediatric emergency department. Ped Clin North Am 1989;36:965–978.

Simon HK. How good are tissue adhesives in repairing lacerations? Contemp Pediatr 1997;14:90–96.

Simon HK, McLario DJ, Bruns TB, et al. Long-term appearance of lacerations repaired using a tissue adhesive. Pediatrics 1997;99:193–195.

Singer AJ, Hollander JE, Quinn JV. Evaluation and management of traumatic lacerations. N Engl J Med 1997;337:1142–1148.

Singer AJ, Hollander JE, Valentine SM, et al. Prospective, randomized, controlled trial of tissue adhesive (2-Octylcyanoacrylate) vs standard wound closure techniques for laceration repair. Acad Emerg Med 1998;5:94–99.

Smith RJ. Emergency care of the injured hand. In: Wilkins ED, ed. Textbook in Emergency Medicine Scientific Foundations and Current Practices. 5th ed. Baltimore: Williams & Wilkins, 1989:810–812.

Templeton JM. Minor trauma. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine, 3rd ed. Baltimore: Williams & Wilkins,

1993:1288–1297.

Zook E. Discussion of “management of acute nail bed avulsions.” Hand Clin 1990;6:57.

CHAPTER 117

Minor Lesions and Injuries

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A variety of minor lesions in children may prompt an emergency department (ED) visit. Most are the result of acute injury or infection (e.g., hair tourniquet, felon). Some formerly quiescent abnormalities (e.g., thyroglossal duct cyst, pyogenic granuloma) become clinically apparent after rapid enlargement secondary to infection or direct trauma. Alternatively, asymptomatic minor lesions (e.g., lipoma, pilomatrixoma) may be noted during the evaluation of an unrelated complaint. Regardless of the presentation, a systematic approach is necessary for proper diagnosis and subsequent management. Although most “lumps and bumps” in children have a benign cause, the examiner should bear in mind the possibilities of associated systemic illness and future complications.

HAND LESIONS

Eponychia and Paronychia

Infections and/or minor trauma are the major sources of hand lesions. By far, the most common infections of the hand involve the eponychium (cuticle). Most of these arise following minor trauma—a traumatized hang nail, finger sucking, or nail biting—that causes a breakdown in the epidermal barrier. In its initial stage, the infection consists of a superficial cellulitis that remains localized to the cuticle and is termed an eponychia. With progression, pus collects in a single thin-walled pocket under the cuticle, forming a paronychia (Fig. 117.1). This may progress, extending under the skin at the base of the nail, and along the nail fold. Less commonly, the pus burrows beneath the proximal nail, forming an *onychia* or subungual abscess. Causative organisms include *Staphylococcus aureus, Streptococcus pyogenes,* and anaerobic species. Treatment of a simple eponychia involves frequent warm soaks and attention to local hygiene. Topical antibacterial ointments may hasten resolution. Incision and drainage is the treatment of choice for a paronychia (see Section VII, Procedures). If an onychia has formed, removal of the proximal portion of nail overlying the abscess is essential to ensure adequate drainage and prevent destruction of the germinal matrix. Occasionally, an onychia may form under the anterolateral aspect of a nail. Here, elevation and excision of the overlying portion of nail suffices. The role of oral antibiotics after incision and drainage has not been clearly established. Most physicians choose a short course of a b-lactamase stable antibiotic (e.g., dicloxacillin, cephalexin). Oral antimicrobial therapy is definitely indicated for patients with associated lymphangitis.

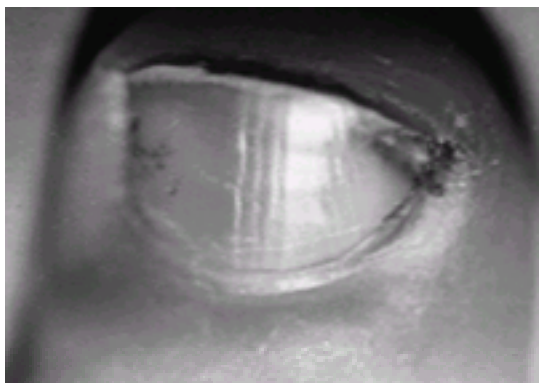

FIGURE 117.1. Small paronychia located along nail fold, noted after minor trauma.

An *herpetic whitlow* involving a finger is sometimes mistaken for a paronychia and is the major differential diagnostic consideration. Clinically, this lesion is characterized by the appearance of multiple, painful, thick-walled vesicles on erythematous bases. Over the ensuing few days, vesicles begin to coalesce and their contents become pustular. A Gram stain of pustular fluid is negative for bacteria. Tczank prep of scrapings from the base of a lesion reveals multinucleated giant cells. Subsequently, ulceration and crusting occur. The process initially results from inoculation of herpes simplex virus into a small break in the skin. The source may be a parent with herpes labialis, or a child with herpetic gingivostomatitis or herpes labialis may inoculate his or her own finger. If the infection is primary, fever and regional adenopathy are seen. With recurrences, these findings are usually absent.

Felon

A *felon* consists of a deep infection of the distal pulp space of a fingertip. Felons are caused by introduction of bacteria into the pulp space usually by punctures (which may be trivial) or splinters. Causative organisms are similar to those found in eponychial infections. A felon typically presents as an exquisitely tender and throbbing fingertip that is swollen, tense, warm, and erythematous. However, its evolution usually is relatively slow, beginning with mild pain and minimal swelling that progresses over a few days. This process is in part caused by the anatomy of the pulp, which consists of multiple closed spaces formed by fibrous septae that connect the volar skin to the periosteum of the distal phalanx. With progression of infection, pressure buildup within these small compartments may cause local ischemia. In some cases, organisms may spread to invade the phalanx, resulting in osteomyelitis. In others, the process may point outward to the center of the touch pad, where the septae are least dense, producing an obvious area of fluctuation. Because the deep septal attachments are distal to the distal interphalangeal (DIP) joint and flexor tendon sheath, there is less risk of spread to these structures.

Treatment consists of incision, blunt dissection, and drainage. A longitudinal incision over the area of maximal tension or fluctulance is the procedure of choice. Care should be taken to extend the incision past the DIP joint to prevent formation of a flexion contracture (see Section VII, Procedures). Older techniques, such as the extended “fishmouth” incision, incision through the touch pad, and the through-and-through incision, should be abandoned to avoid unnecessary complications (e.g., unstable touch pad, persistently painful scar, permanent anesthesia, skin sloughing). After drainage, a course of oral antibiotics is indicated. Close follow-up is essential to assess response to therapy and identify complications, such as septic arthritis and suppurative tenosynovitis. Patients presenting with fever, lymphangitis, or evidence of osteomyelitis should be referred to the hand service for admission, parenteral antibiotics, and definitive care.

Subungual Hematoma

A *subungual hematoma* is a collection of blood located under a nail that arises after trauma to the nailbed. The typical mechanism is a crush injury. Because this mechanism is a common cause of phalangeal fractures, radiographs are advisable. The patient experiences throbbing pain that worsens with increasing pressure as more blood collects. The underlying nailbed injury may be trivial or may involve a laceration that requires repair. If the subungual hematoma involves more than 50% of a nail surface, is associated with a distal phalanx fracture, or the nail or its margins are disrupted, the presence of a significant nailbed injury should be suspected. Nail trephination provides drainage with attendant relief of pressure and pain and suffices for uncomplicated subungual hematomas. This procedure also reduces risk of secondary infection. The trephined opening should be large enough (larger than 3 to 4 mm) to allow for ongoing drainage without risk of closure by new clot (see Section VII, Procedures). When the hematoma is large, the nail or its margins are disrupted, and/or a phalangeal fracture is present, the nail should be removed and the nailbed repaired. Antimicrobial prophylaxis for these injuries remains a source of controversy but is prescribed by most practitioners for patients with underlying fractures and those with severe soft-tissue injuries.

Subungual Foreign Body

Foreign bodies such as a wood splinter or metallic shaving become embedded under the nail and may be the source of pain and/or infection. When the foreign body is only partially embedded, the nail can be trimmed close to the nailbed, and the object's projecting end grasped with splinter forceps and gently extracted. If a portion remains, or the foreign body is deeply embedded from the outset, a digital block should be performed. Then the part of the nail overlying the object can be shaved down with a scalpel until the foreign body is exposed. Alternatively, the nail can be lifted, and the object removed (see Section VII, Procedures). After splinter removal, the finger should be soaked in warm, soapy water and an antibiotic ointment and protective dressing applied. Soaks should be repeated three times daily at home for the ensuing 3 to 5 days. In the unusual case of a child with multiple subungual splinters or fragments, it is best to remove the

nail, clean out the foreign material, irrigate thoroughly, and then replace the nail (after trephining it to allow drainage).

Hair Tourniquet

A *hair tourniquet* injury is an entity unique to pediatrics. It involves strangulation of a digit (or occasionally the penis) by a hair or fine thread. It is seen most commonly in young infants and can be the cause of unexplained irritability or crying. The mechanism involves entwinement of the hair around an infant's digit. This may occur during a bath, during subsequent toweling, or as a result of wiggling of the toes in a sock, bootie, or mitten that inadvertently has a hair or loose thread in it. A hair shed from a parent during diapering is the probable source of penile tourniquets. As the hair or thread becomes more tightly entwined, it produces a tourniquet effect, impairing blood flow with resultant ischemic pain and distal swelling. When noted early, the hair is often visible in a crease just proximal to the swollen area. If seen later, the hair may have cut through the skin, making it difficult to visualize (Fig. 117.2). In rare cases, frank ischemic necrosis of the distal digit may be seen on presentation. Removal requires a fine-tipped forceps and the aid of a thin loupe or probe that is inserted proximally under the constricting hair. Usually the hair can be unwound from the digit intact or cut with scissors. When the hair is deeply embedded or there is any question of a remaining constricting band, a nerve block should be performed and a perpendicular incision made over the hair. To avoid damage to neurovascular structures, such an incision should be made on the lateral aspect of a finger or toe at 3 or 9 o'clock or at 4 or 8 o'clock along the penile shaft. When the entire hair cannot be removed with certainty, plastic surgical consultation is indicated.

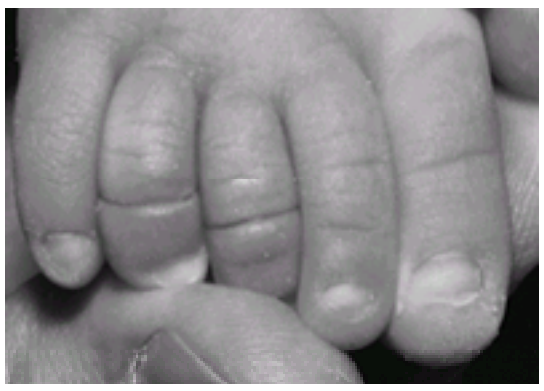

FIGURE 117.2. Hair tourniquets of third and fourth toes.

Ganglion

A *ganglion* is a cystic outgrowth or protrusion of the synovial lining of a tendon sheath or joint capsule. Common locations of such lesions include the dorsal or volar surface of the wrist (usually on the radial side), the dorsum of the foot, or near the malleolus of an ankle (Fig. 117.3). Occasionally, a flexor tendon sheath ganglion may present on the palmar surface of the hand at the base of a digit. The cause is thought to involve prior trauma that causes partial disruption of the synovium and subsequent herniation of synovial tissue. The cysts are soft, slightly fluctuant, and transilluminate. Most are painless or only mildly uncomfortable. However, those on the foot or ankle may cause pain when shoes are worn. Elective surgical excision with obliteration of the base is indicated only if function is impaired or the lesion is of cosmetic significance. Even then, up to 20% recur. The old folk remedy of striking the cyst with a large book or against a hard surface should be strongly discouraged because the cystic fluid may be dispersed through the surrounding soft tissue, inciting diffuse scar formation.

FIGURE 117.3. Ganglion cyst of the tendon sheath of flexor carpi radialis.

FACE AND SCALP LESIONS

Epidermal Inclusion Cyst

Among the most common postpubescent skin lesions is the *epidermal inclusion cyst* (EIC). These have also been termed epithelial, sebaceous, and pilar cysts. Most result from occlusion of pilosebaceous follicles, although some stem from inoculation of epidermal cells into the dermis via needle stick or other trauma. A few may arise from epidermal cells that became trapped along embryonic lines of closure. Lesions consist of firm, slow-growing, 1- to 3-cm, round nodules. Most are found about the scalp and face, although they also may be located on the trunk, neck, and scrotum. Most are solitary.

Histologically, these dermal and subcutaneous nodules consist of epidermally lined keratin-filled cysts. Presentation is that of a slow-growing painless lump that may provoke concerns of malignancy. At times, these cysts become acutely infected, and the patient complains of pain, erythema, and sudden increase in size. Infected cysts are incised and drained and treated with oral antibiotics before elective excision. Noninflamed cysts can be referred for elective excision that must include the entire sac to prevent recurrence.

When a patient presents with multiple large EICs, Gardner's syndrome should be suspected. This autosomal-dominant disorder is characterized by multiple EICs, intestinal polyposis, desmoid tumors, and osseous lesions. Early diagnosis is especially important because of a 50% risk of malignant transformation of the intestinal polyps.

Dermoid Cyst

Dermoid cysts are congenital, subcutaneous nodules derived from ectoderm. They, too, are lined with epithelium, but unlike EICs, they may contain multiple adnexal structures such as hair, glands, teeth, bone, and neural tissue, as well as keratin. The cysts usually present as solitary, round, firm nodules. They have a rubbery or doughy consistency on palpation, a smooth surface and normal overlying skin. Lesions tend to grow slowly, and malignant transformation is rare. Whereas some dermoids may be mobile, many are fixed to overlying skin or underlying periosteum. Some have other deeper attachments. Because these cysts form along areas of embryonic fusion, common sites include the nasal bridge, midline neck, or scalp, the lateral brow (Fig. 117.4), anterior margin of the sternocleidomastoid, and midline scrotum or sacrum. Midline dermoids tend to have especially deep attachments that may extend intracranially or intraspinally along with an accompanying sinus tract. An external ostium may or may not be visible. Because the sinus tract can serve as a conduit for spread of secondary infection, all midline lesions should have appropriate imaging (usually magnetic resonance imaging [MRI]) followed by elective excision.

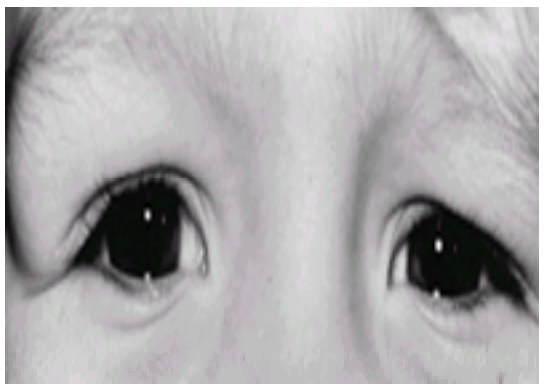

FIGURE 117.4. Dermoid cyst of right lateral brow. (Reprinted with permission from Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997.)

Nasal Bridge Lesions

Midline nasal masses in infants and children may be acquired (e.g., EIC) or congenital, the latter stemming from improper embryologic development (e.g., dermoid cyst, encephalocele, and glioma).

Dermoids are the most common embryologically derived midline nasal lesions (see previous discussion). Clinically, a firm, round, subcutaneous mass is seen in the midline over the dorsum of the nose. Some have an overlying dimple, which may have an extruding hair (Fig. 117.5). Its attachment may extend only to the nasal septum or may go deeper through the cribriform plate into the calvarium. Because of their proximity to the nasopharynx, these dermoids are particularly prone to secondary infection and fistula formation. Hence, prompt excision is indicated after careful MRI or computed tomography (CT).

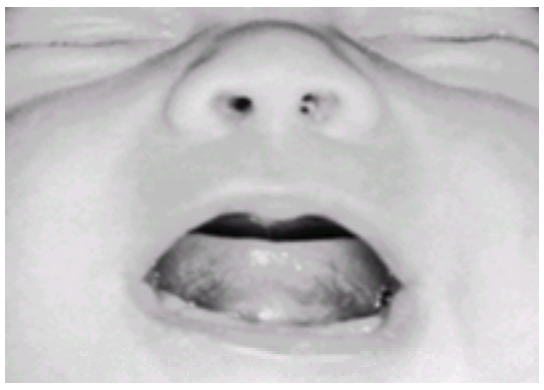

FIGURE 117.5. Midline nasal dermoid with overlying dimple. (Reprinted with permission from Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997.)

Gliomas contain neuroectodermally derived elements. A *glioma* is a benign growth composed of ectopic neural tissue. The lesion usually consists of a firm, gray or red-gray nodule, ranging in size from 1 to 5 cm and can be mistaken for a hemangioma. Most are extranasal (60%), occurring on the bridge of the nose. The remainder are either solely intranasal

masses (30%) or have both intranasal and extranasal elements (10%). By definition, they do not have intracranial communication. They are composed of neural and fibrous tissue, covered by nasal mucosa. To prevent possible distortion of surrounding bone and cartilage, surgical excision is the treatment of choice.

Encephaloceles consist of neural tissue that has herniated through a congenital defect in the midline of the calvarium and thus, always have an intracranial communication. Lesions appear as soft, at times pulsatile, compressible masses that enlarge with crying or straining. Compression of the jugular veins (Furstenberg test) may also cause the mass to expand in size. Some infants with nasal encephaloceles are born with overt craniofacial deformities and a rounded swelling at the base of the nose, whereas in others the mass is confined to the nasopharynx, and external facial features are normal. The latter may present with signs of persistent nasal obstruction. In these patients, a grapelike mass is found on nasopharyngoscopy. MRI is the modality of choice for differentiating encephaloceles from other midline nasal masses and for determining their size and extent. Neurosurgical evaluation and management is indicated for all encephaloceles.

Preauricular Lesions

Preauricular lesions, located just anterior to the tragus, may be the result of imperfect fusion of the first two branchial arches *(sinus tract*, *pit)* or may consist of first arch remnants *(cutaneous tag).* They may be unilateral or bilateral, single or multiple. Usually, they are seen as isolated minor anomalies, but on occasion, they can be found in association with other developmental anomalies involving the first branchial arch or in infants with chromosomal disorders. Most lesions are evident shortly after birth. Some individuals simply have a surface pit or dimple, whereas in others, the overlying dimple represents the entrance to a sinus tract or blind pouch with a small cyst at its base (Fig. 117.6A). The latter may contain hair and other epidermal elements. Sinuses are prone to infection and abscess formation, whereupon the child presents with sudden enlargement of a painful preauricular mass and overlying erythema (Fig. 117.6B). When this occurs, the patient should be treated with appropriate antimicrobial therapy before elective excision of the cyst and fistula tract. Cutaneous tags, also called accessory auricles, are flesh-colored pedunculated lesions that may or may not have a cartilaginous component (Fig. 117.7). Some with narrow bases may simply be tied off with silk sutures. Those with wider bases and those containing cartilage can be referred for elective excision for cosmetic reasons.

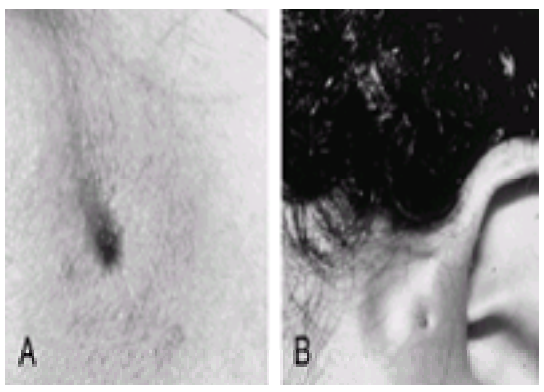

FIGURE 117.6. A. Close-up of preauricular surface pit. **B.** Infected preauricular sinus with overlying pit. (Reprinted with permission from Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997.)

FIGURE 117.7. Preauricular skin tags. (Reprinted with permission from Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997.)

Neck Lesions

Neck lesions in children may be of congenital origin or may be acquired as the result of an inflammatory process (Fig. 117.8). Although malignancy is a much rarer cause of neck masses in children, it must always be considered in the differential diagnosis. Neck masses or lesions are most conveniently divided into those occurring in the midline and those located in the lateral aspects of the neck.

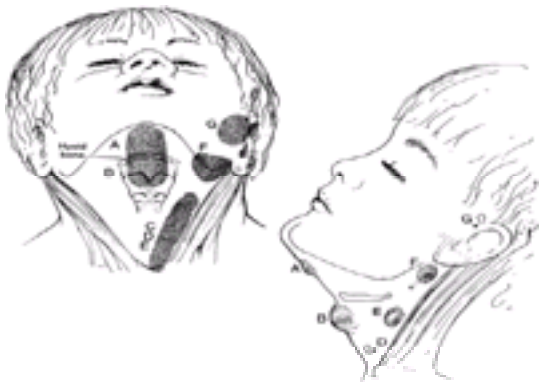

FIGURE 117.8. Head and neck congenital lesions seen in children in frontal and lateral views. The shaded areas denote the distribution in which a given lesion may be found. *A,* Dermoid cyst; *B,* thyroglossal duct cyst; *C,* second branchial cleft appendage; *D,* second branchial cleft sinus; *E,* second branchial cleft cyst; *F,* first branchial pouch defect; *G,* preauricular sinus or appendage.

Midline Neck Lesions

Submental lymphadenitis or *lymphadenopathy* occurs in the midline just beneath the chin. Nodal enlargement stems from drainage of primary infection of the lower lip, buccal floor, or anterior tongue.

Dermoid cysts (see Face and Scalp Lesions) can occur throughout the midline of the neck but usually are found above the area of the hyoid. They also may be found more laterally along the anterior border of the sternocleidomastoid.

Thyroglossal duct cysts are among the more common midline neck masses in children. Approximately 40% present before 10 years of age. They are comprised of an ectodermal ductal remnant that fails to regress after fetal descent of the thyroid gland. They may occur anywhere along the path of descent of the thyroid, from the foramen cecum at the base of the tongue to the sternal notch, although most are found near the level of the hyoid bone. Presentation is usually that of a painless, smooth, mobile, cystic mass that is located in the midline or just slightly off-center (Fig. 117.9). Because of its intimate association with the hyoid, the mass moves with protrusion of the tongue or swallowing. On occasion, an overlying pore is present. Some cysts go unnoticed until infection occurs, causing acute swelling, pain, and erythema of the overlying skin. Patients with asymptomatic thyroglossal duct cysts should be referred for elective surgical excision. If the thyroglossal duct cyst is infected on presentation, excision is deferred until appropriate antimicrobial therapy is completed and inflammation has subsided. If incision and drainage are required during treatment, the patient should be referred to a surgeon comfortable with thyroid anatomy. Elective excision involves removal of the cyst, the entire duct to the level of the foramen cecum, and the midportion of the hyoid bone. On rare occasions, ectopic thyroid tissue in a thyroglossal duct cyst is the patient's only functioning thyroid. Therefore, most authorities recommend ultrasound or radioisotope scanning to confirm the presence of a normal thyroid gland before surgery.

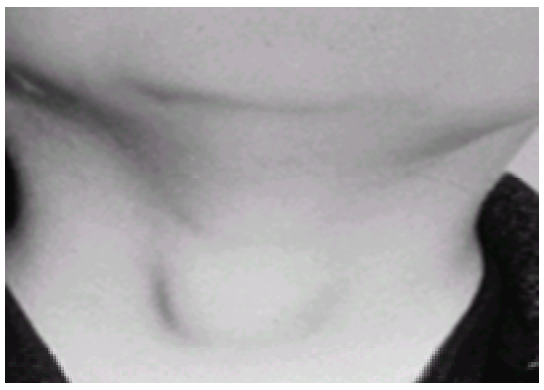

FIGURE 117.9. Thyroglossal duct cyst, which elevated with tongue protrusion. (Reprinted with permission from Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997.)

Diffuse enlargement of the thyroid gland, or *goiter,* may be the result of infiltration, inflammation, or overstimulation of the gland. By far, the most common cause of pediatric thyroid enlargement is chronic *lymphocytic thyroiditis* (also called *Hashimoto's thyroiditis* or *autoimmune thyroiditis*). This disorder is characterized by a defect in cell-mediated immunity that results in lymphocytic infiltration of the thyroid gland. Females are affected predominantly, and peak occurrence is during adolescence. Usual presentation is one of a slow growing, painless midline neck mass. Occasionally, a patient may complain of sore throat. Examination reveals a firm, nontender, diffusely enlarged gland in most affected children, but approximately one-third will have some lobular or nodular enlargement. Evaluation includes assessment of thyroid function and the detection of thyroid autoantibodies in the serum. Most patients with lymphocytic thyroiditis are euthyroid. When thyroid dysfunction is present, it usually takes the form of hypothyroidism. Any degree of nodularity of the gland warrants further investigation to rule out malignancy.

Inflammation of the thyroid gland secondary to infection, *acute suppurative thyroiditis*, is a rare cause of diffuse thyroid enlargement. Presentation usually follows an upper respiratory infection and is characterized by abrupt appearance of a painful, tender, swollen mass in the region of the thyroid. Systemic toxicity in the form of fever and chills and severe dysphagia are often present. Flexion of the neck may alleviate pain, whereas extension worsens it. Etiologic agents

include *S. aureus, Streptococcus hemolyticus, Streptococcus pneumoniae,* and occasionally, anaerobic species. Appropriate parenteral antimicrobial therapy is usually sufficient to eradicate the infection. Abscess formation necessitates incision and drainage by a surgeon comfortable with thyroid anatomy.

Acute immune stimulation of the thyroid gland may also produce diffuse thyroid enlargement. In *Graves' disease,* autoantibody attachment to the thyroid-stimulating hormone (TSH) receptor stimulates an increase in thyroid hormone synthesis and release. This produces glandular enlargement with a soft consistency and symptoms of juvenile thyrotoxicosis, including tachycardia, nervousness, tremor, weight loss, increased appetite, and heat intolerance. An elevated T_4 in the context of a low TSH level and presence of TSH receptor antibodies confirms the diagnosis. A pediatric endocrinologist guides therapy in the form of antithyroid drugs.

Solitary nodular thyroid masses deserve careful attention. Although most are secondary to chronic lymphocytic thyroiditis or consist of a benign adenoma, the incidence of malignant neoplasms is actually higher in children with thyroid nodules than in adults. Hence, every thyroid nodule found in a child merits a complete evaluation that may include ultrasound imaging, radioisotope scanning, and biopsy.

Lateral Neck Lesions

Enlarged cervical lymph nodes constitute the most common lateral neck masses in children. Knowledge of the anatomy of the cervical lymphatics is of fundamental importance to understanding processes that cause enlargement of cervical lymph nodes. This section focuses mainly on local processes that cause nodal enlargement, but it is important to note that many systemic infections and inflammatory disorders can cause diffuse adenopathy that includes the cervical chain. Therefore, any child with a neck mass deserves a complete examination to look for the presence of generalized adenopathy and other signs of systemic disease.

Reactive cervical adenopathy refers to mild enlargement of cervical lymph nodes that accompanies a viral or bacterial upper respiratory infection. Involved nodes are typically located in the upper portion of the cervical chain. They are usually discrete, firm, mobile, and less than 2 cm in diameter. They may be mildly tender but have no overlying erythema, edema, or warmth. Regression within 1 to 2 weeks of resolution of the primary infection is the rule, although occasionally mild enlargement of the node may persist, if fibrosis has occurred.

Local infection of a lymph node itself is termed *acute suppurative lymphadenitis.* The involved node is solitary, typically 2 to 3 cm or larger in diameter, and extremely tender. As the infection proceeds, overlying swelling, erythema, and warmth develop and become more pronounced (Fig. 117.10). Initially the node is firm, but later it may become fluctuant. Acute suppurative lymphadenitis is most often caused by streptococcal or staphylococcal organisms. Because of the high incidence of b-lactamase production by *S. aureus*, b-lactamase stable antibiotics (dicloxacillin, cephalexin) are the treatment of choice. Most patients respond to oral antimicrobial therapy and application of warm compresses. However, if fluctuance develops, incision and drainage are recommended. Other potential causative organisms of acute, subacute, or chronic lymphadenitis include anaerobic bacteria, *Pasteurella multocida* (following animal bites), *Haemophilus influenzae, Streptococcus agalactiae, Francisella tularensis, Brucella* species, *Bartonella henselae* (cat-scratch disease), mycobacteria, and actinomycoses. Kawasaki disease also may present with an acutely enlarged cervical node and should be considered when other clinical criteria are present (fever longer than 5 days, rash, conjunctivitis, extremity and oral changes, and hyperirritability).

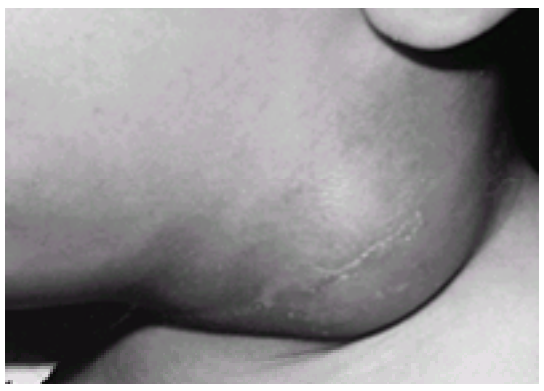

FIGURE 117.10. Suppurative lymphadenitis of an anterior cervical lymph node.

Salivary gland infections, *sialadenitis,* and *parotitis*, may cause lateral neck or submental swelling. When the parotid gland is involved, firm indurated swelling is found extending in an arc from the preauricular area down under the ear and behind it. The degree of swelling is often sufficient to blunt the angle of the jaw, and the mass is usually mildly tender. Patients complain of mild pain in the region of the pinna, which increases with eating. Most salivary gland infections affect the parotid gland, with involvement of the sublingual and submandibular glands being much less common. Viral agents (e.g., mumps virus, parainfluenza types 1 and 3, influenza A, Coxsackie A, and rarely, human immunodeficiency virus) cause most of these infections. Less commonly, parotitis is of bacterial origin. In these cases, patients present with rapid gland enlargement and severe pain, and they often have high fever and signs of systemic toxicity. On examination, overlying erythema and exquisite tenderness are present, and purulent material can often be expressed from Stensen's duct by massaging the gland. Symptomatic treatment of sialadenitis includes close attention to hydration and avoidance of foods that require excessive chewing or induce rapid salivary flow (e.g., citrus fruits, sour foods). If bacterial sialadenitis or parotitis is suspected, b-lactamase stable parenteral antibiotics should be administered. Otolaryngologic consultation should be obtained if surgical drainage is needed because of the proximity of the facial nerve. Much less commonly, parotid gland swelling is of noninfectious origin. Causes include occlusion of Stensen's duct by a calculus and

traumatic insufflation of the gland with forceful blowing.

Cystic hygromas (lymphangiomas) represent malformations of the lymphatic system. They consist of dilated lymphatic channels and may be multiocular or unilocular. They occur most often in the posterior triangle of the neck (Fig. 117.11) but may be found in the axillae, groin, popliteal fossae, or on the chest or abdominal wall (Fig. 117.12). When found in the neck, extension of the mass into the anterior triangle, sublingual space, retropharyngeal space, or mediastinum is possible. Such infiltration can result in airway compromise and/or compression of vascular and neural structures. Most cystic hygromas are present at birth or become apparent shortly thereafter. Patients usually present with a slow-growing, painless neck mass that is soft and compressible, although some are brought for care because of sudden enlargement caused by secondary infection or hemorrhage within the lesion. Anatomic delineation of the mass is best done using MRI or ultrasonography. When lesions are located in the neck, the potential risk to the airway and neurovascular structures, coupled with the possibilities of hemorrhage or lymphangitis, dictates the need for early intervention. Therapy has traditionally consisted of meticulous surgical excision; however, recurrences have been common. Recently, use of a new sclerotherapeutic agent has produced excellent results. OK-432 is a mixture of penicillin G and a low virulent Su strain of type II, group A streptococcus that has lost its ability to produce streptolysin-S. When injected into the lesion, it destroys the endothelial lining of the cyst, and thus accelerates drainage and shrinkage of the mass.

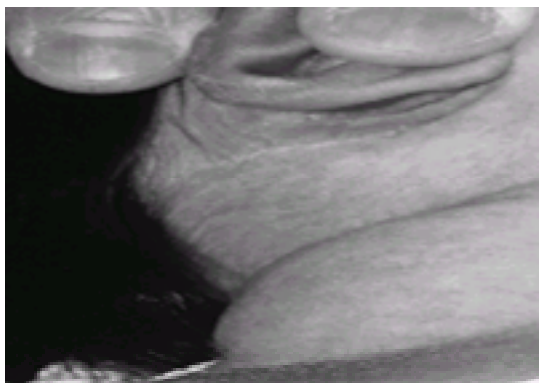

FIGURE 117.11. Lateral neck cystic hygroma (lymphangioma) in an infant.

FIGURE 117.12. Lateral abdominal wall cystic hygroma (lymphangioma).

Branchial cleft anomalies consist of a group of congenital malformations, including subcutaneous cysts, sinus tracts, and cartilaginous remnants. They are caused by persistence of structures derived from the embryonic branchial arches. Of these anomalies, 90% arise from the second branchial arch and are found along the anterior border of the sternocleidomastoid muscle. Sinus tracts of second branchial arch remnants may end in an internal ostium located near the tonsillar fossa. Less commonly, first branchial arch anomalies may be noted as masses or sinus tracts near the mandibular ramus. Some first branchial arch remnants end in an internal ostium located in the external auditory canal. Branchial cleft anomalies may be noted shortly after birth either as a firm, mobile mass with or without an overlying pore or simply as an external ostium or pore without an underlying mass (Fig. 117.13). More commonly, branchial cleft cysts are detected later in childhood when they may present as an asymptomatic mass or with acute painful enlargement as a result of secondary infection. All branchial cleft anomalies should be referred for surgical excision for cosmetic purposes and to avoid potential morbidity, which includes infection and the development of carcinoma in situ. When patients present with infection, excision must be deferred until antimicrobial therapy and incision and drainage (if needed) have quelled all signs of inflammation.

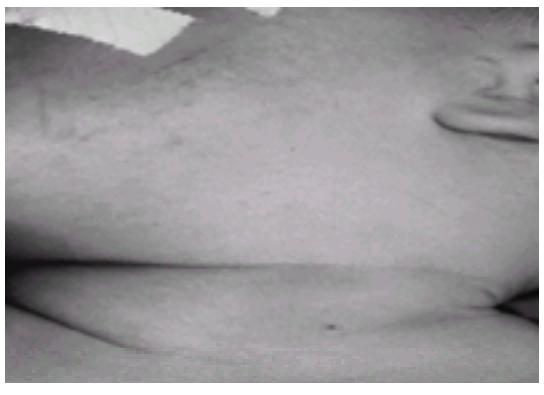

FIGURE 117.13. Second branchial cleft pit that had an underlying sinus tract.

The combination of torticollis and a lateral neck mass in early infancy is highly suggestive of a *sternocleidomastoid tumor.* Clinically, a nontender, firm, ovoid mass is found along the middle third of the sternocleidomastoid muscle. The mass represents local muscle hemorrhage that subsequently undergoes fibrosis. It is thought to be the result of traumatic extraction of the head during delivery, or of fibrous dysplasia secondary to intrauterine positioning. Some are noted at birth, whereas others become apparent within the ensuing few weeks. The head is bent toward the affected side, and limitation of bending to the opposite side and rotation toward the involved side are noted. Initial treatment consists of passive stretching exercises and positioning of the infant so he or she has to turn from the affected side to see others. If this fails, surgical release of the contracture is indicated to prevent secondary facial deformity with growth. Infants with this disorder should be carefully assessed for associated hip dysplasia, which coexists in up to 20% of cases.

The possibility of *malignancy* must be considered in the differential diagnosis of any child with a cervical mass. History regarding the presence of persistent fevers, malaise, night sweats, weight loss, and other constitutional symptoms should be sought, and the child assessed for presence of pallor, petechiae, generalized adenopathy, and hepatosplenomegaly. Primary lymphoid malignancies, such as leukemia and lymphoma, may present initially with a rapidly enlarging neck mass. In contrast to infectious adenopathy, involved nodes tend to be firm, matted, nontender, and poorly mobile. Posterior triangle and supraclavicular masses carry a much higher risk for neoplasm than do anterior triangle masses. Metastatic tumors, such as rhabdomyosarcoma and neuroblastoma, also may initially be manifest as a neck mass. If malignancy is suspected, the ED workup should include complete blood count, electrolytes, uric acid, liver function studies, heterophile antibody titer, and a chest radiograph. Further evaluation, including imaging and biopsy, should be performed in consultation with a pediatric oncologist.

SURFACE LESIONS

Vascular Malformations

Vascular malformations result from errors in vascular morphogenesis. Unlike hemangiomas, they are present at birth, grow only in proportion to the child, and do not undergo regression. They may be of capillary, venous, or arterial origin, or combinations of vessel types may exist within the same lesion. *Port-wine stains* are among the more common capillary vascular malformations. They have a characteristic deep red to purple hue (Fig. 117.14). Children with facial port-wine stains that lie in the distribution of the ophthalmic branch of the trigeminal nerve (which includes the forehead, upper eyelids, and nose) merit careful evaluation for associated anomalies. Specifically, the *Sturge-Weber syndrome* is characterized by ipsilateral vascular angiomatosis of the leptomeninges and ocular vessels. Clinical manifestations may include seizures, mental retardation, hemiplegia, and glaucoma. Serial head CT scans performed on these children often demonstrate evolution of serpiginous calcifications and progressive atrophy of the cerebral cortex underlying the pial vascular malformations. Children with port-wine stains involving an extremity may develop hemihypertrophy of the affected limb because of an unusually rich underlying blood supply, the *Klippel-Tréenaunay-Weber syndrome*.

FIGURE 117.14. Facial port-wine stain that does not involve the distribution of the ophthalmic branch of the trigeminal nerve. (Reprinted with permission from Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997.)

All cosmetically significant port-wine lesions should be referred to a dermatologist. Some can be masked by using camouflage cosmetics (Covermark). Others may warrant definitive treatment using pulsed dye laser therapy.

Salmon patches, the most common form of vascular malformation seen in infancy, occur in 30 to 40% of all newborns. These flat pink lesions, which become more prominent with crying or exertion, are most commonly located on the nape of the neck (stork bites), on the glabella, or over the eyelids (angel kisses). They consist of distended dermal capillaries and almost always fade or disappear by the end of the first year of life, although nuchal salmon patches may persist into adulthood.

Hemangiomas

Hemangiomas, the most common benign neoplasm of infancy, occur in 10% or more of children less than 1 year of age.

Histologically, they are composed of hyperplastic vascular endothelium that develops from angioblastic tissue that has failed to connect normally with the vascular system during gestation. Although only a small portion of hemangiomas are evident at birth (2.5%), most become apparent within the first month of life. Lesions tend to undergo a period of rapid growth over the ensuing 6 to 12 months, then plateau. Subsequently, a slow process of involution begins, usually by 18 months. Approximately 50% of lesions involute completely by 5 years of age, and 95% by 9 years of age. A simple way to categorize hemangiomas is to subdivide them into three types: 1) *Superficial hemangiomas* are confined to the upper dermis. Formerly called capillary or strawberry hemangiomas, these lesions are well demarcated and compressible (Fig. 117.15). 2) *Deep hemangiomas*, previously called cavernous hemangiomas, lie in the lower dermis. They tend to have indistinct margins, and the overlying skin often has a bluish hue (Fig. 117.16). 3) On close inspection, many hemangiomas have a combination of both superficial and deep elements, and thus should be called *mixed hemangiomas* (Fig. 117.17).

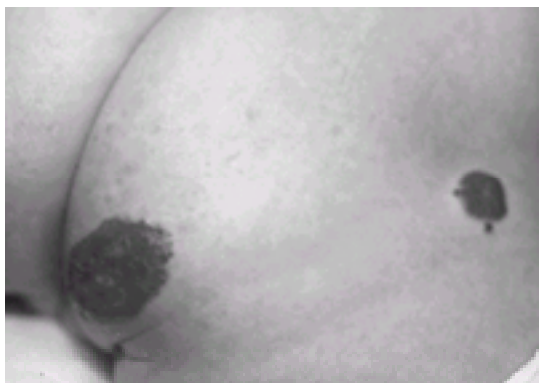

FIGURE 117.15. Superficial hemangiomas of hip and buttock. (Courtesy Joseph Glustein, MD.)

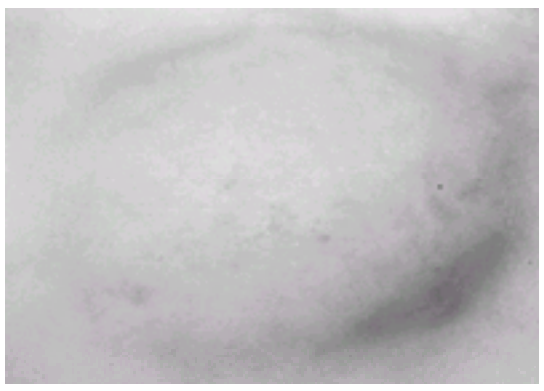

FIGURE 117.16. Deep hemangioma of upper lateral chest.

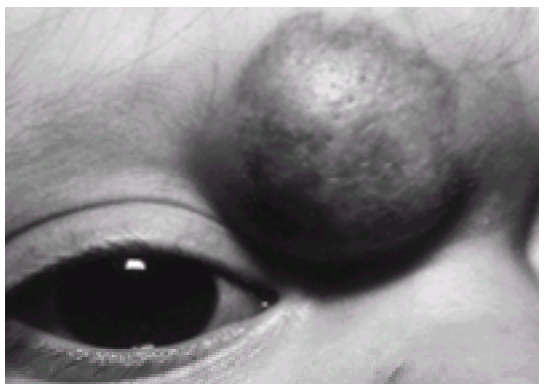

FIGURE 117.17. Mixed hemangioma containing both superficial and deep components. (Reprinted with permission from Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997.)

Because of their natural history of ultimate regression, a combination of watchful waiting and parental reassurance remain the standard of care for most hemangiomas. However, active intervention is indicated for lesions that compromise vital structures (airway, eyes, nose); lesions that are susceptible to trauma, hemorrhage, or infection; and those that grow at an alarming rate. Extremely large cavernous hemangiomas pose a risk for development of *Kasabach-Merritt syndrome,* which is characterized by sequestration of platelets with secondary thrombocytopenia and high-output cardiac failure. For lesions requiring active intervention, new treatment modalities that show promise include pulsed dye laser therapy and parenteral a-interferon. Intralesional steroid injections have also proved successful in some cases. Other options include cryotherapy and grenz ray therapy (which has replaced radiation). Decisions regarding which lesions to treat and how are best made in consultation with either a pediatric dermatologist or a plastic surgeon.

Lipoma

Lipomas are benign subcutaneous tumors composed of mature adipose cells. They often present in adolescence as

painless and usually solitary nodules. Although they may be located anywhere on the body, preferential sites include the neck, shoulders, back, upper chest, abdomen, and arms. Clinically, lipomas are nontender and have a soft, rubbery consistency, often with lobulations. Overlying skin is normal and easily slides across the mass, which helps distinguish lipomas from other skin nodules such as pilomatricomas. *Angiolipomas* are a variant of lipoma that have a component of capillary proliferation. Unlike lipomas, they tend to be painful. Lesions that are cosmetically significant, large, or painful warrant elective surgical excision.

Pilomatricoma

Pilomatricomas (calcifying epitheliomas) are relatively common lesions, accounting for 10% of superficial nodules seen in children. These benign tumors arise from cells of the hair matrix, hair cortex, or inner root sheath. Most are found on the head and neck but some arise on the trunk and extremities. They appear as firm (resulting from calcification), solitary nodules ranging in size from 0.5 to 5 cm. An overlying bluish hue may help distinguish the lesion from other benign nodules such as epidermal or dermoid cysts. When pinched, the overlying skin "tents," providing another distinguishing feature. Multiple pilomatricomas have been associated with myotonic dystrophy. Familial occurrences have been reported but are rare. If the lesion is located in a cosmetically sensitive area, elective surgical excision is the treatment of choice.

Pyogenic Granuloma

A *pyogenic granuloma* (also called lobular capillary hemangioma) is a benign vascular lesion most commonly found on exposed skin surfaces such as the face, hands, and forearms. Occasionally, lesions form on oral or nasal mucosal surfaces. They are comprised of granulation tissue with significant vascular overgrowth and are considered the result of an exaggerated vascular growth factor response after local trauma. Lesions are usually solitary and pedunculated, measuring from 0.5 to 2 cm. At times, multiple satellite lesions are found around a central granuloma. The color and character of a pyogenic granuloma varies according to its stage of growth. Early on, the lesion appears as a glistening, red, polypoid nodule with a friable surface that bleeds easily (Fig. 117.18). Later (weeks to months), the lesion becomes fibrotic and shrinks, taking on a reddish brown hue. The most common reasons for presenting to the ED are bleeding or chronic oozing of an early lesion. Treatment consists of excision followed by silver nitrate cauterization of vessels at the base. Recurrence merits referral to a dermatologist.

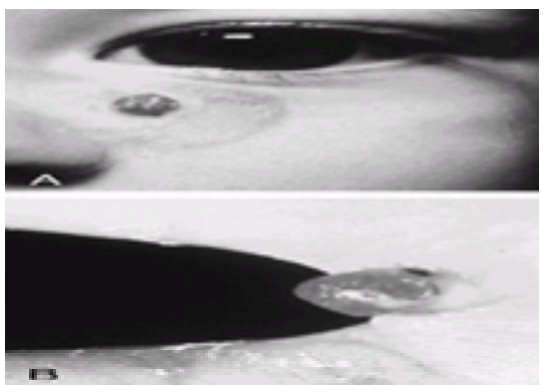

FIGURE 117.18. A. Pyogenic granuloma of the face. **B.** Interdigital pyogenic granuloma. (Reprinted with permission from Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997.)

Umbilical Granuloma

An umbilical granuloma presents as a soft, friable, polypoid mass that is pink or dull red. It arises from the base of the umbilical stump and at times may be pedunculated with a short stalk (Fig. 117.19). It is the product of an exuberant granulation tissue reaction, probably secondary to excessive moisture and/or low-grade infection. Treatment of most lesions consists of cauterization with a silver nitrate stick. During this procedure, care should be taken to cover the skin of the umbilical rim with gauze to protect it from burns. Following cauterization, the lesion should be blotted dry to avoid seepage of excess silver nitrate to surrounding tissue. Home care consists of keeping the umbilicus clean and dry. Large granulomas may require repeated cautery at intervals of several days. Pedunculated granulomas are candidates for suture ligation (3-0 nylon). The parent is then instructed to return for follow-up for cauterization of the base (once the granuloma has necrosed and dropped off) to prevent recurrence. Umbilical granulomas must be differentiated from persistent embryonic remnants such as an *omphalomesenteric duct* or *patent urachus.* The presence of a central lumen or chronic discharge should prompt the clinician to consider these rare umbilical anomalies. The distinction is of great clinical significance because these problems may be associated with other congenital malformations, and surgical excision of the entire remnant is necessary to prevent sequelae, such as infection.

FIGURE 117.19. Large pedunculated umbilical granuloma that responded to suture ligation and repeated silver nitrate applications.

Granuloma Annulare

The lesions of *granuloma annulare* are comprised of infiltrates of lymphocytes and altered collagen within the dermis. They first appear as raised nodules that gradually expand centrifugally to form annular rings ranging from 1 to 5 cm in diameter. They have a firm, fibrous, sometimes lumpy consistency on palpation. Overlying skin is usually normal or slightly hyperpigmented (Fig. 117.20). Although most are asymptomatic, a patient occasionally may report mild pruritus and present with superficial excoriation caused by scratching. The lack of an active microvesicular border, firm consistency on palpation, and the deeper dermal location of these lesions help distinguish them from *tinea corporis.* Lesions are commonly found on the extensor surfaces of the lower legs and the dorsum of the hands and feet and, less often, on the trunk or abdominal wall. Although granuloma annulare may present at any age, more than 40% of cases appear before age 15. Because most lesions undergo resolution within 1 to 2 years, reassurance is usually all that is necessary. In the rare case of a patient with severe or widespread lesions, dermatologic consultation should be sought. Although adult studies have shown an association of granuloma annulare with diabetes mellitus, this is not the case for children.

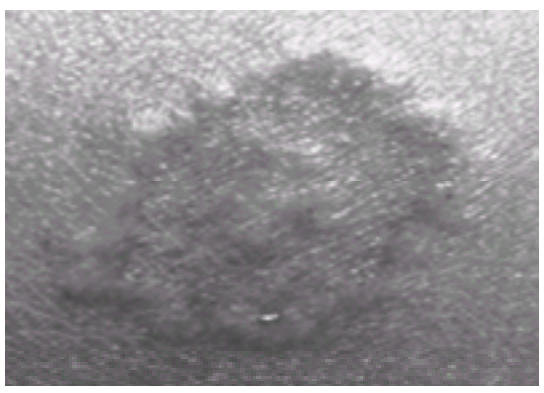

FIGURE 117.20. Granuloma annulare.

Juvenile Xanthogranuloma

Juvenile xanthogranulomas (JXG) present as nodular or plaquelike lesions with a firm or rubbery consistency. Initially reddish in color, they evolve to have a distinct yellow or orange hue (Fig. 117.21). Many are noted at birth, whereas others appear within the first several months. They range in diameter from 0.5 to 4 cm. Like hemangiomas, they tend to grow rapidly in infancy, then spontaneously regress in early childhood. Common sites include the scalp and face, proximal extremities, and occasionally, the subungual area of a digit or a mucocutaneous junction. Lesions may be solitary but are often present in groups. Histologically, xanthogranulomas are comprised of lipid-laden macrophages or histiocytes within a granulomatous matrix whose inciting source is unknown. In rare cases, giant or disseminated lesions may occur. Patients who have multiple or diffuse lesions may have ocular lesions as well. Specifically, lesions of the iris that have been associated with spontaneous anterior chamber hemorrhage and glaucoma. On occasion, ocular lesions have been misdiagnosed as retinoblastoma. A systemic form of JXG exists, and affected patients may or may not have concomitant cutaneous findings. In this variant, noncutaneous lesions may involve the brain, heart, liver, spleen, and lungs. Children who have both JXG and neurofibromatosis are at a much higher risk for unusual forms of leukemia and thus should be appropriately monitored. Last, unlike children with disseminated xanthomas, there is no relationship between JXG and lipid abnormalities. All children with suspected xanthogranuloma should undergo biopsy. An ophthalmologic evaluation is necessary if JXG is confirmed, and careful observation for evidence of systemic involvement is warranted.

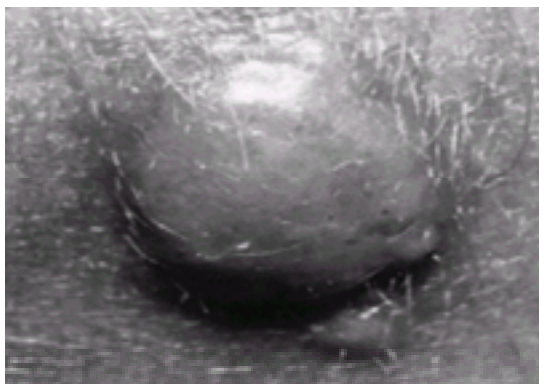

FIGURE 117.21. Yellow nodular lesion typical of juvenile xanthogranuloma.

Neurofibroma

A neurofibroma may present as a solitary lesion in an otherwise normal patient or as a feature of neurofibromatosis type I. Cutaneous neurofibromas arise from nerve sheath cells located in the dermis. They appear as pink or flesh-colored nodules that are soft and range in size from 0.5 to 3 cm. Most do not appear until adolescence. Lesions may be confused with angiolipomas and hemangiomas; however, a distinguishing feature is the tendency of neurofibromas to be especially soft centrally and invaginate with digital pressure, described as “button-holing.” Elective excision is indicated only if the lesion is compressing a nerve, causing nerve root pain, because excision is often followed by recurrence of an even larger lesion.

Keloid/Hypertrophic Scar

Exaggerated proliferation of fibrous connective tissue in the process of cutaneous wound healing results in formation of *hypertrophic scars* and *keloids.* Wounds involving areas of skin that are thick or under high tension (shoulders, back, chest, or chin) are at greatest risk. The ear lobe is another commonly affected site. Individuals with dark skin are much more susceptible to abnormal scarring, which has its highest incidence in adolescence and early adulthood. Hypertrophic scars remain confined to the area of original injury. They are rarely painful and tend to undergo slow regression over 6 to 12 months. In contrast, keloids extend beyond the original wound margins and rarely regress spontaneously. Initially, keloids may be painful and tender or pruritic. They have a rubbery consistency on palpation and a smooth pink surface (Fig. 117.22). Ear piercing, tattooing, and elective cosmetic procedures should be avoided in persons who have a tendency to form keloids. Severe keloids should be referred to a dermatologist or plastic surgeon for optimal treatment, which may include injection of intralesional steroids, cryosurgery, or meticulous excision with steroid infiltration along wound margins.

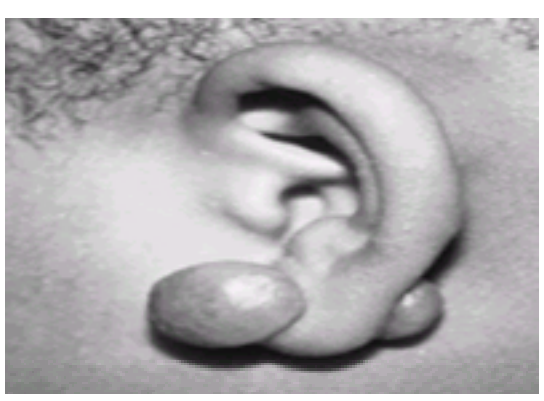

FIGURE 117.22. Large keloid that formed after ear piercing in a susceptible child.

Lumbosacral Lesions

Pilonidal dimples, typically located in the midline in the sacrococcygeal area, are benign lesions of no clinical significance. On close inspection, there is no evidence of a central pore or opening. In contrast, pilonidal sinuses, which are found in the same area, do have a small surface opening to a tract lined by stratified squamous epithelium that extends toward, but not into, the spinal canal. In some instances, sinuses appear to be of embryonic origin, stemming either from an abnormality of midline fusion or invagination of ectodermal elements. The base of such a lesion may consist of a small cyst containing products of skin cells and epithelial appendages, including hair. In other cases, the source may be a distorted hair follicle. Pilonidal sinuses and cysts are asymptomatic until the sinus becomes obstructed and/or infected. This phenomenon is most likely to occur during adolescence or early adulthood. Males are much more commonly affected than females. Excess weight and hirsutism and a sedentary lifestyle or occupation that requires prolonged sitting also appear to be predisposing factors.

Infecting organisms usually gain access through the external sinus tract. Once infection occurs, an abscess forms and tends to enlarge rapidly. Because the overlying skin is thick, expansion tends to occur deep to the skin surface, and acquired sinus tracts may form external to the postsacral facia. Patients typically complain of low back pain, increased on sitting, and local tenderness. On examination, a tender, indurated swelling is noted overlying the sacrococcygeal area with the original sinus at its cephalad end (Fig. 117.23). Treatment consists of incision and drainage with careful probing

to break up loculations and extract any hairs present because these act as foreign bodies. Cultures grow mixed organisms, including staphylococci, anaerobes, and fecal flora. Home care includes sitz baths and oral antimicrobial therapy. Elective excision of the entire cyst and all associated sinus tracts is indicated once inflammation has resolved.

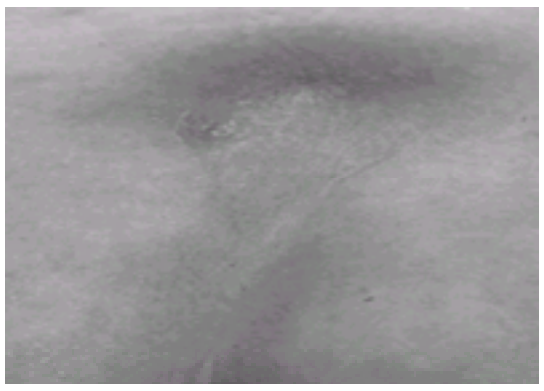

FIGURE 117.23. Infected pilonidal cyst.

Cutaneous Manifestations of Spinal Dysraphism

A number of midline cutaneous abnormalities found in the lumbosacral area are associated with underlying vertebral or spinal cord defects that are the result of defective closure of the caudal neural tube, *occult spinal dysraphism.* Skin findings include *hairy patches* (Fig. 117.24), *skin tags*, *port-wine stains*, *hemangiomas,* and *congenital dermal sinuses.* The latter tend to be more cephalad than pilonidal sinuses, and their sinus tracts often extend to the spinal column. Underlying intraspinal lesions include dermoid tumors, lipomas, and diastematomyelia. In the latter condition, the lower cord is divided sagittally by an osseous or fibrocartilaginous septum, which tethers the cord at that level, impeding its normal ascent within the spinal canal as the child grows. Patients with tethering may present with lower extremity neurologic deficits at birth or may insidiously develop symptoms later in infancy or childhood, especially during a period of rapid growth. Complaints may include back or leg pain or stiffness, buttock pain, weakness or numbness, and bowel and bladder complaints.

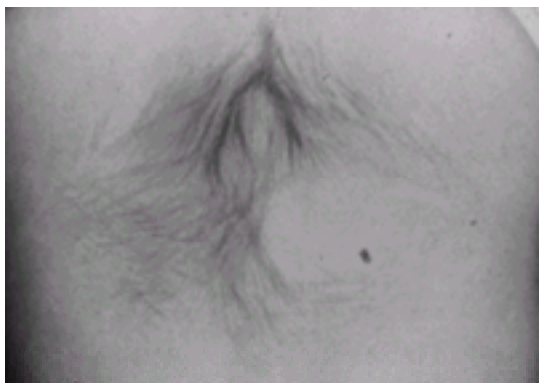

FIGURE 117.24. Lumbosacral hairy patch in a patient with diastematomyelia.

Any child found to have one of the midline cutaneous findings just described should undergo neuroimaging to detect and delineate underlying vertebrospinal defects because early neurosurgical intervention enables substantial reduction in morbidity.

Perineal Lesions

Urethral prolapse is a phenomenon seen primarily in obese prepubescent girls. Two-thirds or more are African-American. In this condition, the urethra prolapses through the urethral meatus and is seen as a red or purplish red, friable, edematous mass overlying the anterior portion of the introitus (Fig. 117.25). It often has a donut shape, and close inspection reveals a central orifice. The prolapsed mucosa is usually mildly painful and tender and tends to bleed easily. Presenting complaints may include perineal pain, dysuria, and blood spotting on underwear. Urination is not impaired. The precipitating event is characterized by increased intra-abdominal pressure, usually severe straining with constipation, a severe coughing spell, or prolonged crying. Because the red friable mass often overlies the hymenal orifice, it can be mistaken for traumatized hymenal folds, raising suspicion of sexual abuse. Correct diagnosis is made by examination under magnification after applying a topical anesthetic. This enables visualization of the central orifice and elevation of the mass to visualize the hymen. Management consists of treating the predisposing condition, oral analgesics and topical antibiotic/anesthetic creams for symptomatic relief, and twice daily application of estrogen cream.

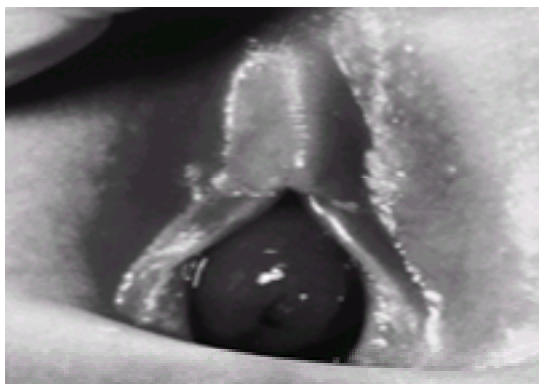

FIGURE 117.25. Typical doughnut appearance of urethral prolapse.

Perianal skin tags are common sequelae of anal fissures and thus tend to be seen in children with a history of large, hard stools. They consist of pedunculated masses on short stalks that form during the process of healing of an anal fissure, probably in part caused by frictional forces common to this area. They are usually asymptomatic. They also can be seen in association with hypertrophic scars, another common sequela of the healing of an anal fissure. Although most patients with these lesions are otherwise normal, a small percentage have them as manifestations of perianal disease, internal fissures and/or fistula, which may be the primary problem or one manifestation of inflammatory bowel disease. Management is directed at treating the predisposing or underlying condition. Bothersome pedunculated lesions can be tied off with silk suture.

Rectal prolapse, herniation of the rectum through the levator and then the anal orifice, is a phenomenon typically seen in children between 1 and 2 years of age. The most common predisposing conditions, severe constipation and severe diarrhea, are characterized by repeated straining on defecation, which stretches pelvic suspensory structures, facilitating herniation. Patients with spina bifida may have prolapse as a consequence of deficits in perineal innervation with attendant atrophy of the supporting perineal muscles. In children with undiagnosed cystic fibrosis, passage of large to voluminous stools and poor muscle tone secondary to malnutrition appear to predispose. Occasionally, an apparent rectal prolapse represents the lead end of a sigmoid intussceptens. In these cases, patients have a history of antecedent, intermittent, abdominal pain or irritability and may have vomiting, lethargy, and/or rectal bleeding as do other infants and children with intussusception. Clinically, a cylindrical mass with a central orifice and a glistening red surface is seen protruding through the anus. Acutely, the mass can be reduced with gentle pressure. Attention is then directed at identifying and treating the underlying condition to prevent recurrences. The need for operative intervention for persistent recurrences is rare and is largely limited to neurodevastated patients with intractable constipation.

Suggested Readings

Ashcraft KW, Holder TM. Acquired anorectal disorders. In: Ashcraft KW, Holder TM, eds. Pediatric Surgery. 2nd ed. Philadelphia: WB Saunders, 1993:410–415.

Bellinger MF. Urologic disorders. In: Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997: 421–440.

Bethel CA. Incision and drainage of a felon. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:1211–1215.

Canales FL, Newmeyer WL, Kilgore ES. The treatment of felons and paronychias. Hand Clin 1989;5:515–523.

Cohen BA, Davis HW, Mallory SB, et al. Dermatology. In: Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997:211–264.

Davis HW, Karasic RB. Pediatric infectious disease. In: Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997:343–396.

Derkay CS, Tunnessen WW. Picture of the month: nasal glioma. Arch Pediatr Adolesc Med 1994;148:954–955.

Dunmire SM, Paris PM. Atlas of Emergency Procedures. Philadelphia: WB Saunders, 1994.

Eberlein R. Hand and finger injuries. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:1047–1061.

Freyer DR, Kennedy R, Bostrom BC, et al. Juvenile xanthogranuloma: forms of systemic disease and their clinical implications. J Pediatr 1996;129:227–237.

Frieden IJ. Which hemangiomas to treat—and how? Arch Dermatol 1997;133:1593–1595.

Henretig FM. Incision and drainage of a paronychia. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:1205–1210.

Hinson RM, Biswas A, Mizelle KM, et al. Picture of the month: persistent omphalomesenteric duct. Arch Pediatr Adolesc Med 1997;151:1163–1164.

Hurwitz S. Clinical Pediatric Dermatology. 2nd ed. Philadelphia: WB Saunders, 1993.

Johnson CF. Prolapse of the urethra: confusion of clinical and anatomic characteristics with sexual abuse. Pediatrics 1991;87(5):722–725.

Knight PJ, Reiner CB. Superficial lumps in children: what, when, and why? Pediatrics 1983;72:147–153.

Loiselle J, Cook RT Jr. Hair tourniquet removal. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:1183–1187.

McBride TP, Davis HW, Reilly JS. Otolaryngology. In: Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997:683–728.

McCann J, Voris J, Simon M, et al. Perianal findings in prepubertal children selected for non-abuse: a descriptive study. Child Abuse Negl 1989; 13:179–193.

Mooney MA, Janniger CK. Pyogenic granuloma (Review). Cutis 1995; 55:133–136.

Murray PJ, Davis HW, Hamp M. Pediatric and adolescent gynecology. In: Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997:525–562.

Nakayama DK. Surgery. In: Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997:487–524.

Paller AS, Pensler JM, Tomita T. Nasal midline masses in infants and children. Arch Dermatol 1991;127:362–366.

Prendville JS. Diseases of the dermis and subcutaneous tissues. In: Schachner LA, Hansen RC, eds. Pediatric Dermatology. 2nd ed. New York: Churchill Livingstone, 1995.

Sebastian MW. Pilonidal cysts and sinuses. In: Sabiston DC Jr, ed. Textbook of Surgery. 15th ed. Philadelphia: WB Saunders, 1997:1330–1334.

Simon RR, Wolgin M. Subungual hematoma: association with occult laceration. Am J Emerg Med 1987;5:302–304.

Tallman B, Tan OT, Morelli JG, et al. Location of port-wine stains and the likelihood of ophthalmic and/or CNS complications. Pediatrics 1991;87:323–327.

Wahrman JE, Honig PJ. Hemangiomas. Pediatr Rev 1994;15:266–271.

Ward WT, Bisignani G, Davis HW, et al. Orthopedics. In: Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997:625–682.

Warden TM, Fourré MW. Incision and drainage of cutaneous abscesses and soft tissue infections. In: Roberts JR, Hedges JR, eds. Clinical Procedures in Emergency Medicine. 2nd ed. Philadelphia: WB Saunders, 1991:591–610.

Wessel HB. Neurology. In: Zitelli BJ, Davis HW, eds. Atlas of Pediatric Physical Diagnosis. 3rd ed. St Louis: Mosby, 1997:441–466.

CHAPTER 118

Abdominal Emergencies

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Although some acute intra-abdominal diseases may affect both adults and children, other conditions occur specifically in children and are extremely uncommon in adults. Appendicitis is similar in both age groups. Disorders such as malrotation and volvulus, Hirschsprung's disease, and congenital bowel stenoses affect the younger age group. The emergency physician must be aware of the unique problems that occur in infancy and childhood and gear his or her thinking to an understanding of the clinical manifestations of these entities. There must also be an awareness and sensitivity to the special techniques for evaluating a child with a potential “surgical abdomen.”

The abdominal surgical problems most commonly found in infants and children are discussed under the following categories: 1) diseases that produce peritoneal irritation; 2) acute intestinal obstruction; 3) chronic, partial intestinal obstruction; 4) problems that produce rectal bleeding; 5) intra-abdominal masses; 6) abdominal wall defects; and 7) foreign bodies of the gastrointestinal (GI) tract. Nonsurgical GI emergencies are covered in Chapter 93. Chapter 108 deals with major trauma to the abdominal contents. Chapter 50 reviews the diagnostic approach to the child with abdominal pain.

EVALUATION

History

Neither an infant nor a young child can give a complete history, but it is useful to obtain whatever history possible from the child. Younger children, for example, are likely to recall recent events. Events somewhat more remote in time may be

better remembered by an adult, although the adult's accuracy may be clouded by his or her emotional state. If a child has been toilet-trained for a few years, neither the child nor the parent may be sufficiently attentive to give an accurate history of bowel habits. In general, however, once a child reaches school age, taking a history becomes considerably easier.

In obtaining the history, the emergency physician should seek an accurate chronology of events, with direct questions: When did the pain start? Where was it felt at first? What happened next? The physician should also obtain a careful history of other symptoms. By asking when the child was last perfectly well, one may get a more accurate assessment of the child's pathophysiology. In this way, one may better evaluate a child with multiple symptoms, such as fever, vomiting, abdominal distension, and any changes in bowel habits, appetite, feeding pattern, or general activity level. The physician then develops a feeling for the patterns of different diseases in children.

For example, if there is high fever at the onset of the problem, it is more likely infectious in origin (i.e., viral gastroenteritis or a urinary tract infection rather than appendicitis). The child with acute appendicitis usually has a low-grade fever, which develops after the onset of the first vague midepigastric pain. If the appendix has perforated and the child is dehydrated and septic, the temperature may reach as high as 39° to 40°C (102.2° to 105.8°F).

Vomiting is often associated with acute or chronic abdominal pain, especially in the younger child. Infants with a history of colicky pain and vomiting should cause suspicion of intussusception or malrotation with volvulus. The younger child with acute appendicitis often has vomiting, whereas a teenager does not. Voluminous bilious vomiting is often the first symptom in a child who has had previous abdominal surgery and has intestinal obstruction.

There must be concern about a possible underlying surgical condition in any child whose first presenting symptom is abdominal pain. It is important to determine the type of pain, its location, and whether it is constant or intermittent and colicky. Visceral pain has a colicky character and is usually generalized in the upper, middle, or lower abdomen, depending on which level of the GI tract is involved. A child who complains of a sharp pain, often in the right lower quadrant, that comes and goes, does not have appendicitis. Peritoneal pain reflects secondary features of the abdominal disease process such as inflammation, bowel perforation, or infarction. Often, a child can perceive the actual change from visceral-type pain to peritoneal pain. A catastrophic event such as infarction of the entire small intestine from a volvulus or a closed loop obstruction can cause lethargy and a shocklike picture rather than extreme abdominal pain in a short period.

If the child's illness begins with nonbloody diarrhea, it is less likely to be surgical in nature, in contrast to a child with a 3-day history of abdominal pain and fever in whom the onset of diarrhea may be secondary to a perforated appendix with pelvic abscess formation. Bloody diarrhea may or may not be surgical in nature, but it warrants a very thorough assessment (see Chapter 19 and Chapter 30).

In addition to detailing the chronology of events and the characteristics of the symptoms, the physician should inquire about any treatments which have been attempted at home, such as cathartics, enemas, or drugs. Often, these home remedies will have modified the symptoms and their chronology in some way.

It is important not to overlook the history. For example, a child known to have sickle cell disease may have an acute abdominal crisis as the cause of abdominal pain. The presence of this underlying hematologic abnormality forces the examiner to be more precise in the interpretation of the physical examination. A child with known cystic fibrosis or inflammatory bowel disease may also commonly have abdominal pains that do not require surgery.

Examination

Children vary a great deal in their ability to cooperate during the abdominal examination, and often the physician must make patient, repeated efforts to perform an adequate evaluation. A few minutes spent gaining the child's confidence is important to the child and helpful to the examiner. The best abdominal evaluation is done before the child becomes disturbed by the drawing of blood samples or the performance of a rectal examination. On some occasions, it is helpful to let the distraught, crying child actually go to sleep; a second examination while the child is sleeping may determine whether real tenderness is present. It may also be helpful to relax the child by having a parent hold him or her for the examination. The use of narcotics should be avoided because they might mask findings of peritonitis. All infants and children should have an examination that is not hindered by the child's clothes. Small incarcerated indirect inguinal hernias can easily escape detection if the entire abdomen is not observed and palpated. Some children need reassurance that being undressed for the examination is important. The child's sense of modesty must be respected when possible.

Once the child relaxes, the physical examination should follow an orderly progression: inspection and observation, nontouching maneuvers, abdominal palpation, auscultation, examination of nonabdominal areas, and rectal examination (Table 118.1).

Physical Findings	Meaning
Abdominal distension	Peritonitis, intestinal obstruction, ileus with gastroenteritis
Visible bowel loops	Intestinal obstruction, intussusception
Asymmetry	Appendiceal abscess
	Tumor
Point tenderness	Appendicitis, cholecystitis
Guarding	Peritonitis, appendicitis, abscess
Rigidity	Cholecystitis
Rebound	Infarcted bowel
Palpable mass	Tumor or cyst, intussusception
	Chronic constipation
Gas palpable in cecum	Not appendicitis
High-pitched bowel sounds	Intestinal obstruction
No bowel sounds	Peritonitis, infarcted bowel, perforated appendix or bowel
Rectal examination: tenderness or bogginess on right	Appendicitis

Table 118.1. ***Abdominal Physical Findings and What They Mean***

The initial step of inspection should focus on the presence of distension, visible bowel loops, and asymmetry. Inspection is easily done because of the relative prominence of the child's abdomen and thinness of the overlying muscles and subcutaneous tissue. Sometimes, observation during a feeding gives important information. The child should be observed in motion—for example, behavior when getting on and off the examination table and walking. Nontouching maneuvers include having the child cough or jump up and down while noting any change in symptoms.

Palpation is the most difficult, yet most informative, aspect of physical examination. When possible, the child can initiate his or her own examination by self-palpation. Ask the child to point to the place where the pain is most severe. Avoid the tender area initially but return to palpate it last. A number of “tricks” can be used to facilitate the palpation, such as diverting the child with conversation, palpating with the head of the stethoscope in hand, and palpating while asking the child to take a deep breath. For example, quick withdrawal of a palpating stethoscope may confirm rebound tenderness and the presence of an underlying peritoneal irritation.

Auscultation of the abdomen indicates the state of bowel motility. The presence of normal bowel sounds may not rule out surgical pathology, but a silent abdomen or one with high-pitched tinkles and rushes suggests the possibility of sepsis or obstruction. Auscultation and palpation of nonabdominal areas is important. In addition, evaluation of the acute abdomen requires careful examination of the lung fields, testes and scrotum, inguinal regions, femoral triangles, and costovertebral areas. Failure to examine these areas carefully will result in missing important diagnostic clues to nonabdominal problems that may present with abdominal symptoms.

The final step in the evaluation should be a rectal examination. The important technical aspects include use of generous amounts of lubricant, slow insertion of the examiner's finger as the patient takes slow deep breaths, and the palpation of the presumed painful area last. If done carefully and gently, the rectal examination is usually well tolerated by the child. Any stool on the examiner's glove should be tested for the presence of blood.

Laboratory

Laboratory and radiographic studies vary, depending on the diagnoses that are being considered. Most children accept radiographic studies as “having your picture taken.” A urinalysis is an important part of the evaluation of any abdominal symptoms in children. A complete blood count (CBC) and differential is essential when considering surgical conditions. The need for other tests—blood urea nitrogen (BUN), electrolytes, serum amylase, sickle cell preparation, and so on—depends on the child's condition and the diagnostic considerations.

Assessment

Once the history, physical examination, and laboratory data are all available, the emergency physician must synthesize them into an overall assessment. If the physical findings are reproducible and consistent in location and character, an accurate diagnosis can often be made and appropriate treatments started. If the findings are both worrisome and equivocal, and do not fit into a comprehensive picture, the patient should be admitted for observation and reassessment. The following sections detail the cardinal symptoms and signs of the common acute surgical problems in children. Initial emergency department (ED) management is discussed, but in all cases of presumed surgical disease, the definitive treatment requires consultation with a surgeon.

DISEASES THAT PRODUCE PERITONEAL IRRITATION

The physician must perform a careful examination to elicit accurate signs of peritonitis. Tenderness is not necessarily an indication of an intra-abdominal surgical problem in a child. A child with localized peritonitis may have only minimal findings, while a patient with a nonsurgical condition may have severe pain and generalized tenderness. For example, on examination, a child with severe colic caused by gastroenteritis may appear to have genuine peritoneal signs. In fact, such a child may have only exaggerated voluntary guarding of the abdominal wall, mimicking the true rigidity of peritonitis.

The well-known features of peritonitis—rigidity, spasm, involuntary guarding, and rebound—are as valid for a child as they are for an adult. In a child with abdominal pain, however, one has to be patient and gentle enough in the initial phases of the examination so that signs of peritonitis can be checked repeatedly without breaking rapport with the child. Reproducible anterior peritoneal tenderness in the same location is much more suggestive of peritonitis than deep abdominal tenderness that shifts in location with each reexamination of the child.

Acute Nonperforated Appendicitis

Background

Acute appendicitis is one of the most common abdominal surgical conditions seen by the emergency physician or surgeon who cares for children. It occurs in all age groups and is particularly difficult to diagnose in its early states and in infants and toddlers. The emergency physician must accurately evaluate the child and promptly consult a surgeon when the diagnosis is clear or when appendicitis cannot be safely ruled out. Such consultation is especially urgent in younger children, in whom perforation can occur within 8 to 24 hours of the onset of symptoms.

Clinical Manifestations

Usually the child with appendicitis complains initially of poorly defined and poorly localized midabdominal or periumbilical pain. Unfortunately, this symptom is common to many other intra-abdominal, nonsurgical problems. In the young and, to a lesser extent, the older child, vomiting and a low-grade fever often occur soon thereafter. Characteristically, the pain then migrates to the right lower quadrant (Table 118.2).

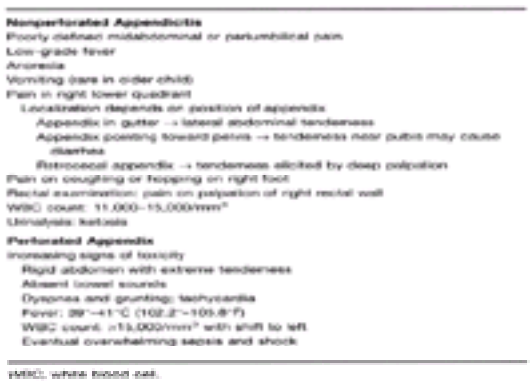


Table 118.2. ***Progression of Symptoms and Signs of Appendicitis***

Because the position of the appendix may vary in children, the localization of the pain and the tenderness on examination may also vary. An appendix that is located in the lateral gutter may produce flank pain and lateral abdominal tenderness; an inflamed appendix pointing toward the left lower quadrant may produce hypogastric tenderness. An inflamed low-lying, pelvic appendix may not cause pain at McBurney's point but instead may cause diarrhea from direct irritation of the sigmoid colon. Often, the child with appendicitis is anorexic, suggesting the presence of nausea even if he or she is unable to verbalize this complaint. Because motion aggravates peritoneal irritation, a child with appendicitis typically prefers to lie still or, when moving, splints toward the painful area.

When obtaining the history, the physician needs to consider other causes of abdominal pain which may appear as appendicitis but, in fact, are nonsurgical. Concurrent GI illness in other family members or friends suggests the possibility of an infectious gastroenteritis. Chronic constipation, urinary tract disease, or an emotional disturbance may also point to an explanation other than appendicitis for the abdominal pain. Cough, fever, and rapid breathing suggest the possibility of a lower lobe pneumonia.

On examination, palpation is usually reliable in demonstrating focal peritoneal signs at the site of the inflamed appendix. If the appendix is in the pelvis or retrocecal area, however, typical anterior peritoneal signs may be absent. The physician can confirm his or her impression of point tenderness by pressing gently in each quadrant and asking the child to indicate which area is most tender. When the inflamed appendix is not close to the anterior abdominal wall, as in the case of retrocecal appendix, tenderness may be more impressive on deep palpation of the abdomen or by palpating in the flank. This impression of focal tenderness can sometimes be confirmed by shaking the child's abdomen or getting him or her to cough, which often produces a wince of pain at the involved area. Finally, a well-done rectal examination may make the difference between deciding to operate or observe the child. The examining finger should be inserted as fully as possible without touching the area of presumed tenderness. Then, when the child is relaxed and taking deep breaths, the examiner can gently stroke or indent an area high on the right rectal wall. A sudden involuntary reaction of pain confirms the presence of inflammation. In a child with a history of probable appendicitis for more than 2 or 3 days, a boggy, full mass may also be in this location, suggesting an abscess.

A CBC in a child with appendicitis usually shows an elevated white blood cell (WBC) count in the range of 11,000 to 15,000/mm^3 in the first 12 to 24 hours of the illness. As the appendix becomes more gangrenous, the WBC count rises further, and the differential demonstrates more and more neutrophils and an increasing number of bands. Urinalysis often shows ketosis. If the inflamed appendix lies over the ureter or adjacent to the bladder, a few WBCs may be found in the urinary sediment. The presence of numerous WBCs and bacteria on a freshly spun specimen may indicate an acute urinary tract infection. However, a child with a chronic urinary tract infection may also develop appendicitis.

If the clinical and laboratory diagnosis of acute appendicitis is convincing, no further studies are indicated.

As far as the abdominal roentgenogram is concerned, many consider it a peripheral study valid only for the demonstration of a fecalith or free air. Abdominal roentgenograms are normal in many cases of acute appendicitis. In 8 to 10% of cases, however, a calcified appendiceal fecalith can be identified on abdominal roentgenograms. Because the incidence of subsequent appendicitis with perforation is significant, it has been suggested that prophylactic appendectomy be performed on the asymptomatic child in whom a calcified fecalith is detected.

Most children with an acute nonperforated appendix show diminished air in the GI tract. This is a result of anorexia, nausea, vomiting, and diarrhea. Other roentgenographic signs of appendicitis are thickening of the cecal wall and mucosal folds with air–fluid level, indistinct psoas margins with scoliosis concave toward the right, focal obliteration of the adjacent properitoneal fat pad, or presence of air in the appendix. However, a retrocecal appendix may be filled with gas in the normal person. Ileus secondary to peritoneal irritation or an inflamed appendix crossing the terminal ileum may be seen. Rarely, a perforated appendix may produce pneumoperitoneum.

Barium enema in the diagnosis of appendicitis is no longer used because in 8 to 10% of normal children, barium does not

fill the appendix. However, partial or nonfilling of the appendix with local impression on the cecum or terminal ileum, or associated with other evidence of a pelvic or right lower quadrant mass, is indicative of appendicitis. In a child with leukemia, a tender right lower quadrant mass may indicate typhlitis, an inflammatory reaction of the cecum. Differentiation of typhlitis and an appendiceal abscess in a child with leukemia, therefore, may be difficult. Today, for evaluation of appendicitis Doppler ultrasound or computed tomography (CT) is preferred.

Routine chest radiographs are not indicated unless the patient has a history of pulmonary problems, such as asthma, or unless the symptoms and signs indicate atypical right abdominal pain with splinting respirations. A chest radiograph in such a case may reveal subtle evidence of a right lower lobe pneumonia. With good hydration, these subtle findings on radiograph may develop into an early visible infiltrate in a few hours.

Unfortunately, laboratory findings do not differentiate mesenteric adenitis, which closely mimics acute appendicitis. A complete approach to the child with abdominal pain is covered in Chapter 50.

Management

The preoperative preparation of a patient with acute appendicitis should include BUN and electrolytes if the patient has been vomiting or has had poor fluid intake for more than a few hours. If an unexpected anemia is discovered on the CBC, a crossmatch should also be done. Intravenous (IV) fluids should be started, with emphasis on replacing the child's deficits as quickly as possible. Protracted GI losses, as with vomiting, may lead to potassium depletion. Therefore, the initial IV fluids should contain at least 0.5 n saline with 10 mEq of potassium chloride per 500 mL. These fluids can then be altered, if necessary, once the serum chemistries are known. During this period, the patient and the family should be psychologically prepared for surgery.

The emergency physician must keep in mind the many variations in the way appendicitis can present. As a good rule of thumb, a patient should at least be admitted for observation if there are positive findings in two of the three classical modes of assessment: history, physical examination, or laboratory. For example, a child with an excellent history of appendicitis and a high WBC count should be admitted even if the abdominal examination is normal. The appendix may have perforated just before the examination, causing temporary cessation of physical findings. In equivocal situations, the key element is follow-up assessment. A child who is admitted can, if necessary, be reexamined every 1 to 2 hours. As for a child in whom the possibility of appendicitis seems more remote, the physician may elect to have the child return for reexamination in the office or in the ED. Such follow-up arrangements work best when the parents are reliable and do not live too far from the hospital. Arrangements for follow-up should be specific, including time of appointment, the specific doctor to be seen, and a contact telephone number in case the child does not return as scheduled.

Perforated Appendicitis

Ideally, once the diagnosis of appendicitis is considered seriously, the patient will have surgery before the appendix has perforated. Unfortunately, some patients, particularly younger children and infants, may arrive for emergency care with an already perforated appendix because of a delay in seeking treatment or in making the diagnosis. Once the appendix has perforated, there are usually signs of generalized, rather than localized, peritonitis. In a young child, the omentum is flimsy and often incapable of walling off the inflamed appendix. As a result, perforation occurs more quickly, and secondary dissemination of the infection occurs more widely. Although the mortality from appendicitis has decreased, the incidence of perforation in children has remained the same over the last several decades.

Clinical Manifestations

Within a few hours after perforation has occurred, the child begins to develop increasing signs of toxicity. First, the lower abdomen and then the entire abdomen becomes rigid with extreme tenderness. Bowel sounds are sparse to absent. In addition, there are signs of prostration such as pale color, dyspnea, grunting, significant tachycardia, and higher fever (39° to 41°C [102.2° to 105.8°F]). Rarely, the patient may develop septic shock (see Chapter 3) from the overwhelming infection caused by bowel flora.

Initially, the findings may be confused with those of pneumonia because the extreme abdominal pain may cause rapid shallow respirations and decreased air entry to the lower lung fields. There is impaired excursion of the diaphragm and respiratory failure in some cases. In infants and toddlers, the findings may also be confused with meningitis because any movement of the child (even flexion of the neck) produces pain and irritability. The high fever and other signs of prostration may be indications for a lumbar puncture. After the spinal fluid analysis is found to be normal, the suspicion of perforated appendicitis may be heightened.

The laboratory findings in the child with perforated appendicitis often suggest this diagnosis. The WBC count is significantly elevated, usually above 15,000/mm^3, with a marked shift to left; leukopenia may be seen when perforation has resulted in overwhelming sepsis and septic shock.

The radiologic evaluation of suspected perforated appendicitis should include both plain abdominal radiographs and pelvic ultrasound. The plain film of the abdomen may show free air or evidence of peritonitis (Fig. 118.1). The ultrasound of the pelvis may show a complex mass with or without a calcified fecalith or free fluid within the abdominal cavity (Fig. 118.2).

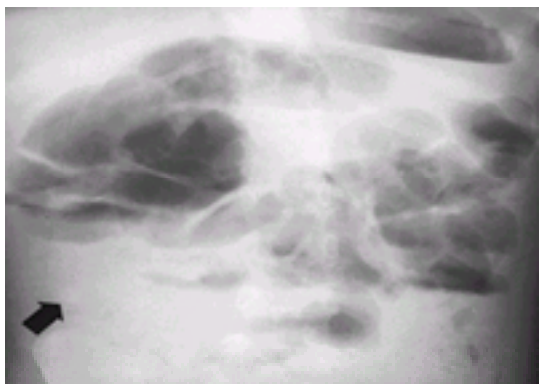


FIGURE 118.1. Perforated appendicitis with abscess and fecalith. The upright abdominal roentgenogram shows numerous dilated loops of bowel and a calcified fecalith *(arrow).* Note that the space between the individual loops indicates the presence of intraperitoneal fluid.

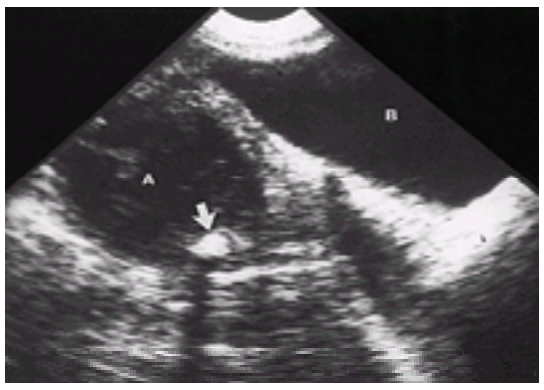


FIGURE 118.2. Perforated appendicitis with abscess and fecalith. Ultrasonography of the pelvis shows a complex mass *(A)* with a fecalith *(arrow)* producing characteristic acoustic shadowing to the right of the bladder *(B).*

Management

Initially, the focus of therapy should be on resuscitation. There must be careful attention to airway, breathing, and circulation (see Chapter 1). The severely septic child may need positive-pressure ventilation with high concentrations of oxygen to overcome ventilatory insufficiency. Hypovolemia should be corrected with 5% dextrose in either normal saline or Ringer's lactate solution. An alternative volume expander is 5% albumin solution in normal saline. An initial bolus starting at 20 mL/kg is given over 30 minutes or less until vital signs are restored and the patient produces urine. Metabolic acidosis should be treated with sodium bicarbonate, the dose being determined by the results of the serum electrolytes or, more precisely, the base deficit from an arterial blood gas.

Hypovolemia and acidosis must be treated vigorously until adequate circulation has been reestablished, as measured by the strength of the pulse, blood pressure, skin perfusion, capillary refill in the extremities, and urine output. If it is difficult to reestablish an adequate circulation, a central venous pressure catheter may be helpful in determining the amount of volume needed. Once adequate circulation has been attained, much of the acidosis may correct spontaneously as homeostatic mechanisms are restored.

Additional steps in the management include treatment of infection and further preparation of the patient for surgery. The child should receive broad-spectrum antibiotics (e.g., ampicillin 200 mg/kg per day, gentamicin 6 to 7.5 mg/kg per day, and clindamycin 25 mg/kg per day) for additional anaerobic coverage. Once the emergency physician is certain that the airway can be controlled and the circulation is adequate, relief of pain can be accomplished by using narcotic agents (e.g., morphine 0.1 mg/kg). The patient's fever can usually be controlled by the use of a hypothermia mattress and/or acetaminophen (10 mg/kg per dose rectally). A nasogastric tube should be placed to evacuate the contents of the stomach and to drain ongoing gastric secretions. Several units of blood or packed red cells, must be prepared for the operative procedure.

Children with perforated appendicitis can deteriorate quickly. Moreover, the fever, often 40°C (104°F) or higher, may not be controlled until the intra-abdominal infection is drained. Therefore, emergency resuscitation should be quickly followed by operative intervention. At surgery the appendix is removed, the area is drained, and other appropriate treatments are given. The child will recover more rapidly if this approach is taken than if prolonged medical management is instituted and surgery delayed. Rarely, the abscess may be drained with the expectation that an appendectomy will be performed later.

Meckel's Diverticulitis with and without Perforation

Most patients with a Meckel's diverticulum are asymptomatic or, if symptomatic, have rectal bleeding from ulceration at the junction of the ectopic gastric mucosa and the normal ileal mucosa, where the diverticulum is attached. A preoperative diagnosis of an inflamed or a perforated Meckel's diverticulum is rarely made but, nevertheless, should be considered in the differential diagnosis of a perforated viscus leading to generalized peritonitis. The diagnosis is usually made in the operating room by the surgeon who finds a normal appendix, and then an exploration of the bowel finds a

diseased diverticulum 20 to 25 cm of the ileocecal valve.

Primary Peritonitis

Primary peritonitis is a bacterial infection of the peritoneal cavity, usually secondary to a bloodborne or lymphborne infection. Although rare, it can occur in children with nephrosis or cirrhosis, ascites, or other etiology, and may mimic appendicitis. Primary peritonitis is usually caused by pneumococcus, group A Streptococcus, or Gram-negative organisms. When peritonitis is a Gram-negative infection, the portal of entry is often the vagina. This occurs in girls from 5 to 10 years old in whom the cervix is usually open and the vaginal fluid is not yet acidic enough to retard the ascent of infection.

The clinical manifestations include fever, vomiting, and abdominal pain. The physical examination includes findings of peritoneal irritation. An elevated WBC count (greater than 15,000/mm ^{3}) and left shift are also seen. Often, the symptoms, signs, and laboratory findings are indistinguishable from those for perforated appendicitis; thus, the diagnosis may be made at laparotomy. If the diagnosis is suspected before surgery, the patient should undergo paracentesis. The diagnosis may be confirmed by a Gram stain showing Gram-positive cocci, followed by a positive culture. It is important to remember that children with nephrosis or cirrhosis may have appendicitis unrelated to their underlying disease.

Pancreatitis

Although acute pancreatitis is common in adults, it occurs rarely in children. The most common cause is abdominal trauma. Pancreatitis produces upper abdominal or periumbilical pain, often radiating to the back. Occasionally, the presentation is that of a patient in shock. Findings that support the diagnosis include paralytic ileus, distension, and ascites. Serum or urine amylase is usually elevated. When severe, the serum calcium is also decreased. When pancreatitis occurs in a child without a history of trauma, the physician should evaluate the patient for possible congenital abnormalities of the biliary tree or pancreatic ducts, such as abnormal insertion of the main pancreatic duct or the presence of a choledochal cyst. Surgical intervention is rarely indicated in the acute phase. However, active surgical consultation from the beginning is essential, in case the patient deteriorates in spite of maximal medical therapy (see Chapter 93). Signs of deterioration or nonresponse to therapy include persistently low serum calcium, a falling hematocrit, increasing toxicity, and deterioration of the patient's coagulation profile.

ACUTE INTESTINAL OBSTRUCTION

In any child who vomits persistently, particularly if the vomitus becomes stained with bile, the diagnosis of acute intestinal obstruction must be considered. If the obstruction is high in the intestinal tract, the abdomen does not become distended; however, with lower intestinal obstruction there is generalized distension and diffuse tenderness, usually without signs of peritoneal irritation. Only if the bowel perforates or vascular insufficiency occurs will signs of peritoneal irritation be found. If complete obstruction persists, bowel habits may change, leading to complete obstipation of both flatus and stool. All patients with suspected bowel obstruction should have radiographs of the abdomen in supine, upright, and prone cross-table lateral views. In patients with acute mechanical bowel obstruction, multiple dilated loops are usually seen. Fluid levels produced by the layering of air and intestinal contents are seen in the upright or lateral decubitus radiographs (Fig. 118.3).

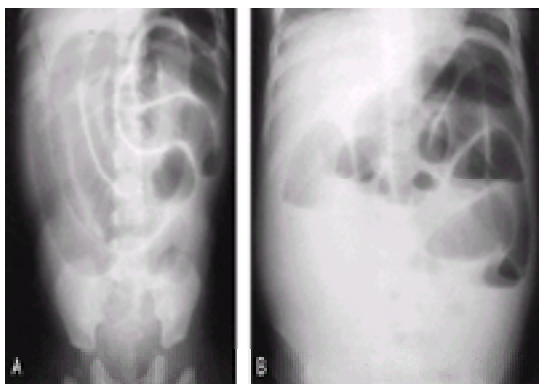


FIGURE 118.3. A. Small bowel obstruction. Numerous dilated small bowel loops occupy the midabdomen and have a stepladder configuration. Minimal air is seen in the rectum. **B.** Same patient as in **A.** The upright abdominal roentgenogram shows numerous dilated loops in the small bowel with differential fluid levels in one loop indicating mechanical bowel obstruction.

Intussusception

Background

Intussusception occurs when one segment of bowel telescopes into a more distal segment. This is the leading cause of acute intestinal obstruction in infants and occurs most commonly between 3 and 12 months of age. The most common intussusception is ileocolic but the small bowel may intussuscept into itself. Often, it will be ileoileal at a location close to the cecum. Typically, this small bowel intussusception then prolapses through the ileocecal valve (Fig. 118.4). The intussusception continues through the colon a variable distance, occasionally as far as the rectum, where it can be palpated on rectal examination. Colocolic intussusceptions are very rare. In infants the lead point for the intussusception may be hypertrophied Peyer's patches. In children older than 2 years of age, a specific lead point, such as a polyp, a Meckel's diverticulum, a duplication, or a tumor, is much more likely. A diarrheal illness, viral syndrome, or

Henoch-Schönlein purpura may be a preceding illness several days to a week before the onset of abdominal pain and obstruction.

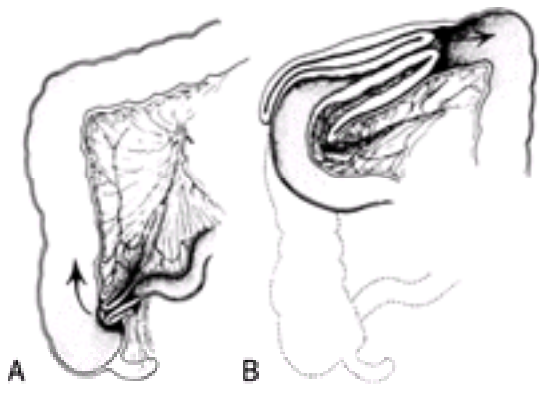


FIGURE 118.4. Ileocolic intussusception. **A.** Beginning of an intussusception in which terminal ileum prolapses through ileocecal valve. **B.** Ileocolic intussusceptum continuing through the colon. This can often be palpated as a mass in the right upper quadrant.

Clinical Manifestations

The main manifestation of intussusception is crampy abdominal pain. This symptom may have been preceded by the symptoms and signs of a viral gastroenteritis or even an upper respiratory infection. Gradually, the child becomes more irritable and anorectic and may vomit. The pattern of pain in a child with an intussusception is often consistent and characteristic, and the diagnosis is suggested strongly if a history of episodic pain is obtained. The pain may cause the infant to cry out with intermittent spasms, but the child then appears to be perfectly comfortable or only slightly irritable between these episodes. More often, lethargy is a typical sign that occurs between the episodes of pain. The infant may become still and pale and exhibit an almost shocklike state because of the intense visceral pain. At times, patients with intussusception have been misdiagnosed as being in the postictal state.

The localized portion of the intussusception leads to either partial or complete obstruction and generalized abdominal distension. In some cases, the intussuscepted mass can be palpated as an ill-defined sausage-shaped structure if the abdomen is not too distended. This mass is most often palpable in the right upper quadrant.

When children arrive in the ED early in the course of intussusception, there is often no history of having passed a currant jelly stool, although blood may be seen on the examining finger. However, the absence of bloody stools should not preclude making the diagnosis of a possible intussusception, and a barium enema should be obtained.

As the bowel becomes more tightly intussuscepted, the mesenteric veins become compressed, while the mesenteric arterial supply remains intact. This leads to the production of the characteristic currant jelly stool, which may be passed spontaneously or found on the rectal examination. However, development of melena may occur fairly quickly, and this fact reinforces the need for a rectal examination in a child with unexplained abdominal symptoms. Once the intussusception has become tight, even the arteries are occluded by the pressure of entrapment. At this point, the bleeding lessens, but the bowel can become gangrenous and even perforate, leading to peritonitis.

Management

The patient should be prepared by inserting an IV line and a nasogastric tube. IV fluids should be given to correct dehydration. Nasogastric suction minimizes the risk of vomiting and aspiration. Once blood has been sent to the laboratory for CBC, BUN, serum electrolytes, and a crossmatch, the patient should be taken to the radiology department.

Plain film roentgenographic findings of intussusception are variable and depend primarily on the duration of the symptoms and the presence or absence of complications. In early cases, a normal gas pattern is seen. In the patient with symptoms longer than 6 to 12 hours, flat and upright films often show signs of intestinal obstruction, including distended bowel with air–fluid levels (Fig. 118.3). Occasionally, the actual head of the intussusception can be seen on a plain film as a soft-tissue mass. If the plain film shows the presence of feces in the right colon, it is likely that the child has intussusception.

In recent years, hydrostatically controlled barium enema reduction has been a successful therapy in more than 50% of cases (Fig. 118.5). Strict guidelines must be adhered to so that perforation is avoided when this method is attempted. The barium column should be no higher than 3 feet above the abdomen, and manual palpation of the abdomen during the study is contraindicated. Often an IV dose of morphine 0.1 mg/kg or the use of another sedative is helpful to relax the child during the study or if there is difficulty in accomplishing the reduction. The full reduction of the intussusception is confirmed only when there has been adequate reflux of barium into the ileum. Otherwise, only the ileocolic component of an ileocolic intussusception may be reduced, leaving the ileoileal intussusception unreduced. A 24-hour follow-up film should be taken to determine whether the intussusception has recurred.

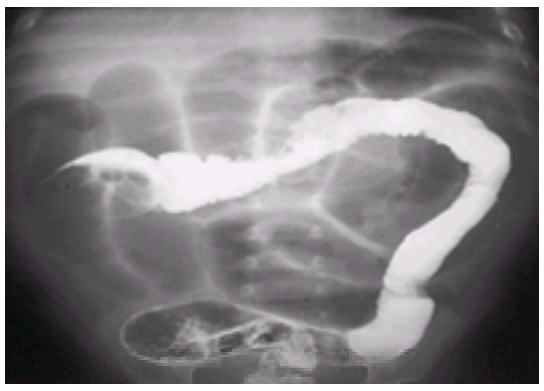


FIGURE 118.5. Ileocolic intussusception. Barium enema shows the intussusception as the filing defect within the hepatic flexure surrounded by spiral mucosal folds. Significant distended small bowel represents distal small bowel obstruction.

Recent series have described reduction of intussusception by air insufflation rather than barium. This may be a safer method because if a perforation is present the peritoneal cavity is not contaminated with barium. Many pediatric radiology departments in this country are now evaluating this method of air reduction.

A barium enema reduction is contraindicated if free peritoneal air is present. In the seriously ill infant with signs of peritonitis or a frank small bowel obstruction, the diagnosis of intussusception should be made with isotonic water-soluble contrast media with no attempt at reduction. The reduction in such infants should be performed surgically to prevent the complication of barium peritonitis after perforation of the colon.

Many youngsters with intussusception require emergency surgery, especially if the intussusception has been of long duration or the child shows evidence of gangrenous bowel, including high fever, leukocytosis, significant distension, and general toxicity. If a barium enema seems safe and appropriate, the operating room should be placed on standby and the operating team should be ready to commence immediate surgery if complications develop or if the hydrostatic reduction by barium enema is unsuccessful. Preoperative preparation and resuscitation begins in the ED and continues during the course of the barium enema. Undue delay may well result in gangrene of the entrapped bowel. The moment surgery is decided on, the patient should receive broad-spectrum IV antibiotics.

The recurrence rate after barium enema reduction ranges from 1 to 3%. When there is a recurrence, a second attempt at reduction may be done by barium enema. This is usually successful in most cases, but with a third episode of intussusception, an exploratory laparotomy must be done. Recurrences are more common in older children and may be caused by a lead point such as a Meckel's diverticulum, an intestinal polyp, or an intraluminal tumor such as lymphoma. Therefore, it may be wise in an older child to operate with the first recurrence.

Incarcerated Inguinal Hernia

Incarcerated inguinal hernia is a common cause of intestinal obstruction in the infant and young child. Approximately 60% of incarcerated hernias occur during the first year of life. Incarceration occurs more often in girls than in boys but usually involves the ovary rather than the intestine. Often, the patient or family has no previous knowledge of the presence of a congenital hernia. Incarceration does not necessarily mean that the nonreducible portion of intestine is compromised or gangrenous. However, strangulation can occur within 24 hours of a nonreduced incarcerated hernia because of progressive edema of the bowel caused by venous and lymphatic obstruction. This obstruction then leads to occlusion of the arterial supply with resulting necrosis of the bowel and perhaps perforation.

The clinical presentation of a child with an incarcerated hernia is usually irritability, crying because of pain, vomiting, and occasionally abdominal distension. A firm, discrete mass can be palpated at the internal ring and may or may not extend into the scrotum. Occasionally, the testicle may appear dark blue because of pressure on the spermatic cord causing venous congestion, and in a prolonged incarceration, the testicle may be infarcted. Intestinal obstruction may develop quickly and an abdominal radiograph shows gas-filled loops of intestine in the scrotum.

It is often difficult to differentiate a tense hydrocele in the scrotum from an incarcerated hernia. If the child has had a hydrocele, a sudden increase in fluid in the tunica vaginalis may produce discomfort and the concern is that the child has developed an incarcerated hernia. However, it is uncommon for a hernia to appear in the presence of a communicating hydrocele because of the narrowness of the patent processus vaginalis that is associated with the hydrocele. The acute hydrocele presents only in the scrotum but may extend somewhat up into the inguinal canal. However, no mass can be felt in the area of the internal ring, indicating that no intestine is exiting from the ring.

Unless the child is extremely ill with signs of intestinal obstruction or toxic from gangrenous bowel, a manual reduction of the incarcerated hernia should be attempted. The child should be sedated with morphine 0.1 mg/kg intravenously. The mother should then cuddle the baby until it relaxes and falls asleep. An older child may be placed in the Trendelenburg position to allow gravity to facilitate the reduction. Once the child is asleep, gentle manipulation of the incarcerated mass should be attempted. Mild pressure should be exerted at the internal ring with one hand, while the other attempts to squeeze gas or fluid out of the incarcerated bowel back into the abdominal cavity. If the reduction is unsuccessful, the child should be taken immediately to the operating room.

After the hernia has been reduced manually, the child may be admitted for observation but not immediate repair. The hernia sac and spermatic cord are edematous after a reduction, making the repair difficult. Usually, it is done 24 hours after admission. In a youngster who vomits persistently after a manual reduction of an incarcerated hernia, the possibility that the bowel has not been entirely reduced, causing continued obstruction, must be considered. Rarely should a child

be sent home after a manual reduction unless the parents are properly informed concerning signs of recurrence or intestinal obstruction and they are thoroughly reliable (see Inguinal Hernias, p. 1535).

Incarcerated Umbilical Hernia

Incarceration of an umbilical hernia is rare. If present, there is a persistent and tender bulge in the umbilical hernia sac. If the incarceration is of short duration, a gentle effort might be made to reduce it manually, but it often is necessary to prepare the child for urgent surgery. At the time of surgery, the loop of incarcerated bowel should be inspected, rather than letting it drop back into the abdominal cavity, to be certain there has been no vascular impairment (see Umbilical Hernias, p. 1536).

Malrotation of the Bowel with Volvulus

Background

Malrotation of the bowel is a congenital condition associated with abnormal fixation of the mesentery of the bowel (Fig. 118.6). Therefore, the bowel has a proclivity to volvulize and obstruct at these points of abnormal fixation. Although malrotation with volvulus usually occurs either in utero or during early neonatal life, malrotation can be unrecognized until childhood. This is an extraordinarily dangerous situation because a complete volvulus of the bowel for more than an hour or two can totally obstruct blood supply to the bowel, leading to complete necrosis of the involved segment. When a volvulus involves the midgut, the entire small bowel and ascending colon may be lost, making the patient dependent on IV hyperalimentation for survival. The only way to prevent such a catastrophe is to have a high index of suspicion for malrotation in any child with signs of obstruction and to be prepared to get a child with a presumed volvulus to the operating room immediately.

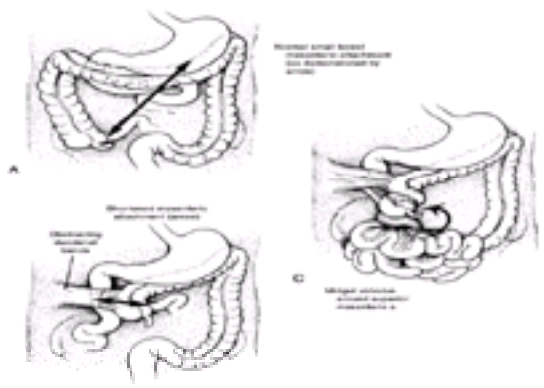


FIGURE 118.6. Malrotation with volvulus. **A.** Normal small bowel mesenteric attachment (as demonstrated by the *arrow*). This prevents twisting of small bowel because of the broad fixation of the mesentery. **B.** Malrotation of colon with obstructing duodenal bands. **C.** Midgut volvulus around the superior mesenteric artery caused by the narrow base of the mesentery.

Clinical Manifestations

Any child with bile-stained vomiting and abdominal pain may have malrotation with volvulus. The pain is usually constant and not crampy. Blood may appear in the stool within a few hours and suggests the development of ischemia and possible necrosis of the bowel. Clinically, malrotation can present in several different ways: first, and most dangerous, is the sudden onset of abdominal pain with bilious vomiting with no prior history of GI problems; a second is a similar abrupt onset of obstruction in a child who previously seemed to have “feeding problems” with transient episodes of bilious vomiting; third is a child with failure to thrive because of alleged intolerance of feedings. This last example may involve a child who has been fed a dozen different formulas and in whom the vomiting is chronic and generally not associated with pain.

On physical examination, there may be only mild distension of the abdomen inasmuch as the obstruction usually occurs high in the GI tract. On palpation, the physician may discern one or two prominently dilated loops of bowel. The abdomen may be diffusely tender and yet not have clear signs of peritonitis. On rectal examination, the presence of blood on the examining finger is an alarming sign of impending ischemia and gangrene of the bowel.

Management

The key to management is to be suspicious of malrotation and to obtain flat and upright roentgenograms of the abdomen immediately. The presence of loops of small bowel overriding the liver shadow is suggestive of an underlying malrotation. When complete volvulus has occurred, there may be only a few dilated loops of bowel with air–fluid levels. Distal to the volvulus there may be little or no gas in the GI tract. A “double-bubble sign” is often present on an upright film because of partial obstruction of the duodenum causing distension of the stomach and first part of the duodenum (Fig. 118.7A).

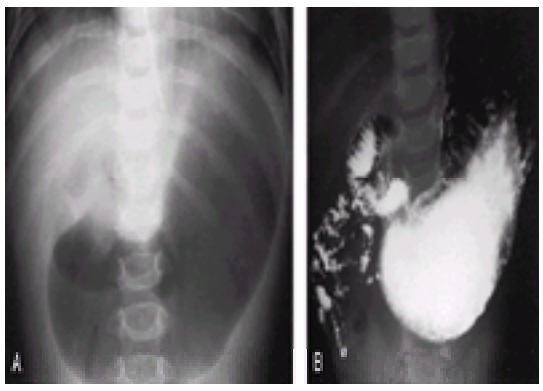


FIGURE 118.7. A. Malrotation of the bowel. Supine plain roentgenogram of the abdomen shows distended stomach and proximal duodenal loop. **B.** Same patient as in **A.** Upper gastrointestinal series shows dilated proximal duodenum with abrupt transition to normal caliber of small bowel. Abnormally placed ligament of Treitz. Proximal jejunum in the right abdomen.

When a child is being assessed for possible malrotation, an upper GI series is the study of choice. The ligament of Treitz is absent in the malrotation anomaly, so the C-loop of the duodenum is not present, the duodenum lies to the right of the spine, and the jejunum presents a coiled spring appearance in the right upper quadrant (Fig. 118.7B and Fig. 118.8). The cecum is not fixed and usually assumes a position in the right upper quadrant. However, because of its mobility, the cecum on barium enema may be seen in its normal position in the right lower quadrant. Therefore, a barium enema is not the most reliable study to rule out malrotation. In the neonate, the cecum sometimes takes a high position and this could give a false impression of malrotation.

FIGURE 118.8. Malrotation. Upper gastrointestinal study showing absence of the ligament of Treitz and coiled spring appearance of jejunum.

As in the case of a child with an unreduced intussusception, a child with a possible volvulus should be prepared for immediate surgery. The operating room and operating team should be on standby. IV fluid and electrolyte replacement should begin immediately. A nasogastric tube should be inserted and blood crossmatched. Because this entity can present even in adulthood, every physician should understand the pathogenesis and the need for surgical therapy of malrotation. If immediate transfer to a children's hospital cannot be accomplished within an hour, a laparotomy should be performed without delay.

Pyloric Stenosis

Narrowing of the pyloric canal as a result of hypertrophy of the musculature often occurs in the first-born male of a family. A familial incidence has been shown, particularly if the mother had hypertrophic pyloric stenosis as an infant. There is a male:female ratio of 5:1. The age of onset is usually 2 to 5 weeks. Rarely, the onset may be late in the second month of life. The cause of the muscle hypertrophy is unknown, but the symptoms, diagnosis, and therapy are well defined.

Clinical Manifestations

Characteristically, the infant does well, without vomiting, for the first few weeks of life and then starts regurgitating, either at the end of feedings or a few minutes later. The infant is hungry and will eat heartily immediately after such a regurgitation. The vomiting becomes more prominent and eventually becomes more forceful, called projectile vomiting. The vomitus ordinarily contains just the feeding that has been given and does not contain bile or blood. Occasionally, some mucus is in the vomitus. Infants with pyloric stenosis may also become jaundiced with the onset of the other symptoms. The hyperbilirubinemia usually improves or abates postoperatively for reasons that are unknown.

The examination of an infant is best accomplished after the infant's stomach has been emptied. With the child lying on his or her back, the examiner holds the infant's ankles and flexes the thighs at a right angle to the trunk as the mother feeds some warm sugar water with the infant's head turned to the right but not elevated. Once the infant starts to suck, the upper abdominal musculature will relax and the examiner's opposite hand can then gently palpate the upper right abdomen. Palpating under the edge of the liver in an up-and-down direction, the physician may discern a firm, fusiform, ballotable mass in the shape of an olive. If this “olive” is not felt, the stomach can be emptied with a nasogastric tube to allow easier palpation and the examination is repeated. Another diagnostic clue is the presence of prominent gastric

peristaltic waves that course from left to right across the abdomen.

If the child has vomited for an extended period, he or she will show signs of growth failure. There may be loose, hanging skin and an absence of subcutaneous tissue. The infant may take on an “old man” appearance, with wrinkled skin on the face and body. Weight gain is inadequate, which may be calculated by knowing that the average child regains birth weight by 10 days of age and thereafter 15 to 30 g (0.5 to 1 ounce) per day.

Serum electrolytes may be abnormal because of gastric losses. The potassium and chloride are low, and serum bicarbonate is high. This hypochloremic alkalosis may be profound with serum chlorides in the 65 to 75 mEq/L range. When dehydration becomes severe, the patient may then develop acidosis, indicating an advanced and even more dangerous metabolic imbalance (see Chapter 86).

Management

Infants should be hospitalized and rehydrated with appropriate fluid and electrolyte replacement. Initially, IV fluids should be 5% dextrose in normal saline. Potassium chloride (3 to 5 mEq/kg) should be added once urine output has been established. If hypotonic solutions are used, there is significant risk of causing hyponatremia (see Chapter 86). A volume of fluid should be used appropriate to the patient's level of dehydration.

If a pyloric “olive” or mass is palpable and clear gastric waves are visible, roentgenogram confirmation of the diagnosis is not needed. If the history of vomiting is not typical and a mass cannot be felt, real-time ultrasound is the first study to confirm the diagnosis. The real-time ultrasound scanning not only increases the accuracy of the diagnosis of pyloric stenosis but can also localize the “olive.” The hypertrophic pyloric muscle is seen as a thick hypoechoic ring surrounding a central echogenic mucosal and submucosal region (Fig. 118.9). The quantitative criteria for the sonographic diagnosis of hypertrophic pyloric stenosis are 1.4 cm or longer length of the pyloric canal with 0.3 cm or greater thickness of the circular muscle.

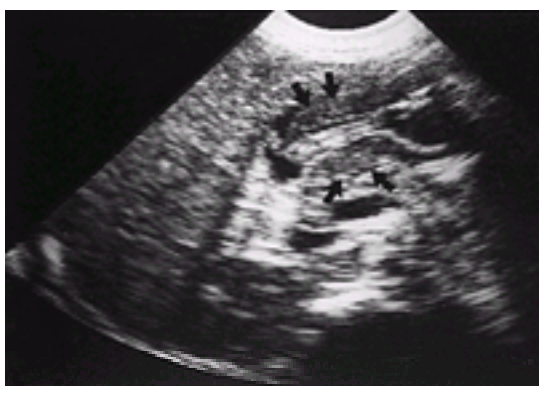


FIGURE 118.9. Hypertrophic pyloric stenosis. Ultrasonography of the abdomen shows thick pyloric muscle surrounding a centered echogenic mucosal and submucosal region *(arrows).*

If the ultrasound study does not show a hypertrophic pylorus, an upper GI series should then be done to eliminate gastroesophageal reflux, malrotation, and antral web as diagnostic possibilities. In general, pyloric stenosis can be identified by the presence of a “string sign” in the pyloric channel, seen best on oblique projections on the upper GI series (Fig. 118.10).

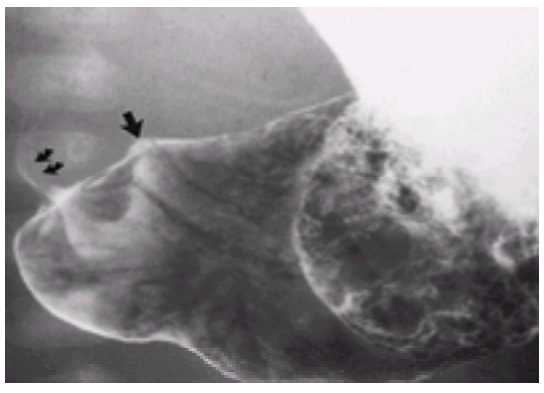


FIGURE 118.10. Pyloric stenosis. Long, narrowed, and tilting upward antropyloric canal. Parallel streaks of barium producing typical string sign with complete obstruction *(arrows)* and eccentric lesser curvature indentation pyloric tilt *(arrow).* The tilt is performed when the peristaltic wave meets the muscle mass

To lessen the risk of vomiting and aspiration, the barium should be evacuated from the stomach after the upper GI series has been completed. Surgical pyloromyotomy is a most successful form of therapy, and such infants can usually be discharged from the hospital 2 days after surgery. Some infants will have some regurgitation postoperatively as a result of a temporary relaxation of the gastroesophageal sphincter.

Postoperative Adhesions

Prior abdominal surgery or peritonitis places a child at risk for intestinal obstruction from adhesions (Fig. 118.11). Such obstruction can occur relatively early in the postoperative course or months or even years later. Suddenly and without warning, the child develops abdominal cramps, nausea, vomiting, and abdominal distension. Although most intestinal obstructions from adhesions do not jeopardize the vascularity of the bowel, occasionally a loop of intestine, caught under a fibrous band, can become gangrenous. Therefore, the diagnosis should be made quickly. All such patients need to be admitted to the hospital and evaluated by a surgeon who should direct the complete management.

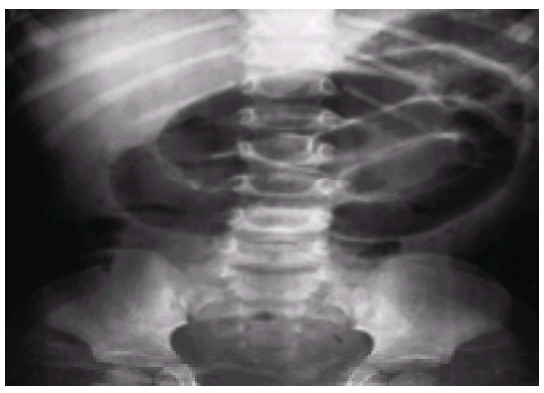


FIGURE 118.11. Dilated loops of small intestine and absence of air in lower abdomen indicating a high intestinal obstruction caused by postoperative adhesions.

CHRONIC PARTIAL INTESTINAL OBSTRUCTION

Any child with intermittent abdominal distension, nausea, anorexia, occasional vomiting, or chronic constipation or obstipation may have partial intestinal obstruction. A number of diagnostic considerations exist.

Chronic Constipation

Chronic constipation is probably one of the most common causes for abdominal pain, distension, and vomiting in children. The history, if available from a reliable parent, may attest to chronic constipation; however, occasionally, such a child is diagnosed only by palpating a large fecaloma through the intact abdominal wall or a hard fecal mass blocking the anal outlet on rectal examination. Such youngsters may have a history of encopresis and appear malnourished. Chapter 14 covers the diagnostic approach to the child with constipation.

These children should be disimpacted by instilling a generous amount of warm mineral oil into the rectum, followed by copious saline lavages using a large-caliber rectal tube with extra holes. Often, a gloved finger is necessary to break up a hard fecal mass and allow its evacuation. If the process has reached this stage, it is unlikely that the mother or father can manage it at home, and either ED or in-hospital management is necessary to clean out the bowel adequately. Once the bowel has been cleaned, it is important to institute an appropriate regimen to retrain the child and produce better bowel habits.

Aganglionic Megacolon (Hirschsprung's Disease)

In patients with Hirschsprung's disease, the parasympathetic ganglion cells of Auerbach's plexus between the circular and longitudinal muscle layers of the colon are absent. The involved segment varies in length, from less than 1 cm to involvement of the entire colon and small bowel. The effect of this absence of ganglion cells produces spasm and abnormal motility of that segment, which results in either complete intestinal obstruction or chronic constipation.

These children have a lifelong history of constipation, so it is important to obtain an accurate account of the child's stool pattern from birth. A child with Hirschsprung's disease typically has never been able to stool properly without assistance (e.g., enemas, suppositories, stimulation with the finger or thermometer). Normal stooling is not possible because of the failure of the aganglionic bowel and interval anal sphincter to relax. The child usually has no history of encopresis, as one would find in chronic functional constipation. These youngsters have chronic abdominal distension and are often malnourished. Vomiting is uncommon, as are other symptoms. Complete intestinal obstruction in Hirschsprung's disease is more likely to occur in early infancy and only rarely in the older age groups.

Table 118.3 summarizes the pertinent diagnostic features differentiating functional constipation from Hirschsprung's disease.

	Functional Constipation	Hirschsprung's Disease
Onset	≥2 years	Birth
History	Coercive training	Enemas necessary
	Colicky abdominal pain	No abdominal pain
	Periodic volume stools	Episodes of intestinal obstruction
Encopresis	Present	Absent
Abdominal distension	Absent or minimal	Present
Rectal examination	Feces-packed rectum	Empty rectum
Barium examination	Dilated rectum	Narrow segment
Motility	Normal	Abnormal
Biopsy	Ganglion cells	No ganglion cells

Table 118.3. ***Differential Diagnosis of Functional Constipation and Hirschsprung's Disease***

After flat and upright abdominal roentgenogram radiographic studies have been obtained, a properly performed barium enema with a Hirschsprung's catheter is the best initial diagnostic procedure. There should be no preparation of the bowel. Ideally, the rectum should not be stimulated by enemas or digital examination for 1 to 2 days before the procedure. The key to diagnosis is seeing a “transition zone” (Fig. 118.12) between the contracted aganglionic bowel and the proximal dilated ganglionated bowel. Stimulation of the rectum shortly before the study may result in decompression of the proximal bowel, with loss of definition of the transition zone. When a clear-cut transition zone is seen, it is not necessary to fill the colon with barium more than 12 to 18 inches above the transition point. It is important, however, not to empty the colon of barium at the end of the study. The presence of retained barium above the transition point 24 hours later strongly suggests the diagnosis of Hirschsprung's disease.

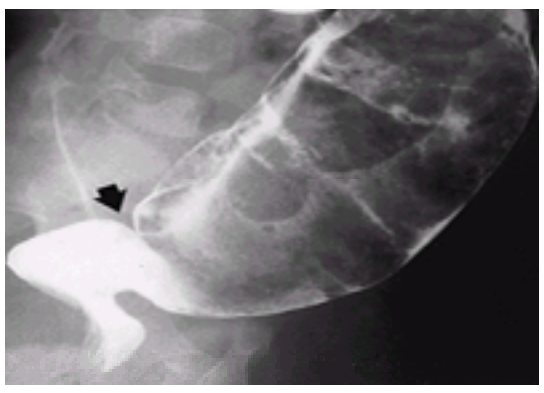


FIGURE 118.12. Hirschsprung's disease. Barium enema studies in lateral view show transition zone *(arrow)* with narrow rectum but dilated sigmoid colon.

Anorectal manometry to determine the presence or absence of relaxation of the internal anal sphincter is helpful in establishing the neurogenic dysfunction of the bowel. Barium enema studies and manometry are clearly complementary in the diagnosis of Hirschsprung's disease. However, rectal manometric studies are more reliable than radiologic methods for short aganglionic segments that are usually not apparent on barium enema studies (Fig. 118.13). Manometric studies are not dependable in infants less than 3 weeks of age. If the barium enema and anal manometry studies indicate Hirschsprung's disease, rectal biopsy is not necessary to confirm the diagnosis.

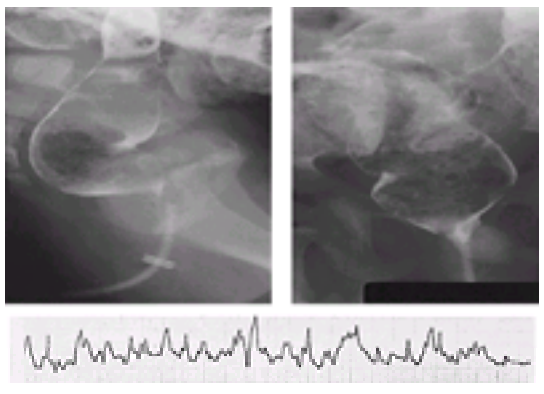


FIGURE 118.13. Ultrashort Hirschsprung's disease. Barium enema and rectal manometry studies in a 2-year-old child. Barium enema study in anteroposterior and lateral view. There is no transition zone but the sigmoid is somewhat dilated, suggesting idiopathic megacolon **(A).** Rectal manometry 2 days later shows no relaxation of the internal sphincter pressure after rectal stimulation **(B).** The tracing **(C)** shows no decrease in anal sphincter pressure, indicating Hirschsprung's disease. At surgery, low-segment Hirschsprung's disease was found.

In children of all ages, an adequately performed suction mucosal biopsy of the rectum 2 cm or more above the dentate line can be reliable in diagnosing Hirschsprung's disease. Because of the complicated evaluation and management of

this disease, referral to a pediatric surgeon is recommended.

Duplications

Duplications occur anywhere from the mouth to the anus and produce a variety of symptoms. In the abdomen, there may be a noncommunicating cyst that gradually fills up with secretions and compresses the adjacent normal bowel, producing a palpable abdominal mass or chronic intestinal obstruction. An occasional duplication has a communication, particularly at its distal end, that produces a large mass that may be confused with the fecal mass felt with aganglionic Hirschsprung's disease. Rarely, a marginal ulcer resulting from ectopic gastric mucosa may occur, and this produces painless bleeding. After appropriate radiographic diagnosis, surgery is indicated; the surgical procedures vary, depending on the locations, size, and communications of the anomaly.

Inflammatory Bowel Disease

The older child or adolescent may develop either Crohn's disease or ulcerative colitis (see Chapter 93), and this must be included in the differential diagnosis of chronic intestinal obstruction. Usually, the child has a history of changing bowel habits, with mucus or blood in the stools, chronic abdominal pain, and weight loss. Chapter 93 covers inflammatory bowel disease in detail.

DISEASES THAT PRODUCE RECTAL BLEEDING

Rectal bleeding is an alarming symptom. It is important to determine the quantity of bleeding and whether the blood is on the outside of the stool or mingled with it. A “tarry” stool suggests a source of bleeding in the proximal portion of the GI tract and bright red blood a more distal origin (Fig. 118.14). Occasionally, the child will have an episode or two of blood with his bowel movements. All patients with rectal bleeding should have a rectal examination. Those with significant hemorrhage require flexible colonoscopy and a contrast enema. In some patients, no definite diagnosis may be reached despite extensive studies. In any patient with significant bleeding, however, surgical consultation is indicated. Chapter 30 and Chapter 93 further discuss the diagnosis and management of patients with GI bleeding.

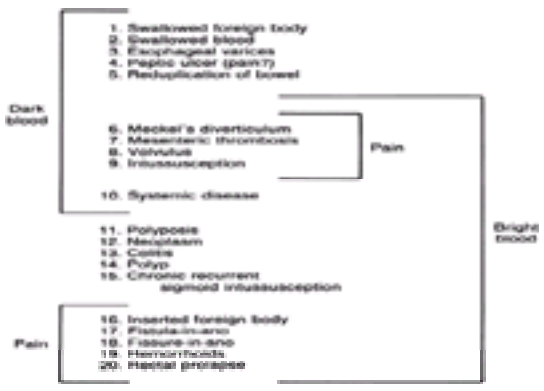


FIGURE 118.14. Causes of rectal bleeding in children.

Fissures

An anal fissure is probably the most common cause of bleeding, especially in infants. However, fissures may occur at any age. The child usually has a history of passing a large, hard stool with anal discomfort. Often, the child has a history of chronic constipation with progressive reluctance to pass stool because of the associated discomfort. If bleeding occurs, it usually involves streaking of bright red blood on the outside of the stool, or red blood on the toilet tissues. The diagnosis can easily be made by inspection or anoscopic examination and appropriate measures taken to relieve the chronic constipation (see Chapter 14). Children may be helped by sitz baths and the application of local anesthetic ointments such as Nupercainal. Rarely does a child require hospitalization or surgery.

Juvenile Polyps

Older infants and children can develop either single or multiple retention polyps. Usually, the polyps occur in the lower portion of the colon and can often be palpated on rectal examination. Polyps bleed, but they rarely cause massive hemorrhage. They may intermittently prolapse at the anus or on occasion come free and be passed as a fecal mass associated with bleeding. Colonic polyps may be lead points for intussusception. Usually, however, polyps are asymptomatic except for the associated bleeding. These are not premalignant lesions, and they tend to be self-limiting (Fig. 118.15).

FIGURE 118.15. Juvenile polyp. Double air-contrast barium enema shows a single polyp with long stalk in transverse colon *(arrow).*

If a polyp can be felt on rectal examination or viewed through the sigmoidoscope, it may be safely removed from below. Polyps beyond the reach of the sigmoidoscope should be removed by colonoscopy.

Familial Polyposis

Families with multiple adenomatous colonic polyps are rarely encountered. Bleeding is rare. More often, a colitis type of mucous discharge is present. Rectal examination and endoscopy reveal multiple “cobblestone” sessile polyps. These individuals are at risk for neoplasia because these are premalignant adenomatous polyps. The child should be referred to a pediatric surgeon and gastroenterologist for evaluation and long-term management.

Meckel's Diverticulum

Two percent of the population is born with a Meckel's diverticulum. This is the most common omphalomesenteric duct remnant. The diverticulum is usually located 50 to 75 cm proximal to the terminal ileum. Only 2% of persons with a Meckel's diverticulum manifest any clinical problems. The most common complication of a Meckel's diverticulum is a bleeding ulcer. Ectopic gastric mucosa in such patients is usually present in the diverticulum. The acid secretion produces an erosion at the junction of the normal ileal mucosa with the ectopic mucosa. Currant jelly stools are supposed to be classic for this type of bleeding, but it can be severe at times. Other modes of presentation include diverticulitis, perforation with peritonitis, or intussusception as a result of the diverticulum's serving as a lead point.

Barium studies usually fail to outline a Meckel's diverticulum. The imaging modality of choice for detection of ectopic gastric mucosa in a bleeding Meckel's diverticulum is nuclear scintigraphy. A well-defined focal accumulation of radionuclide (99m-technetium pertechnetate) usually appears at or about the same time as activity in the stomach and gradually increases in intensity (Fig. 118.16). A duplication cyst with gastric mucosa shows the same focal accumulation of radionuclide. Preoperative differentiation between two lesions as a cause of GI bleeding is not important. The accuracy of scintigraphy in detection of ectopic gastric mucosa in Meckel's diverticula is approximately 95%. False-negative results may rarely occur in patients with rapidly bleeding Meckel's diverticula and with those diverticula that do not contain gastric mucosa. Ultrasound and magnetic resonance imaging (MRI) are of no use in finding a Meckel's diverticulum.

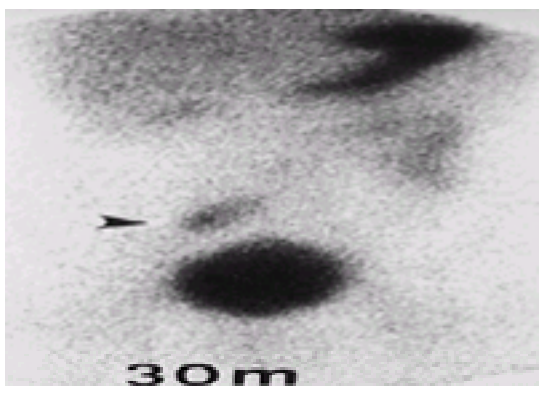


FIGURE 118.16. Meckel's diverticulum. Anterior image at 30 minutes shows an oval focal accumulation of ^{99m}Tc-pertechnetate in the right lower quadrant of the abdomen *(arrowhead).*

In any child with a major rectal bleed and a negative scan, further workup, including an arteriogram if the bleeding continues to be active or colonoscopy when the bleeding is not active, is required.

Henoch-Schönlein Purpura

Henoch-Schönlein purpura (see Chapter 87) is a vasculitic disorder that can produce asymptomatic rectal bleeding or abdominal pain and hematuria. Usually there is visible evidence of vasculitis on the skin surface, but a patient initially may have only abdominal pain. Occasionally, a child will develop a small bowel intussusception from a submucosal hemorrhage that is acting as a lead point. Other common manifestations are purpuric rash, fever, and joint pains (see

Chapter 87).

Other Causes of Rectal Bleeding

Other causes of rectal bleeding include intestinal vascular malformations, intussusception, duplications, inflammatory bowel disease, peptic ulcer with bleeding, and portal hypertension with bleeding varices. These topics are covered elsewhere in this chapter, in Chapter 93 and in Chapter 30.

INTRA-ABDOMINAL MASSES

Background

Intra-abdominal masses in children may be benign, but unfortunately, there are also a number of malignant ones. Abdominal masses may be silent, even after the tumor has reached a large size. Often, they are first recognized by the parent or the child's caretaker while the child is being undressed or bathed.

It is difficult to feel an intra-abdominal mass and outline its limits and its degree of mobility if an infant or child is crying. If the child is fed a bottle with his or her head turned to one side, the abdominal musculature will relax. The physician should then make an effort to palpate the intra-abdominal contents carefully. These masses are fragile and prone to rupture. Therefore, palpation of the mass should be done gently and should be strictly limited to as few examiners as possible.

Retroperitoneal masses tend to be fixed, whereas masses attached to the mesentery or omentum are mobile and may be shifted to different locations by the examiner. Pelvic masses are commonly fixed and often can best be felt by rectal examination. A presacral mass may narrow the rectum and produce constipation. Abdominal masses present with various characteristics and may be smooth, nodular, cystic, or rock hard.

Initial evaluation in the ED may include flat and upright abdominal films. If, after such an examination, the origin of the mass is unclear or suggests a neoplasm, the patient should be admitted and a workup done without delay. Observation has no place in dealing with unexplained abdominal masses in children.

The role of diagnostic imaging is to identify the precise anatomic location and extent of the pathologic process with a minimal number of procedures. It should be stressed that ultrasonography, nuclear scintigraphy, CT, and MRI have dramatically changed the traditional approach to pediatric imaging. The general location of a mass, with or without calcification, can be confirmed by plain abdominal roentgenograms. Real-time ultrasonography has become increasingly important initial imaging because it does not use radiation and yet is diagnostically accurate. Ultrasonography can differentiate a cystic flank mass (Fig. 118.17) that could be a hydronephrotic kidney from a solid tumor such as an adrenal neuroblastoma and thus facilitate the proper referral of the child to either a urologist or a pediatric surgeon. CT is superior to other modalities for anatomic detail and obtains an entire anatomic section of tissue, which aids in determining the precise extent of disease. CT provides anatomic and physiologic information about organs and vascular structures despite overlying gas and bones. Nuclear scintigraphy has a limited role in the evaluation of pediatric abdominal masses. Renal scintigraphy is superior to excretory urography for quantitating renal function. Angiography is indicated for an abdominal mass only if a precise knowledge of segmental vascular anatomy is required or if interventional techniques are contemplated.

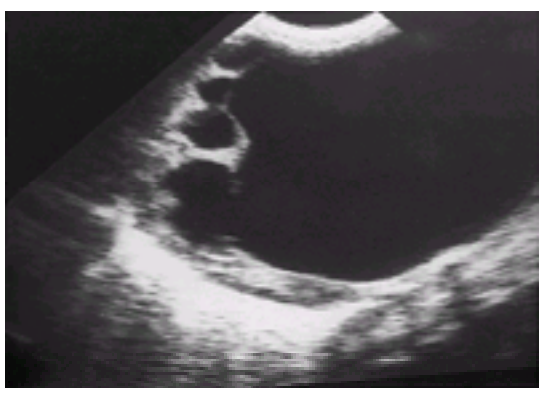


FIGURE 118.17. Ureteropelvic junction obstruction. A newborn with left flank mass. Ultrasonography of the left flank shows dilated pyelocalyceal system. The communicating dilated collecting systems are seen in the periphery of the significantly dilated renal pelvis.

Excretory urography has traditionally been the radiologic choice for abdominal masses in children. However, ultrasound and CT have forced reassessment of this classic approach. In approaching abdominal masses in infants and children, the plain film with either real-time ultrasonography and excretory urography or CT can define the origin of abdominal masses.

Sacrococcygeal Teratoma

The presacral sacrococcygeal teratoma is the most common tumor of the caudal region in children and is more common in females than in males (4:1). Most tumors are benign and are noted at birth. However, tumors in patients beyond neonatal age have a higher incidence of malignancy. Radiography shows a soft-tissue mass that arises from the ventral

surface of the coccyx. Calcifications are present in 60% of presacral sacrococcygeal teratoma and are more common in benign tumors. Ultrasound confirms whether presacral sacrococcygeal teratomas are cystic, solid, or mixed. Also, ultrasonography is valuable to determine impingement on the urinary tract. CT is helpful in confirming the diagnosis, particularly in older children, and demonstrates the content of a tumor as well as its extent and bone anomalies. Tumors with more solid components are more often malignant than those with more cystic components (Fig. 118.18).

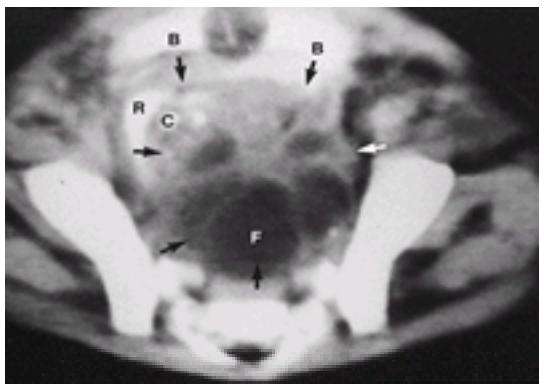


FIGURE 118.18. Presacral teratoma. Computed tomographic section of pelvis with contrast medium enhancement shows a large cystic mass *(arrows).* The mass contains both fat and calcification, and displaces the rectum anteriorly and laterally, and the bladder anteriorly. *B,* bladder with Foley catheter; *C,* calcification; *F,* fat; *R,* rectum.

Nonmalignant Intra-Abdominal Masses

Fecaloma

A lower abdominal mass, particularly one on the left side, is most often related to retained stool and is more often associated with chronic functional constipation than with Hirschsprung's disease. If a mass is found, a careful review of the bowel habits is important. If an abdominal mass is a fecaloma, a large bolus of stool usually can be felt on rectal examination just inside the anus. The evaluation of the impaction, and irrigation of the upper sigmoid colon, should cause the mass to disappear. See Chapter 14 for the causes of constipation.

Ovarian Masses

Simple ovarian cysts and solid teratomas are not uncommon and may be asymptomatic even though they have reached a large size. Occasionally, the child presents with urinary complaints from the pressure on the bladder or urethra. Granulosa cell tumors of the ovary produce precocious puberty because they are hormonally active tumors. They may be malignant. The sudden onset of severe abdominal pain may indicate a torsion of an ovarian mass on a slender pedicle, with resultant infarction. Granulosa cell tumors tend to give the impression of rising up from the pelvis and on occasion lie in the midabdomen or even the upper abdomen. When they are still partially within the pelvis, they usually can be felt by rectal examination. Most ovarian masses are smooth and nontender. Radiographs may show calcification in about half of patients with teratomas (Fig. 118.19). Because an occasional ovarian tumor is malignant in children, children with ovarian masses should be promptly evaluated and prepared for surgery (see Chapter 94).

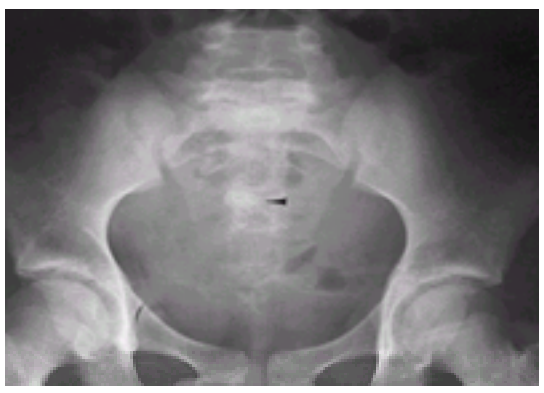


FIGURE 118.19. Ovarian dermoids. Note calcification *(arrows)* in superior aspect of a large pelvic mass in a 12-year-old girl.

Omental Cysts

Omental cysts are rare, are usually asymptomatic, and can reach gigantic size. It is often difficult to differentiate an omental cyst from ascites. There are a number of cases on record in which omental cysts have been tapped, on the assumption that they were ascitic fluid. Smaller cysts are more mobile and can be pushed freely into all quadrants of the abdomen. If a cyst volvulizes on its pedicle or has bleeding within it, it may cause abdominal pain or tenderness. Elective surgical excision is indicated.

Mesenteric Cysts

Mesenteric cysts can occur anywhere in the mesentery but are most common in the mesentery of the colon. They tend to be multilocular and are often discovered during a routine examination or after an episode of abdominal trauma with enlargement from bleeding. They are benign, but surgical therapy is indicated, both to confirm the diagnosis and to prevent complications. They can usually be removed with sparing of the bowel, or they can be marsupialized into the general peritoneal cavity where the fluid is absorbed.

Duplications

GI duplications within the abdomen can occur anywhere along the greater curvature of the stomach, the lesser curvature of the duodenum, or the mesenteric side of either the small or large intestines. They can also be pararectal, rising up out of the pelvis. Duplications that produce abdominal masses are either noncommunicating, and hence gradually enlarge, or communicating in that their secretory lining has a distal communication with the true lumen of the bowel. Except for the rare occurrence of massive rectal bleeding in a child with a communicating duplication, most duplications do not present as emergencies. Instead, they present in children either as unexplained abdominal masses or with symptoms of intermittent colic, resulting from partial obstruction of the true lumen of the adjacent bowel. The exact diagnosis is often unclear until the time of laparotomy.

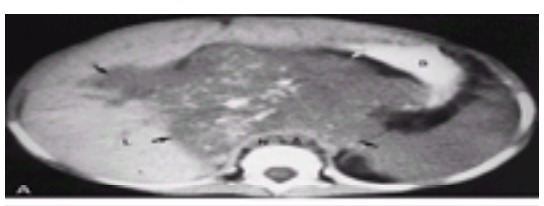
Malignant Intra-Abdominal Masses

About 50% of the solid malignant tumors seen in children occur within the abdominal cavity. They generally occur in the retroperitoneum. The most common is neuroblastoma, followed by Wilms' tumor and rhabdomyosarcoma. Other unusual tumors, such as embryonal cell carcinomas (yolk sac tumor) and lymphosarcoma, also occur in young children. Chapter 100 covers oncologic emergencies. As with most malignant tumors, early diagnosis and treatment provide the best prospects for a cure. Therefore, the physician must have a high index of suspicion for malignancy in any child with a mass or unexplained GI or genitourinary symptoms.

Neuroblastoma

Neuroblastoma most often occurs as a tumor in the left or right adrenal gland, but it can develop anywhere along the sympathetic chain or in the pelvis. It has even been found intrarenally. It has the ability to grow extensively, often crossing the midline of the abdomen, and enveloping key vascular and visceral structures. The best cure rates are generally in children who are less than 1 year of age at the time of diagnosis and in whom the tumor is still localized to the point of origin. In such favorable cases, the tumor can be totally excised. When widespread dissemination occurs, complete resection is unwarranted because of the risk to other vital structures.

CT is superior to ultrasonography for clearly defining morphologic details of neuroblastoma, such as calcifications, and precise extent of tumor by direct spread or lymphatic metastasis. CT with contrast enhancement demonstrates precise anatomy, as well as renal function and organ vascularity. The CT characteristics of neuroblastoma include irregular shape, irregular margins, lack of well-defined capsules, and mixed low-density center. Neuroblastoma often displaces surrounding organs and encases vessels. Prevertebral midline extension is common. There are calcifications in at least 75% (Fig. 118.20). Ultrasonography has limitations in accurately determining tumor margins or local extension. Therefore, CT is the modality of choice for imaging neuroblastoma.



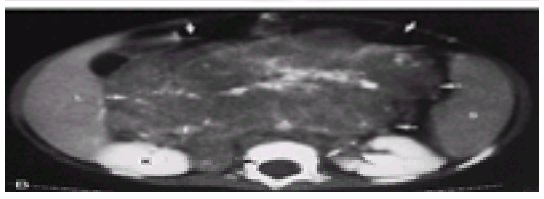


FIGURE 118.20. A. Celiac axis neuroblastoma. Computed tomographic (CT) section of the abdomen shows a large lobulated mass with multiple flake of calcification displacing stomach and the liver *(white arrows).* Note presence of retrocrural node *(black arrows). A,* aorta; *L,* liver; *N,* node; *S,* stomach. **B.** Celiac axis neuroblastoma. Enhanced CT section of the abdomen at level of the kidney shows a large lobulated mass with irregular margins and calcification displacing the right kidney inferoposterior and laterally. Note encased inferior vena cava (IVC) and aorta. The IVC is displaced laterally and ventrally and to the right the superior mesenteric artery and celiac axis are completely surrounded by the mass. *A,* aorta; *I,* IVC; *K,* kidney; *L,* liver; *S,* spleen; *white arrows,* mass.

Wilms' Tumor

Wilms' tumor is the most common intrarenal tumor seen in children. Great progress has been made in the last several decades in its management. The tumor can reach a gigantic size before its discovery. Wilms' tumor should be considered

in any child who has hematuria even if he or she has a history of trauma.

A solid renal mass demonstrated by ultrasound in infants and children is usually a Wilms' tumor. Because of the high frequency of tumor extension into the renal veins and inferior vena cava, these vascular structures should be examined by real-time ultrasound. Venous extension is diagnosed when echogenic filling defects are identified within a renal vein, the inferior vena cava, or the right heart. Because Wilms' tumors are usually large and expansive, the inferior vena cava often is extrinsically displaced by the tumor mass. CT with bolus contrast enhancement may be required for confirmation of equivocal invasion in a patient suspected of having Wilms' tumor. After abdominal real-time ultrasound, excretory urography should be performed to evaluate the function of the kidneys. Angiography seldom provides additional diagnostic information. CT defines the presence of an intrarenal mass and extent of tumor, visualizes vascular structures, identifies nodal involvement, defines internal hemorrhage and necrosis, evaluates the presence or absence of liver metastases, and images the opposite kidney. Also, CT can define whether a tumor is initially nonresectable or bilateral (Fig. 118.21). CT may be extremely helpful in following the response to chemotherapy. Chest CT is also performed at the initial evaluation to identify pulmonary metastases. Bone scintigraphy is helpful in diagnosing metastases. Surgical removal of Wilms' tumor should not be delayed. Postoperative management with chemotherapeutic agents and selective irradiation is important. These youngsters should be cared for by a team of specialists with thorough knowledge of ancillary forms of therapy.

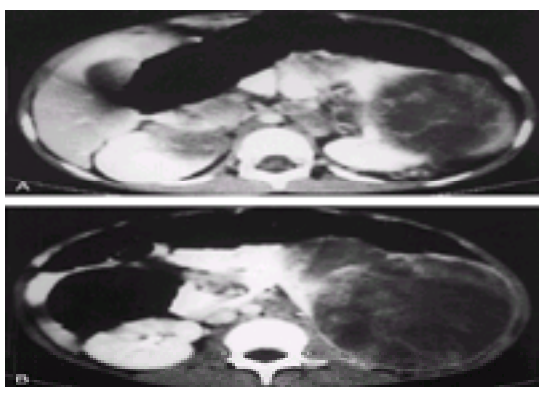


FIGURE 118.21. A. Bilateral Wilms' tumor. A 5-year-old girl with left flank mass. Computed tomographic (CT) sections of the upper abdomen with contrast medium enhancement show a necrotic mass arising from superior aspect of the left kidney. Note a small mass in the superior medial aspect of the right kidney. **B.** Bilateral Wilms' tumor (same patient as in **A** CT section of the abdomen with contrast medium enhancement shows extent of the large necrotic left Wilms' tumor with periaortic adenopathy.

Rhabdomyosarcoma

Rhabdomyosarcoma can occur anywhere in the abdomen or pelvis where there is striated muscle. Tumors are particularly common in the pelvis, involving the prostate, uterus or vagina, and retroperitoneal structures, but they have also been found in the common bile duct and other unusual sites. These tumors can reach a large size before they become symptomatic and each must be managed individually, depending on the site of origin, extent of growth, and the degree of spread. Modern selective therapy has greatly improved the survival rate of this highly malignant tumor.

Hepatoma

Hepatomas are fortunately rare. They are usually seen in older infants and young children. Hepatoblastoma, more common than hepatoma, is often discovered accidentally when the child is undressed for a bath. The child may feel and act well, yet the tumor is already of a formidable size when first noted. Differential diagnosis should include hemangioendothelioma, hamartoma, and renal and adrenal tumors, especially if they occur on the right side.

An important application of CT in the examination of patients with hepatomas involves a complementary role with MRI and angiography in the assessment of potential resectability. The ability of CT to show the location and extent of a tumor's relation to the intersegmental fissure makes it a useful procedure to determine the feasibility of this operation. MRI without IV contrast is more sensitive than dynamic CT for the detection of liver lesions. It possesses the imaging qualities of both ultrasound and CT scan in that it can image the liver in any plane and displays the vascular structures without the need for contrast media (Fig. 118.22A). Angiography is required before surgery to provide the surgeon with knowledge of the vascular supply of the tumor and the arterial anatomy of the liver. More recently, a new tool for angiographic imaging is magnetic resonance angiography (Fig. 118.22B). This method eliminates the need for arteriography and gives the surgeon an excellent view of the extent of the tumor with its vasculature and possibility of resection. CT and MRI are reliable methods to observe the response or progression of tumors in patients who undergo nonoperative therapy (Fig. 118.23).

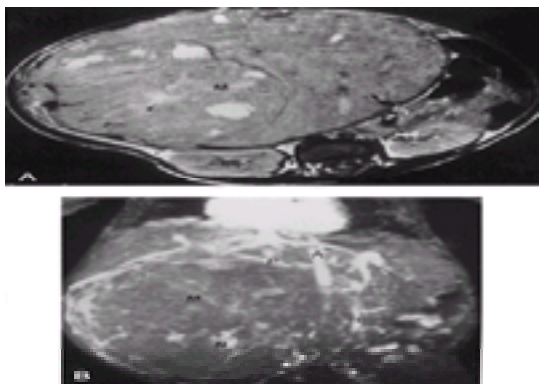


FIGURE 118.22. A. Hepatoblastoma in a 2-month-old boy. Axial T1 magnetic resonance imaging (MRI) shows a solid mass *(M)* occupying entire liver, gallbladder *(arrow),* and right kidney *(K).* **B.** Coronal magnetic resonance angiography shows liver mass *(M),* with stretching of the hepatic vessels and multiple area of neovascularity *(N).* Note marked stretching and displacement of the inferior vena cava *(I)* with patent portal vein *(P). A,* aorta.

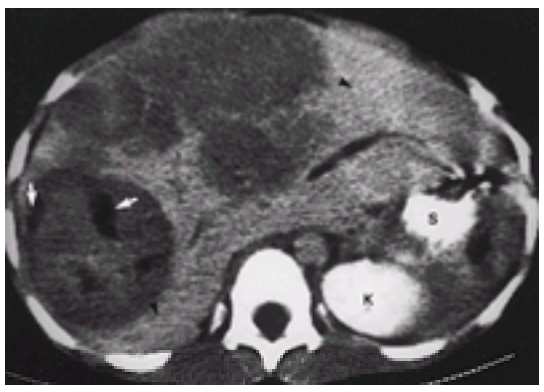


FIGURE 118.23. Hepatocellular carcinoma. Computed tomographic (CT) section of the abdomen with contrast medium enhancement shows multiple area of nonuniform lower density than the adjacent normal liver occupying most of the liver. The area of more lower density is representative of focal necrosis *(arrows).* Only small part of the left lobe and right lobe of the liver is normal *(arrowhead). K,* kidney; *S,* stomach

Survival depends on both complete resection of the primary tumor before metastases have occurred and intensive prolonged chemotherapy postoperatively. As much as two-thirds of the liver may have to be removed.

ABDOMINAL WALL DEFECTS

Inguinal Hernias and Hydroceles

Indirect inguinal hernia is the most common congenital anomaly that is found in children. It is approximately 10 times more common in males than in females. There is a strong familial incidence.

Clinical Manifestations

The child with a hernia may present in different ways. The presentation is determined by the extent of obliteration of the processus vaginalis before birth. A child may have a completely open hernia sac, which extends from the internal ring to the scrotum, or a segmental obliteration producing a sac that is narrow at its proximal end, creating a hydrocele of either the tunica vaginalis or the spermatic cord. The narrowing of the processus allows the abdominal fluid to seep into the distal portion of the sac. It then becomes entrapped and produces what is clinically recognized as a hydrocele. It is often difficult for this fluid to egress through the narrow patent processus vaginalis back into the abdominal cavity.

At the time of the embryologic closure of the processus vaginalis, many fetuses will have some fluid trapped around the testicle in the tunica vaginalis. This is called a physiologic hydrocele, which is a normal newborn finding. In such cases, the fluid gradually is absorbed in the first 12 months of life. If, however, an infant or child develops a hydrocele along the cord in the tunica vaginalis sometime after birth, it must be assumed that the processus vaginalis is still patent and in communication with the peritoneal cavity. This patent processus vaginalis represents a hernia sac. Surgical closure of the sac and drainage of the hydrocele are then indicated.

Many infants and children manifest the classical bulge in the inguinal canal that occurs during straining or crying. This is caused by a loop of intestine distending into the hernia sac. Usually, the hernia sac contents reduce into the abdominal cavity when the straining ceases. If the prolapsing loop of intestine becomes entrapped in the hernia sac, an incarceration has occurred. This is a true emergency that could eventually lead to intestinal obstruction and possibly strangulation of the bowel (p. 1521).

Elective herniorrhaphy should be done shortly after the hernia is diagnosed. One always worries about incarceration. It is an outpatient procedure in all term infants at 44 weeks' gestational age. If the operation is urgent and the infant is less than 44 weeks' gestational age, the child should be admitted for overnight monitoring. All premature babies are admitted

for 1 night because of the high incidence of postoperative apnea.

Hydroceles of the spermatic cord with associated communicating hernias are sometimes difficult to differentiate from an incarcerated hernia. If an empty hernia sac can be felt above the hydrocele, the physician can be assured that this is an asymptomatic hernia with an associated hydrocele. However, if there is a fullness above the hydrocele and the mass cannot be reduced, the child should be taken to the operating room on the assumption that it probably is an incarcerated hernia that needs to be managed surgically.

Management

Fortunately, strangulation of the entrapped loop of bowel in an incarcerated hernia occurs relatively late so that, contrary to adult practice, efforts to reduce the incarceration without surgery are usually warranted. When a child with an incarcerated hernia presents in the ED, the child should be given nothing to eat or drink, sedated if necessary with morphine 0.1 mg/kg, and placed in a Trendelenburg position. Often, this alone will reduce the incarceration. If it does not, bimanual reduction should be attempted. If the child is warm and, preferably, asleep as a result of the sedation, the reduction is facilitated. The fingers and thumb of one hand should compress the internal ring area while an effort is made with the other hand “to milk” either gas or fluid out of the entrapped bowel back into the abdomen. This relieves the pressure and usually allows the entire loop of bowel to reduce back into the abdominal cavity. Once the incarcerated hernia is reduced, the child should be admitted or scheduled for elective surgery at the preference of the surgeon. A day or two should be allowed to pass to lessen the edema of the area and allow an easier and safer elective herniorrhaphy.

Epiploceles (Epigastric Hernias)

If a discrete mass occurs intermittently about one-third of the distance from the umbilicus to the xiphoid, it is usually the result of a weakness of the linea alba through which properitoneal fat protrudes. This defect is called epiplocele. Such defects are fairly common in infants and usually close spontaneously. In older children, the mass may occasionally be tender. If it becomes excruciatingly tender, it is a sign that fat has become incarcerated in the hernia. Although there is no great urgency, these small midline defects should be repaired surgically when they become symptomatic.

Umbilical Hernias

Umbilical hernias are common in small infants, particularly in African-Americans. Fortunately, most of the hernias tend to close spontaneously and only rarely does incarceration occur. Umbilical hernias can be large and unsightly and families need reassurance that watchful waiting is the best course. However, if the umbilical hernia fails to close by the age of 5 to 6 years, surgical repair is indicated. Umbilical hernias may be repaired earlier if there is a large ring that shows no signs of diminishing in size over 1 to 2 years, if there is a thinning of the umbilical skin, or if an incarceration has occurred. Hernias that have a supraumbilical component tend not to close spontaneously and may be operated on at an earlier time of life.

Other Umbilical Defects

Omphalomesenteric duct remnants may persist in either of two forms. When the duct is patent from the ileum to the umbilicus, there is a release of small bowel contents via an opening in the umbilicus. A second form involves a remnant of the omphalomesenteric duct that contains a secreting mucosal patch that is attached to an opening in the center of the umbilicus. Passage of a sterile blunt probe or instillation of contrast dye under fluoroscopy via the umbilical opening will usually confirm either of these conditions. Once identified, these remnants must be excised surgically. By contrast, some infants present with umbilical granuloma in which an excessive amount of granulation tissue has built up after separation of the umbilical cord. In these patients, no opening in the granulation tissue can be seen or felt by means of a probe. These granulomas are usually best treated by application of silver nitrate to the granulation tissue. Occasionally, two or more treatments are required. After each treatment, the area should be rinsed thoroughly to prevent burning of adjacent skin. If the granuloma is allowed to persist, it will eventually epithelialize and become an umbilical papilloma (Fig. 118.24).

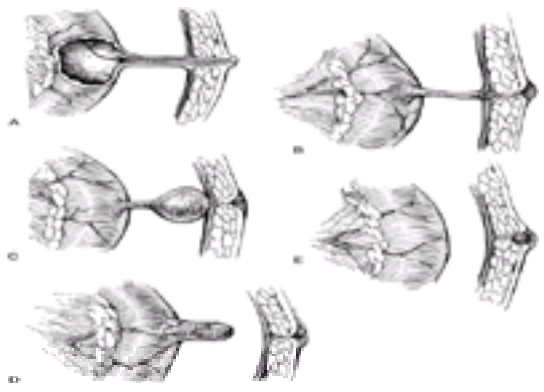


FIGURE 118.24. Omphalomesenteric remnants. Patent omphalomesenteric duct from terminal ileum to umbilicus **(A);** closed omphalomesenteric duct with mucosal patch at umbilicus **(B);** omphalomesenteric cyst below umbilicus **(C);** Meckel's diverticulum **(D);** umbilical granuloma **(E).**

If the urachus persists after birth, it can form a urinary fistula that drains at the umbilicus. This problem is ordinarily noted in the newborn period. Older infants or children may present with drainage at the umbilicus caused by persistence of part of the urachus even though connection with the bladder may be obliterated. These urachal remnants also require

surgical excision.

FOREIGN BODIES OF THE GASTROINTESTINAL TRACT

When a child ingests a foreign body, it causes great family concern. Most swallowed foreign bodies move through the GI tract without complication. Occasionally, a foreign body lodges in the esophagus, necessitating removal. Plain film roentgenograms for suspected foreign body should focus on the suspected area initially but then expand to include the base of the skull to the anus if the object is not seen. Once an esophageal foreign body has been identified, it should be removed promptly to prevent complications such as edema, ulceration, aspiration, pneumonia, or perforation.

Foreign bodies that reach the stomach, whether pointed or sharp-edged, usually pass completely through the intestinal tract and are evacuated. Cathartics and other efforts to hurry their transit should not be used.

Occasionally, a long, thin foreign body such as a bobby pin may not be able to traverse the turn where the duodenum joins the jejunum at the ligament of Treitz. If a foreign body is trapped in this area, perforation with local or generalized peritonitis may occur. When entrapment occurs anywhere beyond the pylorus, surgical removal is indicated either to prevent or to treat local perforation. Occasionally, objects such as straight pins, toothpicks, and broom straws become entrapped in the appendix. When this occurs, the appendix should be removed. Coins may remain in the child's stomach for considerable time, and if they do not become imbedded in the gastric mucosa, they eventually pass, even after a month or more. With the aid of modern flexible endoscopic equipment, foreign bodies in the stomach can usually be removed with ease. Chapter 121 covers pharyngeal foreign bodies.

Suggested Readings

APPENDICITIS

Curtin KR, Fitzgerald SW. CT diagnosis of acute appendicitis. Am J Radiol 1995;164:905.

Doherty GM, Lewis FR. Appendicitis: continuing diagnostic challenge. Emerg Med Clin North Am 1989;7:537–553.

Hahn HB, Hoopner F, Kallet T. Sonography of acute appendicitis in children: seven years experience. Pediatr Radiol 1998;28:147.

Harland RN. Diagnosis of appendicitis in childhood. J R Coll Surg Edinb 1991;36:89–90.

Hollerman JJ, Bernstein MA, Kottamasu SR, et al. Acute recurrent appendicitis with appendicolith. Am J Emerg Med 1988;6:614–617.

Lim HK, Lee WJ, Kim TM. Appendicitis: usefulness of color Doppler ultrasound. Radiol 1996;201:221.

Mollitt DL, Mitchum D, Tepas JJ III. Pediatric appendicitis: efficacy of laboratory and radiologic evaluation. South Med J 1988;81:1477–1479.

Poole GV. Anatomic basis for delayed diagnosis of appendicitis. South Med J 1990;83:771–773.

INTUSSUSCEPTION

Bonadio WA. Intussusception reduced by barium enema. Outcome and short-term follow-up. Clin Pediatr 1988;27:601–604.

Campbell JB. Contrast media in intussusception. Pediatr Radiol 1989; 19:293–296.

Heldrich FJ. Lethargy as a presenting symptom in patients with intussusception. Clin Pediatr 1986;25:363–365.

Mahboubi S, Sherman NH, Ziegler MM. Barium peritonitis following attempted reduction of intussusception. Clin Pediatr 1983;23:36–38.

Schuh S, Wesson DE. Intussusception in children 2 years of age or older. Can Med Assoc J 1987;136:269–272.

Shiels WE II, Bisset GS III, Kirks DR. Simple device for air reduction of intussusception. Pediatr Radiol 1990;20:472–474.

Young DG. Intussusception. In: O'Neill JA, et al, eds. Pediatric Surgery. St Louis: Mosby, 1998:1185–1198.

MALROTATION

Long FR, Kramer SS, Markowitz RI. Intestinal malrotation in children: tutorial or radiographic diagnosis in difficult cases. Radiology 1996;198:775.

Powell OM, Othersen HB, Smith CD. Malrotation of the intestines in children: the effect of age on presentation and therapy. J Pediatr Surg 1989;24:777–780.

Spigland N, Brandt ML, Yazbeck S. Malrotation presenting beyond the neonatal period. J Pediatr Surg 1990;25:1139–1142.

Touloukian RJ, Smith EI. Disorders of rotation and fixation. In: O'Neill JA, et al, eds. Pediatric Surgery. St Louis: Mosby, 1998:1199–1214.

HIRSCHSPRUNG'S DISEASE

Badner JA, Sieber WK, Garver KL, et al. A genetic study of Hirschsprung's disease. Am J Hum Genet 1990;46:558–580.

Teitelbaum DH, Coran AG, Weitzman JJ, et al. Hirschsprung's disease and related neuromuscular disorders of the intestines. In: O'Neill JA, et al, eds. Pediatric Surgery. St Louis: Mosby, 1998:1381–1424.

PYLORIC STENOSIS

Lund Kofoed PE, Host A, Elle B, et al. Hypertrophic pyloric stenosis: determination of muscle dimensions by ultrasound. Br J Radiol 1988;61:19–20.

Ravitch MM. The story of pyloric stenosis. Surgery 1960;48:1117.

Shaw A. Physical examination for ‘initial evaluation' of hypertrophic pyloric stenosis (Letter; Comment). Am J Dis Child 1990;144:139.

Stanley-Brown EG. Use of ultrasound and upper gastrointestinal (GI) x-rays to diagnose pyloric stenosis (Letter). J Pediatr Surg 1990;25:924.

DUPLICATIONS

Bower RJ, Siber WK, Kiesewetter WB. Alimentary tract duplications in children. Ann Surg 1978;188:669.

Holcomb GW, Gheissari A, et al. Surgical management of alimentary tract duplications. Ann Surg 1989;209:167.

HERNIAS

Holder TM, Ashcraft KW. Groin hernias and hydroceles. In: Holder TM, Ashcraft KW, eds. Pediatric Surgery. Philadelphia: WB Saunders, 1980:594.

Lassaletta L, Fonkalsrud EW, Tovar JA, et al. The management of umbilical hernias in infancy and childhood. J Pediatr Surg 1975;10:405.

Lloyd DA, Rintala RJ. Inguinal hernia and hydrocele. In: O'Neill JA, et al, eds. Pediatric Surgery. St Louis: Mosby, 1998:1071–1086.

Rescoria FJ, Grosfeld JL. Inguinal hernia repair in the perinatal period and early infancy. J Pediatr Surg 1984;19:832.

INTRA-ABDOMINAL MASSES

Davey, MS, Cohen MD. Imaging of gastrointestinal malignancy in childhood. Radiol Clin North Am 1996;34:717.

Grosfeld JL. Neuroblastoma. In: O'Neill JA, et al, eds. Pediatric Surgery. St Louis: Mosby, 1998:405–419.

King DR. Liver tumors. In: O'Neill JA, et al, eds. Pediatric Surgery. St Louis: Mosby, 1998:421–430.

Otherson HB, Tagge EP, Garvin AJ. Wilm's tumor. In: O'Neill JA, et al, eds. Pediatric Surgery. St Louis: Mosby, 1998:391–403.

FOREIGN BODIES IN THE GASTROINTESTINAL TRACT

el-Barghouty N. Management of disc battery ingestion in children. Br J Surg 1991;78:247.

Hodge D, Tecklenberg F, Fleisher G. Pediatric coin ingestion: does every child need an x-ray? Ann Emerg Med 1985;14:443.

Mahboubi S, McLean G. Emergency interventional radiology in infants and children. In: Neal MP, ed. Emergency Interventional Radiology. Boston: Little Brown, 1989:395.

Votteler TP, Nash JC, Rutledge JC. The hazard of ingested akaline batteries in children. JAMA 1983;249:2504.

CHAPTER 119

Thoracic Emergencies

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INTRODUCTION

Thoracic emergencies in children often result in life-threatening alterations in cardiorespiratory physiology. A rapid, yet organized approach to the child with a thoracic emergency may represent the difference between life and death. This chapter is aimed at guiding the emergency physician toward evaluation and stabilization of children presenting with surgical diseases involving the thorax. Congenital abnormalities usually diagnosed at birth are not included. Thoracic trauma is discussed in Chapter 107.

This chapter reviews the pathophysiology and clinical manifestations of thoracic emergencies. The general principles of physical and laboratory assessment are detailed. Subsequent sections cover specific entities within the following categories: 1) airway obstruction, 2) violations of the pleural space, 3) circulatory impairment, 4) intrinsic pulmonary lesions, 5) mediastinal tumors, 6) diaphragmatic defects, and 7) chest wall tumors.

PATHOPHYSIOLOGY

Thoracic conditions of surgical significance usually present because of a mechanical or infectious complication of an anatomic abnormality. Responding to these problems requires thinking about the patient on a gross anatomic level. These anatomic distinctions may be conveniently grouped into conditions resulting in airway compromise, violations of the pleural space, circulatory compromise, intrinsic lesions of the lung, mediastinal masses, and diaphragmatic defects.

Within each of these categories, it is particularly useful to consider *fluid pressure* changes and their effect on normal physiology. Fluids, including air, move in the body down pressure gradients. Air moves through the oropharynx, through the trachea, and into the lungs *only if the pressure in the lungs is less than atmospheric pressure.* Accumulation of air or fluid around the lungs in the pleural space may need to be removed if it is preventing lung expansion. Obstructions to flow caused by masses compressing the airway or esophagus may make it impossible for fluids to flow down the pressure gradient.

Infectious problems requiring surgical care usually have an underlying *anatomic abnormality.* Examples include an infected bronchogenic cyst or pulmonary sequestration, and an H-type tracheoesophageal fistula producing aspiration

pneumonia. Empyema, the accumulation of infected pleural fluid, which complicates pneumonia in childhood, is an example of an exception to this rule. The pathophysiology of this condition and its predilection for younger children remain poorly defined.

Whereas in most conditions of surgical significance, it is important to view the patient at the anatomic level, in some cases masses are important because of their *cellular* makeup rather than their compressive or displacing effects. Because the cellular morphology of tumors often guides therapy and relates to prognosis, biopsy is often indicated for thoracic masses. Appropriate referral of such patients is imperative.

The emergency physician evaluating the child with a thoracic problem must attempt to determine whether the patient has evidence of airway compromise, circulatory compromise, or components of both.

Airway Compromise

Airway compromise can occur anywhere in the respiratory tract from the nose to the alveoli. Obstructive emergencies relating to the oropharynx, larynx, and proximal trachea are discussed in Chapter 112 and Chapter 121.

Compromise of the more distal tracheobronchial tree may be caused by lesions in the lumen, in the wall, or outside the wall of the bronchus. Examples of intrinsic obstruction include tumor within the bronchial lumen (e.g., carcinoid tumor), foreign body, and a mucous plug. Obstruction from lesions in the wall of the bronchus include collapse from tracheomalacia and stenosis after tracheostomy. Extrinsic lesions make patients symptomatic by producing impingement on a bronchus by some adjacent structure such as a bronchogenic cyst or inflamed lymph nodes. Table 119.1 lists intraluminal, mural, and extrinsic conditions that produce airway obstruction.

Intraluminal
Foreign bodies
Aspiration (esophageal reflux, tracheoesophageal fistula, bronchial fistula, biliary fistula, or esophageal fistula)
Mucous plugs (cystic fibrosis)
Granuloma (chronic intubation, tuberculosis)
Hemoptysis (vascular malformations, cystic fibrosis, tuberculosis, sarcidosis, hemosiderosis, lupus)
Acute infection (tracheitis)
Mural
Tracheomalacia
Lobar emphysema
Bronchial atresia
Bronchial tumors
Extrinsic
Lymphadenopathy
Bronchogenic cyst
Cystic hygroma
Esophageal duplication
Mediastinal tumors

Table 119.1. Tracheobronchial Conditions Associated with Airway Compromise

The anatomic level of the obstruction correlates with its effects: an obstruction of the distal tracheobronchial tree may lead to segmental lung overdistension, or segmental infection. An obstruction of the proximal trachea affects both lungs, with a much greater likelihood of catastrophe for the patient. Similarly, greater degrees of obstruction, as a rule, lead to greater effects on gas exchange. Infection commonly follows obstruction of bronchial drainage because the clearance of bacteria or inhaled foreign materials by the mucociliary elevator is prevented.

Circulatory Impairment

Hemorrhage has somewhat different effects on the circulation in children than in adults. The ability of the child to support blood pressure in the face of significant blood loss has particular implications in the chest. Significant amounts of blood loss may be hidden in the large volume of the chest. It is important to recognize the early signs of shock before significant decreases in blood pressure occur, as this may represent a loss of 20% or more of the blood volume. Fortunately, non-traumatic causes of intrathoracic major blood loss are uncommon in children.

Collections of fluid in the pleural space and mediastinum, whether the result of bleeding or other causes, may produce obstruction of the venous return by *tension phenomena:* the child's mediastinum is mobile, and kinking of the great veins occurs much more easily than in adults. In a patient who requires positive-pressure ventilation, the positive inspiratory pressure inside the chest may be greater than the venous pressure returning blood to the heart. Thus, major intrathoracic bleeding may produce more than one difficulty: the central venous pressure and systolic arterial pressure are decreased because of loss of blood volume, and in addition, the pressure inside the chest of a ventilated patient may collapse the veins returning blood to the heart. Both of these problems require rapid administration of volume to the patient.

Rarely, the heart itself can be obstructed by primary tumors such as rhabdomyosarcoma or metastatic Wilms' tumor. Tamponade of the heart can be caused by pericardial effusion, hemopericardium, or, even more rarely, by pneumopericardium or pneumomediastinum. These topics are addressed in Chapter 82.

CLINICAL FINDINGS

Physical Examination

Evaluation of the child with a thoracic emergency requires a calm, orderly assessment of the ABCs. The physician must first address whether the patient's problem is currently causing or likely to cause imminent impairment of the airway,

breathing pattern, or circulatory status.

In assessing the airway, the physician must evaluate the adequacy of air movement and gas exchange. Pulse oximetry should be done upon the patient's arrival. Anxiety or confusion in a patient with a thoracic emergency may be evidence of hypoxemia. The work of breathing can be evaluated by assessing the use of intercostal, subcostal, and supraclavicular accessory muscles.

Breathing is best evaluated by palpation and auscultation of the chest. The trachea should be palpated to make sure that it is midline. Any lateralization of the trachea is evidence suggestive of a pneumothorax. The neck and chest should be palpated for signs of subcutaneous emphysema, suggestive of an ongoing air leak. Finally, breath sounds should be assessed via auscultation for symmetry, and adequacy of inspiratory and expiratory air flow.

Evaluation of the cardiovascular system should include an assessment of the patient's pulse for quality, rate, and regularity. The peripheral skin should then be assessed for color, temperature, and capillary refill. Signs of poor perfusion often precede that of pressure instability. The neck should be assessed for signs of jugular venous distension. Finally, the heart should be examined for signs of displacement of the point of maximal impulse (PMI); shift or alteration in the heart tones; or new murmurs, gallops, or friction rubs.

Laboratory Studies

The most important study when evaluating any patient with a thoracic emergency is a good quality chest radiograph. A radiograph of the chest should include posteroanterior (PA) and lateral views done in an upright position, unless contraindicated by the patient's condition (e.g., possible injury to the spine). The width of the mediastinum and the degree of mediastinal shift are much better seen in the upright chest radiograph. Moreover, abnormalities in the lung, pleural cavity, and diaphragm are also best appreciated in this view. For example, in the patient with a traumatic rupture of the diaphragm, the true nature of the patient's respiratory distress may not become clear until an upright chest radiograph is obtained. When a pulmonary effusion exists, lateral decubitus anteroposterior (AP) views of the chest can be obtained to determine whether the effusion layers freely or is loculated.

In interpreting the chest radiograph, the physician should distinguish between a diffuse pulmonary problem and a focal lesion. Hyperaeration of one portion of the lung suggests air trapping in the involved lobe. Hyperaeration of the entire lung field on one side is usually the result of compensatory enlargement of the lung because of atelectasis and loss of lung volume on the opposite side.

Other studies that should be considered in a patient with a thoracic emergency include a complete blood count (CBC), blood urea nitrogen (BUN), serum glucose, electrolytes, CO_2 concentration, and an arterial blood gas or measurement of oxygen saturation. Depending on the patient's specific problem, a crossmatch, blood cultures, and an assessment of any sputum by Gram stain and bacteriologic culture may be helpful. Clinical evidence of a bleeding problem, but not need for operation alone, mandates evaluation of platelet count, prothrombin time (PT) and partial thromboplastin time (PTT). Other more involved studies, such as pulmonary function tests, barium contrast studies, sonograms, computed tomography (CT) scans, and magnetic resonance imaging (MRI), can be utilized as needed.

AIRWAY OBSTRUCTION

Tracheal Obstruction

Tracheal obstruction may be produced by lesions within the lumen of the trachea, in the wall of the trachea, or extrinsic to the tube. Intrinsic obstruction most commonly occurs in children because of an aspirated foreign body. Intrinsic obstruction may also occur because of a subglottic stenosis after tracheostomy. A hemangioma may also occur, but is rare. Tracheomalacia, sometimes complicating lung disease of prematurity, is characterized by a floppy trachea that collapses during expiration, when the intrathoracic trachea is compressed by the positive intrathoracic pressure. Laryngomalacia, or tracheomalacia outside the thoracic inlet, may produce obstruction during inspiration, when the negative intraluminal pressure transmitted from the chest causes the floppy wall to collapse. Tracheomalacia often occurs in infants born with tracheoesophageal fistula. Extrinsic compression may occur both from mass lesions (Table 119.1) and as a result of anomalous arteries. Bacterial tracheitis may produce sufficient inflammation that the mucosa effectively obstructs the airway.

Clinical Findings

Tracheal compromise produces symptoms that vary from mild to severe, depending on the amount of obstruction present. When symptoms are mild, the underlying cause may not be evident. Occasional episodes of respiratory infection that are thought to result from croup or bronchitis may be the only symptom. Stridor, wheezing, or cough occur in patients with more significant obstruction, and a history of previous hospitalizations for treatment with mist tent, antibiotics, and chest percussion may be given.

Severe tracheal compromise usually is manifested by a history of stridor at rest. Progressive cyanosis and apneic episodes occur. On examination, a child with obstruction caused by extrinsic compression often has wheezing or stridor throughout the respiratory cycle. In contrast, a patient with the floppy trachea of tracheomalacia often wheezes only during expiration.

Management

If the patient has a life-threatening airway obstruction, he or she should receive airway management as outlined in Chapter 1 and Chapter 5. Intubation of the airway to within a short distance of the carina supports most patients with

lesions extrinsic to the trachea or in the tracheal wall with a critical obstruction. Such a patient requires admission to an intensive care or other unit with ventilator capability. Lesions within the lumen will likely require endoscopic management in an operating theater.

Radiographic evaluation of the stable patient should begin with PA and lateral chest radiographs, ideally obtained at full inspiration and again at full expiration. Mass lesions will usually require CT to evaluate them. Bronchoscopy is often indicated to evaluate obstructive lesions, whether in the lumen, the wall, or extrinsic to the wall of the trachea (Fig. 119.1).

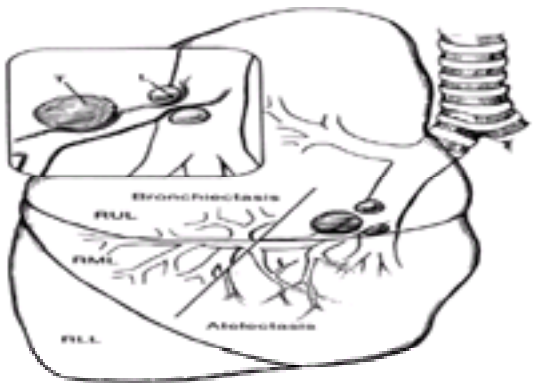

FIGURE 119.1. Acute and chronic obstruction of a bronchus owing to tumor or cyst *(T)* or lymph nodes *(L).* When the obstruction is acute, there may be bronchiectasis caused by recurrent pneumonia. The right middle lobe as shown here is particularly prone to bronchial obstruction caused by pressure from encircling lymph nodes. *RUL, RML, RLL,* right upper, middle, and lower lobes, respectively.

Vascular Rings

Vascular rings are developmental anomalies of the aorta and great vessels. They may produce obstruction of the esophagus, trachea, or both. Many anatomic types of rings are produced by failure of the normal involution of the appropriate segments of the six embryologic aortic arches. The number of possible variants is at least 36; 16 or more have been seen in humans. The level of obstruction is usually at the trachea, but compression of a bronchus by the ductus arteriosus, or by a pulmonary artery sling may produce compression more distally. The reader is referred to standard texts of pediatric or thoracic surgery for further details.

Clinical Findings

Vascular rings should be suspected in infants with stridor, dysphagia, failure to thrive associated with difficult feeding, or recurrent pneumonia. The wide variety of anomalies produce varying degrees of symptoms. Esophageal obstruction produces difficulty swallowing, designated *dysphagia lusoria* by Bayford in 1794. Often, diagnosis is delayed by failure to consider these anatomic obstructions. Chest radiographs may be supplemented by a variety of diagnostic tests: angiography, echocardiography, MRI, and digital subtraction angiography are needed in some combination to define the anatomy.

Management

Although a few patients with constricting anomalies improve as they grow, the usual situation is for a poor prognosis with medical therapy. Surgical treatment is usually indicated to relieve the obstruction. This is accomplished by dividing the vascular ring and preserving the blood supply to the aortic branches. This is usually accomplished by a left thoracotomy.

Bronchial Lesions

Bronchial Atresia

Congenital bronchial atresia is a rare anomaly characterized by a bronchocele caused by a mucus-filled, blindly-terminating segmental or lobar bronchus, with hyperinflation of the obstructed segment of lung. Hyperaeration is thought to result from communication via the pores of Kohn and the channels of Lambert with the normally aerated lung. First reported in 1953, a 1986 review of the literature reported a total of 86 cases.

Clinical Findings

Neonates and infants with the lesion usually are seen for respiratory distress. In older patients, a history of episodic upper respiratory infection and wheezing may be elicited. Some older patients may complain of dyspnea on exertion or unilateral chest pain. Physical findings seldom suggest the diagnosis, but often unilaterally decreased breath sounds are evident.

Management

Chest radiographs make the diagnosis most of the time. Chest CT scan is indicated to help define the anatomy. Bronchoscopy is the most efficient way to identify the atretic opening to the involved bronchus. Bronchography has been

used in the past, but high-resolution CT scan can often provide the same anatomic information non-invasively.

Right Middle Lobe Syndrome

The right middle lobe is anatomically predisposed to compression of its bronchus by the lymph nodes in its vicinity, which tend to encircle it. Because the right middle and lower lobes are favored sites for aspirated material, recurrent inflammation caused by pneumonia leads to adenopathy. Previously, especially in the era before antituberculous chemotherapy, this tended to result in compression of the right middle lobe bronchus alone, which produced eventual bronchiectasis. Often, right middle lobectomy was necessary. Presently, it is more common for the right middle and lower lobes to be involved together.

Clinical Presentation

Recurrent episodes of pneumonia and associated atelectasis in the right middle (and often lower) lobes occur in these patients, and are not responsive to chest percussion, postural drainage, or antibiotic treatment. The mechanical compression of the bronchus leads to a sequestered infection, which may require resection of the right middle or right middle and lower lobes. Although the need for resection is far less common than in the past, acute pneumonia in these anatomic locations should prompt a discussion of previous pneumonias and treatment. Close follow-up is indicated in such patients.

Esophagus-Related Causes of Airway Difficulties

Tracheoesophageal Fistula

Tracheoesophageal fistula (TEF) occurs in children both as a congenital lesion and as an acquired problem after suppuration of mediastinal nodes. The congenital fistula is accompanied by atresia of the esophagus in more than 85% of patients. However, in about 3% of all patients with TEF, the connection between the tracheal tube and the esophagus creates the shape of the letter “H” (Fig. 119.2). In these, there is no accompanying esophageal atresia.

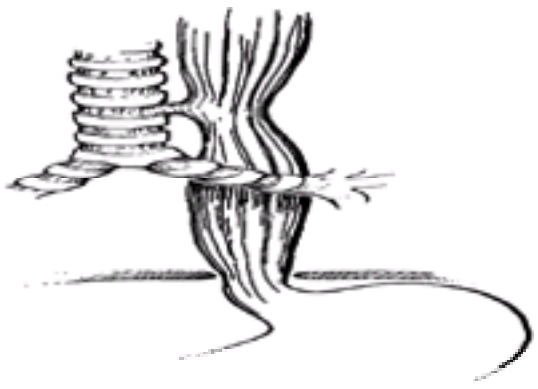

FIGURE 119.2. H-type tracheoesophageal fistula.

It is this “H-type” fistula that is most likely to be seen in the ED. The acquired form is usually in the distal trachea or proximal bronchial tree, and is extremely uncommon.

Clinical Findings

These fistulae are notoriously difficult to diagnose. Children generally develop recurrent pulmonary infections for which no source is evident. The characteristic history of choking or gagging when swallowing that accompanies esophageal atresia with TEF may not be present.

Management

Contrast esophagram may identify the lesion. Most of these fistulae are small in diameter (much less than 1 cm), and short (also less than 1 cm), making radiographic identification difficult. Even when contrast appears in the tracheobronchial tree, it may be difficult to know whether primary aspiration of orally administered contrast is responsible. Placing a feeding tube in the esophagus and injecting contrast while pulling the tube from the lower esophagus up under fluoroscopic observation may be helpful. High-resolution CT scanning may identify the anatomy. Bronchoscopy and esophagoscopy may be both diagnostic and may aid the repair if a small catheter can be passed across the fistula to aid its identification by enabling palpation at operation. Most such fistulae are cervical, and can be repaired without a thoracotomy.

Gastroesophageal Reflux

Introduction/Pathophysiology

The mucosal lining of the trachea and bronchial tree tolerates periodic soilage relatively well; even witnessed aspiration of gastric contents does not reliably produce infection or pneumonia. Ongoing irritation by gastric acid, bile, or contaminated secretions will eventually overcome the mucociliary elevator that transports contaminants to the pharynx, and the other barriers to infection of the tracheobronchial mucosa, and bronchitis or pneumonia will result. Common

causes of repetitive soilage of the tracheobronchial tree include primary aspiration of oropharyngeal secretions, often in children with impaired swallowing mechanisms, and gastroesophageal reflux (GER). GER is universal in babies, and is usually outgrown. Particularly in the neurologically impaired patient, however, GER may require medical or surgical treatment.

Clinical Presentation

GER presents with symptoms of spitting up or vomiting after eating. Aspiration may lead to presentation with recurrent pneumonia. Complications that follow prolonged GER include failure to thrive because of inadequate nutrition, esophagitis, esophageal ulceration and esophageal stricture (Fig. 119.3). Some patients present with an acute life-threatening event (ALTE) in which laryngospasm or bronchospasm precipitated by aspiration of gastric contents produces profound hypoxia and even respiratory or cardiac arrest.

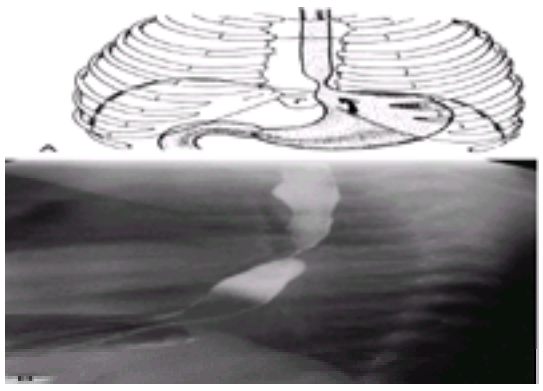

FIGURE 119.3. A. Distal esophageal stricture caused by prolonged reflux esophagitis. Note the loss of the normal angle between the esophagus and the stomach and propensity for gastric contents to reflux into the esophagus. **B.** Esophageal stricture. Lateral barium esophagram shows narrowing of midesophagus in an infant with gastroesophageal reflux.

Management

Management of GER begins with establishing the diagnosis. If this is evident clinically, and the child responds to medical management, no further evaluation may be needed. Recalcitrant GER may be an indication for an upper gastrointestinal contrast study (UGI series) to establish that there is no anatomic obstruction to gastric emptying such as a duodenal web or annular pancreas. Recording the esophageal pH over a 24-hour period with a pH probe may help quantify the severity of the problem.

GER is managed by a three-tiered approach. Initially, elevating the head of the bed, thickening the feeds, and decreasing the volume of individual feeds are useful to allow gravity and mechanical effects to help. Medical management (second tier) of this problem includes efforts to decrease gastric acidity, including antacids, H_2-receptor antagonists such as ranitidine, and proton-pump inhibiting drugs. Many clinicians add prokinetic medications to improve the gastric motility such as metoclopramide or cisapride. Recently, concerns regarding the association between cisapride and ventricular tachycardia have led to a decrease in the utilization of this medication. Surgical indications, the third tier, are failure of nonoperative management or occurrence of a complication that cannot be tolerated, such as esophageal stricture or repeated ALTEs without other evident cause. Presently, the favored operative treatment in North America is fundoplication: wrapping the fundus of the stomach either partially (a Thal operation if anterior to the esophagus, or Toupet operation if posterior) or completely (the Nissen operation) around the esophagus just above the gastroesophageal junction. The procedure may be performed laparoscopically in appropriate patients.

Esophageal Web

Introduction/Pathophysiology/Management

Rarely, a patient presents with GER that is caused by an esophageal web (Fig. 119.4). The membranous, congenital narrowing of unclear origin is usually able to transmit liquids, and symptoms often arise when the child begins to eat solid food. Recurrent aspiration pneumonia may also develop. An esophagram is usually diagnostic. Often, a thin membranous web may be split by a hydraulic balloon placed endoscopically across the stenosis. If this approach is unsuccessful because the lumen is too small to transmit the dilator or the tissue is unyielding, segmental esophageal resection may be necessary via thoracotomy.

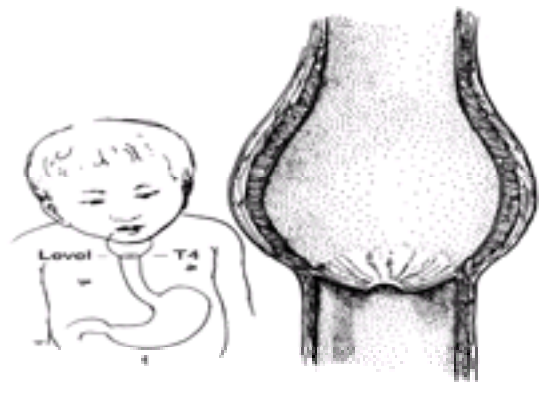

FIGURE 119.4. A child with chronic partial obstruction of the esophagus caused by a congenital web. Similar bulbous enlargement of the proximal esophagus can occur with any type of stricture and result in pressure on the trachea and recurrent regurgitation with aspiration.

CIRCULATORY IMPAIRMENT

Circulatory collapse, or shock, is often classified as hypovolemic, cardiogenic, neurogenic, or septic. Hypovolemic shock results from massive loss of blood or fluid. Cardiogenic shock follows impairment of the pump function of the heart. Neurogenic shock follows spinal cord transection or spinal anesthesia and results from loss of vascular tone with attendant pooling of blood in the lower body, reducing cardiac preload. Similarly, septic shock is, in large part, caused by decreased peripheral resistance. Trauma-producing shock is reviewed in Chapter 3, Chapter 103, and Chapter 107. Thoracic causes for each of these categories may be enumerated, but the important task for the emergency physician is to classify the cause of the patients problem in a way that will direct management.

Cardiogenic shock results from decreased cardiac preload, failure of the heart as a pump, increased afterload, or loss of heart rate. Preload may be reduced by tension pneumothorax causing kinking of the great veins bringing blood to the heart in the child's mobile mediastinum; by massive intra-abdominal hemorrhage compressing the inferior vena cava; by cardiac tumor obstructing one of the atria; or by tamponade of the heart from mediastinal pressure caused by blood, pericardial fluid, or air. Hypertension resulting from coarctation of the aorta, pheochromocytoma, or other causes, may increase afterload so greatly that cardiac output is impaired. Failure of the heart as a pump occurs in viral myocarditis, or after infarction. Drug ingestions and hypoxia may slow the heart sufficiently to decrease cardiac output.

The causes, presentation, and management of cardiogenic shock are reviewed in detail in Chapter 3.

Clinical Findings

Findings are dependent on the type of shock and the primary lesion. Whether caused by tension pneumothorax, intracardiac tumor, or tamponade, hypovolemic (or decreased preload) shock is accompanied by tachycardia. Usually blood pressure is maintained until perhaps 20% of the blood volume is lost to hemorrhage; predicting the onset of hypotension with tension pneumothorax or compressive phenomena is difficult. Characteristically, the extremities are cold and poorly perfused as peripheral vasoconstriction compensates for loss of central venous pressure. Tamponade is accompanied by muffled heart tones, often difficult to recognize in a noisy ED, especially in the setting of trauma. Distended neck veins are often present. Once tamponade has reached a critical compression pressure, it often does not respond to intravenous (IV) fluid. Pulsus paradoxus may not accompany acute tamponade.

Clinical findings suggestive of cardiogenic shock are discussed in Chapter 3. Septic and neurogenic shock are “warm shock” in which the extremities are well-perfused because of loss of vascular tone; tachycardia commonly accompanies them. Fever in septic shock, and flaccid extremities with loss of bladder control and rectal tone in neurogenic shock may aid diagnosis.

Management

IV support with two large-gauge peripheral catheters, electrocardiogram monitoring, pulse oximetry, and oxygen supplementation are indicated for any type of circulatory collapse. Stable patients should undergo a chest radiograph immediately. Afterward, management is directed to relieving the condition that produced shock.

If acute cardiac tamponade is suspected, emergency pericardiocentesis is indicated immediately (see Section VII, Procedures). If the patient is not improved by pericardiocentesis, the pericardium may be filled with clotted blood that will not drain through the inserted needle. In this circumstance, pericardial drainage will require a larger opening in the pericardium. In a patient with shock and incipient cardiac arrest, a vertical subxiphoid incision should be made in the ED. After opening the linea alba, the pericardium can be opened widely enough to digitally clear hematoma from the pericardium.

PLEURAL DISEASE

The lung is covered by the densely adherent visceral pleura, which moves smoothly over the parietal pleura of the chest wall because of a thin film of pleural fluid, allowing lubricated motion of the chest during respiration, and contributing to the full expansion of the lung mechanically. When air, excess fluid, or pus comes between the two layers of the pleura, the lung tends to collapse, and consideration needs to be given to removing the interloper.

Pneumothorax

Air can collect in the pleural space acutely or chronically, statically or progressively. Because atmospheric pressure is always greater than intrapleural pressure, any mechanism that allows even momentary communication between the atmosphere outside the chest wall or the atmosphere within the tracheobronchial tree, can result in a rapid shift of air into the pleural space. Penetrating wounds of the chest are the most common cause for pneumothorax. The penetrating object (a knife, a bullet, or a doctor's needle) may cause injuries of both the parietal pleura and often the lung parenchyma. Therefore, many patients with penetrating trauma to the chest will have not only an initial pneumothorax but also an expanding pneumothorax, as more and more air leaks from the surface of the lung.

Nonpenetrating trauma to the thorax can also result in a pneumothorax. For example, a fracture of one or more ribs may result in puncture of the visceral pleura and lung, causing an escape of air from the lung into the pleural space. If the intrapleural pressure increases, air may leak out through the hole in the parietal pleura and into the chest wall tissues, resulting in subcutaneous emphysema. Another form of nonpenetrating trauma is barotrauma, which can occur in infants and children who have been ventilated with high inflating pressures via a tight-fitting endotracheal tube. A particularly hazardous form of pneumothorax occurs when severe blunt trauma to the chest results in partial or complete tear of a bronchus or the trachea. Usually, patients with more peripheral bronchial tears will immediately develop symptoms of a pneumothorax. If the tear is more central, the patient may first develop mediastinal and even cervical emphysema before a secondary rupture occurs into the pleural cavity.

Seemingly spontaneous episodes of pneumothorax may occur in children or adolescents. For example, a patient with one or more emphysematous blebs on the surface of the lung may develop spontaneous rupture, resulting in an acute pneumothorax often associated with nearly complete collapse of the involved lung (Fig. 119.5). In patients with cystic fibrosis, spontaneous pneumothorax is the second most common pulmonary complication of this condition. It usually occurs in teenage or young adult patients with far-advanced, diffuse bilateral cystic fibrosis. Another group of children with a high incidence of spontaneous pneumothorax are those with pulmonary metastases, for example, those with osteogenic sarcoma. Many of the metastases occur just below the pleural surface of the lung and, thus, may be the foci for the pneumothorax. Children with staphylococcal pneumonia are especially prone to develop unilateral or bilateral pneumothorax.

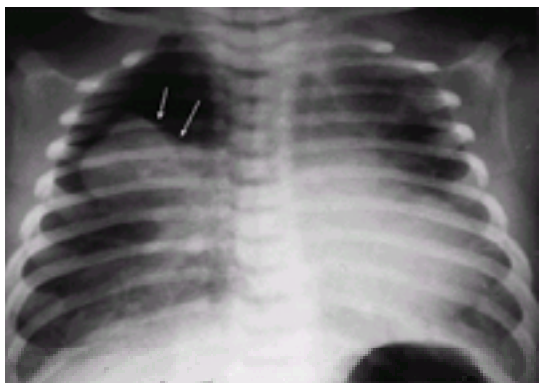

FIGURE 119.5. Radiograph revealing obvious pneumothorax. Note the collapsed right upper lobe segment borders *(arrows).*

If the site through which air enters the pleural cavity seals quickly, and no fluid or blood collects in the pleural space, a small or moderate pneumothorax will resolve spontaneously. However, some patients may have what appears to be a chronic or static pneumothorax. This usually occurs when there is a slow, persistent leak of air from the surface of the lung. A patient with osteogenic sarcoma metastases to the lung, for example, might continue to have a small, but clinically significant, separation of the lung from the parietal pleura.

Two special forms of pneumothorax require emphasis because these conditions may result in the death of the patient if not recognized early and attended to rapidly. The first is a tension pneumothorax, which results not only in total collapse of the lung but also in progressive tension across the mediastinum (Fig. 119.6). The development of a progressive tension pneumothorax is a result of air accumulating in the hemithorax with each inspiration. Whether the site of entry of the air into the pleural space is through the chest wall, a torn bronchus, or an injured portion of lung, the physiologic result is a one-way valve effect, whereby air continues to accumulate in the pleural cavity with inspiration but cannot be extruded on expiration. This phenomenon continues until the intrathoracic pressure on the involved side is so high that no further air can enter the pleural space. This is often the point at which venous return from below the diaphragm is impeded and circulatory failure ensues.

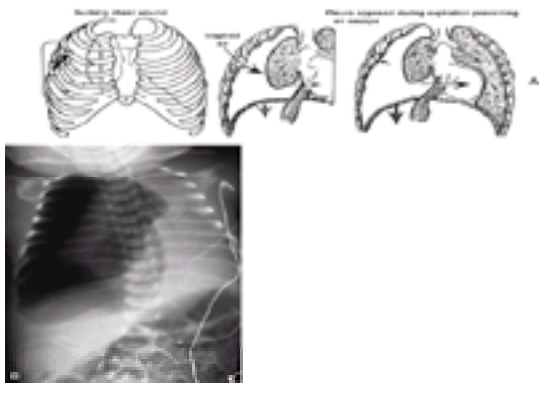

FIGURE 119.6. A. The development of a progressive tension pneumothorax as a result of air accumulating in the hemithorax with each inspiration. **B.** Severe pneumothorax causing marked hyperaeration of the right lung field and shift of the mediastinum to the left side.

The second life-threatening form of abnormal collection of air in the thorax is massive pneumomediastinum with or

without an associated pneumothorax. In extreme cases, the tension produced in the mediastinum can be great enough to impair both circulation and ventilation. This phenomenon is particularly likely to occur in a patient who is receiving positive-pressure ventilation, which enhances escape of air from the bronchial tree into the mediastinum. See Chapter 107 for trauma-related causes such as rupture of a major bronchus or the trachea.

Clinical Findings

The symptoms and signs of pneumothorax depend on the size of the pneumothorax and how rapidly it occurred. A patient with spontaneous rupture of an emphysematous bleb may complain of sudden acute pain on the involved side of the chest followed by tachypnea, pain at the tip of the ipsilateral shoulder, and a sense of shortness of breath. Such patients usually have a small to moderate pneumothorax (less than 20% of the lung volume).

In general, a patient with a pneumothorax has signs and symptoms of ventilatory impairment: dyspnea, tachypnea, pain, splinting on the involved side, agitation, increased pulse rate, diminished breath sounds, and increased resonance on the involved side, and possibly, displacement of the trachea and heart away from the involved side. See Chapter 107 for evaluation of traumatic pneumothorax.

Management

The essential components of management involve confirmation that a pneumothorax exists and reexpansion of the lung. If the patient's condition is not severe, an immediate upright PA and a lateral chest radiograph should be taken. These radiographs are important to determine not only the site and extent of the pneumothorax but also any complicating features such as tumor; fluid within the pleural space; or abnormalities of the lungs, diaphragm, or mediastinum.

Once a pneumothorax is diagnosed on radiograph, the urgency for treatment of the pneumothorax depends on the patient's signs and symptoms, clinical circumstances, and the extent of the pneumothorax. If the pneumothorax is greater than 20% of the lung volume or if the patient is symptomatic, a controlled, sterile chest tube insertion should be considered (see Section VII, Procedures). In the ED, the percutaneous, guidewire “pigtail” catheters (sometimes called “Fuhrman tubes”) are ideal for pneumothorax not associated with blood in the chest or empyema. In the case of a patient who requires surgery but who has only a small pneumothorax, a chest tube should be inserted because of the risk that the pneumothorax will expand under anesthesia and therefore complicate the course of surgery.

If the child's condition is so severe that there is not time for a chest film and if a pneumothorax is suspected, immediate therapy includes 1) tamponading and obliterating any sucking or open chest wound, and 2) inserting an angiocath, percutaneous central line, or pigtail catheter into the pleural space and evacuating air. Placement of whichever tube is used is usually best done in the midaxillary line over the top of a rib about level with the nipple. Once the plastic cannula enters the pleural space, it can be advanced further inside and then attached to sterile IV tubing and placed to underwater seal. Alternatively, a stopcock can be attached to the same setup and an attempt made to aspirate enough air and/or fluid to improve the patient's pulmonary dynamics. In a patient with a tension pneumothorax, the insertion of the needle and catheter will immediately result in release of the tension on the mediastinum and diaphragm.

Many infants can be effectively managed in this way if the amount of air present in the pleural space is small. However, these temporary catheter devices are small gauge and thus tend easily to develop fibrin plugs. Therefore, in any infant or older child who requires a tube within the pleura for more than 1 hour, it is best to proceed with a standard or pigtail chest tube insertion. Patients with pneumothorax should be admitted to a hospital, even if no chest tube is thought necessary, so that the stability of the air collection can be monitored in a setting where tension pneumothorax may be rapidly treated should it develop.

Pleural Effusion

Pleural fluid in excess amount is not a disease per se, but it indicates the presence of pulmonary or systemic illness. The classification of the fluid into *transudate,* which accumulates when the normal pressure relationships between the capillary pressure in the lung, the pleural pressure, and the lymphatic drainage pressure are disturbed, or *exudate,* an inflammatory collection, has less utility today than in previous years because of other diagnostic tools presently available. Nevertheless, an awareness that an increased pulmonary capillary pressure (as in congestive heart failure), a decreased colloid osmotic pressure (as in renal disease), increased intrapleural negative pressure (as in atelectasis), or impaired lymphatic drainage of the pleural space (e.g., from surgical trauma to the thoracic duct) may result in transudative effusion is important. In children, the inflammatory cause of effusion is often evident as a pneumonia. The accumulation of blood in the pleural space because of trauma is discussed in Chapter 107. Hemothorax may result from non-traumatic conditions as well. Necrotizing pulmonary infections; tuberculosis; pulmonary arteriovenous malformation; torn pleural adhesions with spontaneous pneumothorax, hemophilia, thrombocytopenia, systemic anticoagulation; and pleural tumors all have been reported to cause hemothorax. Chylothorax, or the accumulation of lymphatic fluid in the pleural space, has become more common as thoracic, especially complex cardiac, surgical operations, have become more common in children.

Clinical Findings

Pleuritic chest pain is a sharp, intense pain on deep inspiration that is often not present on quiet breathing. Small, sterile collections, as well as large, chronic collections, tend to be asymptomatic. Acute collections produce symptoms by compressive effects on the lung, with resultant atelectasis and right-to-left shunting, which produces oxygenation and ventilation compromise. Respiratory distress may follow, with attendant dyspnea, tachypnea, increased use of accessory muscles of respiration, and even cyanosis. Except for huge effusions, the examination, using auscultation and percussion to define the amount of fluid, is not nearly as useful as a chest radiograph. Almost all effusions are detected on this basis. Bilateral decubitus chest radiographs help define the presence of pleural fluid in patients in whom it is difficult to see because of adjacent parenchymal disease. This examination also demonstrates whether the fluid is free to move

about in the chest.

Management

If the presence of a significant effusion is evident by examination and radiograph, no further radiographic studies may be needed. A CT scan, or in some institutions, an ultrasound examination of the chest helps determine whether opacity seen on a chest radiograph is parenchymal disease or pleural fluid. All patients should have a CBC and differential and blood culture. Analysis of the fluid itself is the most useful diagnostic test. The technique for thoracentesis is given in Section VII.

Aspirated fluid should be sent for cell count, differential, Gram stain, acid-fast bacillus (AFB) stain, total protein, lactate dehydrogenase (LDH), protein, specific gravity, and a complete set of cultures (aerobic, anaerobic, AFB, and fungal). The normal protein concentration is 1.5 gm/dL. Classically, an exudate was said to have a total protein of more than 3.0 gm/dL and a specific gravity of more than 1.016. An accuracy rate of more than 99% in classification is obtained by noting that fluid is an exudate if any one of the following criteria are present: 1) Pleural fluid protein divided by serum protein is greater than 0.5; 2) pleural fluid LDH divided by serum LDH is greater than 0.6; or pleural fluid LDH greater than two-thirds of the upper limit of normal for serum LDH. The studies ordered should clearly be tailored to the clinical setting: in patients with hemothorax with an evident cause, little is to be learned by studies of the pleural fluid. Suspected chylothorax may be identified by measurement of triglycerides and cholesterol; a fat stain such as Sudan black or oil red “O” may be done on the fluid. Empyema can appear similar to chylothorax. Centrifuging the specimen can differentiate the two because in empyema the supernatant is clear.

Draining the pleural fluid must then be considered. Thin fluid may sometimes be managed by intermittent thoracentesis. If the underlying medical problem can be managed, the effusion may take care of itself. If not, a small-diameter tube, such as an 8-Fr pigtail percutaneous tube, may suffice. Thick fluid, such as blood, pus, and sometimes chyle, requires a large-diameter chest tube to drain it. Either tube must be attached to a pleural drainage system. When the drainage decreases significantly, to approximately 1 mL per pound of body weight per day, the drain may be removed. The drain should not be removed in the presence of an accompanying “air leak” caused by a bronchopleural connection.

Empyema

Empyema or pus within the pleural cavity is a particularly serious and, at times, life-threatening situation. The predominant organism is *Streptococcus pneumoniae* with *Staphylococcus aureus* and group A streptococcus also meriting consideration. Empyema is usually the result of septicemia or direct or lymphatic extension from an associated pulmonary infection. When empyema follows accidental trauma or surgery, other bacterial organisms may be involved.

Clinical Findings

Empyema is most common in children 2 to 9 years of age. Presentation with a pneumonia that does not respond to antibiotic treatment for many days should lead to consideration of decubitus chest radiographs or CT scan for diagnosis. High fever is common, as are the symptoms of pneumonia: cough, pleuritic chest pain, and lassitude.

Management

Empyema in healthy children may respond to prolonged (3 to 4 weeks) IV antibiotic therapy and chest tube drainage. Recovery may be hastened to less than a week in most cases by thoracoscopic debridement of the pleural space of the infected fibrinous peel that encases the lung and prevents its full expansion in many cases. Under a general anesthetic, a fiberoptic high-resolution camera placed within the pleural space via a short (2-cm) incision between the ribs allows removal of the purulence and the fibrinous peel that often encases the lung, restricting its expansion. The peel may be removed under direct visualization via other thoracoscopic instruments placed through two or three small incisions. A chest tube is then placed to drain the pleural cavity through one of these incisions and is left in place for a few days. Because sedation approaching the depth of general anesthesia is needed for placement of a chest tube, many surgeons and infectious disease consultants recommend thoracoscopy as the initial approach to a child with empyema. Seldom is open thoracotomy now necessary to resolve empyema.

Solid Pleural Lesions

Solid lesions in the pleural space occur uncommonly in children. A localized pleural-based mass should suggest neoplasm, which may be primary or metastatic. Diffuse collections usually are the sequelae of bleeding into the pleural space in the distant past or of empyema. They may encase the lung and produce restrictive lung disease.

Clinical Presentation/Management

It is impossible to generalize on the mode of presentation of such rare processes. Focal lesions may be expected to be found in investigation of symptoms caused by local compression or erosion; because of the large functional pulmonary reserve of children, restrictive lung disease caused by a diffuse process is distinctly uncommon. A full radiographic evaluation, including CT scan, should be obtained; admission to the hospital should be strongly considered, and appropriate consultation sought. Focal lesions should be considered malignant until proven otherwise, so operation for biopsy or excision will likely be required.

LUNG LESIONS

The lung is often affected in childhood illness. Asthma, pneumonia, and other conditions that do not require surgical management are addressed elsewhere in this text. Mass lesions and cystic lesions of the lung include congenital cystic

adenomatoid malformation (CCAM), congenital lobar emphysema, bronchogenic cyst, congenital pulmonary arteriovenous fistula, and bronchopulmonary foregut malformations. Acquired conditions of the lung that require surgical management are distinctly uncommon because of the control of tuberculosis in North America. Bronchiectasis—the chronic dilation of the bronchi resulting from the chronic infection of the lung in cystic fibrosis, tuberculosis, or other chronic pneumonic infection—may require pulmonary resection.

Bronchogenic Cyst

Bronchogenic cysts are thought to result from aberrant budding from the primitive foregut or tracheobronchial tree. They may occur along the trachea, along the bronchi, in the lung substance, or adjacent to the esophagus (Fig. 119.7).

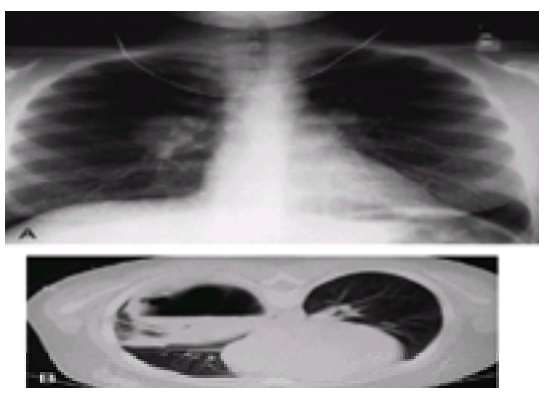

FIGURE 119.7. A. Plain film of patient with bronchogenic cyst arising off the right mainstem bronchus. **B.** Computed tomography scan of similar lesion reveals large fluid-filled cyst compressing adjacent lung tissue.

Clinical Presentation

Centrally located cysts may present with symptoms caused by compression of an airway. Wheezing, cough, fever, and recurrent pneumonia may result in such children. In contrast, patients with peripherally located cysts develop respiratory symptoms only 50% of the time. Physical examination is often unrewarding.

Management

Detection of bronchogenic cysts almost always occurs radiographically. Chest radiograph often suggests the process, but CT scan is usually indicated to clarify the anatomy. Plain-film findings include a homogeneous, water-density mass without sharply-defined borders. CT scanning usually shows a water-density mass as seen by Hounsfield or other density units. Cysts with turbid, mucoid fluid may appear solid on CT scan.

Treatment of bronchogenic cysts is by surgical resection. Active infection should be brought under control. Thoracoscopy may be used for some lesions, depending on the location of the mass. Asymptomatic cysts should be removed to establish the diagnosis and to prevent the complications of secondary bronchial communication, bleeding, or perforation into the pleural cavity. Carcinoma and fibrosarcomas have been reported to arise in benign-appearing bronchogenic cysts.

Congenital Cystic Disease of the Lung (Congenital Cystic Adenomatoid Malformation and Sequestration)

Grouping the several pathologic entities included in congenital cystic disease of the lung makes particular sense for the emergency physician. From a single giant unilocular cyst, to a mixed lesion composed of multiple cysts and solid tissue, or a lesion composed predominantly of solid tissue with only an occasional small cyst, these lesions are all congenital processes that present with pulmonary infection, an abnormal chest radiograph, or possibly, a mass or tension effect. Cystic adenomatoid malformations are the result of an excessive overgrowth of bronchioles (Fig. 119.8) and an increase in terminal respiratory structures and mucous cells lining the cyst walls. Pulmonary sequestrations arise from an accessory bronchopulmonary bud of the foregut. Histologically they are portions of pulmonary tissue; however, they are not connected with bronchi or vessels to the rest of the lung (and hence the pulmonary tissue is “sequestered”). Usually, there is a systemic rather than pulmonary blood supply. Sequestration can be intralobar (like cystic adenomatoid malformation) or extralobar.

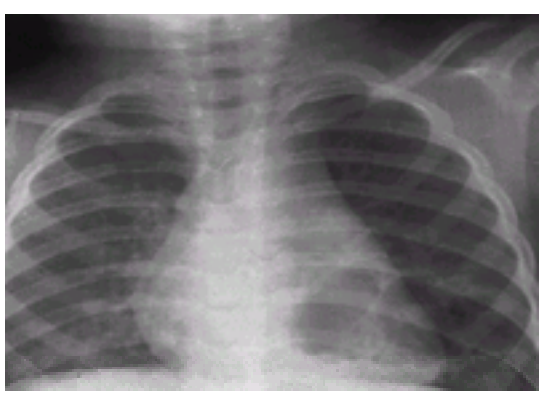

FIGURE 119.8. Cystic adenomatoid malformation in a 12-month-old girl with recurrent episodes of apparent left lower

lobe pneumonia.

Clinical Findings

Recurrent respiratory infections often lead to the chest radiograph, which confirms the condition. Clinical findings may be identical to those of a lobar pneumonia. Occasionally, a lesion is discovered after failure of resolution of an empyema by chest tube placement.

Management

Chest radiographs in the PA, lateral, and bilateral decubitus positions should be obtained to evaluate any areas with air-fluid levels. Any pathogens identified in the sputum should be treated with appropriate antibiotics (see Chapter 84). After control of superimposed infection, the lesion should be resected to prevent recurrent infection. Attempted aspiration of the cystic lesions or placement of a chest tube is to be avoided because it may lead to spread of infection into the pleural space. When the lower lobe seems to be involved, a CT scan with IV contrast should be obtained to identify any possible systemic blood supply. Because the blood supply may arise from below the diaphragm, the scan should include both the chest and abdomen. Arteriography is seldom necessary with currently available imaging techniques. The CT scan will likely exclude other conditions that may be misdiagnosed, such as a diaphragmatic hernia, postpneumonic pneumatoceles, or esophageal duplication.

Congenital Lobar Emphysema

Congenital lobar emphysema, also known as infantile lobar emphysema or congenital segmental bronchomalacia, is caused by overexpansion of the air spaces of a segment or lobe of the lung (see Fig. 119.9). There is no significant parenchymal destruction. This entity accounts for about half of all congenital lung malformations. Bronchial obstruction caused by a variety of entities produces the condition.

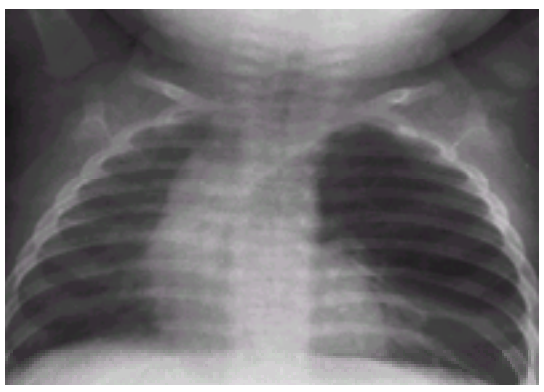

FIGURE 119.9. Congenital lobar emphysema of the left upper lobe in a 3-month-old girl who presented with decreased breath sounds and rales in this area. Note the secondary compression atelectasis of the left lower lobe.

Clinical Findings

Infants with congenital lobar emphysema are often normal in appearance at birth, but develop tachypnea, cough, wheezing, dyspnea, and/or cyanosis within a few days. The onset of symptoms may be more gradual; nevertheless, 80% of patients are symptomatic by 6 months of age. The upper lobes are involved in about two-thirds of patients, and in less than 1% are the lower lobes involved. Chest radiographs show striking radiolucency in the involved lobe with mediastinal shift to the opposite side. The diaphragm is usually flattened on the affected side. It can be difficult to tell whether pulmonary markings are present at all in the involved lobe, and pneumothorax may be suspected. The compressed normal lung may be erroneously thought to be atelectatic with the emphysematous lobe compensatory.

Management

Treatment should be given to patients with life-threatening pulmonary insufficiency from compression of normal pulmonary tissue. If a bronchial obstruction such as a mucous plug or foreign body can be relieved, no further treatment may be necessary. Pulmonary lobectomy may be needed acutely if symptoms are progressive. The diseased lobe is evident at thoracotomy because of its overdistended state, which often pushes this part of the lung out of the chest. Lobectomy is curative if the cause of the obstruction is also relieved.

Congenital Pulmonary Arteriovenous Fistula

Congenital pulmonary arteriovenous (AV) fistula, a congenitally occurring communication between a major pulmonary artery and a vein within the lung, is usually an aneurysmal sac. Fistulae vary in size from a few millimeters to several centimeters, and can be multiple. At times, a systemic artery may also be involved. Direct right-to-left shunting leads to hypoxemia, and the size of the fistula correlates with the degree of desaturation.

Clinical Findings

As the initial presentation of this disorder is frequently that of wheezing and desaturation, the child may be misdiagnosed as having asthma. Clubbing and cyanosis may suggest chronic hypoxemia. Examination of the chest may demonstrate a palpable thrill or murmurs. If there are symptoms of hemoptysis and epistaxis, one may find telangiectasias or hemangiomas of the skin and mucous membranes. Evaluation of the family may also reveal the presence of hereditary hemorrhagic telangiectasis (Rendu-Osler-Weber disease), which is present in more than half the patients with congenital pulmonary AV fistula.

Management

Children who are symptomatic from this condition should be evaluated by means of CT scan, contrast echocardiography, perfusion scintigraphy, and arteriograms of the pulmonary artery and aorta. Chest films may demonstrate the aneurysmal areas as rounded or lobulated discrete lesions in the parenchyma. Often, tortuous vessels trace from these rounded areas to the hilum. Resection of the fistula, often involving lobectomy, is indicated if the lesion is localized. Unfortunately, some patients have such diffuse disease that this is impossible.

Rare Lesions

There are a variety of rare lesions of the lungs, including rare tumors and uncommon infections. Rare tumors, often identified on radiographs obtained for nonspecific symptoms, include primary sarcoma, pulmonary blastoma, hamartomas, and teratomas. Fungal infections, including actinomycosis, histoplasmosis, mucormycosis, and coccidioidomycosis, may look like tumors on chest radiograph. Atresia of the bronchus or pulmonary artery rarely occurs and produces differences in the lucency of the two lungs. The reader is referred to texts of pulmonary medicine or thoracic surgery for further discussion.

MEDIASTINAL TUMORS

Mediastinal Mass

At least one-third of all mediastinal masses occur in children younger than 15 years of age. Half of these masses are symptomatic, and 50% of the symptomatic masses are malignant tumors. More than 90% of the asymptomatic masses are benign. More than 95% of biopsied mediastinal masses in children are secondary to cysts or tumors. The mediastinum is commonly divided into anterior, superior, middle, and posterior compartments (Fig. 119.10). If only the anterior and middle mediastinal compartments of children are included, between 40 and 90% of the masses are malignant or cystic in origin. Neurogenic tumors are the most common cause of mediastinal masses, with lymphomas and germ cell tumors being second and third in frequency. Infection is an uncommon cause of mediastinal node enlargement, but, when present, is largely caused by histoplasmosis. Thymic enlargement may mimic an anterior mediastinal mass.

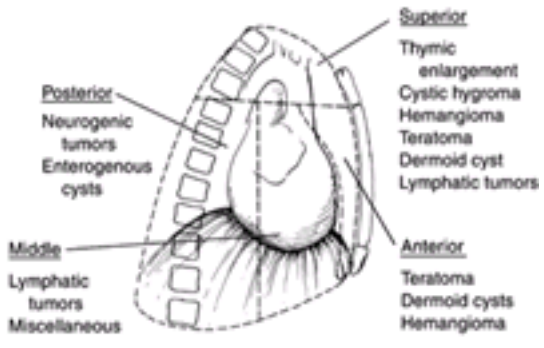

FIGURE 119.10. Mediastinal tumors in children. Differential diagnosis is based on anatomic location within the mediastinum.

Clinical Presentation

Mediastinal masses usually present with respiratory symptoms secondary to airway obstruction or erosion. As a result, patients may present with cough, wheezing, recurrent respiratory infections, bronchitis, atelectasis, hemoptysis, chest pain, or sudden death. Dysphagia and hematemesis may occur with compression of the esophagus. Superior vena cava syndrome is a rare complication, usually in association with a rapidly growing tumor. If the recurrent laryngeal nerve is compressed as a result of the mass, hoarseness and inspiratory stridor may result. Spinal cord compression syndrome and vertebral erosion can be seen with a posterior mediastinal tumor.

Management

Children with tumors of the anterior or superior mediastinum should be admitted to a hospital to undergo urgent evaluation. CT scan of the chest is almost always needed to supplement plain radiographs.

When biopsy of a large mediastinal mass is necessary, the logistics of biopsy require careful, thoughtful evaluation, ideally involving the pediatrician, surgeon, and anesthesiologist. Airway compression by large mediastinal masses is

often significant. If a general anesthetic is administered, the thoracic trachea may be occluded by tumor because the anesthetic eliminates the negative intrathoracic pressure caused by expansion of the chest wall. This situation can be difficult to manage; passage of a rigid bronchoscope may be necessary to stent the trachea open to allow gas exchange. Large mediastinal masses should be evaluated by CT scans to assess the likelihood of tracheal compression. MRI may be a better diagnostic tool for posterior mediastinal masses, because many of them are neurogenic in origin and extradural with extension into the spinal canal. Consideration should be given to the feasibility of biopsy under local anesthesia. The anesthesiologist should be apprised of the nature of the tumor, and a bronchoscope should be at hand if a general anesthetic is needed. Tissue may be obtained in a number of ways: 1) with a mediastinoscope, inserted via an incision above the sternal notch and passed behind the sternum; 2) a video-assisted thoracic surgery (thoracoscopy); or 3) a thoracotomy, usually a limited thoracotomy, often through an anterior interspace. In some cases, mediastinal masses can be accurately diagnosed by biopsy of supraclavicular or other extrathoracic adenopathy.

DIAPHRAGMATIC PROBLEMS

Congenital Diaphragmatic Hernia

Congenital diaphragmatic hernia (CDH) is the presence of intestinal viscera in the chest through an opening in the diaphragm not caused by trauma. About 90% occur on the left. They occur through the foramen of Bochdalek, which is at the back of the thoracic cavity. Herniation may occur through the foramen of Morgagni, which lies just posterior to the sternum and is even more rare than Bochdalek's hernia, comprising 2 or 3% of all diaphragmatic hernias. Traumatic diaphragmatic rupture may occur through any portion of the diaphragm and may present in a delayed time frame.

Pathophysiology

Most babies with congenital diaphragmatic hernia become symptomatic as newborns, when profound respiratory compromise leads to diagnosis. Until recent years, it was thought that the respiratory difficulties of babies with CDH were caused by mechanical compression of the lung by the intestinal viscera extruded through the diaphragmatic opening into the chest. It has become clear that this is not the case: pulmonary hypertension; surfactant deficiency; and a vicious cycle of hypoxia, acidosis, and intrapulmonary shunting lead to the death of about half of newborns with this diagnosis. CDH may be also identified after the neonatal period. These pathophysiologic situations are not present in older infants and children with the condition, who present with features of bowel obstruction, visceral ischemia, or pleural inflammation arising from sudden shift of abdominal viscera into the chest.

Clinical Presentation

When found in older babies and children, identification usually is by a chest radiograph obtained for nonspecific symptoms such as fever, cough, chest or abdominal pain, or vomiting. The presence of loops of intestine on the chest radiograph may be confirmed by passing a nasogastric tube, which will often end up with its tip in the thorax. The chest radiograph may suggest pneumonia with pneumatocele formation; in fact these “pneumatoceles” may be loops of bowel (Fig. 119.11). A gastrointestinal contrast study or chest and abdominal CT scan may clarify confusing findings. Potential intestinal or visceral ischemia caused by strangulation obstruction is one of the reasons operative repair is undertaken, and this possibility should be considered.

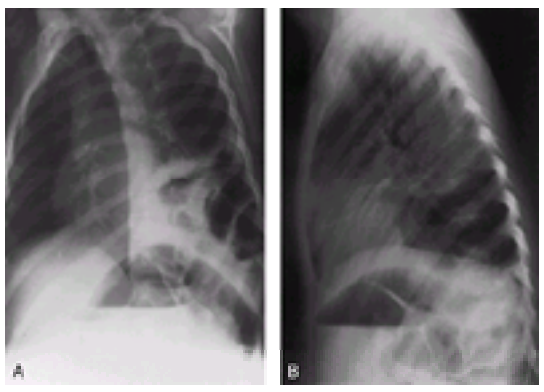

FIGURE 119.11. A 4-year-old boy admitted with 1-day history of recurrent severe upper abdominal colicky pain with dyspnea and decreased breath sounds in left base. Posteroanterior **(A)** and lateral **(B)** chest films demonstrate multiple bowel loops in the lower, posterior left chest, indicative of a foramen of Bochdalek hernia that was subsequently repaired without difficulty.

Management

Surgical repair should be undertaken soon after the diagnosis is made but may be elective in the asymptomatic patient. Because diagnosis is often made incidentally during evaluation for a condition such as pneumonia, which would increase risk of elective operation, the timing of surgery must be tailored to the individual situation. Certainly the pediatric surgeon should be consulted as soon as the diagnosis is suspected. If a patient is symptomatic from an acute ischemia of the herniated viscera, an urgent operation may be required. Usually, a subcostal abdominalincision is used, because it permits easier manipulation or resection of compromised intestine or other abdominal viscera, and allows for correction of the malrotation that usually accompanies this condition.

Foramen of Morgagni Hernias

Usually asymptomatic, an opening in the diaphragm just behind the sternum allows protrusion of abdominal viscera, usually including the colon, into the pericardium (Fig. 119.12). Described by Morgagni in 1769, this defect was also noted and repaired by Larrey, Surgeon General to Napoleon, and is sometimes called Larrey's hernia. Substernal or epigastric pain and bowel obstruction resulting from the narrow neck of the sac may occur spontaneously or be precipitated by any condition that increases intra-abdominal pressure.

FIGURE 119.12. Diaphragmatic defects in infants and children. The nature of these defects are often better appreciated on a lateral view of the chest. Eventration of the diaphragm **(A);** foramen of Morgagni hernia **(B);** and left foramen of Bochdalek hernia **(C).**

Clinical Findings/Management

A lateral chest radiograph should define the herniation as anterior, and suggest that the protrusion is not at the esophageal hiatus. A barium enema in stable patients should be considered. Surgical repair, indicated to prevent incarceration of bowel even in asymptomatic patients, may be performed through an upper abdominal incision.

Diaphragmatic Eventration

Often presenting to the emergency physician as an unexpected finding on a chest radiograph obtained for another reason, eventration of the diaphragm may be congenital or acquired. Acquired forms result from some form of phrenic nerve paralysis, which may be caused by birth, operative, or other trauma. Absent movement of the attenuated muscle produces atelectasis by decreasing the volume of the hemithorax.

Clinical Findings

Symptoms vary from mild wheezing to profound respiratory distress and are directly correlated in most cases with the size of the defect. Examination showing findings of non-aerated lung, including absent breath sounds and dullness to percussion, are investigated by chest radiograph. Chest radiographs usually confirm the presence of a high, rounded diaphragm shadow filled with bowel and other viscera involving more than 50% of the thoracic cavity on the involved side (Fig. 119.13). This study may be elucidated by observation of a diaphragm that does not move with respiration, which may be confirmed by fluoroscopy or ultrasound. Mediastinal shift may occur. Eventrations may be bilateral, but are more commonly on the left.

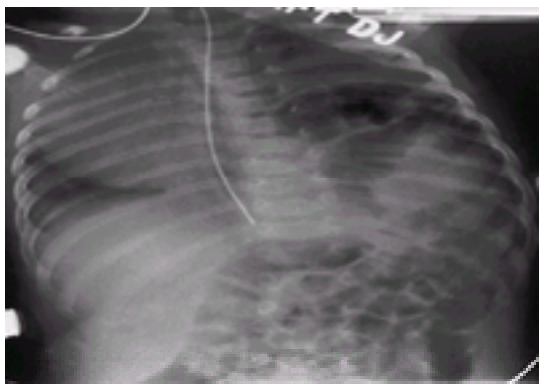

FIGURE 119.13. This 2-month-old girl was well until 4 days before admission. She developed congestion and an apparent upper respiratory tract infection. She slowly developed increasing dyspnea and was admitted in acute respiratory distress. A chest radiograph revealed a high left diaphragmatic eventration with a significant mediastinal shift to the right.

Management

Small degrees of eventrations incidentally identified and asymptomatic may be observed. Major diaphragmatic eventrations should be referred for repair, even in asymptomatic patients, to avoid collapse of the ipsilateral lung.

Acquired Diaphragmatic Malfunction

Clinical Findings

Paralysis of the diaphragm can occur as a result of injury to the phrenic nerve during birth or during a cervical or thoracic operation. In most cases, the paralysis is complete and the symptoms present shortly after the injury occurs. In the

newborn, a paralyzed diaphragm is particularly debilitating as air exchange is greatly impaired. The mediastinum is so mobile that a pressure differential allows the paralyzed diaphragm to rise paradoxically on inspiration, resulting in a shift of the mediastinum toward the normal side. Infants with only partial paralysis of the diaphragm may develop respiratory distress or pulmonary infections.

Management

As in a child with eventration of the diaphragm, an upright PA and lateral radiograph indicate the degree of diaphragmatic compromise (Fig. 119.14). Fluoroscopy may demonstrate the paralyzed portion of the diaphragm. If a trial of keeping the child in an infant seat at 60- to 75-degree elevation does not significantly improve the symptoms, resection and plication of the attenuated portion of the diaphragm should be performed. In selected cases, an implanted pacemaker can be used to stimulate the phrenic nerve and produce diaphragmatic motion.

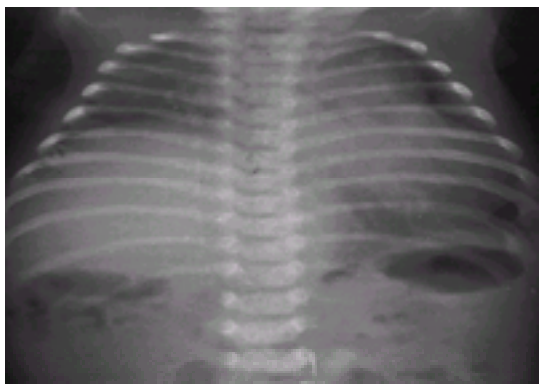

FIGURE 119.14. A 2-month-old girl with a brachial palsy at birth. She initially did surprisingly well but subsequently developed respiratory distress with cyanosis when out of oxygen. A chest radiograph showed a high attenuated right diaphragm.

Paraesophageal Hernia

Paraesophageal hernia is the protrusion of the stomach through an opening in the diaphragm that is not the diaphragmatic esophageal hiatus. It is extremely uncommon in children. When a part of the stomach migrates into the chest, it may become strangulated or kinked. Symptoms of vomiting and upper abdominal pain, tachypnea, and tachycardia may accompany the condition as the herniated stomach distends with swallowed air inside the chest.

Clinical Findings/Management

Chest radiographs show an air and fluid-filled mass in the left lower chest. A nasogastric tube may not pass into the stomach because of angulation of the gastroesophageal junction (Fig. 119.15). Surgical consultation should be sought because immediate laparotomy is likely necessary for reduction of a potentially strangulated stomach.

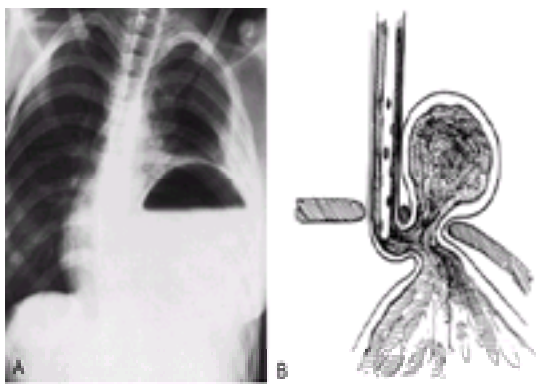

FIGURE 119.15. A. A 13-year-old girl developed first right and then left epigastric pain with retching but little or no vomitus. She had grunting respirations. A radiograph revealed a large air- and fluid-filled mass in the left lower chest. **B.** As shown in the diagram, a nasogastric tube would not pass into the stomach.

CHEST WALL TUMORS

Tumors of the chest wall occurring in childhood are likely to be malignant. Tumors at this site are uncommon in adults, and rare in children. Benign tumors, arising from the ribs in many cases, include aneurysmal bone cysts, chondromas, lipoid histiocytosis, osteochondromas, and osteoid chondromas. If the clinical and radiologic picture clearly indicates a benign, self-limited process, observation may be appropriate. However, if this is unclear and there is any concern that the lesion is not benign, even a small chest mass in a child should be considered malignant. Many malignant tumors may be present at birth and have been identified early in the first year of life.

Clinical Findings

Benign tumors of the chest wall are usually asymptomatic until trauma or fracture brings them to light. Malignancy is signaled by a rapid increase in size, pain, tenderness, or local inflammation. The site of the lesion may give a clue (Fig. 119.16). Ewing's tumor typically involves the lateral aspects of the ribs. Chondrosarcoma typically involves the costal cartilages between the sternum and the distal rib end. The sternum is a favored site for anaplastic sarcomas. These last two tumors may extend intrathoracically, as well as outside the chest cage. Chest radiographs may show pleural effusion, a mass adjacent to the pleura, or direct involvement of the lung.

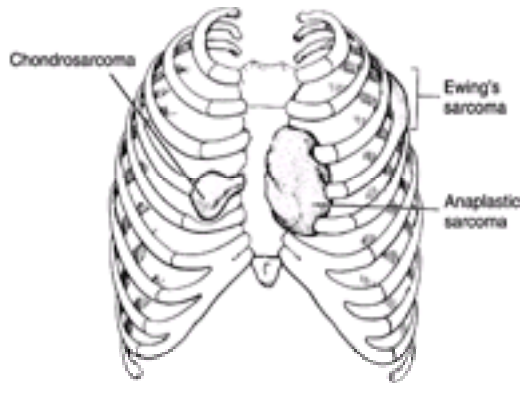

FIGURE 119.16. Malignant chest wall tumors in children. Most common lesions and their usual site of origin.

Management

Radiographic evaluation of these areas should include a CT scan of the pertinent area, bone scans of the entire body, and a metastatic bone survey. Multiple modality, coordinated treatment is usually required involving surgery, chemotherapy, and radiotherapy. If biopsy is anticipated before definitive wide resection, the route of the biopsy should be designed to avoid compromise of the subsequent chest wall reconstruction. Preoperative chemotherapy and radiotherapy may be useful to shrink particular lesions. Resection of the tumor and even subsequent recurrences have resulted in disease-free survivals of 15 years or more.

Suggested Readings

GENERAL

Chernick V, ed. Kendig's Disorders of the Respiratory Tract in Children. 5th ed. Philadelphia: WB Saunders, 1990.

Coran A, Fonkalsrud EW, et al, eds. Pediatric Surgery. 5th ed. Chicago: Year Book, 1998:1.

Greenfield LJ, Mullholand MW, Oldham KT, et al. Surgery: Scientific Principles and Practice. 2nd ed. Philadelphia: Lippincott-Raven, 1997.

Sabiston DC, Spencer FC, eds. Surgery of the Chest. 5th ed. Philadelphia: WB Saunders, 1990.

TRACHEAL OBSTRUCTION

Donnelly BW, McMillan JA, Weiner LB. Bacterial tracheitis: report of eight new cases and review. Rev Infect Dis 1990;12:729-735.

Schuster SR, Fellows KE. Management of major hemoptysis in patients with cystic fibrosis. J Pediatr Surg 1977;12:889-896.

Wenig BL, Abramson AL. Tracheal bronchogenic cyst. Ann Otol Rhinol Laryngol 1987;96:58-60.

BRONCHIAL OBSTRUCTION

Augustin N, Hofmann V, Kapherr S, et al. Endotracheal tumors in childhood. Prog Pediatr Surg 1987;21:136-144.

Bailey PV, Tracy T Jr, Connors RH, deMello D, Lewis JE, Weber TR. Congenital bronchopulmonary malformations. Diagnostic and therapeutic considerations. J Thorac Cardiovasc Surg 1990;99:597-603.

Eskenasy A, Eara-Iorgulescu L. Pathology of the middle lobe syndrome. Med Intern 1982;20:73-80.

Gupta DK, Rohatgi M, Chandna S, Bajpai M. Lobar emphysema in infancy. Indian Pediatr 1988;25:632-635.

Lazar RH, Younis RT, Bassila MN. Bronchogenic cysts: a cause of stridor in the neonate. Am J Otolaryngol 1991;12:119-121.

Livingston GL, Holinger LD, Luck SR. Right middle lobe syndrome in children. Int J Pediatr Otorhinolaryngol 1987;13:11-23.

Ramenofski ML, et al. Bronchogenic cyst. J Pediatr Surg 1979;14:219-224.

Senyuz OF, Danismend N, Erdogan E, Yeker D, Buykunat C, Oruc N. Congenital lobar emphysema: a report of 5 cases. Jpn J Surg 1989; 19:764-767.

Tapper D, Schuster S, McBride J, et al. Polyalveolar lobe: anatomic and physiologic parameters and their relationship to congenital lobar emphysema. J Pediatr Surg 1980;15:931-937.

Torres AM, Ryckman FC. Childhood tracheobronchial mucoepidermoid carcinoma. J Pediatr Surg 1988;23:367-370.

Wesley JR, Heidelberger KP, DiPietro MA, Cho KJ, Coran AG. Diagnosis and management of congenital cystic disease of the lung in children. J Pediatr Surg 1986;21:202.

ESOPHAGUS-RELATED CAUSES OF AIRWAY DIFFICULTIES: FISTULAE

Ghandour KE, Spitz L, Brereton RJ, Kiely EM. Recurrent trache-oesophageal fistula: experience with 24 patients. J Pediatr Child Health 1990;26:89-91.

Leithiser RE Jr, Capitanio MA, Macpherson RI, Wood BP. “Communicating” bronchopulmonary foregut malformations. Am J Roentgenol 1986;146:227-231.

Risher, WH, Arensman RM, Ochsner JL. Congenital bronchoesophageal fistula. Ann Thorac Surg 1990;49:500-505.

GASTROESOPHAGEAL REFLUX

Guzzetta PC, Rouse TM, Randolph JG. Gastroesophageal reflux and hiatal hernia in infants and children. In: Scott HW Jr, Sawyers JL, eds. Surgery of the Stomach Duodenum, and Small Intestine. Cambridge: Blackwell Scientific, 1992.

Mansfield LE. Gastroesophageal reflux and respiratory disorders: a review. Ann Allerg 1989;62:158-163.

Rode H, Stunden RJ, Millar AJ, Cywes S. Esophageal pH assessment of gastroesophageal reflux in 18 patients and the effect of two prokinetic agents: cisapride and metoclopramide. J Pediatr Surg 1987; 22:931-934.

ESOPHAGEAL WEB

Ibrahim NB, Sandry GJ. Congenital esophageal stenosis caused by tracheobronchial strictures in the esophageal wall. Thorax 1981;36:465-468.

Scherer LR, Grosfeld JL. Congenital esophageal stenosis, esophageal duplication, neurenteric cyst and esophageal diverticulum. In: Pediatric Esophageal Surgery. Orlando: Grune & Stratton, 1986:53-71.

CIRCULATORY COMPROMISE

Blalock A. Acute circulatory failure as exemplified by shock and hemorrhage. Surg Gynecol Obstet 1934;58:551-555.

Perkin RM. Shock States. In: Fuhrman BP, Zimmerman JJ, eds. Pediatric Critical Care. St. Louis: Mosby, 1992.

PLEURAL DISEASE

Beg MH, Reyazuddin, Faridi MM, Ahmad SH, Shahab T. Spontaneous pneumothorax in children: a review of 95 cases. Ann Trop Paediatr 1988;8:18-21.

Gustafson RA, Murray GF, Warden HE, Hill RC. Role of lung decortication in symptomatic empyemas in children. Ann Thorac Surg 1990; 49:940-947.

Light RW, MacGregor MI, Luchsinger PC, Ball WC Jr. Pleural effusions: The diagnostic separation of transudates and exudates. Ann Intern Med 1972;77:507.

McLaughlin FJ, Goldmann DA, Rosenbaum DM, Harris GB, Schuster SR, Strieder DJ. Empyema in children: clinical source and long-term follow-up. Pediatrics 1984;73:587-593.

Ramnath RR, Heller RM, Ben-Ami T, et al. Implications of early sonographic evaluation of parapneumonic effusions in children with pneumonia. Pediatrics 1998;101:68-71.

Singla R, Bhagi RP, Singh P, Anand E. Pulmonary metastases from Ewing's sarcoma presenting as spontaneous pneumothorax in a young child. Indian J Chest Dis Allied Sci 1988;30:148-153.

Stiles QR, Lindesmith GG, Tucker BL, Meyer BW, Jones JC. Pleural empyema in children. Ann Thorac Surg 1970;10:37-44.

Stovroff M, Teague G, Heiss KF, et al. Thoracoscopy in the management of pediatric empyema. J Pediatr Surg 1995;30:1211-1215.

LUNG LESIONS

Bailey PV, Tracy T Jr, Connors RH, deMello D, Lewis JE, Weber TR. Congenital bronchopulmonary malformations. Diagnostic and therapeutic considerations. J Thorac Cardiovasc Surg 1990;99:597-603.

Bentur L, Canny G, Thorner P, Superina R, Babyn BP, Levinson H. Spontaneous pneumothorax in cystic adenomatoid malformation. Chest 1991;99:1292-1293.

Berlinger NT, Porto DP, Thompson TR. Infantile lobar emphysema. Ann Otol Rhinol Laryngol 1987;96:106-111.

Dhamash NS, Chen JTT, Ravin CE, Reed JC, Pratt PC. Unusual radiologic manifestations of bronchogenic cysts. South Med J 1984;77:762.

Hendren WH, McKee DM. Lobar emphysema of infancy. J Pediatr Surg 1966;1:24-39.

Ribet M, Pruvot FR, Dubos JP, et al. Congenital cystic adenomatoid malformation of the lung. Eur J Cardiothorac Surg 1990;4:403-406.

Wagenvoort CA, Zondervan PE. Polyalveolar lobe and congenital cystic adenomatoid malformation type II: are they related? Pediatr Pathol 1991;11:311-320.

Wolf SA, Hertzler JH, Philippart AI. Cystic adenomatoid dysplasia of the lung. J Pediatr Surg 1980;15:925-930.

MEDIASTINAL TUMORS

Azizkhan RG, Dudgeon DL, Buck Jr. Life threatening airway obstruction and a complication to the management of mediastinal masses in children. J Pediatr Surg 1985;20:816-822.

Grosfeld JL, Weinberger M, Kilman JW, Clatworthy HW Jr. Primary mediastinal neoplasms in infants and children. Ann Thorac Surg 1971; 12:179-190.

Kelly CS, Kelly RE Jr. Lymphadenopathy in children. Pediatr Clin North Am 1998;45:August (in press).

Kenny JB, Carty HM. Infants presenting with respiratory distress due to anterior mediastinal teratomas. Br J Radiol 1988;61:241-244.

Mullen B, Richardson JD. Primary anterior mediastinal tumors in children and adults. Ann Thorac Surg 1986;42:338-345.

Silverman NA, Sabiston DC Jr. Primary tumors and cysts of the mediastinum. Curr Probl Cancer 1977;Feb.

ENTEROGENOUS CYSTS

Egelhoff JC, Bisset GS III, Strife JL. Multiple enteric duplications in an infant. Pediatr Radiol 1986;16:160-161.

Ildstad ST, Tollerud DJ, Weiss RG, Ryan DP, McGowen MA, Martin LW. Duplications of the alimentary tract. Clinical characteristics, preferred treatment, and associated malformations. Ann Surg 1988;208:184-189.

Robinson RJ, Pavlina PM, et al. Multiple esophageal duplication cysts. J Thorac Cardiovasc Surg 1987;94:144-147.

DIAPHRAGMATIC DEFECTS

Brouillette RT, Marzocchi M. Diaphragm pacing: clinical and experimental results. Biol Neonate 1994;65:265-271.

Dalvi R, Chari G, Fernandez AR. Congenital eventration of diaphragm. Indian J Pediatr 1990;57:125-127.

Malone PS, Brain AJ, Kiely EM, Spitz L. Congenital diaphragmatic defects that present late. Arch Dis Child 1989;64:1542-1544.

Obara H, Hoshina H, Jwai S, Ito H, Hisano K. Eventration of the diaphragm in infants and children. Acta Pediatr Scand 1987;76:654-658.

CHEST WALL TUMORS

Joseph WL, Fonkalsrud EW. Primary rib tumors in children. Am Surg 1972;38:338.

King RM, Pairolero PC, Trastek VF, Piehler JM, Payne WS, Bernatz PE. Primary chest wall tumors: factors affecting survival. Ann Thorac Surg 1986;41:597.

McAfee MK, Pairolero PC, Bergstralh EJ, et al. Chondrosarcoma of the chest wall: factors affecting survival. Ann Thorac Surg 1985;40:535.

CHAPTER 120

Ophthalmic Emergencies

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There are a number of ocular disorders in children that may be seen by the emergency physician. Although ophthalmology consultation may be necessary in some cases, many problems can be treated by the emergency physician. The goal of this chapter is to provide the emergency physician with a basic foundation for the diagnosis and treatment of some common pediatric eye emergencies.

EXAMINATION

Many children regard eye examinations and eye drops with the same fear that they harbor for injections. Therefore, it is important to gather as much information as possible before touching the patient or instilling eye drops. This can be accomplished by turning the initial parts of the eye examination into games, using toys and distracting stimuli, and exploiting the full potential of the direct ophthalmoscope, the main instrument for ophthalmic examination available to the pediatric emergency physician.

A detailed history can be an invaluable tool in focusing the examination and making a diagnosis. Questions regarding the unilaterality/bilaterality, acute/chronic onset of symptoms, and prior ophthalmic care are particularly helpful. Perhaps the child is known to have an eye with poor vision. Even if the parent does not know that this is the case, a history of having one eye patched for a visual problem suggests that the unpatched eye had amblyopia. However, if a child has previously passed his or her visual screening examination at school, this does not necessarily imply that the vision was normal because, false-negative tests are known to occur. The child may also be unaware of having poor vision in one eye because the pediatric brain is able to suppress recognition of the blurred image, allowing the child to proceed with normal activity unaware of the unilateral visual deficit (see the following).

The examiner may start the evaluation either by testing visual acuity in children who are readily verbal and interactive, or by using other techniques in children who need to be “warmed up.” In the latter case, it is often useful to start with assessment of the extraocular muscle movements. This procedure is discussed in Chapter 25. By using a toy or another interesting hand-held object, the physician can distract the child to look in the direction in which the object is placed. Both eyes should move equally, quantitatively and qualitatively, in all directions. The examiner should test gaze in all directions: up, down, left, and right.

After the eye movements have been assessed, the examiner can then use the direct ophthalmoscope as a tool to accomplish several tasks without touching the child. The direct ophthalmoscope light may be useful as a fixation target in testing eye movements. It can also be used to assess whether the eyes are aligned (Hirschberg light reflex test, see Chapter 25) and to test for a red reflex (see Chapter 111). In addition, the ophthalmoscope can be used as a simple hand-held magnifier by viewing through the ophthalmoscope and dialing the focusing wheel in the direction of the black or green numbers. This allows the eyeball to come into focus regardless of the distance between the examiner and the patient. The closer one gets, the greater the magnification.

Visual acuity testing is usually performed at a distance of 20 feet (6 meters) or 10 feet (3 meters). Most standard wall charts are calibrated to be read at 20 feet. However, if space does not permit this distance to be used, the patient can be placed 10 feet in front of the chart and the results interpreted with an adjustment for this system. For example, the line marked 20/60 on the chart (a line that a person with normal sight can see at 60 feet, but a person with that visual acuity would need to stand at 20 feet to see) becomes a 20/30 line at a distance of 10 feet.

Chart selection is important when trying to obtain an accurate visual acuity. Letter charts should be used only for patients who clearly can recognize all of the alphabet. If there is any question by parental report, a “tumbling E” or picture chart (Fig. 120.1) should be used. The patient may be asked to walk right up to the chart to identify the letters or pictures. If the patient can perform this task, the chart may be used for distance testing. Some children give remarkably unique interpretations of the pictures (e.g., calling the birthday cake a bag of French fries). When using picture charts that have colored figures, the examiner should avoid using those figures that are yellow because the bright illumination of the emergency department (ED) lessens the contrast between these figures and the white chart background, thus making recognition more difficult.

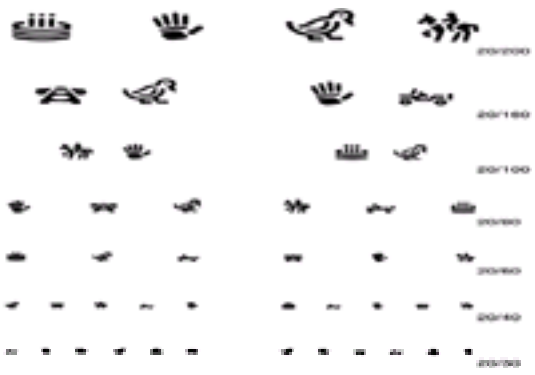

FIGURE 120.1. Picture visual acuity chart.

When using the tumbling E chart, it is important to give clear directions to the child. Otherwise, a falsely low visual acuity may be obtained. The child should be asked to point his or her fingers in the direction of the E (or call it table legs). Do not ask the child to use the words left and right because this can sometimes be confusing. The examiner may ask the child to perform this test up close to be sure that the instructions are clearly understood.

When using any visual acuity chart, it is not necessary to start with the largest symbol and have the patient read every symbol on every line thereafter. Doing so risks losing the child's attention. Rather, one can start with the 20/20 line and then go to larger lines if the child is having trouble. The child needs to recognize only a few letters on each line. Minor errors such as the substitution of the letter F for the letter P, or the letter C for the letter O, may be tolerated.

It is almost an instinct for young children to use the better eye and suppress the vision in the lesser eye. Therefore, if the good eye is covered inadequately, the patient will naturally try to read the chart with what the examiner thought was the covered eye. Children should never be allowed to cover their eye with their own hand because the small cracks between the fingers can actually allow vision out of the “covered” eye and even improve that vision by the pinhole effect (see Chapter 111). Children may also look around commercially available occluders for the same reasons. Perhaps the best way to obstruct the vision in the eye not being tested is to use a broad piece of tape, being sure that the tape also covers the depression at the bridge of the nose (Fig. 120.2). To help ensure that the patient is not “cheating,” the examiner should stand by the chart indicating the letters or pictures while simultaneously looking back at the child.

FIGURE 120.2. A broad piece of tape can be used to obstruct the vision of the eye not being tested. However, if the tape is not adherent to the bridge of the nose, the child can peek out by turning the face to the side *(right frame)*.

Any child who shows a reduced visual acuity should be offered the pinhole test as described in Chapter 111. If the patient is unable to identify any object or picture on the chart, the examiner should at least try to determine whether vision is present (see Chapter 111). After external examination and visual acuity are completed, the examiner can then proceed with other procedures as indicated, such as upper lid eversion and dilating the pupil. These techniques, along with the proper methods of examining the retina and optic nerve using the direct ophthalmoscope, are also discussed in Chapter 111.

One circumstance that may present an obstacle to proper examination of the eye is the situation in which the eyelids are swollen or the patient refuses to voluntarily open the eyelids. The techniques described in Chapter 111 for opening the traumatized eye may be useful. Commercially available speculums, when used in association with a topical anesthetic, are a painless and efficient way of opening the eyelids (see Fig. 111.5). If these are not available, either a Desmarres retractor (see Fig. 111.6) can be used or a similar device can be fashioned out of paper clips (Fig. 120.3). These types of single-blade retractors are most helpful when used on the upper eyelid (see Fig. 111.6). A retractor also can be applied simultaneously to the lower eyelids, although this often requires an extra assistant if there is a problem holding the child still while the eyelids are retracted. When paper clips are used, it is important to inspect the paper clip after bending the “blade.” Some paper clips have a coating that may become fragmented, potentially causing particles that could be dispersed on to the conjunctiva or cornea. Eyelid speculums and retractors should be sterilized between patients. Paper clip speculums are designed for single usage and may be prepared by cleansing with an alcohol swab.

FIGURE 120.3. Paper clips can be bent into a tool to open the eyelids.

In infants, the eyelids may be separated by using cotton swabs. The swabs should be placed at the midbody of the upper and lower eyelids. As they are separated, pressure should be applied against the eyelid and the swab should be rotated inward toward the eyelashes. This will keep the eyelids in place so that they do not spontaneously evert and further obstruct the examiner's view. When long cotton swabs are used, the stick should be grasped close to the patient to prevent breakage when pressure is applied. The cotton swab technique should not be used in patients being evaluated for eye trauma because pressure on the eyeball from this technique could cause further injury.

COMMON EYE EMERGENCIES

The pediatric emergency physician is called on to care for a number of eye problems. The reader is referred to other sections of this book for discussions of eye injuries (see Chapter 111), disorders of the eye muscle movement and strabismus (see Chapter 25), iritis (see Chapter 24 and Chapter 111), and unequal pupils (see Chapter 26).

Periorbital and Orbital Cellulitis

Clinical Manifestations

The primary concern when making the diagnosis of periorbital cellulitis (preseptal cellulitis) is to rule out the possibility of orbital cellulitis. The cardinal signs of orbital cellulitis include decreased eye movement, proptosis, decreased vision, and papilledema. Both orbital and periorbital infection may be associated with fever, pain, swollen eyelids, and red eye. If orbital cellulitis is suspected, computed tomography (CT) scanning of the orbit is indicated. However, the diagnosis of orbital cellulitis can be made clinically if the initial CT scan taken in the first 24 to 48 hours of infection is normal and clinical signs are present. Ophthalmology consultation is indicated in all cases of suspected or proven orbital cellulitis. Surgical intervention may be required. Otorhinolaryngology consultation should also be considered when orbital cellulitis is secondary to contiguous sinus infection.

Historical and clinical information can also be helpful in establishing the probable bacterial source of the infection. Infection secondary to a bug bite or other skin lesion that may have served as a route of entry for local bacteria is more often caused by staphylococci or streptococci. The bluish hue of the periorbital skin that is sometimes attributed to *Haemophilus-influenzae* is not a specific or sensitive indicator. In fact, since the introduction of the *H. influenzae* type b vaccine in 1985, the incidence of this pathogen as a cause for periorbital or orbital cellulitis has dropped substantially. It is now an uncommon cause.

One must rule out other conditions that can simulate a periorbital cellulitis. Insect bites and allergic reactions can cause dramatic acute periorbital swelling. However, these conditions are not usually associated with fever. Often, close inspection of the skin with magnification (using the direct ophthalmoscope) can localize the site of an insect bite. Allergic swelling is often bilateral, whereas periorbital cellulitis is rarely bilateral. Underlying sinusitis can also cause periorbital swelling. Some authors have argued that CT scan evaluation of the sinuses is indicated in all cases of presumed periorbital cellulitis. Severe conjunctivitis, especially adenoviral infection and neonatal gonorrhea conjunctivitis, can also result in significant lid swelling. The presence of conjunctival discharge is helpful in making these diagnoses. Contiguous spread of conjunctival infections to the periorbital tissues can occur and one must be careful about falsely eliminating the diagnosis of periorbital infection based on the presence of conjunctivitis.

Management

There is some controversy about the appropriate route of antibiotic administration in periorbital cellulitis. When *H. influenzae* was more common, with the risk of hematogenous spread, it seemed prudent to use intravenous (IV) antibiotics. Some clinicians are now becoming more liberal with oral antibiotic treatment now that the risk of *H. influenzae* has declined. In otherwise well children who are beyond infancy and have mild periorbital cellulitis and no systemic signs or symptoms, particularly when the cause of the cellulitis is believed to be a skin wound, intramuscular and/or oral antibiotics may be tried. The patient should be seen again (or with phone follow-up) within 24 to 48 hours, at which time improvement should be documented. If no improvement occurs, the patient should then be admitted for IV antibiotics. Periorbital cellulitis is a potentially fatal disease because complications such as meningitis may develop if inadequately treated. All cases of orbital cellulitis must be treated with IV antibiotics.

The choice of antibiotics should reflect the probable causative organism. Antibiotic coverage that would be used for presumed sepsis in an immunocompetent host with an unknown organism is usually appropriate. Before starting IV antibiotics, blood culture should be taken. Other systemic cultures (e.g., cerebrospinal fluid, urine) may be indicated if signs of systemic toxicity are present. Percutaneous aspiration from the area of cellulitis is not recommended.

Conjunctival cultures do not necessarily identify the causative agent of the cellulitis, but it may be reasonable to treat a predominant organism, particularly if purulent conjunctivitis is present. The patient should be monitored daily for signs of orbital cellulitis if improvement is not occurring.

Chalazions and Styes

Clinical Manifestations

Chalazions (internal hordeolum) and styes (external hordeolum) represent blocked glands within the eyelids. Both may present acutely with localized lid swelling, erythema, and tenderness. Styes are associated with swelling and purulent drainage at or near the lid margin (Fig. 120.4). More than one lesion may occur simultaneously and more than one lid may be involved. Acute chalazions cause swelling and redness in the body of the eyelid and may be associated with drainage on the conjunctival surface of the eyelid with or without a red eye. They may also point and drain via the skin (Fig. 120.5). Chalazions may enter a chronic phase in which there is a nontender, noninflamed, mobile pea-sized nodule within the body of the eyelid (Fig. 120.6). History can be helpful in establishing these diagnoses because patients often have had recurrent lesions at varying sites.

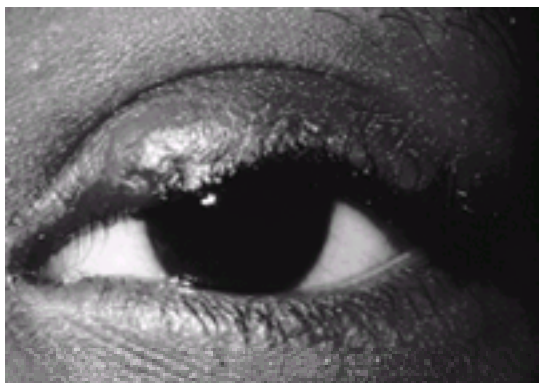

FIGURE 120.4. Acute sty (external hordeolum). *Please see the color-tip insert (Color Plate 120.4).*

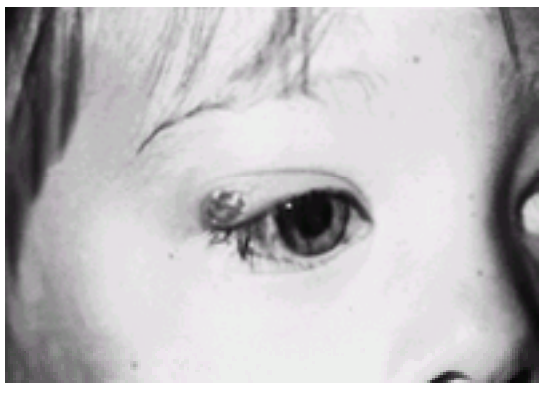

FIGURE 120.5. Chalazion draining spontaneously via skin. *Please see the color-tip insert (Color Plate 120.5).*

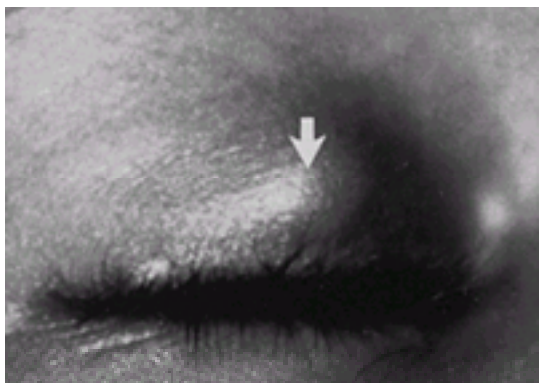

FIGURE 120.6. *Arrow* indicates chronic chalazion within upper lid.

Management

The treatment for both chalazions and styes is essentially the same. Eyelash scrubs with baby shampoo once or twice daily are helpful in mechanically establishing drainage. Baby shampoo is applied to a washcloth and then used to scrub the base of the eyelashes. Warm compresses are also useful, but rarely tolerated well by younger children. Optimally, warm compresses should be applied four times daily for 10 to 20 minutes at each sitting. The eyelid should be closed for both lid hygiene and warm compresses. Antibiotics probably play a minimal role in the treatment of styes and chalazions. If desired, a topical antibiotic ointment (Table 120.1) can be given twice daily following eyelash scrubs. If medical treatment has failed to cause adequate resolution after at least 4 weeks, surgery can be offered either for cosmesis or for

uncomfortable lesions. In severe recurrent cases, particularly when associated with red eye, oral erythromycin or tetracycline (8 years old) may be tried, although at this stage, ophthalmic consultation should be obtained.

Use	Avoid
Dilating drops	
Phenylephrine 2.5%	Scopolamine
Tropicamide 1%	Atropine
Cyclopentolate 1%	Homatropine
	Cyclopentolate 2%
Antibiotics	
Bacitracin	Neomycin
Erythromycin	Sulfacetamide
Polysporin	Aminoglycosides (except neonate)
Polytrim (trimethoprim/polymyxin B)	Quinolones
Lubricants	
Artificial tears/ointment	
Vasoconstrictors/antihistamines	
Albalon A, Naphcon A, Vasocon A	
Diagnostic Agents	
Topical fluorescein	
Anesthetic Agents	
Proparacaine, tetracaine	Cocaine
AVOID ALL ANTIVIRALS, MIOTICS (see Chapter 26), STEROIDS,* and ANTI-GLAUCOMA AGENTS	

*Including steroid-containing preparations, such as combination antibiotic-steroids.

Table 120.1. Pediatric Emergency Department Ophthalmic Drug Guidelines

Chemical Injury

Clinical Manifestations

When the child has a clear history of a noxious substance coming in contact with the ocular surface, it is important to determine whether this substance is an acid or an alkali. Alkali injuries tend to be much more severe. It is also important to determine whether particulate matter may have been deposited on the ocular surface. Smoke can also cause chemical conjunctivitis, particularly in house fires when chemicals are liberated into the air from the burning of plastics and other substances. The examiner must also assess the degree of exposure. If a child has no symptoms (pain, photophobia) or signs (red eye, epiphora, conjunctival swelling) and a weak history of actually getting the chemical into the eye, it may be acceptable to avoid lavage.

Management

Chemical injury to the eyeball is a true ocular emergency. Immediate intervention by ED personnel is essential to improving the patient's prognosis. Any patient with sufficient history should be immediately placed in the supine position so that ocular lavage may be started. Although a drop of topical anesthetic can make this procedure more comfortable, the physician should not wait for this to become available if it is not immediately handy. Usually, the irrigating solution itself will induce cold anesthesia. If a speculum, Desmarres retractor, or paper clip (see previous discussion) is readily available, this may be used to help obtain optimal exposure of the ocular surface. Again, the physician should not wait for these to become available. Virtually any IV solution can be used for ocular lavage, although normal saline solution or Ringer's lactate is perhaps preferable. A standard IV bag and tubing set is used without a needle on the end. Rather, the solution is allowed to flow, with the system at its maximum flow rate, across the surface of the open eye from medial to lateral. If both eyes have been exposed, they should both be lavaged simultaneously with two separate setups. Lavage should be continued until the involved eye(s) has received either 2 L of fluid or until approximately 20 minutes has elapsed. Lid eversion should be performed (Fig. 111.12) and lavage should be continued with the lid in this position so that the conjunctiva under the upper lid may also be cleansed. Mechanical debridement should be limited to the removal of visible particles from the ocular surface.

It is useful to have a strip of standard litmus paper available in the ED. The litmus paper is touched against the surface of each conjunctiva before beginning lavage. The pH is noted and the lavage is continued if, after the required minimum time/volume, the pH has not become normal (6.5 to 7.5) or equal between the two eyes. The end point of equality should only be used if one eye has not been exposed to chemical injuries. The conjunctiva under the upper lid may also be tested separately because noxious material can be harbored in the recess above the eye under the lid.

Ocular lavage can often be frightening to a child. If sedation can be administered promptly, it may be helpful. However, the physician should never wait for the effects of sedation before proceeding with lavage.

Ophthalmology consultation is usually indicated in cases of significant chemical injury. The consultant should be notified while lavage is ongoing. In cases of minor exposure to substances that are clearly not alkaline or strongly acidic, and when the eye is not injected, an ophthalmology consultation may be deferred. However, the physician must be cautious about the absence of conjunctival injection because alkali burns can cause blanching of the conjunctiva, which is a poor prognostic sign.

Conjunctivitis

Clinical Manifestations

Chapter 24 provides an approach for eliminating other causes of red eyes from the differential diagnosis. Table 120.2 is designed to give some additional help in differentiating causes of conjunctivitis. The patient's age is often useful in determining a diagnosis. Neonates presenting in the first 3 days of life can have a chemical conjunctivitis caused by silver nitrate used for ocular prophylaxis perinatally. Most hospitals have now discontinued this practice and many are using erythromycin ointment. However, this is not completely effective in eliminating subsequent gonorrheal or chlamydial conjunctivitis in the neonatal period. These two forms of conjunctivitis, as well as bacterial conjunctivitis secondary to enteric organisms, can be difficult to distinguish clinically. Each can present as either a mild purulent form or chronic

purulent conjunctivitis. A dramatically hyperacute conjunctivitis with significant lid swelling and copious purulent ocular discharge is more characteristic of gonorrhea (Fig. 120.7). In view of the risk of spontaneous corneal perforation associated with gonorrheal conjunctivitis, infants should be presumed to have this infection until proven otherwise. Immediate Gram stain should be performed looking for Gram-negative diplococci. If present, treatment for gonorrheal conjunctivitis should be started while awaiting culture results. In this age group, *Chlamydia* studies may be useful as well. Conjunctival scrapings are useful to look for inclusion bodies of chlamydial conjunctivitis. However, the sensitivity of this test depends on sampling, and the techniques may not be readily available or properly performed. Other methods to detect *Chlamydia* must always be used. Although rapid slide methods are approved for conjunctival samples (Syva), *Chlamydia* cultures are preferred because they increase diagnostic sensitivity. Even if *Chlamydia* is detected by Giemsa staining, this does not rule out the presence of concomitant gonorrheal infection.

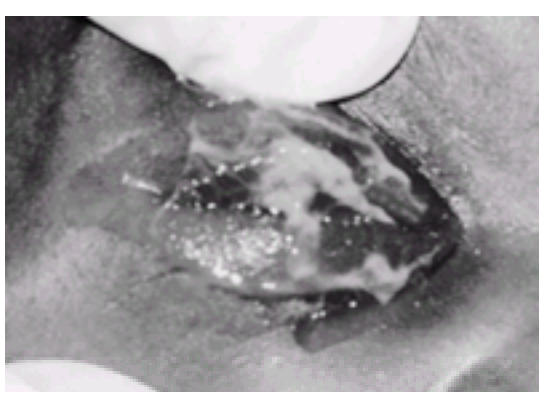

FIGURE 120.7. Neonatal gonorrheal conjunctivitis. Note the dramatic lid swelling and severe purulent discharge. *Please see the color-tip insert (Color Plate 120.7).*

	Bacterial	Viral (Nonherpetic)	Herpetic	Chlamydial	Allergic
Discharge—purulent	+++	+/-	-	+/-	-
Discharge—clear	-	+++	+++	+/-	++
Swollen lids	+++	+/0 to +++	+/0 to +++	+	+/0 to +++
Acute onset	++	++	+++	Chronic	Usually
Red eye	+++	+/0 to +++	Focally or diffuse +++	++	+
Cornea staining fluorescein	Nonspecific	Nonspecific	Dendrite	-	-
White cornea infiltrates	-	-	Possible	Multiple peripheral	-
Unilateral or bilateral	Unilat	Uni/bi	Uni	Usually bi	Usually bi
Contact history	-	+++	-	"STD"	-
Preauricular node	++	+++	-	+/-	-
Other associations	Otitis media? (H. influenzae)	Otitis media? Malaise, fever, pharyngitis	Prior or current skin lesions Recurrent	Genital discharge	Chronic

STD, sexually transmitted disease symptoms or contact.
Adapted from Lyons AM. Ophthalmology. In: Krugman SP, ed. The NYDC Handbook of Pediatrics. 18th ed. Toronto: Medsby; 1987.

Table 120.2. Differential Diagnosis of Conjunctivitis

In children beyond the neonatal period, a wide range of organisms, both viral and bacterial, as well as *Chlamydia,* can cause conjunctivitis. Clinically, these entities may appear to be similar. In general, purulence is more characteristic of bacterial infections, whereas clear serous discharge is more characteristic of viral infection. Although both viral and bacterial conjunctivitis may be unilateral or bilateral, a history of multiple infected contacts argues in favor of a viral etiology. Likewise, dramatic lid swelling, associated with preauricular adenopathy; mucoid or serous discharge; and perhaps an uncomfortable, sandy, foreign body sensation that affects one eye followed closely by the other, is strongly suggestive of epidemic keratoconjunctivitis secondary to adenovirus. This fulminant viral infection is easy to recognize (Fig. 120.8).

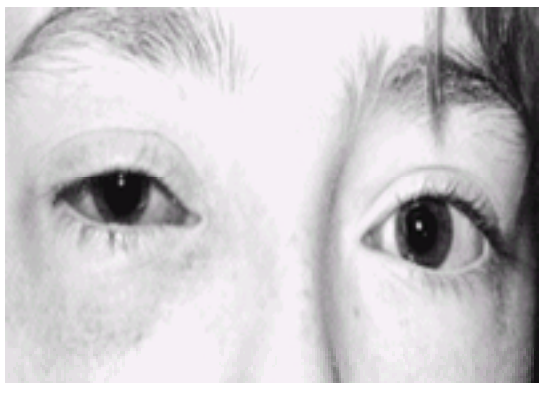

FIGURE 120.8. Patient with right epidemic keratoconjunctivitis infection. Note the lid swelling, red eye, and absence of purulent discharge. Patient also has right preauricular adenopathy (not visible). Note the early injection of left eye, representing sequential involvement. *Please see the color-tip insert (Color Plate 120.8).*

Viral culturing is rarely necessary. Bacterial cultures should be considered in all cases of purulent conjunctivitis, particularly when antibiotic treatment is going to be instituted. Gonorrheal conjunctivitis has been reported in prepubertal

children and in sexually active adolescents. Gram stains should be done in children with severe or persistent purulent conjunctivitis to rule out this infection.

Allergic conjunctivitis is usually a hyperacute conjunctival injection associated with tearing and a blister-like swelling of the conjunctiva (chemosis) (Fig. 120.9). Itching is often a prominent symptom, although this may also be a symptom of blepharitis (see Chapter 24). Conjunctival smears stained with Gram or Wright methods may reveal abundant eosinophils.

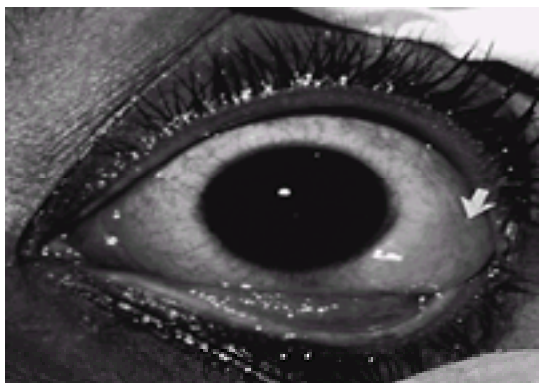

FIGURE 120.9. Allergic conjunctivitis.

Nasolacrimal duct obstruction is often confused with conjunctivitis because discharge may be present. However, the conjunctiva is rarely inflamed, indicating the absence of true conjunctivitis. The eye is white. The discharge is mostly mucus that has precipitated out of the tear film because of stagnation of tear flow. Patients are usually less than 1 year old, with a history of symptoms dating back to the first weeks of life. The discharge is usually worse on waking. Crusts may form on the lashes. Tearing may become more prominent after the first few months of life. Older children often have epiphora without discharge. The diagnosis can be confirmed by placing pressure on the lacrimal sac, which lies under the skin against the lacrimal bone between the medial canthus and bridge of the nose. This maneuver may cause an increase in the amount of discharge as it is forced out of the sac back onto the surface of the eye.

Management

Until proven otherwise, and in the presence of Gram-negative diplococci, neonatal purulent conjunctivitis should be treated as gonorrheal conjunctivitis, pending the results of cultures. The patient should be admitted for IV antibiotic therapy with cephalosporin (ceftriaxone 25 to 50 mg/kg, maximum 125 mg, intramuscularly or intravenously as single dose, or cefot1axime 100 mg/kg intramuscularly or intravenously as single dose), particularly in areas where penicillinase-producing strains are common. Ophthalmology consultation is indicated. Saline ocular lavage on an hourly basis may be helpful in decreasing the amount of organisms having access to the cornea. Topical erythromycin ointment is helpful because it will also treat *Chlamydia.* However, topical treatment alone is insufficient for either organism. If *Chlamydia* is laboratory proven, then the child must receive a 14- to 21-day course of oral erythromycin as well. This is necessary to eradicate carriage of *Chlamydia* in the nasopharynx, which can subsequently lead to pneumonia. The mother and father should be tested.

Any of the local antibiotics suggested in Table 120.1 would be appropriate for empiric coverage in treating a presumed bacterial conjunctivitis other than gonococcal while awaiting culture results. Gram stain can be helpful when narrowing down the possible causes, particularly when sheets of one predominant type of organism are seen. In the first 3 months of life, topical aminoglycosides might be a reasonable choice because Gram-negative and enteric organisms are more common.

If the patient clearly has a viral conjunctivitis, antibiotic treatment is probably not needed. Some physicians use antibiotics to “prevent secondary infection”; however, this is not a clinically significant problem in immunocompetent children. Rather, these patients are best soothed with cool compresses and artificial tear preparations. Depending on the virus, symptoms may last for weeks. Patients with symptoms that appear to be getting worse or persisting for longer than 1 week may benefit from ophthalmology consultation.

Allergic conjunctivitis is also helped by topical lubricants and cool compresses. The combination vasoconstrictor/antihistamine preparations listed in Table 120.1 may also be prescribed. Patients with recurrent allergic conjunctivitis, atopy, or asthma may benefit from long-term or seasonal topical mast cell stabilizers. Ophthalmology consultation may be useful, especially when symptom relief is not obtained or pain and red eye are present.

Any patient with a history of herpetic ocular infection and any patient who wears contact lenses and has conjunctivitis should be referred immediately for ophthalmology consultation. Herpetic corneal infection is usually painful. Patients may or may not have a history of skin lesions. Characteristic fluorescein dendritic staining patterns can be seen on the cornea or conjunctiva (Fig. 120.10). However, even if there is no staining but a history of herpetic (varicella-zoster or simplex) corneal infection, urgent ophthalmology consultation is essential. However, skin lesions on the lids without any conjunctival injection does not require ophthalmology consultation.

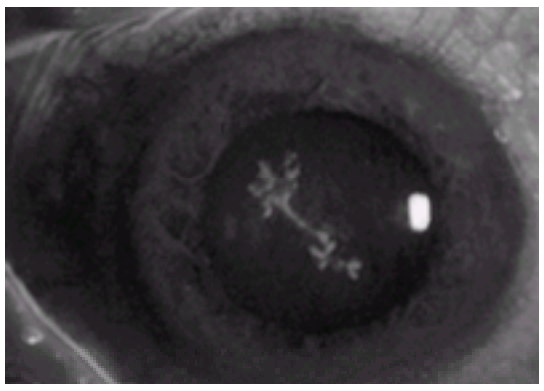

FIGURE 120.10. Fluorescein staining pattern of herpes simplex virus corneal infection. Eye is illuminated with blue light to demonstrate yellow/green branching staining pattern of herpetic dendrite. *Please see the color-tip insert. (Color Plate 120.10)*

DRUGS

Table 120.1 is designed to give emergency physicians some guidelines regarding the prescription and use of ophthalmic medications. Those drugs that should be avoided are listed because of problems with ocular toxicity, systemic toxicity, undesirable selection of resistant organisms, or the need for ophthalmology consultation and management regarding the problem that those drugs are designed to treat. In addition, the following guidelines should be adhered to:

1. No local drugs should be prescribed to patients who wear contact lenses without the supervision and consultation of an ophthalmologist.
2. Topical anesthetics must never be prescribed for outpatient use. These are strictly diagnostic agents. Prolonged use of topical anesthetics may result in corneal ulceration.
3. Steroids should never be prescribed by the emergency physician. Inappropriate use of steroids may lead to glaucoma, cataract, increased severity of corneal viral infection, or rebound symptoms when the drug is discontinued.

Instillation of eye drops can sometimes be difficult because of swollen eyelids or noncompliance from the patient. It will sometimes be noted that some of the drop is expelled upon blinking after drop instillation. This is not an indication for repeat instillation because only approximately 20% of an eye drop is actually absorbed for use. Most ophthalmic solutions are designed for a one-drop dose. Drops are most efficiently delivered by pulling down the lower eyelid and placing the drop in the inferior fornix. In patients who are extremely resistant, forced eyelid opening is needed to expose just a small strip of palpebral conjunctiva. The same techniques as described previously and in Chapter 111 for opening the eyelids may be used for the administration of eye drops in the ED. The eyeball itself need not be visualized. An alternative technique involves placing the eye drop in the sulcus between the medial canthus and the bridge of the nose while the patient is in the supine position. Every child must eventually open his or her eyes and when this happens the eye drop will naturally flow onto the conjunctiva.

Topical anesthetics do sting for approximately 10 to 20 seconds before taking effect. This may still be more desirable than the severe sting associated with dilating drops. In addition, the placement of a topical anesthetic before instillation of dilating eye drops increases the effectiveness of the latter.

When an eye drop and ointment are to be used simultaneously, the solution should always be instilled before the ointment. Ophthalmic ointments are applied by placing a strip of ointment along the conjunctiva of the lower lid. When treating styes and chalazions located within the eyelid, ointment can be placed on the lashes or conjunctiva.

Suggested Readings

Gigliotti F, Williams WT, Hayden FG, et al. Etiology of acute conjunctivitis in children. J Pediatr 1981;98:531–536.

Hammerschlag MR. Conjunctivitis in infancy and childhood. Pediatr Rev 1984;5:285–290.

Lessner A, Stern GA. Preseptal and orbital cellulitis. Infect Clin North Am 1992;6:933–952.

Levin AV. Eye emergencies: acute management in the pediatric ambulatory care setting. Pediatr Emerg Care 1991;5:367–377.

Matoba A. Ocular viral infections. Pediatr Infect Dis 1984;3:358–368.

Sadow KB, Chamberlain JM. Blood cultures in the evaluation of children with cellulitis. Pediatrics 1998;101:e4.

Siegel JD. Eye infections encountered by the pediatrician. Pediatr Infect Dis 1986;5:741–748.

Stenson S, Newman R, Fedukowicz H. Laboratory studies in acute conjunctivitis. Arch Ophthalmol 1982;100:1275–1277.

CHAPTER 121

Otolaryngologic Emergencies

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The ear, nose, and throat are common sites for infection and neoplasms and may be the sources of acute pain. This makes the head and neck region one with which the emergency medicine specialists must be familiar because they are called on to evaluate this area frequently. Although the presenting complaints may seem extremely distressing to the patient and cause considerable anxiety for the parents, the diseases prompting the visit are rarely life-threatening. This chapter includes discussion of disorders of the ear, nose, nasal sinuses, oral cavity, pharynx, esophagus, larynx, trachea, and neck.

EAR

Methods of Examination

Every emergency department (ED) should have an otoscope with good illumination, wax loop (curette), illuminated head light, and otologic forceps.

Examination of the ear begins by inspection of the auricle and surrounding areas. The external meatus should be visualized directly with a bright light after it is fully opened by pulling the pinna posteriorly and superiorly. The tragus may be displaced forward by traction on the skin in front of the ear with the examiner's other hand (Fig. 121.1). The ear canal can then be examined with a pneumatic otoscope, using the largest speculum that will fit in the meatus without discomfort. Wax or debris occluding the ear canal should be removed with a curette or by repeated irrigation with body-temperature water (see Procedure 5.3 in Section VII). Irrigation of the canal should not be done if a ventilating tube is in place or if a perforation of the tympanic membrane (TM) is suspected.

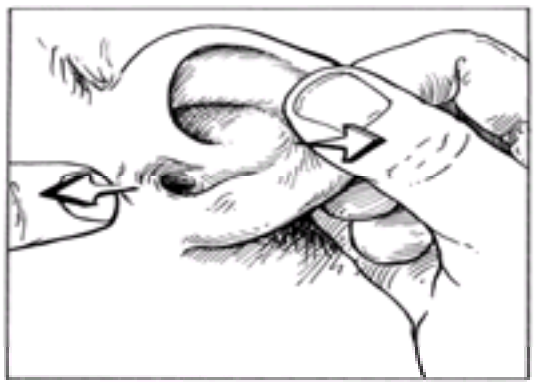

FIGURE 121.1. The external meatus is opened by pulling the auricle in the posteriorsuperior direction and placing traction on the skin immediately in front of the tragus.

The TM should be evaluated for its appearance, but the examination should not stop there. Part of the middle ear contents can often be seen through a translucent ear drum (Fig. 121.2). Mobility should be assessed with the pneumatic otoscope. Pneumatic otoscopy is performed by applying positive and negative pressure to the TM with the pneumatic otoscope fitted snugly into the ear canal. The ear pressure can be varied by squeezing a rubber bulb or blowing through a tubing connected to the otoscope head (see Procedure 5.1 in Section VII).

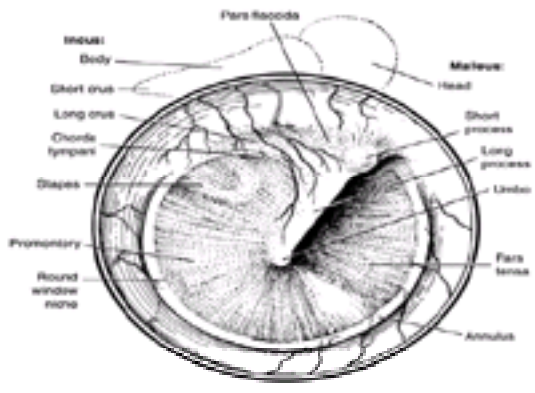

FIGURE 121.2. Right tympanic membrane.

The ear of a neonate requires special attention to perform an adequate otologic examination. The ear canal itself is narrow and collapsible. Often, the otoscopic speculum can only be inserted as positive pressure from the pneumatic bulb distends the canal ahead of the advancing speculum. The canal may be filled with vernix caseosa, which must be removed or irrigated out of the canal to permit visualization of the TM. The neonate's TM lies at a more oblique angle to the ear canal (compared with older children) and may make recognition of the TM and its landmarks more difficult.

Crude hearing acuity can be tested with a ticking watch or a 512-Hz tuning fork. The sound should be heard equally in each ear. However, this does not rule out a symmetric bilateral hearing deficit. If the tuning fork is applied to the forehead, it should be heard equally in both ears. If it is heard only in one ear, it signifies either a conductive loss in the ear that hears the tone or a sensorineural loss in the opposite ear. Audiometry (behavioral or evoked response) is required for an accurate evaluation of hearing.

Infections

Acute Otitis Media

Acute otitis media (AOM) is one of the most common head and neck infections in children and is the second most common diagnosis in the ED. It may occur as an isolated infection or as a complication of an upper respiratory infection (URI). Otitis media with effusion (OME), which is noninfected fluid in the middle ear (also called serous otitis media or secretory media), and immunodeficiency states predispose children to recurrent AOM.

The most common organisms causing acute otitis at all ages are *Streptococcus pneumoniae, Haemophilus influenzae,* and less often, *Moraxella catarrhalis,* group A b-hemolytic streptococcus, and various upper respiratory viruses. Gram-negative organisms may occur in hospitalized patients who are younger than 8 weeks of age or immunosuppressed.

Clinical Manifestations

AOM should be suspected in any child who is irritable or lethargic. The pain develops rapidly and is often severe. Spontaneous perforation of the TM with serosanguineous drainage may occur in less than 1 hour after the onset of pain. AOM is best diagnosed by pneumatic otoscopy. The TM is hyperemic, and mobility is decreased. As the drum becomes more edematous, it bulges outward and the landmarks may become unrecognizable. Infection with *Mycoplasma pneumoniae* and other bacteria may cause blebs on the lateral surface of the drum. The vesicles of bullous myringitis are filled with clear fluid and are painful. The appearance of the TM in AOM secondary to bacterial pathogens does not differ

significantly from AOM of viral etiology.

Complications

The following complications of AOM may be encountered in the ED:

1. The purulent exudate that fills the middle ear space causes a conductive hearing loss. The congealed exudate may organize and stimulate hyalinization and calcification, leading to tympanosclerosis (white patches on the undersurface of the TM).
2. Spontaneous perforation of the TM usually produces a small hole that heals rapidly; however, large perforations may occur that do not heal even after the infection has cleared.
3. Ossicular necrosis may also occur, causing a persistent conductive hearing loss.
4. As the TM heals after a perforation, skin from the lateral surface of the TM may be trapped in the middle ear to form a cyst (cholesteatoma) that can expand and destroy the structures of the middle ear and surrounding bone.
5. Facial nerve paralysis may occur suddenly during AOM. The nerve paralysis may be partial or complete when the child is first examined. The facial nerve usually recovers complete function if appropriate systemic (intravenous [IV] followed by oral) antibiotic therapy is administered and a wide myringotomy for drainage is carried out as soon as possible.
6. AOM may cause inflammation in the inner ear (serous labyrinthitis). This causes mild to moderate vertigo without a sensorineural hearing loss.
7. Bacterial invasion of the inner ear (suppurative labyrinthitis) causes severe vertigo and sensorineural hearing loss.
8. Pus always fills the mastoid during AOM, causing radiographic opacification , but acute suppurative mastoiditis (acute coalescent mastoid osteomyelitis) may develop, causing destruction of the mastoid air cell system. As the infection spreads to the postauricular tissues, subperiosteal collection of purulent material displaces the auricle laterally and downward from its normal position. The pus may extend through air cells to the medial portion of the temporal bone, causing sixth cranial nerve paralysis, deep retro-orbital pain, and otorrhea (Gradenigo's syndrome). Pus may also break through the mastoid tip and extend into the neck (Bezold abscess).
9. The most common intracranial problem associated with AOM is meningitis, which may be associated with severe sensorineural deafness and irreversible vestibular damage. Less commonly associated problems are cerebritis, epidural abscess, brain abscess, lateral sinus thrombosis, and otitic hydrocephalus. The child with overt or impending intracranial complications should be stabilized and have a computed tomographic (CT) or magnetic resonance imaging (MRI) scan.

Management

The treatment of uncomplicated AOM is oral antibiotic therapy with amoxicillin (25 to 50 mg/kg per 24 hours in three divided doses for 10 days). Systemic or local antihistamine–decongestant preparations are of no proven value. Patients with complicated AOM should also be treated with a wide inferior myringotomy for drainage, usually best performed by an otolaryngologist. Alternatively, in neonates, in immunosuppressed patients, and in cases in which antibiotic therapy is not effecting resolution of the infection, tympanocentesis (see Procedure 5.2 in Section VII) may be performed for Gram stain and culture. Patients treated on an emergency basis for AOM should be referred for follow-up examination in 2 weeks after therapy is started. Children with persistent OME, complications of middle ear disease, or recurrent bouts of AOM that do not respond to a 6- to 8-week course of antimicrobial treatment/prophylaxis should be referred to an otolaryngologist for evaluation for possible surgical treatment.

External Otitis

External otitis usually follows swimming and is often called swimmer's ear. Ear canal trauma or foreign bodies may also contribute to the development of external otitis.

Otitis externa may be localized or diffuse. Localized external otitis is the result of an abscessed hair follicle in the outer two-thirds of the ear canal. These abscesses are caused by *Staphylococcus aureus.*

Diffuse external otitis is caused by *Pseudomonas aeruginosa,* staphylococci, fungi, or a mixture of Gram-negative and Gram-positive organisms. Viral external otitis is usually caused by herpes simplex or herpes zoster.

Clinical Manifestations

External otitis usually begins with itching and fullness that progress to severe pain. The pain is worsened by chewing or by touching the ear. The external canal is red, edematous, and narrowed. The diagnosis of external otitis is usually readily made by external inspection and otoscopy. Otoscopy may be painful, and visualization of the eardrum may be impossible because of edema of the canal walls. A foul-smelling, purulent discharge is usually present. Surrounding cellulitis and regional cervical adenitis may also be present. Malignant external otitis occurs rarely in debilitated patients who have diabetes or are immunosuppressed. It may cause extensive tissue necrosis and can be rapidly fatal if not treated immediately with antibiotics and surgical debridement.

Management

If the abscess in localized external otitis is about to drain spontaneously, it should be opened where it is pointing with an 18-gauge needle, or a No. 11 scalpel blade. Drainage results in immediate relief of pain. Antibiotic therapy with an antistaphylococcal antibiotic (e.g., erythromycin, dicloxacillin, or a cephalosporin) should be administered for 10 days. The treatment of diffuse external otitis is to use antibiotic ear drops containing neomycin, polymyxin, and hydrocortisone (4 drops, three times daily) in the affected ear for 10 days. Before the drops are started, the pus and debris should be cleaned from the ear canal with gentle suction, a curette, or cotton-tipped applicators. If the meatus is so swollen that drops can not enter the external canal, a wick of gauze or Merocel sponge should be gently advanced into the ear canal

with a forceps (Fig. 121.3) to facilitate instillation of the topical medicine. The wick should be left in place for 24 to 48 hours, by which time the canal swelling should resolve to permit entrance of the drops. Broad-spectrum systemic antibiotics should be used if cellulitis or regional cervical adenitis is present. No water should be allowed to enter the ear canal during the 10 days of therapy.

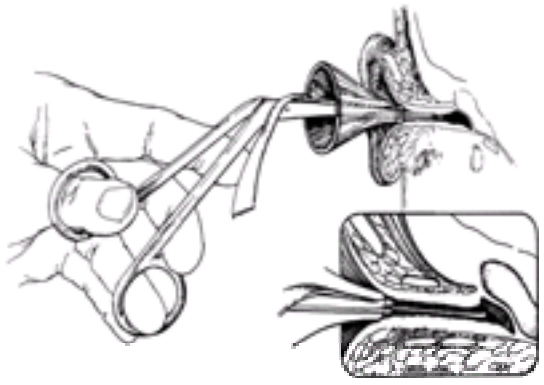

FIGURE 121.3. Gauze wick (¼ × 1½ inch) being placed in ear canal to facilitate topical treatment of otitis externa.

Chronic Otitis Media

Chronic otitis media (COM) is a persistent perforation of the tympanic membrane of more than 3 months' duration; the perforation may be acquired (from AOM or trauma) or iatrogenic (by tympanostomy tube) and may or may not be associated with active infection. When infection is present, the causative organism is usually *P. aeruginosa* or *S. aureus,* and it presents with a profuse, foul-smelling discharge. Any perforation may be associated with a cholesteatoma (white skin-lined cyst) that can destroy the structures of the ear as it expands.

Clinical Manifestations

COM is usually diagnosed by otoscopy. A perforation of the eardrum is readily seen and the white, pearly, flaky debris from a cholesteatoma may also be present.

Management

Dry perforations require no active treatment. When otorrhea is noted, antibiotic-containing ear drops (4 to 5 drops, three times daily) should be placed in the ear canal. Systemic antibiotics are of limited value unless regional cellulitis or cervical adenitis is present. In those cases, an antistaphylococcal antibiotic (e.g., erythromycin, dicloxacillin, or cephalosporin) should be administered for 10 days. Chronic perforation and cholesteatoma require surgical correction. All cases should be referred to an otolaryngologist for definitive management.

The complications of COM with infection are the same as those that occur in AOM, including intracranial spread of the infectious process. In addition, with repeated infection, a progressive high frequency sensorineural hearing loss may develop.

Infection of the Pinna

The pinna may become infected in a fashion similar to skin surfaces anywhere else on the body (see Chapter 84). Preauricular cysts and sinuses may occasionally be infected with *S. aureus* and should be treated with antistaphylococcal penicillin or cephalosporin for 10 days. If an abscess forms, it should be drained surgically. Infected preauricular sinuses/cysts require surgical excision once the acute infection has been treated.

Sudden Hearing Loss

Sudden hearing loss is not a common complaint in the ED, but it requires prompt attention, especially if the loss is determined to be sensorineural. Sudden conductive losses almost never occur without a known antecedent event such as head trauma, ear infection, or wax occlusion of the ear canal. History and otoscopy can usually establish the cause of the conductive hearing loss. However, the cause of sensorineural sudden hearing loss is obscure when the history is unrevealing and otoscopy is normal. Tuning fork testing helps confirm the presence of a sensorineural hearing loss.

Sudden sensorineural hearing loss that occurs after an airplane trip, scuba diving, straining, or head trauma is highly suggestive of a perilymph fistula. A perilymph fistula occurs when inner ear fluid leaks out into the middle ear through a rupture in the round window or stapes footplate (oval window). The leaking fluid causes a fluctuating sensorineural loss and vertigo. Urgent surgical exploration of the middle ear is required for repair.

Sudden sensorineural deafness may occur without a history suggestive of a fistula and without otoscopic abnormalities. This is often secondary to a viral infection of the cochlear labyrinth. Measles, mumps, and cytomegalic viral illnesses are common causes of sudden sensorineural deafness. Other viruses may injure the cochlea as well. There may be no systemic symptoms or signs of such a viral infection. These patients may have partial or complete recovery of hearing over several weeks. There is no proven effective treatment for sudden hearing loss. Aspirin has been recommended (in older children) to decrease platelet aggregation and to maintain patency of the cochlear blood vessels, and corticosteroids have been recommended by some authors. Other treatments have been proposed (e.g.,

cyclophosphamide, hyperbaric oxygen, inhaled CO_2), but are of uncertain efficacy. Antivertigo medications may be prescribed for patients experiencing dizziness. All patients with a sudden sensorineural hearing loss should be referred to an otolaryngologist.

Vertigo

Sudden vertigo is a disturbing and sometimes confusing symptom. A child may be brought to the ED because the parents think he or she is having a seizure. Vertigo may follow dysfunction of any part of the vestibular system from the labyrinth to the vestibular cortex.

Vertigo may be associated with a number of conditions affecting the middle ear:

1. Serous labyrinthitis may develop in a child with OME, AOM, or COM. Pressure and infection in the middle ear may cause inner ear inflammation and vestibular dysfunction. The conductive hearing loss and the dizziness resolve when the middle ear pressure is normalized or the inflammation subsides.
2. Suppurative labyrinthitis may occur when bacteria invade the inner ear. Severe vertigo and a profound sensorineural hearing loss result.
3. When a cholesteatoma arises in association with COM, it may invade the bony wall of the labyrinth. Pneumatic otoscopy may produce the sensation of vertigo by transmitting the pressure directly to the inner ear.
4. A common cause of sudden vertigo is vestibular neuronitis. The origin of this entity is uncertain, and the vertigo resolves spontaneously over several weeks. There are no accompanying signs or symptoms.
5. Trauma can be associated with vertigo in several ways. Perilymph fistulae, which occur most often after barotrauma, blunt head trauma, or straining, produce vertigo that fluctuates in severity. However, head trauma may also cause labyrinthine concussion or hemorrhage (hemorrhagic labyrinthitis), resulting in vertigo. Cerebral injuries involving the temporal lobe (with or without temporal bone fracture) can also cause vertigo.
6. Measles and mumps may also infect the inner ear and cause vertigo.
7. Meniere's disease (endolymphatic hydrops) is rare in children. Its origin is unknown. The symptoms are intermittent vertigo, tinnitus, a feeling of fullness in the ear, and fluctuating hearing.
8. Miscellaneous causes of sudden vertigo in children include benign paroxysmal vertigo of childhood and retrolabyrinthine lesions such as tumors, demyelinating diseases, and temporal lobe seizures.

The emergency physician should be reminded that vertigo is only a symptom of an underlying disease. Emergency treatment should consist of searching for the underlying disease, as well as providing symptomatic relief. Vertigo is rarely associated with a life-threatening illness. Because sensorineural hearing loss usually accompanies serious causes of vertigo, its absence can provide some level of confidence that no life-threatening disease is present. (See Chapter 21 for further discussion.)

Neoplasms

Neoplasms of the external ear are as varied as the tissue types of the auricle and are not difficult to diagnose because they are so visible. Neoplasms of the middle and inner ear are rare, but bear mentioning because they are often missed until they are far advanced. External canal and middle ear tumors are most often brought to the physician's attention because of painful secondary infection that does not respond to conventional treatment of topical and systemic antibiotics. The examiner may overlook a tumor, assuming that it is granulation tissue caused by an infection or related to a ventilating tube. If an ear infection does not respond to appropriate treatment or is associated with any abnormal-appearing tissue, a tumor should be suspected; otolaryngologic consultation should be made to obtain a biopsy of the abnormal tissue.

Inner ear tumors are deceptive in their early stages and are rarely detected until they cause either hearing loss, vertigo, or focal neurologic signs. The most common of these tumors are neural sheath tumors of the eighth nerve that cause progressive sensorineural hearing loss, vertigo, and fifth nerve anesthesia.

Facial Nerve Paralysis

Facial nerve paralysis is a frightening occurrence in children. Bell's palsy (idiopathic facial paralysis) is the most common cause of facial paralysis. (See Chapter 83 for management of this presumed viral infection.) A child presenting with facial paralysis must have a careful examination to detect any other treatable cause for the nerve dysfunction. Facial paralysis secondary to AOM requires a course of systemic (24 to 48 hours of IV followed by oral) antibiotics and an urgent wide-field myringotomy for drainage. Temporal bone or facial trauma and neoplasms of the middle ear and parotid area can also present with facial nerve paralysis. A child with a facial nerve paralysis should be referred to an otolaryngologist for a complete evaluation of the head and neck, audiogram, and radiographic imaging.

NOSE AND PARANASAL SINUSES

Methods of Examination

The external nose and anterior portion of the nasal cavities can be examined by direct visual inspection. A nasal speculum and directed light source are necessary to permit good visualization of the anterior septum and inferior and middle turbinates. In younger children, the examiner's thumb can elevate the mobile nasal tip to allow adequate inspection of the anterior nasal structures. Vasoconstrictors such as 0.25% phenylephrine or 0.05% oxymetazoline (2 or 3 drops) can be applied to the nose to shrink the mucosa and allow a more complete examination. The posterior nasal structures and nasopharynx can be seen with the aid of a flexible fiberoptic endoscope placed in the nose or the posterior oropharynx (see Procedure 7.5 in Section VII). Patency of the nasal cavities in the neonate can be assessed by the passage of small rubber catheters through the nose and into the pharynx. Palpation is also important in the

evaluation of nasal and facial trauma. Tenderness to palpation over the sinuses is a common sign of acute sinusitis.

A careful examination of adjacent areas is important when evaluating a child with sinus disease. Dental pathology must be detected in the search for a possible cause of a bacterial maxillary sinusitis. An examination of the orbit with assessment of visual acuity and ocular mobility should be performed to detect possible orbital complications of sinus disease.

Radiographs are indispensable in evaluating diseases of the nose and sinuses. Plain films (sinus or facial series) are used as screening devices to evaluate a mass or fluid in a sinus, but CT scans are indicated for more precise and detailed evaluation of sinusitis or tumors of this area.

Infections

Infections of the nose and paranasal sinuses are most often a component of the common URI. The symptom complex of fever, nasal congestion/rhinorrhea, and headache is most often caused by a viral agent. Physical examination often reveals swollen, erythematous nasal turbinates. The rhinorrhea can be clear or white in color. Facial tenderness is usually absent. Viral rhinitis requires little more than supportive care with hydration, rest, and antipyretics; oral antihistamines and/or decongestants are thought by some to provide additional relief. Topical decongestants are to be avoided because of their tendency to cause rebound congestion as their vasoconstricting effect on the nasal mucosa wears off.

Bacterial infection of the nose and paranasal sinuses is a more serious condition and requires a careful examination and prompt treatment. Bacterial sinusitis should be suspected when the nasal discharge lasts more than 7 days and is thick yellow to yellow–green. Tenderness over the face may indicate clinical involvement of one or more of the paranasal sinuses. The diagnosis is usually confirmed radiographically. Gram stain of the material reveals many polymorphonuclear leukocytes (PMNs) and the causative organism. Because the most common organisms responsible for bacterial rhinosinusitis are *H. influenzae* and group A streptococcus, amoxicillin 25 to 50 mg/kg per day for 10 days is the treatment of choice.

Complications of acute sinusitis, such as orbital cellulitis, facial cellulitis/abscess, and meningitis, require admission to the hospital for appropriate IV antimicrobial therapy and possible operative intervention. Otolaryngologic consultation should be obtained in the evaluation of these patients with complicated acute sinusitis; surgical drainage may be needed.

One complication of chronic sinusitis, the sinus mucocele, can cause a child to present to the ED with acute symptoms. Mucoceles are expansile cystic lesions that occur secondary to a long-standing blockage of a sinus ostia. Although the lesion evolves over several months or even years, the child usually presents with the sudden onset of signs and symptoms usually related to an acute infection of the mucocele. These include pain and swelling secondary to osteomyelitis of the frontal bone, inferior and lateral displacement of the globe with proptosis, limitation of ocular mobility, and chronic nasal/postnasal discharge. Radiographs (plain films and CT) are often needed to determine the presence and extent of a mucocele. The patient should be referred to the otolaryngology service for appropriate IV antimicrobial therapy and surgical drainage.

Any child presenting to the ED with a history of persistent (after what would appear to have been an adequate course of appropriate treatment) rhinosinusitis should have a careful search for predisposing causes. Foreign bodies, choanal atresia, neoplasms, septal deviation, dental disease, adenoid hypertrophy, allergic polyps, or immunodeficiency states may all present with recurrent or persistent rhinosinusitis.

Chronic Nasal Obstruction

Obstruction to the normal passage of air can occur with a variety of conditions, and gives the sensation of a blocked or “stuffy” nose. Temporary partial obstruction of one nasal cavity at a time occurs normally in the nasal respiratory cycle. However, prolonged blockage, is not physiologic, and the physician should search for a cause.

Although most instances of nasal obstruction cause only mild feelings of discomfort, some children may present with a history of obstructive apnea (Pickwickian syndrome—see Adenotonsillar Hypertrophy, p. 1578) and even cor pulmonale. A history of trauma or foreign body may lead one to the reason for the obstruction. A careful examination of the nasal cavities and pharynx is necessary to determine the cause of the obstruction. Septal deviation, nasal tumor, and turbinate hypertrophy related to allergy and/or infection are common causes. Adenoid hypertrophy, nasopharyngeal tumor (lymphoma, rhabdomyosarcoma), and choanal atresia (unilateral or bilateral) can all present with nasal obstruction. Flexible fiberoptic examination (see Procedure 17.5 in Section VII) and radiographs (usually CT scan) of the nose and nasopharynx may be useful in the evaluation of the blocked nasal airway. If the source of the obstruction is not apparent after these maneuvers, referral should be made to an otolaryngologist to perform a complete examination of the nose and nasopharynx.

Epistaxis

Epistaxis is relatively common in children, and may cause significant anxiety in both the child and the parent. Although bleeding occasionally occurs secondary to the mucosal maceration caused by URIs, nose-picking accounts for most cases of recurrent epistaxis. (A more complete discussion on the differential diagnosis is presented in Chapter 23.) The usual site of bleeding is the anterior nasal septum, Kiesselbach's or Little's area (Fig. 121.4).

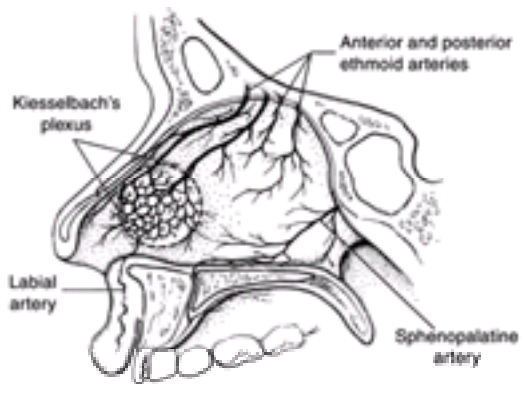

FIGURE 121.4. Vascular supply of nasal septum. Note confluence of vessels that forms Kiesselbach's plexus.

A complete history is an important step in the proper management of epistaxis. Site of bleeding (one or both sides of the nose), frequency, bleeding from other places, history of trauma, and family history of bleeding are all important factors to know in this evaluation. Figure 121.5 presents an algorithm for the management of epistaxis. A careful examination of the nose should be performed to identify the site and cause of the bleeding. Good lighting, suction, and material for cauterization and packing should be readily available (see Procedure 7.1 in Section VII). Topical vasoconstrictors such as phenylephrine (0.25%), oxymetazoline (0.05%) or epinephrine (1:1000) on a cotton pledget can be placed in the nose to shrink the nasal mucosa, allowing better visualization of the nasal cavity; vasoconstrictors may slow or even stop the bleeding. Simple pressure by squeezing the nostrils together is usually sufficient to stop most epistaxis. Occasionally, a roll of cotton placed under the upper lip will stop bleeding by compression of the labial artery. If pressure is not successful, cauterization with silver nitrate sticks or packing of the nose is performed (see Procedure 7.1 and Procedure 7.2 in Section VII). Absorbable packing such as oxycellulose (Surgicel) or gelatin (Gelfoam) is usually adequate for most epistaxis and has the advantage of not requiring removal.

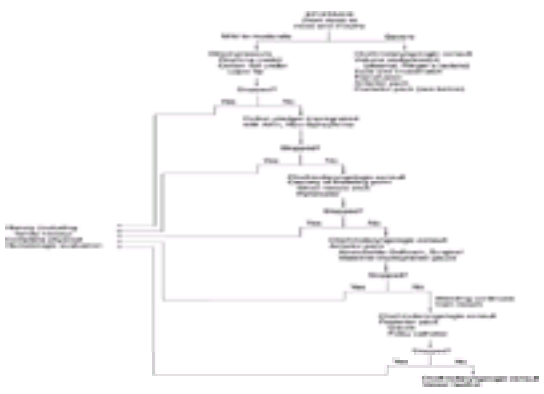

FIGURE 121.5. Algorithm for management of epistaxis.

Further treatment should also be directed toward preventing the child from continuing to traumatize his or her nose, resulting in further bleeding. Using a vaporizer to increase the humidity in the child's room and applying petroleum jelly to the anterior septal areas twice daily can aid in healing the irritated nasal mucosa and preventing recurrent epistaxis. Fingernails should be cut short.

Severe or recurrent episodes of epistaxis require the assistance of an otolaryngologist in their diagnosis and management. Epistaxis that does not stop with simple pressure or oxycellulose or gelatin packing may require a more substantial anterior nasal pack of petroleum jelly-impregnated gauze. A posterior nasal pack (using gauze or a Foley catheter) may be necessary in managing severe epistaxis that originates in the posterior nasal cavity or nasopharynx (see Procedure 7.2 in Section VII).

If the epistaxis recurs despite the above treatment, an otolaryngologist should be consulted to look for other causes for the epistaxis. Nasal septal deviation or perforation, sinusitis, tumor (nasal, nasopharyngeal or sinus), Rendu-Osler-Weber disease (hereditary hemorrhagic telangiectasia), and nasal foreign body can all present with epistaxis. Blood dyscrasias such as hemophilia, idiopathic thrombocytopenia purpura, von Willebrand's disease, and those hematologic conditions associated with leukemia or the administration of chemotherapeutic agents may lead to severe epistaxis. Treatment consists of correcting the underlying hematologic problem in addition to the previously described local measures. Recurrent or severe bleeding may require more extensive cauterization or even ligation of dilated vessels on the septum.

Neoplasms

Neoplasms of the nose and sinuses are uncommon in children. They may present as mass lesions or as chronic/recurrent rhinosinusitis. When a neoplasm is suspected, the child should be referred to an otolaryngologist for a complete evaluation of the nose and sinuses and appropriate radiographic imaging that is a prerequisite to the proper treatment of these lesions.

Hemangiomas are the most common benign neoplasms of the head and neck in children and often occur on the skin near or on the nose. Because hemangiomas often go through a period of rapid growth for the first 12 to 18 months of life

before they begin to involute, a period of observation is recommended before corticosteroids or surgical excision is considered. Recurrent bleeding, thrombocytopenia, skin breakdown, obstruction to vision, respiratory distress and cardiac failure are some indications for early intervention. Papillomas are viral-induced verrucous growths that are the most common neoplasms of the aerodigestive tract. When they appear in the nose, they are most often found on the nasal septum. Simple excision or fulguration is the preferred treatment for these lesions. In addition to these conditions, there are a variety of benign and malignant mass lesions of the nose. Early consultation with an otolaryngologist should be obtained for any tumor of the nose, especially one with recent changes in size or character.

ORAL CAVITY, PHARYNX, AND ESOPHAGUS

Methods of Examination

The oral cavity and oropharynx are directly visible with the aid of a tongue blade. A headlight or brightly lighted flashlight is required for this examination. The tongue should be displaced down and forward with the tongue blade placed on the anterior two-thirds of the tongue to avoid gagging. The examination of the nasopharynx, hypopharynx, and esophagus requires special instrumentation. The nasopharynx and hypopharynx can be examined using a flexible nasopharyngoscope. Nasopharyngoscopy requires special skills and may be best left to the otolaryngology consultant (see Procedure 17.5 in Section VII). Examination of the esophagus requires direct visualization with an esophagoscope, under general anesthesia. Palpation of the hypopharynx and nasopharynx should not be performed because it is uncomfortable to the child and potentially dangerous.

Radiography contributes minimally to the examination of the oral cavity and oropharynx because these areas are visible by direct examination. The lateral neck radiograph is useful to evaluate the nasopharynx and hypopharynx because there are good air–tissue interfaces. The esophagus can be coated with barium contrast.

Infections

Stomatitis

The most common infectious lesion of the oral cavity is the aphthous ulcer. The ulcers are often recurrent, may appear as a single lesion or a confluence of many lesions, and can cause severe stomatitis. The exact cause of aphthous ulcerations is unknown, but it is believed to be infectious.

Herpes simplex can cause severe gingivostomatitis, whereas the pharynx is relatively spared. On the other hand, Coxsackievirus infection (herpangina) causes severe ulcerative lesions of the pharynx, but not the anterior mouth. These viral infections cause severe oral pain and inability to eat. They are self-limited and require only symptomatic relief (see Chapter 84 and Chapter 124).

Candida albicans oral infection (thrush) usually appears as white patches with surrounding inflammation on the oral mucosa. It often occurs in newborns, immunosuppressed patients, and patients receiving antibiotic therapy. Nystatin is an effective treatment. The dosage is 200,000 units (2 mL) four times a day for 14 days.

Acute necrotizing, ulcerative gingivitis (trench mouth) causes painful, bleeding gums. Vigorous brushing of the teeth and gums with a soft brush promotes rapid healing. Antibiotics are of limited value.

Pharyngitis/Tonsillitis

Pharyngitis/tonsillitis (pharyngotonsillitis) may be caused by viral or bacterial organisms. Differentiating viral pharyngotonsillitis from an infection of bacterial origin is difficult on clinical grounds. A throat culture may be helpful in identifying an infection of bacterial origin. The degree of erythema and exudate may vary on the pharynx and tonsils. Bacterial pharyngotonsillitis is treated with a 10-day course of penicillin or amoxicillin. Patients with repeated debilitating bouts of pharyngotonsillitis that do not respond to a 6- to 8-week course of antimicrobial therapy/prophylaxis should be referred to an otolaryngologist for consideration for tonsillectomy and adenoidectomy.

Pharyngeal infections may spread to the peritonsillar area, causing cellulitis. The affected tonsil bulges forward and medially to touch the uvula. If pus localizes in the peritonsillar space, a peritonsillar abscess is formed. The peritonsillar abscess causes trismus. Suspected abscess formation requires immediate consultation with an otolaryngology specialist. Acute treatment of peritonsillar abscess requires systemic (24 to 48 hours of IV followed by oral) antibiotics and needle aspiration or incisional drainage (if possible) of the abscess. Occasionally, a “hot” or quinsy tonsillectomy may be required to treat the acute infection. Later elective tonsillectomy is indicated if there is a previous history of tonsillar or peritonsillar infections.

Retropharyngeal and Parapharyngeal Infection

Retropharyngeal and parapharyngeal lymph nodes may also be involved during pharyngitis and progress to abscess formation. Retropharyngeal abscess is usually easily seen on the lateral neck radiograph (Fig. 121.6). Peritonsillar, retropharyngeal, and parapharyngeal abscess must be treated with IV antibiotics and surgical drainage. (See also Infections under the section on Neck and Associated Structures.)

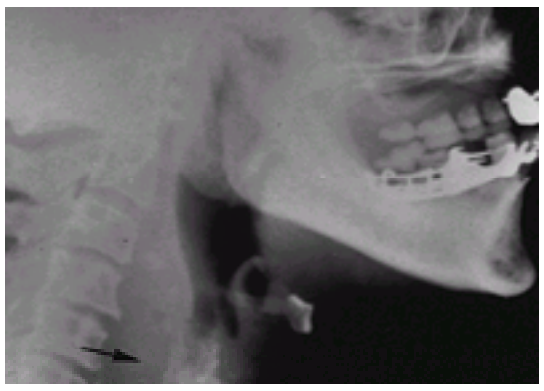

FIGURE 121.6. Lateral neck radiograph demonstrating retropharyngeal abscess *(arrow).*

One must always keep in mind that unusual infections like actinomycosis, mucormycosis, and syphilis may appear in the oral cavity. Actinomycosis appears with oral–cervical fistulas, and *Mucor* causes necrosis of the palate. Syphilis is visible in many ways (e.g., ulceration or raised lesion), and has no one characteristic appearance.

Adenotonsillar Hypertrophy

Lymphoid hyperplasia (enlarged tonsils and adenoids) can cause airway obstruction that can range from mild snoring to severe sleep apnea with right heart strain. Young children with obstructive sleep apnea are often in the lower 25th percentile by weight (failure to thrive). Older children with severe obstructive sleep apnea are often obese, and present with daytime somnolence (Pickwickian syndrome). If right heart strain or daytime somnolence is present, a tonsillectomy and adenoidectomy may be required urgently.

Neoplasms

Benign and malignant neoplasms occur in the oral cavity, pharynx, hypopharynx, and esophagus. Benign neoplasms in the oral cavity may rise from the mucosa or underlying tissues. Minor salivary gland tumors, hemangiomas, lymphangiomas, pyogenic granulomas, and neurofibromas are found in the oral cavity, but they are rarely an emergency.

Nasopharyngeal angiofibromas occur in prepubescent males and cause nasal obstruction. They may appear in the ED with massive epistaxis. Posterior packing is usually required to control the hemorrhage that may be life-threatening (see Procedure 7.2 in Section VII).

Malignant neoplasms are rare but can occur throughout the oral cavity, pharynx, and esophagus. Rhabdomyosarcoma, lymphoma, and squamous cell carcinoma (lymphoepithelioma) are the most common lesions and are rarely seen as emergencies unless there is extensive hemorrhage or a compromised airway.

Biopsy of oral, pharyngeal, and esophageal tumors is best done in the operating room where adequate exposure and control of hemorrhage is most effectively obtained.

LARYNX AND TRACHEA

Methods of Examination

Examination of the larynx is often difficult in the young child. Commonly, however, the tip of the epiglottis may be visualized when the tongue is protruded during the examination of the oropharynx. Examination of the larynx can be performed using a flexible fiberoptic endoscope. Vocal cord mobility, the structures of the larynx, and the presence of laryngeal masses can usually be assessed in this manner in a cooperative child 4 years of age or older (see Procedure 7.5 in Section VII). The otolaryngologist may need to be consulted to perform this examination for the child presenting in the ED with symptoms related to the larynx.

Lateral and anteroposterior plain radiographs of the neck can provide significant information about the larynx and upper trachea. Although xeroradiographs offer more precise detail of the airway by virtue of their property of edge enhancement, the extra radiation exposure inherent in this study makes this a less desirable imaging modality. CT and MRI scans are useful in examining the fine detail of laryngeal and tracheal structures, but the general anesthetic required to keep the child still for these examinations restricts their use to specific situations. Fluoroscopic examination of the larynx is one method of evaluating the movements of the vocal cords during phonation and respiration. Vocal cord paralysis and laryngomalacia can often be identified in this manner. Contrast studies can also be used in the evaluation of laryngeal function. A barium swallow is useful in detecting aspiration related to vocal cord paralysis, posterior laryngeal cleft, or tracheoesophageal fistula.

Infections

Viral laryngitis is often a component of the common URI described previously in this chapter. Laryngitis is manifest by a hoarse, raspy voice as a result of inflammatory edema of the vocal cords. Airway obstruction is rare in viral laryngitis. Symptomatic treatment with humidification, antipyretics, analgesics, throat gargles, and voice rest are recommended while the disease runs its natural course. When the viral infection involves the subglottic space, a more serious clinical problem appears. Laryngotracheobronchitis (croup) is a common, and potentially life-threatening, infection occurring in early childhood. The diagnosis and management of croup is discussed in other chapters of this book (see Chapter 72

and Chapter 84).

Bacterial laryngotracheobronchitis does occur, but is not nearly as common as its viral counterpart. Children age 3 to 6 years are more commonly affected by bacterial tracheitis compared with the infection of viral origin that usually appears in children less than 3 years of age. It may be difficult to distinguish bacterial laryngitis on clinical grounds from a similar infection of viral origin. Etiologic agents responsible for bacterial laryngitis include staphylococci and *H. influenzae.* Severe airway obstruction is a common symptom of bacterial laryngotracheobronchitis. This is caused by thick, inspissated secretions that fill the trachea and are difficult for the child to clear. Treatment consists of the same measures recommended for viral laryngitis with the addition of the appropriate antimicrobial agents. The otolaryngologist is usually required to perform a direct laryngoscopy and bronchoscopy to confirm the diagnosis and to aspirate the thick secretions for therapeutic and diagnostic purposes.

Diphtheria may involve the larynx, in addition to other areas of the upper aerodigestive tract. The diagnosis is suspected by the presence of a membrane covering the pharynx and larynx that leaves a raw, bleeding surface when it is removed. The diphtheria membrane can obstruct the laryngeal airway to cause respiratory distress. Endoscopic removal of the membrane and/or tracheostomy may be required, in addition to antimicrobial therapy.

Bacterial infection of the supraglottic larynx can cause a symptom complex with potentially life-threatening airway obstruction. Epiglottitis (more appropriately called supraglottitis) is an infection of the supraglottic larynx that is caused most often by *H. influenzae* type b. The diagnosis and management of epiglottitis is discussed in other chapters of this book (see Chapter 72 and Chapter 84).

Neoplasms

Neoplasms of the larynx and trachea are uncommon in children. The otolaryngologist should be consulted to assist the emergency physician in the management of these patients.

The most common neoplasm of the larynx in children is the laryngeal papilloma. This is believed to be a viral-induced neoplasm that has a predilection for the upper aerodigestive tract and the larynx in particular. The disease is usually diagnosed in the child between 2 and 5 years of age and presents with persistent or worsening hoarseness and, occasionally, airway obstruction. If papillomas are suspected as the source of hoarseness in a child, the otolaryngologist should be consulted to perform the indirect or direct laryngoscopy required to confirm the diagnosis. A lateral neck radiograph may demonstrate a soft-tissue mass in the area of the larynx (Fig. 121.7). The course of the disease is characterized by multiple cycles of growth and regression until a spontaneous remission occurs, usually around puberty. The otolaryngologist's goal in managing these patients is to maintain an adequate voice and unobstructed airway by frequent repeated excision (with cup forceps or CO_2 laser) of the papillomas. A tracheostomy may be required in cases of severe airway obstruction.

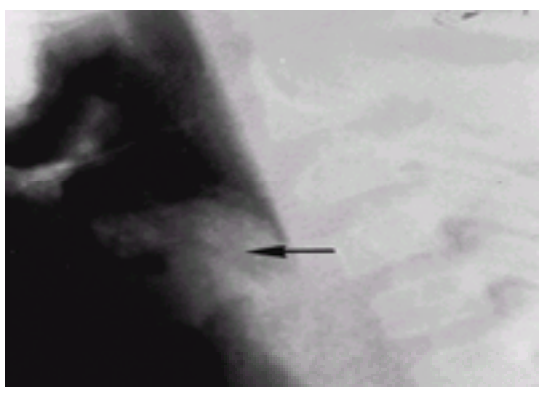

FIGURE 121.7. Lateral neck radiograph demonstrating soft-tissue density *(arrow)* at level of larynx. Direct laryngoscopy revealed this to be papilloma.

Hemangiomas may occur in the larynx, primarily in the subglottic area. As with most juvenile hemangiomas, these lesions present in the second to sixth month of life and can enlarge over several months to cause significant airway obstruction. Episodes of stridor may be precipitated by a URI. Of children with subglottic hemangiomas, 50% have other cutaneous lesions. Therefore, the presence of cutaneous hemangiomas in an infant with stridor should suggest to the emergency physician the possibility of a subglottic hemangioma. Hemangiomas may appear as posterior subglottic masses on lateral neck radiographs, but the diagnosis must be confirmed by laryngoscopy performed by an otolaryngologist. Because most hemangiomas of infancy tend to involute after an initial period of rapid growth during the first 1 to 2 years of life, close observation is the only treatment required for those lesions that are causing minimal symptoms. If there is severe, persistent, or recurrent respiratory distress, intervention is indicated. Systemic corticosteroids, CO_2 laser or direct surgical excision, and tracheostomy are some of the modes of treatment presently being advocated.

Malignant neoplasms of the larynx are uncommon. They include rhabdomyosarcoma, chondrosarcoma, and lymphoma. These tumors are seen with varying degrees of hoarseness and respiratory obstruction. If a laryngeal malignancy is suspected, the otolaryngologist should be asked to perform indirect and direct laryngoscopy to confirm the laryngeal problem and to obtain tissue for histologic identification of the tumor.

Stridor

The differential diagnosis and emergency management of a child presenting with stridor is discussed in detail in Chapter 72.

NECK AND ASSOCIATED STRUCTURES

Methods of Examination

Visual inspection and palpation provide the basis for examination of the neck and its enclosed structures. The head should be erect during the examination, with the normal prominence of the sternocleidomastoid muscle on each side. Anterior projection of the thyroid cartilage or “Adam's apple” is seen in postpubescent males. Palpation of the neck is performed to assess the normal structures in the neck and to detect the presence and nature of any cervical masses. Examination of the two sides is done simultaneously so they can be compared with one another. The examiner should be able to grasp the thyroid cartilage and move it gently from side to side without any discomfort to the patient. Immobility or significant pain may indicate the presence of laryngeal pathology. Crepitance of the neck indicates free air in the tissue planes of the neck from perforation of a hollow viscous. Passive and active range of motion of the neck should be complete in all directions. Restriction in movement may be caused by tender cervical adenopathy, cervical spine disease, spasm or fibrosis of the sternocleidomastoid muscle or meningeal irritation (Brudzinski's sign). Arterial pulses of equal strength should be palpable in the carotid artery on each side of the neck. The carotids can also be auscultated for evidence of bruits.

Radiographs are often invaluable in the examination of the neck. Plain anteroposterior and lateral views provide significant information in the evaluation of cervical problems. The presence of masses projecting into and compromising the airway can be detected. Air between the muscle planes of the neck indicates a perforation of a hollow viscous such as the pharynx, esophagus, larynx, trachea, or pulmonary alveolus. Sinus films, by identifying a neoplasm or sinusitis, are often helpful in determining the cause of a neck mass.

Infections

Cervical adenitis is the most common cause of a neck mass in a child. The lymphatic system of the neck drains the internal cavities of the head and neck (ear, nose, mouth, pharynx, sinuses, and larynx), as well as the skin and associated adnexal structures of the face and scalp. Regional cervical lymph nodes respond when there is a primary infection in any area of the head and neck. Because certain groups of nodes drain specific sites in the head and neck, the location of the swollen and infected lymph node can often help the practitioner to identify the area of the primary infection. Ear infections most often drain to the infra-auricular nodes, pharyngeal infections (e.g., tonsillitis) usually are seen with jugulodigastric node involvement, and posterior cervical nodes often accompany nasopharyngeal infections (e.g., adenoiditis).

Cervical adenitis is uncommon secondary to a brief, uncomplicated viral URI. Tender and enlarged nodes occur more often as a result of bacterial infection of the head and neck, with the ears and throat responsible for a large portion of these. Because *Streptococcus* species are the causative agents in the majority of bacterial infections in the head and neck, the infected lymph nodes usually contain the same organisms. Treatment with oral penicillin (or amoxicillin) usually clears the primary infection and causes regression of the enlarged lymph nodes. Culture of the nasopharynx, throat, or aspirate of the cervical node can assist the physician in the choice of antimicrobial agents.

Although most children will respond to the therapy just described, there is a small group of children whose nodes progress to suppurative cervical adenitis. Studies of children hospitalized with cervical adenitis have shown a predominance of *S. aureus* as the causative agent. This high incidence of staphylococci is probably the result of selecting patients who have not responded to oral antimicrobials effective against the more commonly occurring *Streptococcus* species. Therefore, if cervical adenitis has not responded to the primary antimicrobial treatment, agents should be added that are effective against *S. aureus* (erythromycin, dicloxacillin, clindamycin, or cephalosporins).

A child who has demonstrated rapid enlargement of cervical nodes, poor response to oral antimicrobials, cellulitis of the overlying skin, abscess formation, or signs of toxicity (high fever, malaise, dehydration) should be admitted to the hospital for treatment with IV fluids and antimicrobials. Surgical consultation should be obtained in the management of these complicated cases in which needle aspiration, incisional drainage, or biopsy (for possible neoplasm) may be required.

Retropharyngeal or parapharyngeal nodes are uncommonly involved with inflammatory processes that originate in the pharynx. Sore throat, dysphagia, and stiff neck are some of the symptoms that can accompany significantly enlarged pharyngeal nodes. Retropharyngeal nodes can be overlying the cervical spine during examination of the oropharynx. They also appear as widening of the retropharyngeal soft tissues on lateral neck radiographs. Parapharyngeal nodes are seldom detected clinically unless they enlarge sufficiently to deviate the tonsil and pharyngeal wall medially. Treatment of enlarged pharyngeal nodes consists of IV antimicrobials (usually penicillin) and observation of the child's airway. Biopsy of the mass is indicated if resolution does not occur with treatment or if a malignancy is suspected.

A collection of purulent material within the tissues of the neck, a neck abscess, requires prompt and specific treatment. The most common cause of a neck abscess is breakdown or necrosis of an infected lymph node. Purulent material may be located within a single node or may accumulate between several adjacent nodes. Once the process of cervical adenitis has progressed to the point of abscess formation, treatment involves evacuation of the infected material and the prevention of further spread of the infection. The child is hospitalized, and IV antimicrobials are administered that are effective against *S. aureus* (antistaphylococcal penicillin). Otolaryngologic consultation is obtained to perform a needle aspiration or incision and drainage to evacuate and culture the infected material.

Deep neck abscesses are uncommon in children, but can be extremely dangerous when they occur. Parapharyngeal abscess occurs when purulent material collects in the parapharyngeal space lateral to the pharyngeal constrictors and

medial to the vascular compartment of the neck. Necrosis of parapharyngeal lymph nodes or lateral extension of a peritonsillar abscess are the two main sources of this infection. The child with a parapharyngeal abscess presents with a stiff neck, high fever, malaise, dehydration, and other signs of toxicity. The child usually has dysphagia and may not be able to swallow his or her own saliva. Physical examination reveals diffuse swelling and tenderness of one side of the neck, but fluctuance is seldom appreciated. Intraoral examination may demonstrate medial displacement of the lateral pharyngeal wall and tonsil. Lateral neck radiographs are usually not helpful in evaluating this disease process. CT or MRI scans provide the best evaluation of suspected deep neck abscesses. If left to progress, the parapharyngeal abscess can involve the adjacent vascular structures in the neck, descend into the mediastinum, or spontaneously rupture into the pharynx, causing aspiration of purulent material.

Otolaryngologic consultation should be obtained to assist the emergency physician in the evaluation of a patient with a parapharyngeal abscess. Appropriate treatment consists of hospitalization, IV fluids, antimicrobials effective against *S. aureus* (antistaphylococcal penicillins, clindamycin, cephalosporins), and external drainage of the abscess.

Retropharyngeal abscess occurs as a result of the necrosis of retropharyngeal lymph nodes or secondary to perforation of the pharynx or esophagus. Purulent material collects between the retropharyngeal and prevertebral layers of the cervical fascia, also called the danger space. This potential space extends from the base of the skull to the mediastinum, thus allowing extensive spread of the infection. A child presents with symptoms similar to those associated with parapharyngeal abscess. Lateral neck radiographs demonstrate widening and bulging of the retropharyngeal space (Fig. 121.6). Treatment consists of hospitalization, IV fluids, antimicrobials effective against *S. aureus,* and drainage (either intraoral or external) of the abscess.

Nontubercular mycobacterial (NTM) infection is a common cause of chronic cervical adenitis in children. Also called atypical mycobacteria, the ubiquitous agent is thought to gain access to the cervical lymph nodes through oral mucosal breaks (e.g., teething, minor trauma). The usual presentation of NTM cervical adenitis is that of a non-tender, slightly fluctuant cervical mass with overlying skin that has a characteristic violaceous hue. Chest radiographs are usually normal and purified protein derivative (PPD) tests are most often reported as negative or intermediate in their response. NTM infections do not respond to anti-tubercular antibiotics. The child should be referred to an otolaryngologist to perform surgical excision that is required to cure this condition. Incision and drainage is discouraged because this will lead to a chronic draining sinus.

Salivary gland infections should be considered in the differential diagnosis of a cervical mass suspected to be infectious in origin. Both viral and bacterial agents can be responsible for the infection, with the former being more common. Mumps (endemic parotitis) is the most common salivary infection in children. Although the parotid gland is involved in more than 85% of the cases, the submandibular gland may also be involved with the viral infection. The infection appears with acute painful swelling of the involved gland or glands. There is erythema around the intraoral orifice of the salivary duct and the saliva expressed is generally clear. Treatment is supportive with clear fluids, antipyretics, and analgesics as necessary.

Bacterial infections of the salivary glands are seen with signs and symptoms similar to those associated with cervical lymphadenitis. Neonatal parotitis and, less commonly, submandibular sialadenitis usually occur in a 3- to 4-week-old child after a systemic illness has caused dehydration. The affected gland is swollen and abscess formation may occur. Purulent material may be expressed from either Stenson's or Wharton's duct by massage of the affected salivary gland. Otolaryngologic consultation should be obtained. The child is hospitalized for treatment with IV antimicrobials effective against *S. aureus* (antistaphylococcal penicillin) and surgical drainage of any collection of purulent material. Recurrent or chronic infections of the salivary glands are usually related to some predisposing factor such as stones, ductal stenosis, or secretory immunodeficiency. Management should include the detection and correction of these conditions.

Neoplasms

Neoplasms of the neck, both primary and metastatic, occur in children. If a cervical neoplasm is suspected, an otolaryngologist should be consulted to perform a complete examination of the head and neck, including endoscopy of the nasopharynx, larynx, and hypopharynx.

The hemangioma is the most common neoplasm of the head and neck in children. Although they are more common on the skin of the face and scalp, lesions can occur on the skin of the neck and involve deeper structures, such as the parotid gland. The diagnosis of cutaneous hemangiomas of the cervical skin is usually obvious on physical inspection; the lesions are red to reddish-purple, flat or raised, blanch with pressure, and increase in size with crying or straining. Deep-seated lesions without cutaneous manifestations may require special diagnostic aids such as CT or MRI scans and, rarely, biopsy to confirm the diagnosis.

These juvenile hemangiomas demonstrate a cycle of rapid growth for the first 12 to 18 months of life. Slow regression and even total disappearance occurs over the next year or two. Because of this natural history, once the diagnosis of hemangioma is made, the preferred treatment is close observation. Lesions that grow rapidly to produce complications such as airway obstruction, skin necrosis, hemorrhage, high-output cardiac failure, or thrombocytopenia require more active intervention. The child should be admitted and otolaryngologic consultation obtained. Treatment modalities presently advocated include systemic corticosteroids, cryotherapy, CO_2 laser excision, interferon, sclerosing agents, and surgical excision.

Lymphangiomas are uncommon benign lesions of the neck. Cystic hygroma is the most common type of lymphangioma found in the neck. These lesions consist of multiple cystic spaces filled with lymph and, occasionally, blood. They appear most commonly as large lateral neck masses in neonates. The diagnosis is often obvious on physical examination of a large cystic lesion that transilluminates. The natural history of these lesions is usually one of progressive growth and enlargement. Lymphangiomas can fluctuate in size secondary to a concurrent infection of the head and neck or hemorrhage into a cyst. Small, stable, asymptomatic lesions can be managed by close observation. Surgical excision is the treatment of choice for all large symptomatic lesions, with several staged procedures often being required. Aspiration

of a large cyst (or cysts) can temporarily decompress a lesion, but it is not a substitute for definitive surgical excision. Large cystic hygromas may cause feeding difficulties or respiratory distress in the newborn and may necessitate early surgical intervention, which can include tracheostomy and gastrostomy.

Less common benign neoplasms of the neck in children include teratomas, paragangliomas (carotid body tumors, glomus tumors), neural sheath tumors (neurofibromas, neurolemmomas), and thyroid and salivary gland neoplasms.

The sternocleidomastoid “tumor” of infancy is an unusual lesion that appears as a discrete mass within the substance of the sternocleidomastoid muscle in a child 4 to 8 weeks old. The cause of this localized area of fibrosis is unknown. The lesion usually resolves with range-of-motion exercises. Surgical intervention is indicated in those cases in which the fibrosis progresses to cause torticollis (see the section on Neck Stiffness, Chapter 46) or if there is suspicion of a malignancy.

The most common malignant neoplasm of the neck in children is lymphoma, being almost equally divided into Hodgkin's and non-Hodgkin's types. The disease may be localized in the neck or be a part of a more generalized disorder. Physical examination often reveals multiple firm, rubbery, unilateral, or bilateral nodes. If the diagnosis of lymphoma is suspected, otolaryngologic consultation should be obtained for a careful examination of the oral cavity, pharynx, and paranasal sinuses to look for a primary or associated lesion. This not only aids in the evaluation of the extent of the lymphoma but may also locate a site from which a biopsy can be obtained without the morbidity of a neck exploration.

Cervical lymph nodes may appear as neoplasm metastatic from a nonlymphogenous primary tumor. Thyroid carcinoma, squamous carcinoma (lymphoepithelioma) of the nasopharynx, and malignant melanoma may all be seen first with enlarged cervical lymph nodes. These nodes tend to be hard, singular, and may be fixed to underlying structures. Otolaryngologic consultation should be obtained for a complete examination of the head and neck to search for a primary lesion. Biopsy of the node is usually required for diagnosis.

Rhabdomyosarcoma is the most common soft-tissue sarcoma of the head and neck in children, and its frequency of occurrence in the neck is second only to that in the orbit. The child usually presents with a history of rapid enlargement of a painless neck mass. The mass itself is hard, often diffuse, and poorly mobile. Although the diagnosis of rhabdomyosarcoma may be suspected from the history and physical examination, biopsy is always required for confirmation.

Many other malignant neoplasms can also occur in the neck. These include soft-tissue sarcomas other than rhabdomyosarcoma, malignant fibrous histiocytoma, and neuroblastoma.

Neck Mass

The differential diagnosis and ED management of the child with a neck mass is presented in detail in Chapter 45.

Torticollis (Wryneck)

The differential diagnosis and ED management of the child with torticollis or stiff neck is presented in detail in Chapter 46. For further details, see Chapter 110 and Chapter 112.

Suggested Readings

Bluestone CD, Klein JO. Otitis Media in Infants and Children. Philadelphia: WB Saunders, 1988.

Bodenstein L, Altman RP. Cervical lymphadenitis in infants and children. Semin Pediatr Surg 1994;3(3):134–141.

Britton GH, Block LD. Vertigo in the pediatric and adolescent age group. Laryngoscope 1988;98:139–146.

Fishman SJ, Mulliken JB. Hemangiomas and vascular malformations of infancy and childhood. Pediatr Clin North Am 1993;40(6):1177–1200.

Garfinkle TS, Handler SD. Hemangiomas of the head and neck in children: guide to management. J Otolaryngol 1980;9:439–450.

Guarisco, JL, Graham HD. Epistaxis in children: causes, diagnosis, and treatment. Ear Nose Throat J 1989;68:522–538.

Handler SD, Raney RB. Management of neoplasms of the head and neck in children: I. Benign tumors. Head Neck Surg 1981;3:395–405.

Hopp R, Cooperstock M. Medical management of sinusitis in pediatric patients. Curr Prob Pediatr 1997;27(5):178–186.

Jones JH. Oral Manifestations of Systemic Disease. Philadelphia: WB Saunders, 1980.

Lane RJ, Keane WM, Potsic WP. Pediatric infectious cervical lymphadenitis. Otolaryngol Head Neck Surg 1980;88:332–335.

Marcy SM: Infections of the external ear. Pediatr Infect Dis 1985;4:192–201.

Murray AB, Milner RA. Allergic rhinitis and recurrent epistaxis in children. Ann Allerg Asth Immunol 1995;74(1):30–33.

Northern JL, Downs MP. Hearing in Children. 4th ed. Baltimore: Williams & Wilkins, 1991.

Potsic WP, Handler SD, Wetmore RF, et al. Primary Care Pediatric Otolaryngology. 2nd ed. Andover, NJ: J Michael Ryan Publishing, 1995.

Raney RB, Handler SD. Management of neoplasms of the head and neck in children: II. Malignant tumors. Head Neck Surg 1981;3:500–507.

Suskind DL, Handler SD, Tom LW, et al. Nontuberculous mycobacterial cervical adenitis. Clin Pediatr 1997;36(7):403–409.

Tom LWC, Handler SD, Wetmore RF, et al. Sternocleidomastoid tumor of infancy. Int J Pediatr Otorhinolaryngol 1987;13:245–255.

Tom LWC, Rossiter JL, Sutton LN, et al. Torticollis in children. Otolaryngol Head Neck Surg 1991;105:1–5.

Tusa Rj, Saada AA Jr, Niparko JK. Dizziness in childhood. J Child Neurol 1994;9(3):261–274.

Wahrman JE, Honig PJ. Hemangiomas. Pediatr Rev 1994;15(7):266–271.

CHAPTER 122

Urologic Emergencies

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Early in their lives, children become familiar with the act of voiding and the appearance of their genitals. Disturbances of either are a great source of concern to them and their parents. This may result in an anxious trip to the emergency department (ED), requiring the emergency physician to be familiar with the problems to be discussed in this section (see Chapter 58). This chapter discusses 1) penile problems, 2) testicular problems, and 3) urinary tract infections. Renal trauma is covered in Chapter 108 and Chapter 109.

PENILE PROBLEMS

Penile Care in the Uncircumcised Male

Although the data of Wiswell and Roscelli have suggested that the presence of the foreskin may make ascending urinary infection an increased risk in newborn males, the overall low incidence of problems associated with the foreskin and the benefits from its removal lead us to continue to discourage routine circumcision. This view is common and increasing numbers of uncircumcised children are seen in EDs. Surprisingly, few physicians know how to care for uncircumcised boys. It is important to realize that, in male infants, adhesions between the glans and the foreskin are normal (Fig. 122.1). The foreskin is not normally retractable in this age group. No effort should be made to strip the foreskin back in infants because that not only produces undue pain for the child, but also may result in a raw surface, with consequent inflammation and scarring. Between ages 2 and 4, lysis of the adhesions is spontaneous in 90% of children. It is rare for the young male to have any adverse hygienic consequence from leaving the foreskin in place until spontaneous lysis of the adhesions takes place. The small, whitish lumps that may be seen and felt beneath the foreskin represent only desquamated epithelium and need not be removed. When toilet training has occurred, it is wise to teach a boy to retract the foreskin enough to expose the meatus when he voids. Not only does this facilitate better aiming, but it also avoids leaving the inner foreskin wet with urine. Ammoniacal irritation can lead to inflammatory adhesions and may create a portal of entry for a bacterial balanoposthitis. When a boy is able to retract his foreskin, usually between 4 and 6 years of age but sometimes later, he may be taught to withdraw the foreskin and carry out normal hygiene as part of bathing.

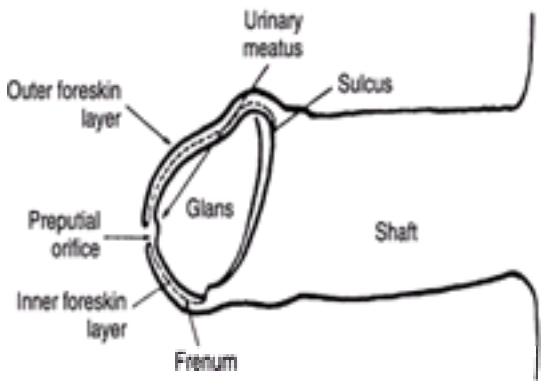

FIGURE 122.1. Anatomy of normal uncircumcised male. Adhesions between inner foreskin layer and glans are normal in newborns and prevent retraction of the foreskin. (Reprinted with permission from Wallerstein E. Circumcision: An American Health Fallacy. New York: Springer, 1980: 201.)

Phimosis and Paraphimosis

Phimosis exists when tightness of the distal foreskin precludes its being withdrawn to expose the glans. Although inflammation of the foreskin from severe chronic ammoniacal rash or infection may lead to scarring and a true phimosis, this is uncommon in children. More often, normal penile adhesions are confused with phimosis.

In the uncircumcised male, if the foreskin is retracted behind the glans and left in that position, venous congestion and edema of the foreskin results, making it difficult to reduce the foreskin to a normal position. This condition of a swollen, retracted foreskin is called paraphimosis (Fig. 122.2). The application of ice and steady local manual compression usually reduces the edema and permits manual reduction of the paraphimosis. A local anesthetic penile block of the dorsal nerve of the penis at the base of the shaft will reduce the discomfort experienced by the child during compression of the edematous foreskin. Once a portion of the edema has been reduced, pressure on glans (like turning a sock inside out) usually permits reduction of the foreskin back to its normal position (Fig. 122.3). If manual reduction fails, a surgical division of the foreskin to permit reduction is indicated (Fig. 122.4). That usually may be accomplished with sedation and local anesthetic. If surgical reduction of the foreskin is required, it should be followed a few weeks later by a circumcision. Education in the care of the uncircumcised male will reduce the incidence of this condition.

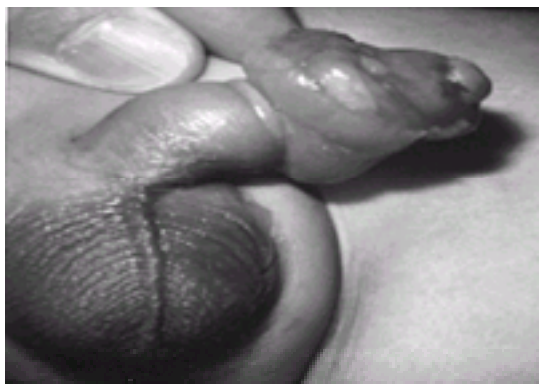

FIGURE 122.2. Paraphimosis: a foreskin that is left in a retracted position leads to venous congestion and edema of the foreskin.

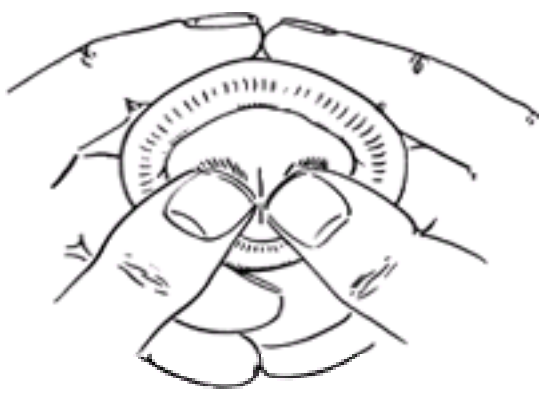

FIGURE 122.3. Manual reduction of paraphimosis. After a local anesthetic block of the dorsal nerve of the penis, the foreskin is manually compressed to reduce edema. The foreskin can be reduced by pressure on glans—like turning a sock inside out. (Reprinted with permission from Klauber GT, Sant GR. Disorders of the male external genitalia. In: Kelalis PP, King LR, Belman AB, eds. Clinical Pediatric Urology. 2nd ed. Philadelphia: WB Saunders, 1985: 287.)

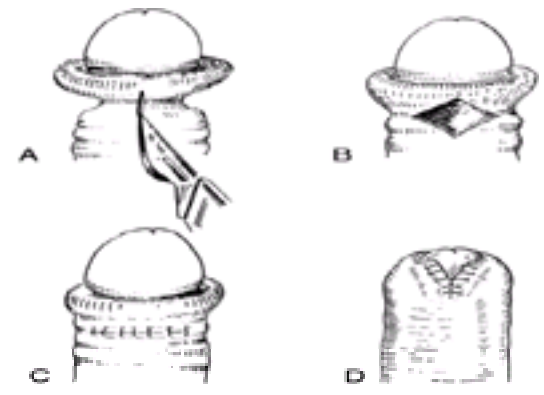

FIGURE 122.4. Surgical correction of phimosis. **A.** Constricting foreskin is incised vertically on dorsum. **B.** The incision opens laterally, relieving constriction. **C.** Incision is closed transversely with chronic catgut sutures. **D.** Foreskin can now be reduced. (Reprinted with permission from Klauber GT, Sant GR. Disorders of the male external genitalia. In: Kelalis PP, King LR, Belman AB, eds. Clinical Pediatric Urology. 2nd ed. Philadelphia: WB Saunders, 1985:827.)

Balanoposthitis

Balanoposthitis is an infection of the foreskin that may extend onto the glans (Fig. 122.5A). It is a form of cellulitis and has its origin from a break in the penile skin, commonly associated with ammoniacal dermatitis. It may be the result of local trauma or may, in the older boy, be associated with poor penile hygiene. Scarring after the inflammatory reaction may lead to true phimosis. The acute infection is dealt with adequately by warm soaks and the administration of an appropriate antibiotic, usually ampicillin (50 to 100 mg/kg every 24 hours in four doses) (Fig. 122.5B). It is unusual for a child to be unable to void as a result of this condition, although he may be more comfortable voiding while in a tub of warm water. After resolution of the acute infection, the youngster should be examined again, and, if true phimosis is present, a circumcision is advisable. One episode of balanoposthitis with a normal retractable foreskin does not indicate a need for a circumcision. However, if a child has recurrent infections, a circumcision is in order.

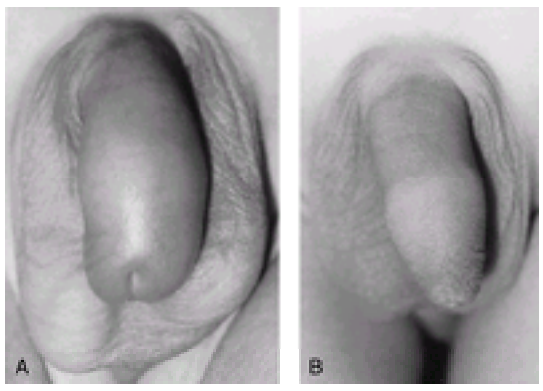

FIGURE 122.5. A. Balanoposthitis: cellulitis of normal foreskin with erythema, edema, and tenderness. **B.** Normal foreskin after treatment of balanoposthitis with antibiotics and warm soaks.

Penile Swelling

Although most penile swelling is painful and the result of either infection, as described previously, or trauma, to be described later, occasionally a child has isolated penile edema that is either nontender or minimally tender. This may result from an insect bite, with local edema secondary to histamine release. A history of a bite or the finding of a small punctate lesion may give the clue to diagnosis. Painless penile edema may be present with a generalized allergic reaction or as part of the manifestation of a general edematous state secondary to renal, cardiac, or hepatic problems. Here, the diagnosis is suggested by evidence of dysfunction in these organ systems on general examination. It is also important to remember that penile swelling may be caused by a strangulation injury (see the following).

Priapism

Prolonged, painful penile erection unaccompanied by sexual stimulation is called priapism. In the pediatric age group, this entity may be caused by trauma or leukemic infiltration, but it is most often seen in African-American males with sickle cell disease. A sickling crisis that involves the corporal bodies does not necessarily need to be related to symptomatic sickling elsewhere in the body. Sickling of the erythrocytes produces sludging and stasis in the erectile tissue of the corporal bodies. This stasis leads to further hypoxia, acidosis, and more sickling. The thick, dark sludge that is formed prevents detumescence of the erectile tissue and thus causes priapism. Pain results from ischemia. It is speculated that an inflammatory reaction to this material may lead to fibrosis of the erectile tissue. Impotence may result.

Although recommendations for treating priapism have ranged from ice or hot packs, estrogens, and spinal anesthesia to radiation therapy, the best treatment for priapism associated with sickle cell diseases now appears to be hydration and irrigation of the corporal bodies with saline in combination with vasoactive substances. This is best carried out with urologic consultation. Although priapism has been documented to lead to impotence in some cases, impotence is rare in priapism related to sickle cell disease, unless the patient has been subjected to a surgical procedure. It may be that the more difficult cases are the ones most likely to come to surgical treatment, and impotence thus may reflect more the basic disease, rather than the type of treatment.

Meatal Stenosis

Meatal stenosis is a problem almost exclusively of circumcised males and follows an inflammatory reaction around the meatus, usually the result of the lower edge of the meatus rubbing against a wet diaper with inflammation of the meatus resulting from mechanical and ammoniacal chemical dermatitis. Meatal stenosis is rare in the boy who has a circumcision after becoming continent. Appearances are often deceiving. The meatus may appear to be stenotic, but may be functioning adequately. Significant meatal stenosis causes spraying of the urinary stream or more commonly, dorsal deflection of the stream. Surgical treatment of the meatus is warranted only if these symptoms are present. Meatal stenosis is not a cause of frequency, enuresis, or urinary tract infection. When it is indicated, we carry out a meatotomy in our office after application of topical penile anesthesia with EMLA Cream and the infiltration of a small amount of Xylocaine with epinephrine into the ventral edge of the meatus. A general anesthetic usually is neither necessary nor indicated.

Penile Trauma

Direct Injury

The most common cause of direct injury to the penis comes from the toilet seat's falling on the penis of a little boy who is learning to stand at the toilet to void. Although the resulting penile edema may be notable, significant injury to the corporal bodies or urethra is rare. Although parents may be concerned that the child will be unable to void, this generally is not a problem, but the child may be more comfortable voiding in a tub of warm water. The only treatment required is warm soaks and expectant observation.

After blunt or sharp trauma, if blood is seen at the urethral meatus, urethral injury must be considered and a retrograde urethrogram carried out. Pediatric urologic consultation is appropriate, as is follow-up for possible stricture formation (see Chapter 109).

If a child is seen for a laceration of the shaft of the penis, it is important to be certain that the corporal bodies and urethra have not been injured concurrently. When a question exists, pediatric urologic consultation, retrograde urethrogram, and exploration under anesthetic may be needed. For simple lacerations of the penile skin, repair with chromic catgut suffices. It should be recalled that a child who has any form of a genital injury may be a victim of sexual abuse (see Chapter 128).

Zipper Injury

Boys often seem to be in a hurry and sometimes fail to get their penis or foreskin completely back in their pants before they pull up the zipper. This results in the entrapment of penile skin or foreskin in the teeth of the zipper. The teeth may be so engaged that it is impossible to simply unzip the zipper. Often, the problem may be dealt with simply, as shown in Figure 122.6. The median bar of the zipper may be cut with a pair of wire cutters, which will permit the two halves of the zipper to fall apart, releasing the entrapped skin. Mineral oil has also been used to allow the tissue to slide free of the metal zipper. Local infiltration of Xylocaine makes this procedure less traumatic to the child. Only rarely is a general anesthetic required. After the zipper is removed, the penis may become edematous, but generally nothing more than warm soaks is required for further treatment.

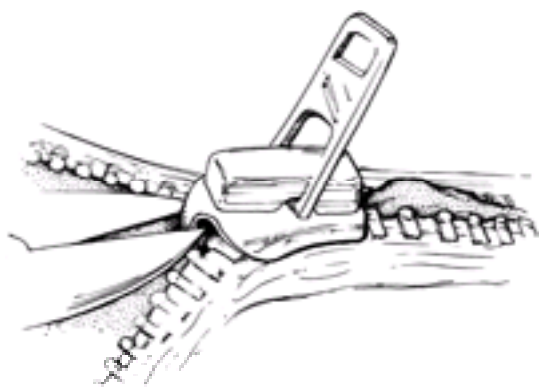

FIGURE 122.6. Penile zipper injury. A wire cutter may be used to cut the median bar of the zipper, releasing the two sides of the zipper and freeing the penis.

Strangulation

The penis may be encircled by a constricting ring formed by hair or a fiber or a thread, just as occurs with digits. Many times the cause of the problem is not immediately evident, because local edema may hide the ring of hair. The edema is produced by venous engorgement, which takes place early, after the development of this type of constriction around the penis. Once the source of the problem has been identified, therapy requires the division of the hair and the release of the constriction. This may require a general anesthetic. Pediatric urologic consultation is advisable. A urethrocutaneous fistula or even the loss of the penis has been reported, but is rare. How the hair comes to encircle the penis is generally unknown, but it should be remembered that such constriction occasionally has been reported as a form of sexual abuse.

TESTICULAR PROBLEMS

Background

Primordial germ cells have their origin in the entoderm of the yolk sac. By the fifth week of intrauterine life, they have reached the ventromedial portion of the urogenital ridge, the portion destined to form the testes. A mesodermal cord, the gubernaculum, becomes attached to the bottom of the testis at the epididymis and runs to the bottom of the scrotum. With rapid growth of the trunk, the testes lie adjacent to the internal ring by the third month of gestation. The testes remain at this location until the seventh month when, preceded by a fold of peritoneum (the processus vaginalis), the testes move down the inguinal canal and reach their final scrotal position shortly before birth. This fact accounts for the higher incidence of undescended testis in premature boys.

The gubernaculum appears to have an important role in testicular descent, although the exact nature of that role remains incompletely understood. Acute conditions involving the testes are discussed in Chapter 58.

Retractile Testis

In the physical examination of the child in the ED, an empty scrotum on one or both sides is a common finding. Although the testis may be found to be truly undescended, more often it is merely a retractile testis. In a boy with a retractile testis, the active cremaster muscle attached to the small prepubertal gonad is able to draw the testis up into a position near the pubic tubercle. There is no evidence that this causes any harm to the gonad. When the testis enlarges at puberty, it will assume a scrotal position permanently because the cremaster is no longer able to draw it out of its more normal position. The diagnosis of a retractile testis is made when one is easily able to milk the testis down into a position in a dependent portion of the scrotum where the testis stays, at least briefly, after overstretch of the cremaster muscle. In an obese youngster, it may be difficult to grasp the testis to pull it down. It is worthwhile putting a youngster in a “catcher's position,” in which the testis is pushed down to where it can be grasped and drawn into the scrotum. If the testis can be pulled into the scrotum but, regardless of how much the cremaster is overstretched, the testis “pops up” when released, this is a low form of a true undescended testis and not a retractile testis. This is a common diagnostic difficulty and pediatric urologic consultation should be sought if the situation is questionable.

Undescended Testis

True undescended testes are seen in 4% of newborn males. That instance decreases to 1.6% by 1 year of age, indicating that some undescended testes do descend after birth. Spontaneous descent rarely occurs after 6 months of age. Although it may be appropriate to continue for a few months to observe an infant who has an undescended testis, the child older than 6 months of age should have urologic consultation.

Testicular malignancy and infertility are increased in the male with an uncorrected undescended testis. By electron microscopy, it is possible to demonstrate degenerative changes in the undescended testis by 1 year of age. Early referral to a urologist for orchiopexy (before age 2 and preferably near age 1) appears advisable because data are now accumulating that indicate early surgery may decrease the incidence of both testicular malignancy and infertility.

Usually, an undescended testis is asymptomatic. However, in a position against the abdominal wall, it may be more subject to trauma than when freely mobile in the scrotum. The undescended testis also is malfixed and may undergo torsion more easily than a normally descended one. The boy who presents with an acutely tender groin mass with an ipsilateral empty scrotum may have a torsion of his undescended testis. The physician must consider the differential diagnosis of an incarcerated inguinal hernia or acute hydrocele of the cord. Prompt surgical treatment is required.

Varicocele

Varicoceles are abnormal dilations of the cremasteric and pampiniform venous plexuses surrounding the spermatic cord (Fig. 122.7). They generally present as an asymptomatic scrotal swelling about the time of puberty and are rare in the prepubertal boy. Almost all are of congenital origin and affect the left testis. The anatomic problem is a defect in the valves of the left spermatic vein that, on the left, drains directly into the left renal vein. Why varicoceles often are not noted until a boy approaches puberty is unclear, but they are common in that age group, affecting about 15% of adolescent boys. If the varicocele does not disappear when the child lies down, it suggests a varicocele secondary to obstruction of the left renal vein, and a renal and bladder ultrasound is appropriate. Varicoceles are rarely symptomatic; a heavy or tugging sensation is occasionally reported.

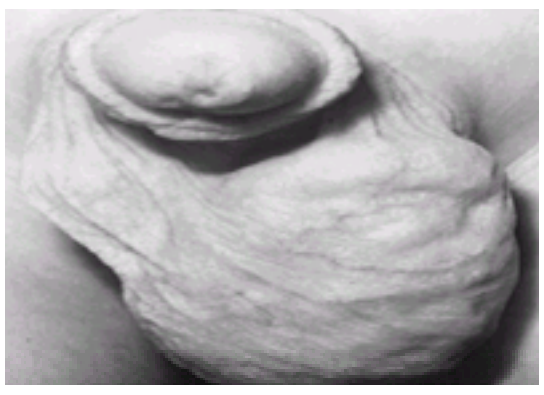

FIGURE 122.7. Varicocele: abnormal dilation of cremasteric and pampiniform venous plexuses surrounding the spermatic cord, giving scrotum the appearance of a “bag of worms.”

Approximately 15% of these boys with a varicocele will have an adult problem with infertility, although the exact mechanism of injury to the spermatogenic elements remains to be defined. Thus, periodic examination of these boys as they progress through pubertal change is recommended. As in the postpubertal testis, more than 80% of testis volume is a result of the spermatogenic elements; testis size is generally accepted as an indication of the effect of the varicocele on testis function. Although testicular asymmetry is common during pubertal change, a progressively smaller ipsilateral testis over 2 years or more of follow-up is an appropriate indication for surgical or radiographic treatment of the varicocele. “Catch-up” enlargement may occur after treatment of the varicocele. In our experience, any form of treatment is needed in only a small minority of cases. However, controversy exists on this point and long-term follow-up of adolescent boys with a varicocele is insufficient to permit firm conclusions.

URINARY TRACT INFECTIONS

Background

Urinary tract infection (UTI) ranks behind upper respiratory problems as the second most common form of bacterial infection in children. Between 1 and 2% of infants and children have bacteriuria at any given time, and 5% of all girls have UTI during their school years. Most UTIs result from fecal bacteria on the perineal skin ascending the urethra. The short female urethra, with resultant ease of bacterial contamination of the bladder, accounts for the higher incidence of UTIs in girls. The uncircumcised male infant appears also to be at increased risk of ascending urinary infection because foreskin bacterial colonization may lead to increased meatal contamination. However, as the absolute risk of UTI in infant males is in the order of 1%, it is questionable to suggest the risk of UTI is an indication for routine circumcision.

It is now recognized that the major risk factor in the development of UTI is the physical nature of the uroepithelium lining the urethra and bladder. In some children and adults, adherence factors in the mucosa lead to recurrent episodes of symptomatic infection. In addition, some bacteria (piliated ones) have increased adherence characteristics that add to the risk of invasive infection. Because voiding dysfunction may also contribute to recurrent infection, this is another reason to consider pediatric urologic consultations, especially in the older child who persists with wetting after appropriate treatment of infection.

A UTI may be defined as the multiplication of bacteria in the urinary tract. Normally, urine from the bladder and upper urinary tract should be sterile. The concept of “significant bacteriuria” (³$10^5$ organisms per milliliter of one colony type) in a cleanly voided midstream specimen is based on the statistical likelihood that this colony count is associated with the actual presence of bacteria in the bladder. A colony count of 10^5 or more organisms per milliliter of a single type suggests infected urine, with an 80% confidence level. Reliability can be increased to 95% if a second culture confirms the presence of the same bacteria type with identical antibiotic sensitivity; 10^4 to 10^5 bacteria per milliliter is an equivocal result and requires repeat culture. Less than 10^4 organisms per milliliter or the presence of several different organisms, suggests no infection or contamination of the specimen (see Chapter 84).

Clinical Manifestations

Particularly in the infant, UTIs may produce nonspecific findings. The urine may be cloudy or have a foul odor. There may be a history of unexplained fevers, general irritability, or failure to thrive and gain weight normally. Gastrointestinal (GI) symptoms are common, and many times the youngster with a UTI is believed to have gastroenteritis or a food allergy. A high index of suspicion is required. If a urine culture is not obtained, the source of the child's problem will be missed.

In the older child, symptoms may point more directly at the urinary tract. Frequency, urgency, and dysuria are produced by inflammation of the bladder and urethra. A previously toilet-trained child may begin to have “accidents.” Particularly in girls, hematuria may be seen. Although symptoms do not provide a completely reliable way of differentiating cystitis from pyelonephritis, the presence of systemic findings such as a high fever and malaise or abdominal/flank pain suggests renal involvement. A UTI, especially when chronic, may also have few or no symptoms. It is important to emphasize that in children, anything that irritates the urethral meatus may produce dysuria and occasionally urgency and frequency (see Chapter 54). The source of the irritation may be a tight or moist bathing suit or underwear or an ammoniacal rash. Bubble bath or other soap in contact with the urethral meatus may not only produce these symptoms, but, by producing inflammation, contributes to the ascent of bacteria up the urethra and the development of true infection. To avoid being confused by a noninfectious cause of symptoms, it is important that UTIs be proven by urine culture and not diagnosed by history and urinalysis alone.

Escherichia coli is the most commonly isolated organism responsible for UTI in children, constituting 80 to 90% of the total. This is because of the prevalence of the organism in GI tract flora, as well as its short mean-generation time, which enables it to multiply rapidly once it has entered the bladder. The other organisms commonly found can be seen in Table 122.1.

Escherichia coli	*Proteus* species
Klebsiella pneumoniae	*Pseudomonas aeruginosa*
Streptococcus faecalis (enterococcus)	*Staphylococcus epidermidis*

Table 122.1. Bacteria Commonly Causing Urinary Tract Infections

Management

The first step in management is to make an accurate diagnosis. The presence of pyuria does not provide an accurate criterion for the diagnosis of UTI. At least 20% of children with pyuria do not demonstrate significant bacteriuria. In any febrile illness, mobilization of the peripheral leukocyte pool may be adequate to produce the presence of white cells in the urine. Conversely, a child with bacteriuria occasionally does not demonstrate pyuria. Bacteria demonstrated by Gram

stain of an unspun urine specimen are more reliably indicative of a UTI. However, it is difficult to determine whether one type of bacteria or several different contaminants are present. Thus, culture of the urine must continue to be the benchmark for the diagnosis of a UTI in children. Obtaining an adequate urine specimen for bacterial culture is the most critical step in diagnosing UTI. A cleanly voided specimen obtained as a midstream catch after washing of the periurethral area is the preferred technique in the toilet-trained child. Simple soap and water washing of the periurethral area is preferred because antimicrobial soaps or solutions may become mixed with a voided specimen and lead to a false-negative result.

In the infant, obtaining an adequate urine specimen is more difficult. Specimens collected in a plastic bag (U-bag) attached to the perineum are rapidly contaminated by perineal bacterial skin flora. If a culture from a bag is sterile, it is acceptable. However, the demonstration of bacterial growth must be confirmed by some other means before a bona fide UTI can be presumed to be present. The most reliable way to obtain a confirming specimen of urine is by suprapubic aspiration of urine from the bladder, a procedure that is not dangerous and that has a reliability approaching 100%. The procedure for performing suprapubic aspiration is covered in Section VII. A specimen obtained by urethral catheterization is an acceptable alternative. If it is essential that the first specimen be the definitive one for diagnosis of UTI, as in the infant undergoing septic workup, the primary use of these techniques is justified. When symptoms strongly suggest the possibility of a UTI, beginning antibiotic therapy as soon as an adequate urine specimen for culture has been obtained is recommended. The matter of just 1 or 2 days before the institution of antibiotics may make a difference in the degree of eventual pyelonephritic scarring. If the urine culture turns out to be negative, the antibiotics may be stopped. Table 122.2 lists the most commonly used outpatient antibiotics for urinary tract infections.

Drug	Oral Dosage	Number of Doses
Trimethoprim–sulfamethoxazole	1 mL suspension/kg/day	2
Sulfisoxazole	120 mg/kg/day	4
Nitrofurantoin	5–7 mg/kg/day	4
Amoxicillin	50–100 mg/kg/day	3
Cephalexin	50–100 mg/kg/day	4

Table 122.2. Antibiotic Agents for Urinary Tract Infections

Although any of these antibiotic choices is acceptable in the initial therapy of a urinary tract infection, trimethoprim–sulfamethoxazole has become most commonly used in recent years because of its acceptance by children and high efficacy. Nitrofurantoin, although effective, can produce GI upset (lessened by taking with meals) and is less well tolerated by most children. Methenamine mandelate is not useful unless there is urinary stasis and acid urine, and accordingly has little role in most childhood UTIs. Tetracycline is not recommended for the child less than 10 years of age because of its potential for discoloration of the teeth. When the organism causing UTI is sensitive to the antibiotic selected, the urine is usually sterilized rapidly. It is advisable to repeat a culture 48 hours after starting an antibiotic. The continued presence of infection suggests inaccuracy of the sensitivity, noncompliance, or obstruction.

If a child is sufficiently toxic to warrant hospitalization, the intravenous administration of antibiotics is appropriate. The drugs of choice while cultures are pending are a cephalosporin or aminoglycoside, singly or in combination.

The duration of therapy has been a subject of recent debate. For uncomplicated cystitis, 1 to 3 days of therapy is usually adequate. For children who have not been radiographically evaluated or for any child with a congenital anomaly, a 10-day course of antibiotics continues to be recommended.

Other factors in the treatment of UTI involve high fluid intake with regular and frequent voidings to promote bladder washout of bacteria. If the child has a history of wetting, infrequent voiding, or frequent urge episodes, the possibility of dysfunctional voiding, which can contribute to recurrent infections, should be considered and appropriate consultation obtained. Avoiding constipation helps ensure better bladder emptying. Good perineal hygiene, including wiping from front to back after a bowel movement, is important. Eliminating pinworms prevents a source of inflammation, excoriation, and secondary increase in perineal skin flora. Bubble bath, by producing inflammation at the meatus, may promote the ascent of bacteria and should be avoided. Acidification of the urine with oral vitamin C or juices high in citric acid content may be useful to produce an acid urine in which bacteria multiply less rapidly.

Urologic Follow-Up and Radiographic Investigation

A suppressive dose of antibiotics should be begun after the acute phase of full-dose treatment. It is customary to use one-third to one-half the dose of antibiotic used for acute treatment, usually administered in a once-a-day evening dose. Suppressive antibiotics reduce the likelihood of recurrent infection, pending urologic consultation and radiographic investigation.

The routine radiographic evaluation of a urinary tract infection is by means of a voiding cystourethrogram (VCUG), followed by an ultrasound examination of the kidneys and bladder. These studies are usually carried out about 2 to 4 weeks after the acute treatment of a UTI; however, failure of a child to respond promptly to appropriate antibiotic therapy should lead to the urgent performance of an ultrasound examination to rule out urinary obstruction. The cystogram must include a voiding phase, or significant pathology may be missed, particularly vesicoureteral reflux, which may be evident

only on voiding films. In the usual child with a UTI, cystoscopy contributes little to the initial investigation; therefore, it is not recommended.

All boys should be investigated after their first UTI. In girls, the usual recommendations have been to wait until a second infection before recommending urographic investigation. However, Kunin's data demonstrate that after one UTI, there is an 80% likelihood of a second episode of bacteriuria and that half of these children will be asymptomatic. Thus, it appears justified to carry out radiographic studies after a first documented infection in girls, as well as boys, or at the least to follow girls who have recovered from a first UTI with repeat cultures at regular intervals.

In approximately 50% of infants and 30% of older children, an anatomic abnormality is found in association with a UTI. The most common finding is vesicoureteral reflux. Reflux permits infected urine to ascend to the kidney, where pyelonephritic damage may occur. With linear growth of the child, many milder cases of reflux may spontaneously resolve, leaving surgical management primarily for the more severe cases. These decisions are best made in consultation with a pediatric urologist.

As for the child who has no abnormality demonstrated by ultrasound and VCUG, the parents can be reassured that although the child may have a symptomatic problem from cystitis, there is little likelihood of renal damage. Occasionally, if a child has frequent episodes of symptomatic cystitis, suppressive antibiotics are justified in order to reduce the morbidity of these infections. The primary factor responsible for the development of urinary infection appears to be an adherent uroepithelium, which leads bacteria that ascend the urethra to stick to the bladder lining and become invasive. Children, like adults, who have such bladder lining may experience several UTIs per year. Surgical manipulations, such as urethral dilation, do nothing to change the basic bladder problem and are no longer performed. When infections recur in rapid sequence, this may indicate the colonization of the GI bacterial flora by organisms with increased adherence characteristics. Fortunately, during 3 to 6 months of suppressive antibiotic therapy, these organisms tend to modulate to less adherent bacteria. It should also be borne in mind that children with dysfunctional voiding patterns also tend to be troubled with frequent UTIs. Thus, if a child has an abnormal voiding pattern when uninfected (wetting, infrequent voiding), a pediatric urologic assessment is in order.

ACUTE URINARY RETENTION

A patient with acute urinary retention is unable to empty the bladder even though it is full. In children, as in adults, the cause may be a urethral obstruction. Congenital lesions, such as urethral valves, or acquired lesions, such as posttraumatic strictures, may lead to urinary retention. In such cases, a careful history often elicits symptoms of a weak stream or difficulty initiating the stream. Children who have any form of urethral irritation and dysuria may voluntarily retain urine. That is a different situation and needs to be separated carefully from organic obstruction causing retention. For the child with voluntary retention, gentle massage of the lower abdomen combined with a soak in a warm tub usually leads to spontaneous evacuation of the bladder. Rarely does a child's bladder become so distended, as after an outpatient surgical general anesthetic, that the child is unable to void. A simple one-time emptying of the bladder by catheterization with a feeding tube usually corrects the problem. It should be remembered that a child is able to hold urine voluntarily for longer periods than would be suspected; up to 12 hours is not unusual. Unless the child has a history suggestive of an organic obstruction or has a palpably enlarged bladder that cannot be emptied by massage and warm tub soaks, instrumenting the child's urethra should not be considered. Urologic consultation would be advisable before undertaking such maneuvers.

Suggested Readings

GENERAL

Brown MR, Cartwright PC, Snow BW. Common office problems in pediatric urology and gynecology. Pediatr Clin North Am 1997:44:1091–1115.

Middleton RG, Matlack ME, Nixon GW, et al. Genitourinary injuries in children. In: Mayer TA, ed. Emergency Management of Pediatric Trauma. Philadelphia: WB Saunders, 1985:341–352.

Shalaby-Rana E, Lowe LH, Blask AN. Imaging in pediatric urology. Pediatr Clin North Am 1997;44:1065–1089.

PENILE PROBLEMS

Baron M, Leiter E. The management of priapism in sickle cell anemia. J Urol 1978;119:610–611.

Kinney TR, Harris MB, Russell MO, et al. Priapism in association with sickle hemoglobinopathies in children. J Pediatr 1975;86:241–242.

Klauber GT, Sant GR. Disorders of the male external genitalia. In: Kelalis PP, King LR, Belman AB, eds. Clinical Pediatric Urology. 2nd ed. Philadelphia: WB Saunders, 1985.

Oosterlinck W. Unbloody management of penile zipper injury. Eur Urol 1981;7:365.

Osborn LM, Metcalf TJ, Mariani EM. Hygienic care in uncircumcised infants. Pediatrics 1981;67:365–367.

Rifkind S, Waisman J, Thompson R, et al. RBC exchange phoresis for priapism in sickle cell disease. JAMA 1979;242:2317–2318.

Seeler RA. Intensive transfusion therapy for priapism in boys with sickle cell anemia. J Urol 1973;110:360–361.

Wallerstein E. Circumcision: An American Health Fallacy. New York: Springer, 1980.

Wiswell TE, Roscelli JD. Corroborative evidence for the decreased incidence of urinary tract infections in circumcised male infants. Pediatrics 1986;78:96–99.

TESTICULAR PROBLEMS

Bierich JR, Giarola A, eds. Cryptorchidism. London: Academic Press, 1979.

Dubin L, Amelar RD. Varicocelectomy as therapy in male infertility: a study of 504 cases. J Urol 1975;111:640.

Fonkalsrud EW, Mengel W. The Undescended Testis. Chicago: Year Book, 1981.

Kass EJ, Lundak B. The acute scrotum. Pediatr Clin North Am 1997; 44:1251–1266.

Oster J. Varicocele in children and adolescents: an investigation of the incidence among Danish school children. Scan J Urol Nephrol 1971;5:27.

Scorer CG, Farrington GH. Congenital Deformities of the Testis and Epididymis. London: Butterworth, 1971.

Skoog JS. Benign and malignant pediatric scrotal masses. Pediatr Clin North Am 1997;44:1229–1250.

URINARY TRACT INFECTIONS

Feld LG, Waz WR, Perez LM, et al. Hematuria: an integrated medical and surgical approach. Pediatr Clin North Am 1997;44:1191–1210.

Kaye D, ed. Urinary Tract Infection and Its Management. St Louis: Mosby, 1972.

Kunin CM. Detection, Prevention and Management of Urinary Tract Infections. 4th ed. Philadelphia: Lea & Febiger, 1987.

Rushton HG. Urinary tract infections in children: epidemiology, evaluation and management. Pediatr Clin North Am 1997;44:1133–1169.

Stahl GE, Topf P, Fleisher GR, et al. Single dose treatment of uncomplicated urinary tract infections in children. Ann Emerg Med 1984; 13:705.

Stamey TA. Pathogenesis and Treatment of Urinary Tract Infections. Baltimore: Williams & Wilkins, 1980.

CHAPTER 123

Orthopedic Emergencies

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ORTHOPEDIC CONDITIONS

Most orthopedic emergencies are related to trauma (see Chapter 115). Anecdotes abound, however, of non-traumatic orthopedic problems that present as injuries. A history of a recent fall is prevalent, perhaps ubiquitous, in active young children. Parents, seeking to understand their child's complaint, assume a painful extremity is the result of a recent mishap. Physicians must not be misled. The non-traumatic orthopedic conditions described in this chapter are especially important to consider when a reported injury mechanism is minor, or onset of symptoms is delayed. Evaluation of children with complaints such as limp (see Chapter 43), joint pain (see Chapter 57), and back pain (see Chapter 51) to name a few, requires consideration of traumatic and non-traumatic causes. Overuse syndromes may occur at any age and can present as trauma. Children train more intensively and at younger ages than in previous years, resulting in overuse syndromes. With knowledge and appropriate suspicion, physicians caring for children can identify non-traumatic orthopedic problems, begin treatment, and make intelligent recommendations about referral.

Osteomyelitis

Background

Osteomyelitis is an inflammation of the bone, most commonly of infectious origin. Infection is confirmed by the presence of two of the following: pus on an aspirate of the bone, clinical findings consistent with the diagnosis, positive blood or bone aspirate cultures, and radiologic imaging. Osteomyelitis is more common in boys, and several studies have found the highest incidence among infants and preschool children. Age and underlying disorders are associated with an increased risk for contracting osteomyelitis, as well as for the particular pathogens involved.

Pathophysiology

Infection occurs by one of three routes: hematogenous, direct spread, or inoculation through a penetrating wound. Hematogenous spread is the most common route in children. A transient bacteremia is believed to be the initiating event in the infection. Bacteria enter the bone at the level of the metaphysis through the predominant vascular supply of the bone. The sluggish blood flow within the microvasculature of the marrow predisposes to infection. Local trauma has been suggested as a possible cause of micro-thrombotic events further predisposing bone to infection. This is supported by the preponderance of infection occurring within the long bones, especially those of the lower extremities. In sickle cell patients micro-infarcts within the more tenuously supplied area of the diaphysis may explain the increased occurrence in this region of the bone. As infection progresses, pressure increases, and organisms penetrate up through the cortex to the subperiosteal space. If left untreated, the infection may spread along this space or rupture through the periosteum into the surrounding soft tissue.

Differences in the underlying bony structure in the neonate and young infant predispose them to a higher incidence of multi-focal osteomyelitis and concomitant septic arthritis. The periosteum is less adherent in these ages and less

effective in limiting the spread of infection. Transphyseal vessels, which are present up to 18 months, allow bacteria to gain access to the adjoining epiphysis and joint space.

A less common source of osteomyelitis in children is penetration of the periosteum by local infections. Inoculation of the bone from stepping on a nail, surgical instrumentation, or intraosseous line placement, provides a third means for infection to gain entrance to the bone. With either mechanism of infection, osteomyelitis can progress to chronic osteomyelitis that may have deleterious effects on growth.

Clinical Findings

Physical signs of osteomyelitis are age dependent. The older child is more likely to have localized infection and is more capable of expressing or identifying a site of localized point tenderness. The neonate or young infant may present with a pseudoparalysis of the affected limb. Another common, although nonspecific, finding in this age group is paradoxic irritability in which the infant exhibits pain or distress upon handling, and is more comfortable when left alone.

Fever and pain are highly sensitive findings but not universally present. Fever is described in up to 90% of children with osteomyelitis upon presentation and may be quite elevated. Pain is expressed through limp, refusal to bear weight, or a decreased range of motion when a limb is involved. Erythema and swelling are less common, but can also be observed at the site and usually suggest more advanced periosteal involvement.

Diagnosis

In addition to clinical findings, the diagnosis of osteomyelitis depends on culture results. Blood cultures and bone aspirates should be obtained in suspected cases of osteomyelitis before the initiation of antibiotics. Isolation of the causative organism is important not only in diagnosis, but also in antibiotic selection and the possibility of eventual outpatient therapy. Reports of positive blood cultures range from 30 to 57%. An organism is recovered from a bone aspirate in 51 to 90% of cases. The combination identifies a pathogen in 75 to 80% of cases. Bone aspirates may remain positive for several days after antibiotic use, whereas blood cultures are often sterile within 24 hours of antibiotics.

Laboratory tests vary in sensitivity. The white blood cell (WBC) count is elevated in only a third of the cases of osteomyelitis, whereas both the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) are elevated in more than 90%. The latter tests are useful both in diagnosis and for monitoring the response to therapy.

The plain radiograph is the initial imaging study of choice. It is useful both in detecting early signs of osteomyelitis and excluding other diagnostic possibilities. The earliest radiograph changes suggestive of osteomyelitis include deep soft-tissue swelling with elevation of the muscle planes from the adjacent bone (Fig. 123.1). These may be seen as early as 3 to 4 days after the onset of symptoms. Lytic bone changes are not detectable until 7 to 10 days. Periosteal elevation, when present, is not generally visible until 10 to 21 days after infection (Fig. 123.1). A negative radiograph in the first 10 days of illness does not rule out osteomyelitis. When suspicion remains high in the setting of a negative radiograph, a bone scan should be obtained. The triple phase technetium bone scan has a reported sensitivity and specificity of more than 90%, and detects osteomyelitis within 24 to 48 hours of symptom onset. A bone aspirate preceding a bone scan will not cause a false-positive result.

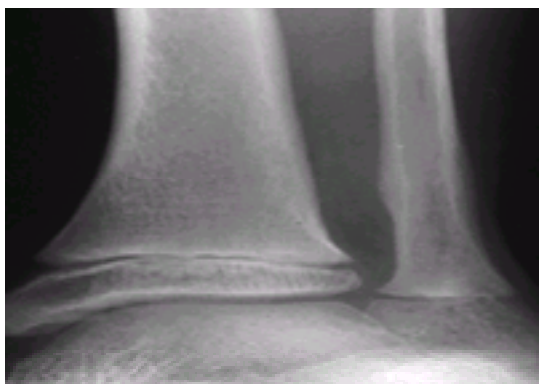

FIGURE 123.1. Periosteal activity in distal fibula in child with *Staphylococcus aureus* osteomyelitis; day 20 of illness.

Other imaging modalities, including magnetic resonance imaging (MRI), computed tomography (CT), and ultrasound, may have a limited role in certain complicated cases or to obtain further details of the infection, but they should not replace the plain film or bone scan in the acute setting.

Microbiology

Organisms responsible for osteomyelitis differ according to the patient's age and the route of infection. *Staphylococcus aureus* is the most common pathogen across all age groups. Epidemiologic data gathered before availability of the *Haemophilus influenzae* type b (Hib) vaccine showed *H. influenzae* to be a common organism in children less than 2 years of age. Although there is evidence of a dramatic decline in the incidence of invasive *H. influenzae* disease since the advent of the Hib vaccination, most experts still recommend that initial antibiotic coverage include *H. influenzae* for children less than 5 years of age. Bacterial isolates from neonates less than 2 months include *S. aureus,* group B streptococcus, and *Escherichia coli,* and antibiotic coverage should reflect this.

Certain groups are at risk for particular organisms. Patients with sickle cell disease have a high incidence of osteomyelitis caused by *Salmonella. Pseudomonas aeruginosa* is the predominant organism found in osteomyelitis of the foot, resulting from a nail penetrating a sneaker.

Management

Initial therapy for osteomyelitis includes intravenous antibiotics. Antibiotic coverage should be based on the predominant organisms in each age group, the mechanism of infection, and Gram stain results. Suggested agents are listed in Table 123.1. Early aggressive antibiotic therapy often prevents the need for surgical intervention.

Age	Pathogens	Antibiotics
Neonate <2 mo	*Staphylococcus aureus*, group B streptococcus, Gram-negative bacilli	Nafcillin and Gentamicin
<5 yr	*S. aureus*, group A streptococcus, *Streptococcus pneumoniae*, *Hemophilus influenzae*	Cefuroxime or Ampicillin/sulbactam Nafcillin and chloramphenicol
>5 yr	*S. aureus*, group A streptococcus, *S. pneumoniae*	Nafcillin or Cefazolin
Special Cases		
Sickle cell disease	*S. aureus*, Salmonella	Nafcillin and ceftriaxone or Nafcillin and chloramphenicol
Foot puncture wound	*Pseudomonas aeruginosa*, *S. aureus*	Nafcillin and ticarcillin or Ticarcillin/clavulanate or Nafcillin and ceftazidime

[a] Vancomycin or clindamycin in penicillin- and cephalosporin-allergic patients.

Table 123.1. Initial Antibiotic Therapy Osteomyelitis [a]

Septic Arthritis

Background

The presence of bacterial pathogens within the articular capsule presents a true surgical emergency. Delay in the identification and treatment of an infected joint in a child can result in severe and permanent sequelae. The urgency associated with this diagnosis has given rise to the maxim, “The sun should never rise or set on an untreated septic hip.” In many cases, the pediatric emergency physician is the initial point of contact, and must maintain a high index of suspicion to recognize and appropriately treat these patients at the time of the initial visit.

Pathophysiology

Bacteria gain entry to the joint space through one of three means. The synovium is most commonly infected through hematogenous seeding. The role of local injury in predisposing joints to infection is unclear. Adjacent areas of infection may invade the joint, or direct inoculation can occur through penetrating injuries. Infection secondary to penetrating objects may be delayed from the actual time of injury. External wounds may be small or healed at the time of presentation. Undetected penetration of the knee joint, for example by a sewing needle in a crawling infant, may give rise to a septic knee. A metatarsal joint infection may be the result of a nail puncture wound to the foot that occurred several weeks earlier.

Eighty to ninety percent of septic joints occur in the lower extremities. The knee and hip are most commonly afflicted. The same distribution is found in the preambulatory child. Infections involve only a single joint in more than 90% of cases. Multifocal infections are more common in neonates.

Once established within the joint, bacteria release proteolytic enzymes that directly damage the intra-articular cartilage. Pressure elevation within the minimally distensible capsule can lead to dislocation of a joint, and compromised vascular flow, resulting in ischemic injury to the bone. This is a particular concern in the hip, where avascular necrosis of the femoral head is a well-described complication of septic arthritis. Prognosis is worse in children less than 1 year old, with involvement of the hip joint, with delay to the initiation of therapy, and with infection by *S. aureus.*

Clinical Findings

Pain is the most common presenting complaint in the child with a septic joint. This may be expressed in many ways. The older child is better able to localize the area of discomfort. Because of the predominance of septic arthritis in the lower extremities, the younger child will often present with a limp, abnormal gait, or inability to bear weight.

Range of motion around the affected joint is dramatically reduced. Any degree of movement causes great distress and is vigorously resisted. Many orthopedic surgeons rely on this aspect of the evaluation more than any other in differentiating infection from alternative causes of joint pain.

Clinical signs are more subtle in the neonate or young infant with a septic joint. Nonspecific findings such as septic appearance, irritability, and pseudoparalysis of a limb are common presenting findings in these ages. Parents may note excessive irritability associated with diaper changes in the infant with a septic hip. The child with a septic hip will typically hold the lower extremity in abduction and external rotation in order to maximize the volume of joint space (Fig. 123.2). A high degree of suspicion, close observation, and isolated manipulation of each extremity help locate the particular area of

involvement.

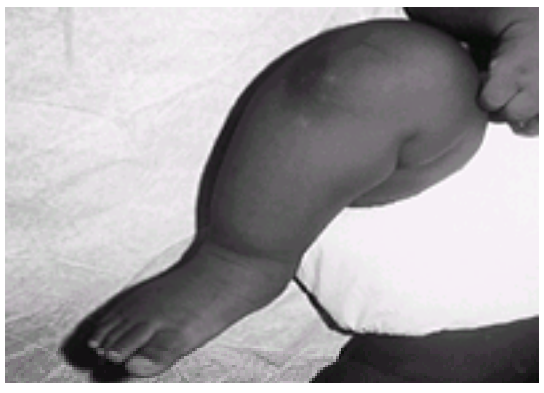

FIGURE 123.2. Five-month-old infant with septic arthritis of the right hip. Hip joint is held in flexion, abduction, and external rotation.

The skin surface should be closely evaluated for local signs of injury. Most involved joints have obvious erythema, warmth, and swelling. The exception is the hip joint because of its deep-seated location. Swelling may be less obvious in the pudgy infant. Fever is a commonly associated sign, but is absent in up to one-third of patients.

Diagnosis

The diagnosis of septic arthritis is confirmed by the presence of purulent fluid within the joint space. Arthrocentesis is a mandatory procedure in all suspected causes of septic arthritis. The level of suspicion and decision to perform this procedure is based on the degree of clinical suspicion in combination with results of laboratory tests and imaging studies. None of these in isolation are 100% sensitive in detecting or excluding septic arthritis from other conditions. A sample of synovial fluid is essential in discriminating septic arthritis from less serious inflammatory processes.

The mean WBC count is elevated in children with septic arthritis; however, more than one-half of patients will have a WBC count less than 15,000/mm^3. The ESR and CRP are more sensitive markers and are elevated in 90 to 95% of patients.

Plain radiographs may demonstrate signs of an effusion ranging from subtle blurring or displacement of fascial planes to complete dislocation of the joint. The main role of the radiograph in the evaluation is to exclude fractures or other bony abnormalities that may mimic septic arthritis. Ultrasound is increasingly used to evaluate questionable joints, especially the hip. It is much more sensitive than the radiograph in detecting a joint effusion. Some have suggested that the absence of an effusion on an ultrasound scan effectively excludes the diagnosis of septic arthritis. However, the ultrasound cannot distinguish between infected and sterile inflammatory effusions. The bone scan localizes areas of inflammation and is unaffected by prior arthrocentesis. It cannot differentiate infection from other causes of inflammation. A bone scan may be helpful in excluding osteomyelitis. Inflammation is found symmetrically across a joint in septic arthritis, whereas in osteomyelitis, it is limited to one side. The reliability of a bone scan in differentiating joint from bone involvement decreases in the neonatal age group.

The isolation of a bacterial pathogen is important in diagnosis and in directing subsequent management. Cultures of joint fluid and blood should be performed on all patients with a possible septic joint. When indicated, cultures from additional sites should be obtained to increase the potential isolation of a pathogen. Cultures of the joint fluid demonstrate the highest yield, and are positive in 50 to 80% of cases. Blood cultures identify an organism in 15 to 46% of patients with septic arthritis, and are positive in many cases in which the organism is not isolated from the joint fluid. Cerebrospinal fluid (CSF) cultures have been helpful in the past in identifying *H. influenzae.* Cervical or urethral cultures in sexually active adolescents with septic arthritis may provide the responsible organism. A causative organism is not recovered in 20% of cases.

A Gram stain should be performed on joint fluid and occasionally provides additional assistance in identifying both an infection and the infecting organism. Although elevation of the WBC count in the synovial fluid above 100,000/mm^3 is considered strong evidence of infection, the actual counts are often much lower.

A differential with greater than 90% neutrophils is also highly suggestive of infection. Presence of purulent fluid, a positive Gram stain, and an elevated WBC count with a left shift in the synovial fluid are often used as indications for operative intervention when there is a concern of a septic hip.

Microbiology

With a few exceptions, the bacteria found in septic arthritis are the same as those in osteomyelitis. *S. aureus* is the most common reported isolate in children older than 5 years, followed by group A *streptococcus* and *Streptococcus pneumoniae.* Before the introduction of the Hib vaccine, *H. influenzae* type b was the leading cause of septic arthritis in the 6 months to 5-year-old age group. *S. aureus* is the predominant pathogen in neonatal patients. Gram-negative coliforms are also found in this age group. *Neisseria gonorrhoeae* is found in the neonatal ages and is a common pathogen in sexually active teenagers. *Kingella kingae* is a fastidious Gram-negative rod, susceptible to b-lactam antimicrobials, which has recently been isolated as a pathogen in a number of childhood bone and joint infections.

Management

The management of septic arthritis consists of parenteral administration of antibiotics (Table 123.2), joint immobilization, and joint irrigation in selected cases. Empiric antibiotic therapy is dictated by the common organisms in the age group and results of the synovial fluid Gram stain. An anti-staphylococcal agent consisting of a b-lactamase–resistant penicillin or a first-generation cephalosporin is effective in most cases. Gram-negative coverage should be added in neonates and adolescents. Until further evidence of the eradication of *H. influenzae* type b from the younger age group, appropriate coverage is recommended for children less than 5 years.

Age	Pathogens	Antibiotics
Neonate	*Staphylococcus aureus*, group B streptococcus, Gram-negative bacilli	Nafcillin and Gentamicin or cefotaxime
≤5 yr	*S. aureus*, *Hemophilus influenzae*, group A streptococcus, *Streptococcus pneumoniae*	Cefuroxime or Ampicillin/sulbactam
>5 yr	*S. aureus*, group A streptococcus	Nafcillin
Adolescent	*S. aureus*, group A streptococcus, *Neisseria gonorrhea*	Nafcillin Ceftriaxone[b]

[a]Common pathogens and empiric antibiotic coverage by age.
[b]Empiric treatment in sexually active adolescent.

Table 123.2. Initial Antibiotic Therapy Septic Arthritis[a]

Surgical intervention for joint irrigation is generally indicated for all cases involving the hip joint, infections in which large amounts of fibrin, debris, or loculations are found within the joint space, or when the patient fails to improve after several days of intravenous antibiotic therapy. Expeditious and aggressive management limits, but does not eliminate, potential sequelae of septic arthritis.

Toxic Synovitis

Background

Toxic or transient synovitis is a benign, self-limiting inflammatory process of the hip. It afflicts males more often than females and is the most common cause of acute hip pain in children 3 to 10 years of age. The underlying cause is unknown, although a post-infectious inflammatory response has been suggested. Its presentation can mimic that of septic arthritis of the hip (Fig. 123.3), a distinction that is as crucial in management as it is difficult in diagnosis.

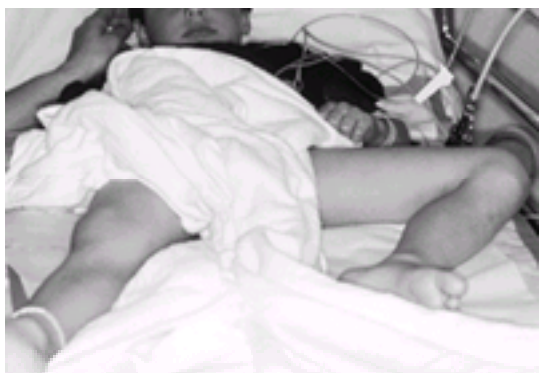

FIGURE 123.3. Seven-year-old child with toxic synovitis of the left hip. Hip joint is held in same position of comfort as in septic arthritis.

Clinical Findings

The onset of symptoms is abrupt with unilateral hip pain and limp (Fig. 123.3). Fever is rare, occurring in less than 10% of cases, and, when present, is usually low grade. Although patients complain of discomfort with movement of the limb, it generally remains possible to put the hip through a full range of motion. This contrasts with the septic hip in which pain and spasm are more extreme, and patients resist a full range of motion. Additional signs of systemic illness are absent and, despite the title, the child is nontoxic appearing.

Laboratory

Laboratory tests are generally useful only in attempting to distinguish toxic synovitis from more serious conditions. The WBC count and ESR are generally normal or only slightly elevated. The mean WBC count and ESR are significantly lower than in septic arthritis; however, sufficient overlap exists between values in toxic synovitis and septic arthritis such that they cannot be relied on to distinguish between them in individual patients.

Radiographs may demonstrate an effusion, but its principal role is in excluding pathologic osseous conditions. Ultrasound is more sensitive than plain films at detecting joint effusions, although accuracy declines in patients under a year. Reports of an effusion of the hip by ultrasound in toxic synovitis vary from 50 to 95%. Although patients often report relief of pain after aspiration, the procedure is unnecessary except to exclude the presence of a bacterial infection. When obtained, synovial fluid is sterile.

Management and Prognosis

Treatment occurs on an outpatient basis and emphasizes rest and analgesics. Traction is of unproven benefit and is potentially harmful. Nonsteroidal anti-inflammatory medications are the first-line therapy for pain. Pain duration is typically 3 to 4 days but may last as long as 2 weeks. Exacerbations can occur if activity is resumed too early.

There is no evidence of serious sequelae resulting from toxic synovitis. The relationship between toxic synovitis and Legg-Calve-Perthes disease is unclear. Studies have been unable to demonstrate cause and effect. Some suggest that these patients are at increased risk for developing Legg-Calve-Perthes, and others only that the clinical presentations are similar.

Penetrating Intra-Articular Wounds

Penetrating intra-articular wounds are not specific to children, but they are injuries that the pediatric emergency physician must recognize and treat on an urgent basis to prevent serious and potentially permanent sequelae. Knees are the most commonly injured joints. Motor vehicle accidents are the cause in the overwhelming number of cases.

An open joint may be detectable on direct visualization or by palpation through a periarticular laceration. Injuries that extend below the skin surface adjacent to a joint effusion should raise a high level of suspicion that the joint space has been violated. The presence of air in the joint on radiograph is diagnostic for joint penetration (Fig. 123.4). In less obvious cases, disruption of the joint capsule can be demonstrated by the saline load test. Arthrocentesis is performed through an uninjured site on the skin surface and saline is injected. Extravasation of saline from the joint into the wound is diagnostic for penetrating injury. A volume of 60 mL of saline is generally adequate to evaluate knee joint integrity, 20 mL for elbow or ankle joints, and 1 to 2 mL for finger joints. The addition of a small amount of methylene blue to the saline may improve visualization of extravasated fluid. Voit et al. found that clinical evaluation had poor sensitivity in identifying penetrating wounds compared with saline injection. If a question still remains regarding the integrity of the joint after such testing, then further imaging studies or surgical exploration is necessary.

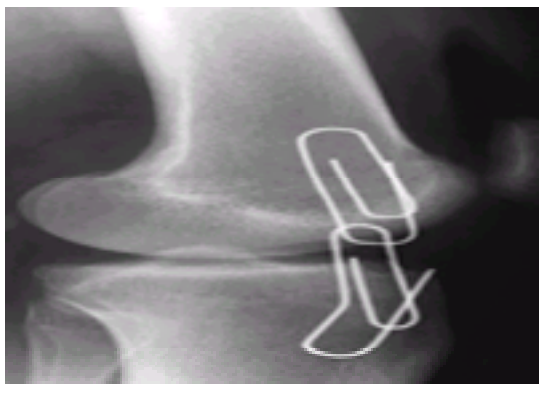

FIGURE 123.4. Intra-articular air in knee joint following penetrating injury sustained in a motor vehicle accident. Center of paper clips mark entrance and exit sites.

The treatment of an open joint wound is similar to that of an open fracture. Open wounds of the joint are considered contaminated and broad-spectrum antibiotics should be administered. Surgical intervention consists of vigorous irrigation and debridement, often in the operating room. Attention should be given to appropriate tetanus prophylaxis, splinting, wound dressing, and pain control while the patient remains in the emergency department.

The prognosis of penetrating intra-articular wounds depends on the degree of overlying soft-tissue injury and the extent of intra-articular damage. Infectious complications are the most common. Septic arthritis with a variety of both Gram-positive and Gram-negative organisms is an early and common outcome of inadequate early intervention. Delayed synovitis, often necessitating synovectomy, has been described after unidentified penetration of small foreign bodies.

Radial Head Subluxation

“Nursemaid's elbow” is the most common joint injury in pediatric patients, usually occurring in children between 6 months and 5 years of age. The left elbow is more often affected because adult caretakers prefer to hold the child's left hand with their dominant right hand. Subluxation of the radial head occurs as a result of abrupt traction on a pronated hand or wrist. The annular ligament slides over the radial head and becomes interposed between the radius and capitellum. The radial head is not abnormal and the annular ligament need not tear for this injury to occur.

In up to half of cases, a history of traction on the arm is not obtained, which may suggest other mechanisms for this injury, or perhaps caretakers who are reluctant to provide self-incriminating information. Astute clinicians should suspect

this injury even in the absence of an appropriate history.

Subluxation of the radial head can be strongly suspected from across the room. The child holds the arm slightly flexed and against his or her body. When left alone, the child does not appear to be in significant pain. Parents may report a problem with the wrist or shoulder because, in their attempts to check these joints, inadvertent movement of the elbow causes pain. Physicians can be similarly fooled, especially when a classic history is not obtained.

Evaluation and Management

The young child must be approached in a slow and non-threatening way. Point tenderness of the clavicle, humerus, radius, and ulna can be excluded with a deliberate examination that does not move the elbow at all. True tenderness and swelling at the elbow are usually absent. When disuse of the elbow is present without pain or bony tenderness, the clinician should perform the reduction maneuver to confirm the diagnosis of radial head subluxation. Radiographs of the elbow are unnecessary unless the physician suspects another injury. Swelling and localized tenderness are usually apparent with supracondylar fractures, the next most common elbow injury in this age group.

Reduction of a subluxed radial head is one of the most gratifying procedures for physicians and parents alike. Several effective maneuvers are described. The clinician holds the elbow with his or her thumb over the radial head (Fig. 123.5). Supination of the affected arm rotates the flared aspect of the radial head, snapping the annular ligament back to its original position with a telltale click. Flexion or extension of the elbow may add to the success rate. If no click is felt, a second attempt can be made, perhaps exerting a little traction to disengage the annular ligament from between the radial head and capitellum. In the absence of a click, the child should be observed for return of function of the arm because many successful reductions occur without a perceptible click. Excessive failed attempts at reduction should be avoided. Radiographs may be useful for patients who fail reduction maneuvers.

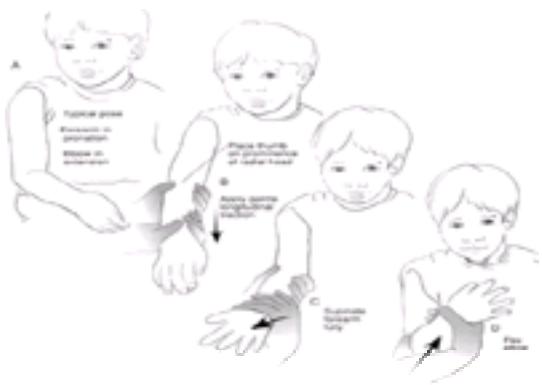

FIGURE 123.5. A–D. Supination–flexion maneuver for reduction of radial head subluxation (nursemaid's elbow).

Return of function after successful reduction is usually prompt, but not immediate. Toys, bottles, or interesting objects can be used to encourage the child to use the affected arm. Voluntary use of the arm will return in less than 15 minutes in almost 90% of patients. Younger children generally take longer to begin reusing the arm. Despite common belief, duration of subluxation was not associated with time to return of function. Many clinicians relate experiences with “failed” reductions in children whose arms are better the following morning. If disuse of the arm persists, and radiographs are normal, a sling should be placed and the child should be seen in follow-up by an orthopedist.

Recurrent radial head subluxations are common, occurring in about one-third of cases. Caretakers should be counseled to lift the child from the axillae, avoiding traction on the extremities.

Shoulder (Glenohumeral) Subluxation/Dislocation

Shoulder dislocation is extremely uncommon in young children. Shoulder dystocia at delivery can lead to displaced Salter I fractures. These injuries may look like dislocations because the unossified proximal humeral epiphysis remaining in the glenoid fossa is not visible radiographically. True dislocations become more common in adolescence and their management is described in Procedures, Section VII.

Glenohumeral subluxation/dislocation may be recurrent, especially if the anterior glenoid rim is avulsed (Bankhart lesion). Patients may report that the shoulder “pops out” and reduces spontaneously. Some disturbed individuals intentionally dislocate their shoulder. Patients with recurrent shoulder dislocation need to be evaluated by an orthopedist. If a rehabilitation program is unsuccessful, a reconstructive procedure may be beneficial.

Slipped Capital Femoral Epiphysis

Background

Slipped capital femoral epiphysis (SCFE) is the most common hip disorder in adolescent patients and should be familiar to all who care for children in this age group. It is twice as common in males as females, and more common in African-American patients. Obesity is a risk factor, although not all patients with SCFE are overweight. For unknown reasons, it is diagnosed far more often in the eastern portion of the United States, with a reported incidence of 3.41 per 100,000 in Connecticut and 0.71 per 100,000 in New Mexico. Cases are usually sporadic, but some familial tendency has been noted. Most children with SCFE are early adolescents in their growth spurt. Boys are most commonly affected

between 13 and 15 years of age; girls are most commonly affected between 11 and 13 years of age because of their earlier pubertal development. SCFE onset after menarche is extremely rare.

Slippage of capital femoral epiphysis is almost always posterior and inferior relative to the proximal femoral metaphysis. Displacement anteriorly or superiorly has been reported. The epiphysis maintains a normal relationship with the acetabulum. The left hip is affected more often than the right. Radiographic evidence of bilateral SCFE is common, even though symptoms are usually unilateral. Plain radiographs document bilateral slippage in about 25% of cases, CT scans in approximately 50%.

Pathophysiology

The pathogenesis and biomechanics of SCFE have been the subject of some research and much reasoned speculation. The perichondrium is primarily responsible for the strength of the proximal femoral physis. SCFE differs from a displaced Salter I fracture in that the perichondrium remains intact in most cases of SCFE and is disrupted with acute Salter I fractures. Collagenous bridges traverse the physeal cartilage and also contribute to the shear strength of the physis. The undulating convexity of the physis toward the epiphysis further stabilizes the interface. It takes an enormous shearing force to produce acute slippage of an initially normal hip joint. SCFE is usually a gradual, chronic process because of the viscoelasticity of the physis. Acute presentations, with radiographic evidence of chronic slippage, are common.

Several factors contribute to slippage of the capital femoral epiphysis in early adolescence. The perichondrial ring thins, probably in response to hormonal changes, reducing its support of the physis. The increased height of the physis lengthens the lever (moment) arm, increasing shearing forces on the growth plate. The geometry of the proximal femur changes during the growth spurt in adolescence such that the vector of body weight is more perpendicular to the femoral neck, adding to the shearing force across the physis. Increasing body weight, especially in obese children, obviously contributes to the forces on the physis. The combination of unfavorable changes in geometry, weakening of the physeal–epiphyseal complex, and increasing weight explains the characteristic age distribution of patients with SCFE.

Most patients with SCFE do not have identifiable endocrinologic problems. However, several hormonal abnormalities have been associated with increased risk. Elevated growth hormone and somatomedin, hypogonadism, hypothyroidism, and secondary hyperparathyroidism from renal failure (renal osteodystrophy) have been associated with SCFE. Short children receiving exogenous growth hormone therapy and tall, thin, rapidly growing children with high levels of endogenous growth hormone are both at increased risk. Children outside of the usual age range for SCFE, and those with other signs and symptoms that suggest possible endocrine abnormalities, should be referred for endocrine evaluation.

Clinical Presentation

Pain and/or limp are the most common chief complaints in patients with SCFE. Physicians may be misled when the pain is referred to the thigh, knee, or groin. It is often dull, vague, intermittent, and chronic in nature. Many patients have had symptoms for weeks or months at the time of presentation. A history of trivial injury is sometimes obtained, perhaps causing the additional slippage that precipitates a medical evaluation. Acute onset of severe symptoms suggests acute or acute-on-chronic slippage. These patients are often unable to bear weight and may be in significant pain. Major trauma can cause SCFE, but these presentations are rare.

Examination findings in patients with SCFE include a resting position with hip flexion and some external rotation. Range of motion of the hip, especially full flexion, medial rotation, and abduction, is decreased and tender. Patients with significant displacement may have evidence of limb shortening. Occasionally, there is tenderness of the hip anteriorly. Patients with more acute presentations should not be forced to walk as part of the evaluation. Testing for full range of motion is potentially injurious and unnecessary once a decision to obtain radiographs has already been reached.

Diagnosis

Plain radiographs of the hip should include two views because SCFE may be missed on an anteroposterior (AP) view alone (Fig. 123.6). On the AP view, widening of the physis is usually seen, even if the displacement is inapparent. The epiphysis is almost always displaced posteriorly; therefore, a frog leg or lateral view is best for documentation of the slippage. External rotation of the hip in the frog view turns the posterior aspect medially. Following the medial margin of the femur proximally reveals a step off between the metaphysis and epiphysis. New bone formation is often visible, suggesting a chronic slip. When radiographic findings are equivocal, comparison with the contralateral, asymptomatic hip should be done with caution, given the frequency of bilateral slippage with unilateral symptoms.

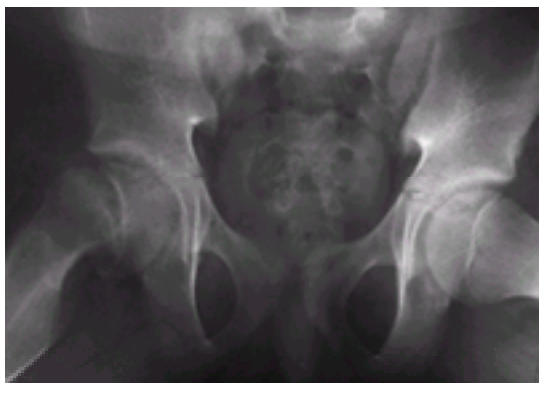

FIGURE 123.6. Slipped capital femoral epiphysis of right hip. Epiphysis is displaced medially on the frog view.

SCFE is classified by symptom duration, stability and degree of displacement. Patients with acute SCFE have symptoms for less than 3 weeks; with chronic SCFE, symptoms are present for more than 3 weeks. Acute-on-chronic SCFE describes patients with symptoms for more than 3 weeks with a recent exacerbation. An acute slip with severe symptoms is unstable. Acute or chronic slips with mild symptoms are stable, and have a more favorable prognosis. The degree of slippage is expressed with a grading system; grade I or preslip with widening of the physis but no displacement; grade II with displacement less than one-third of the width of the metaphysis; grade III with displacement of one-third to one-half of the metaphyseal width; and grade IV with displacement of greater than one-half the metaphyseal width.

Management

Treatment of SCFE is primarily surgical. Screws are usually placed through the femoral neck into the epiphysis. Reduction of the displacement is not performed because there is some evidence that it may increase the likelihood of avascular necrosis of the femoral head and chondrolysis. Chondrolysis is the most common complication of SCFE, occurring in about 8% of patients. Pain and persistent decreased range of motion after pinning are the usual presenting symptoms. If the pins extend into the joint space, the risk of chondrolysis is increased. Two-thirds of patients with chondrolysis have a progressive course. Ankylosis may ensue, leading to long-term disability.

Legg-Calve-Perthes Disease

Legg-Calve-Perthes Disease (LCPD) is a hip disorder that generally has onset between the ages of 4 and 9 years. Males outnumber females by a ratio of 4:1. Genetic factors play a minor role. Most children with LCPD are short, with average or above-average weight. They often have delayed skeletal maturation.

LCPD begins with repeated episodes of ischemia of the femoral head, leading to infarction and necrosis. Theories about the cause of the circulatory insufficiency include increased blood viscosity and elevated intracapsular pressure caused by synovitis, but these theories remain unproven. Patients may remain asymptomatic despite varying degrees of necrosis and resorption of the femoral head. Some children recover completely without developing symptoms. Symptoms generally begin when minor trauma causes stress fracture of the subchondral bone. Rarefaction of the femoral head with subluxation and deformity may ensue. The process of reossification and remodeling takes 2 to 4 years.

The onset of symptoms in LCPD is usually insidious. Presentation as an acute emergency is rare. Mild hip pain and limp have usually been present for weeks to months before diagnosis. Pain is often referred in the distribution of the obturator nerve to the knee, anteromedial thigh or groin. Physical findings include decreased hip abduction and medial rotation. Thigh muscle atrophy, and in advanced cases, limb shortening may also be noted.

The sequence of radiographic changes in LCPD have been described in detail (Fig. 123.7). At diagnosis most patients have widening of the articular cartilage with a small, dense proximal femoral epiphysis. Subchondral fracture may be visible. Irregularity and flattening of the epiphysis develops over time. The differential diagnosis includes various bone tumors and skeletal dysplasias. As the disease progresses, anterolateral subluxation may be quantitated radiographically.

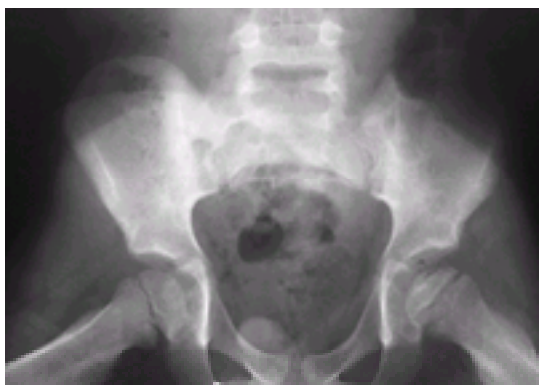

FIGURE 123.7. Legg-Calve-Perthes disease of left hip. Epiphysis is narrowed and radio-dense. A subchondral fracture is also visible.

Management of LCPD requires a pediatric orthopedist who will follow and treat the child through the various stages of the disease. Prompt referral may influence long-term prognosis. Older children, obese children, girls, and those with more severe disturbance of the epiphysis on radiographs have a poorer prognosis.

Discitis

Background

Discitis is an inflammatory condition involving the intervertebral disc space that has also been called acute osteitis of the spine, spondylitis, and spondylarthritis. The variety of diagnostic terms is an indication that the pathophysiology of this condition is poorly understood. Vertebral osteomyelitis with involvement of the disc space is a distinct diagnostic entity with different epidemiology and pathophysiology from discitis.

Discitis is a disease of childhood, with about 75% of patients being less than 10 years of age. No gender or racial predilection has been noted. The involved disc space is usually lumbar or lower thoracic. Most authorities believe discitis results from infection. A history of trauma is obtained in some patients with discitis, but whether the injury plays a role in cause or is a “red herring” is unclear. The vascular anatomy of the disc space supports the notion that organisms reach the disc space via the hematogenous route. In children, the blood supply of the disc space comes from adjacent vertebral body end plates. These vascular connections are absent in older adolescents and adults, which is consistent with the age distribution of discitis.

Bacteria are cultured from a minority of children with discitis. *S. aureus* is the predominant isolate from disc space aspirates and occasionally blood, but other organisms have also been recovered.

Diagnosis

Children with discitis are a diagnostic challenge for clinicians. Symptoms are often nonspecific and vague, especially in the younger child. They have usually been present for more than 1 week at the time of diagnosis. Back pain is not always described. Limp, refusal to walk, leg pain, hip pain, and abdominal pain are common presenting complaints. Low-grade fever and irritability may be reported.

Physical findings suggesting discitis will be missed if this entity is not considered, because careful examination of the spine is not performed routinely by most clinicians. Many children assume a recumbent position of comfort from which they do not want to be moved. Decreased range of motion of the spine and paravertebral muscle spasm are usually present. There is often a change in the lumbar lordosis, which may be decreased or increased. Tenderness to palpation of the disc space can usually be demonstrated. Range of motion of the hips is essentially normal, but inadvertent movement of the lumbar spine during hip examination may cause pain that is misinterpreted to suggest hip pathology. Straight leg raising may be limited by muscle spasm in the hamstrings. Neurologic assessment of the lower extremities is normal. Abnormalities in strength, sensation, and/or deep tendon reflexes suggest a spinal cord lesion, tumor, epidural abscess, or herniation of the disc (rare). Signs of discitis may vary, depending on the location of the inflamed disc. Patients with lesions of the upper spine may have meningismus.

Imaging studies can be useful in the diagnosis of discitis. Plain radiographs are initially normal. Intervertebral disc space narrowing develops after 2 to 3 weeks of illness (Fig. 123.8). Bone scan is the most sensitive imaging modality early in the course of this disease. Increased uptake at the level of the involved disc can confirm the diagnosis. MRI has also proven sensitive in the early phase. CT scanning can demonstrate the degree of bony erosion of the vertebral end plates and paravertebral soft-tissue involvement.

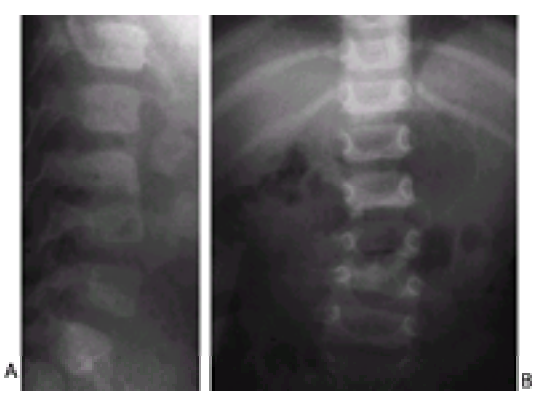

FIGURE 123.8. Discitis. L3–L4 intervertebral disc space is narrowed. Anteroposterior **(A)** and lateral **(B)** views.

Laboratory testing plays a minor role. Elevation of the WBC count is sometimes noted at the time of diagnosis. ESRs of 40 to 60 mm/hour are usually noted in patients presenting with discitis, and decrease with resolution of the disease. Skin testing for tuberculosis, and serologic testing for brucellosis and salmonellosis are often performed. Discitis can usually be diagnosed and treated without biopsy or aspiration of the involved disc space. If the presentation is atypical, signs and symptoms severe, or response to therapy unsatisfactory, obtaining a guided needle aspiration can be helpful.

Management

Discitis is a self-limited disease and need not be treated aggressively. Virtually all children in reported series return to normal function in a few months. Resting the spine usually results in improved symptoms in days to weeks. Immobilization with plaster has not been shown to improve outcome over bed rest alone, but therapeutic decisions should be individualized with input from an orthopedist. Although there are no data to suggest that they speed recovery or improve outcome, antistaphylococcal antibiotics seem prudent, given the frequency of documented staphylococcal infection. When cultures demonstrate particular organisms with known antimicrobial susceptibilities, antibiotic therapy can be individualized.

SPONDYLOLYSIS AND SPONDYLOLISTHESIS

Spondylolysis and spondylolisthesis occur in 2 to 5% of children, but most are asymptomatic. In older children with low back pain, especially adolescents, it is a condition that should be considered. Spondylolysis is a defect in the pars interarticularis of the vertebral body. Spondylolisthesis is displacement of the vertebral bodies, usually involving L5

slipping anteriorly on S1. Spondylolisthesis may result from structural abnormalities of the vertebral bodies (dysplastic type) or acquired defects of the pars interarticularis (isthmic type) that allow slippage. There is a genetic predisposition to spondylolysis and spondylolisthesis. Parents of children with spondylolisthesis are found to have this condition in 28% of cases.

The cause of the defect of the pars interarticularis in spondylolysis is not fully understood. Repeated stress, such as occurs in gymnasts with frequent hyperextension of the spine, causes stress fracture. One side of the pars interarticularis fractures overtly, which adds to the stress on the contralateral side. Fracture becomes bilateral. Displacement may or may not occur. Children who play sports that stress the spine, such as gymnastics, football, rowing, diving, weight lifting and high jumping, are at particular risk.

Patients who develop symptoms generally present during the adolescent growth spurt. Back pain worsens with activity and improves with rest, and usually has an insidious onset. Over time, there may be pain in the buttocks and posterior thighs. Symptoms radiating down the legs suggest significant nerve root irritation. Parents may describe an increase in the lumbar lordosis or a change in the child's gait.

Physical examination shows tenderness with hyperextension of the lumbar spine in the prone position, and with deep palpation. The hamstrings are usually tight, with decreased range of motion on straight leg raise and flexion of the trunk. Children seldom have motor (10%), sensory (15%), or reflex (10%) deficits in the legs.

Plain radiographs should include AP, lateral, and oblique views. The “scotty dog” of the oblique view will have a collar on the neck if spondylolysis is present. Spondylolisthesis can be diagnosed on the lateral view, and the degree of displacement can be quantitated relative to the width of the vertebral body (Fig. 123.9).

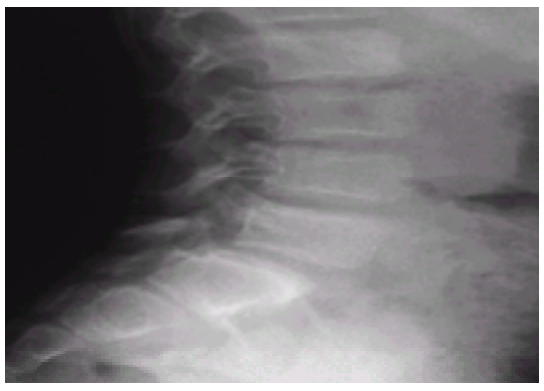

FIGURE 123.9. Spondylolisthesis with slippage of L5 anteriorly on S1.

Treatment varies depending on symptoms and degree of displacement, if any. Most cases of asymptomatic spondylolysis, and spondylolisthesis with mild displacement will not progress. Children with displacement greater than 25% should avoid rough sports. Symptomatic children with displacement may benefit from immobilization. Decisions about treatment should be made in consultation with an orthopedic surgeon.

OVERUSE SYNDROMES

Overuse syndromes is a general term that encompasses a variety of injuries that result from excessive and repetitive forces on susceptible structures. Children are at unique risk for such injuries, which are particularly common in adolescent athletes. There is an increased susceptibility during the growth spurt when skeletal growth exceeds the growth of the muscle–tendon unit. This results in increased stress at the apophysis, the musculotendinous origin or insertion. In children, cartilage is interposed between tendon and bone and is most prone to injury from repetitive forces. Repetitive tensile forces at these sites result in chronic irritation and microfractures or avulsions of the apophysis. If allowed to progress, there is evidence that the repetitive microtrauma may weaken the bone and predispose to major avulsion fractures. Traction apophysitis is unique to the growing child. By adulthood, fusion of tendon to bone leads to tendinitis rather than apophysitis as a result of repetitive forces. The late childhood, early teenage years coincide with increased participation in organized sporting activities. There is a tendency in high-intensity programs to over-train young athletes, and at times to wrongly encourage them to work through or ignore the early warning signs of pain.

General therapy for these injuries must emphasize several areas. Rest is crucial for the specific area involved until pain has completely resolved. The athlete should be actively encouraged to use alternative activities to maintain conditioning during this time. The role of inflammation in overuse injuries is controversial, but the application of ice and use of antiinflammatory agents is generally recommended. Directed stretching exercises are encouraged to reduce tension on affected areas. Biomechanics should be assessed and corrected when necessary. When returning to full activity, an appropriate training regimen should emphasize a slow gradual buildup in intensity and duration and should include explicit limitations. The sudden increase in intensity and duration of training that occurs with a change of sporting seasons is a major culprit in overuse injuries.

A number of overuse syndromes have acquired popular eponyms. Among the most common overuse syndromes in children are Osgood-Schlatter disease, Little Leaguer's elbow, and Sever's disease.

Little Leaguer's Elbow

Little Leaguer's elbow refers to a group of disorders resulting from repetitive valgus stress applied to the skeletally underdeveloped elbow. Its cause is a combination of excessively repetitive pitching and poor throwing biomechanics. Valgus force places tension on the medial collateral ligaments, which is translated to the medial epicondyle. A medial epicondylitis or apophysitis is the most commonly resulting lesion. An avulsion fracture of the medial epicondyle may result from an acute valgus force once the site has become weakened from repetitive microtrauma. As expected, Little Leaguer's elbow occurs most commonly in boys ages 9 to 12. Patients complain primarily of elbow pain that is exacerbated by throwing. Tenderness is localized over the medial elbow. Flexion of the wrist or finger against resistance also elicits pain. In advanced cases, extension of the elbow becomes limited.

Radiographs may reveal nonspecific changes such as an irregular or widened medial epicondylar physis, but in general, an apophysitis is not visible. An avulsion fracture may appear as a bony fragment separated from the medial epicondyle.

Treatment emphasizes rest for at least 1 month, application of ice, and return to activity only after all pain is gone. Once activity is resumed, the athlete must concentrate on limiting the total amount of pitching, as well as minimizing stress on the medial epicondyle by employing an overhand rather than side-arm pitching motion. Routine stretching and range of motion exercises will reduce the risk of recurrence. Displacement of an avulsion fragment may require surgical repair to restore full elbow function.

Osgood-Schlatter Disease

Osgood-Schlatter disease is an apophysitis of the tibial tubercle. Repetitive stress imposed by the patellar tendon on its site of insertion results in a series of micro-avulsions of the ossification center and underlying cartilage. The condition is most common in running and jumping athletes between the ages of 11 and 15. Girls are less commonly affected than boys. Most cases are bilateral, although symptoms are commonly asymmetric.

The physical examination is notable for localized tenderness at the tibial tubercle. Any action that applies tension to the patellar tendon elicits pain. Placing the patient prone and flexing the knee so that the heel contacts the buttocks typically triggers pain at the tibial tubercle. Additional maneuvers likely to cause pain include forced extension of the knee, jumping, or squatting. In advanced cases, callus formation occurs, resulting in further prominence of the tubercle. Some experts have suggested a relationship between Osgood-Schlatter disease and acute avulsion fractures of the tibial tubercle (Fig. 123.10). The diagnosis is based on the clinical features. Radiographs are not indicated in typical cases. Radiographic findings of soft-tissue swelling and irregularities of the tubercle are nonspecific.

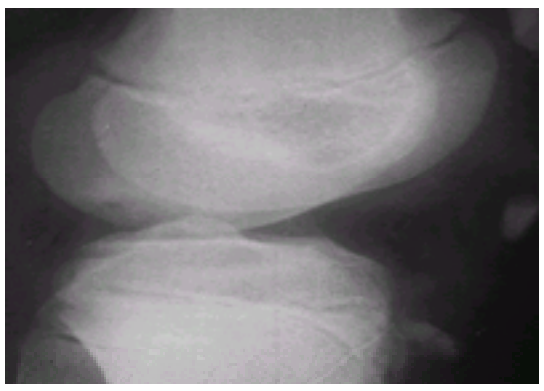

FIGURE 123.10. Acute tibial tubercle avulsion fracture in child with history of Osgood-Schlatter disease.

Management consists first and foremost of avoiding activities that place stress on the tibial tubercle. This is perhaps the most difficult instruction to enforce in young athletes. A brief period of immobilization or non–weight bearing is recommended by some as a means of ensuring compliance. Application of ice reduces pain and swelling. Non-steroidal anti-inflammatory medications are commonly recommended. Activity may be resumed when the patient is free of pain. Flexibility exercises concentrate on stretching the quadriceps and hamstrings to alleviate stress on the tubercle and avoid recurrences. A neoprene sleeve on the knee reduces patellar mobility and reduces forces on the tubercle.

Osteochondritis Dissecans

Background

Osteochondritis dissecans is a lesion involving separation of the osteochondral segment from underlying healthy bone. A variety of articular lesions are often lumped together under this term, including acute osteochondral fractures and epiphyseal dysplasias. This results in confounding descriptions of the natural course and outcome of the true condition. Adults are often diagnosed with osteochondritis dissecans; however, it remains primarily a condition of the adolescent age group, with the highest incidence occurring among male athletes between 12 and 16 years of age. The primary sites of osteochondritis dissecans include the medial femoral condyle in the knee, the posteromedial aspect of the talus in the ankle, and the capitellum in the elbow. It is less commonly reported in the hip, feet, and wrist. Involvement of multiple sites is rare.

Pathophysiology

The underlying cause of osteochondritis dissecans remains controversial and may differ based on the anatomic location

of the lesion. Trauma, vascular insult, genetic predisposition, and abnormalities of ossification have all been proposed as possible causes. The greatest evidence supports repetitive trauma as the sole or major contributory cause of the pathology. Overuse injury is most clearly associated with osteochondritis dissecans of the capitellum, where most cases come from Little League pitchers. Higher incidences of osteochondritis dissecans of the knee and ankle are seen in participants of activities that place increased stress on these areas, such as distance running, ballet, and basketball. Focal necrosis is suspected to follow the initial insult. Spontaneous resolution may occur at this point, or it may progress with the subchondral bone undergoing various degrees of separation from the underlying epiphysis. In advanced stages, complete separation of the osteochondral fragment results in a free floating body within the joint, which can disrupt normal mechanical function.

When the disease occurs in the second decade of life, long-term outcome is generally good. Progression to osteoarthritis or other degenerative joint diseases is rare. A worse prognosis is associated with a diagnosis after skeletal maturity, larger lesions, and complete separation of fragments.

Clinical Findings

The onset of symptoms occurs over several months. Joint pain and swelling typically occur after strenuous exercise and improve over several hours with rest. When a free body is present, patients describe intermittent, abrupt locking of the joint. Locking in the knee or elbow prevents full extension of the extremity. This is in contradistinction to buckling, stiffness, or pain with extended range of motion.

The physical examination of the joint is often normal. Occasionally, a small effusion may be detectable. Lesions in the medial femoral condyle may be directly palpated and pain elicited when the knee is held in 90 degrees of flexion. The typical location of lesions in the talus are not accessible on examination. Osteochondritis dissecans in the femoral condyle may give rise to an abnormal gait with external rotation of the affected limb.

Diagnosis

A plain film of the joint should be obtained, and is often diagnostic when osteochondritis dissecans is suspected (Fig. 123.11). Radiographs reveal a crescentic-shaped defect within the subchondral bone. The avascular segment of subchondral bone may have increased density. A radiolucent line may demarcate the separation from the remainder of the epiphysis. A free body often includes a portion of dead subchondral bone, which appears as a radio-dense object within the joint space. Standard AP and lateral views, as well as tunnel and sunrise views, of the knee are recommended for lesions within the femoral condyle. Lateral, AP, and mortis views of the ankle are adequate when a lesion of the talus is suspected, and AP and lateral views of the elbow are sufficient when the capitellum is involved.

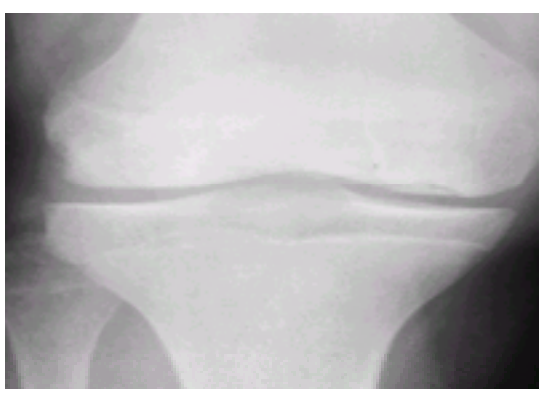

FIGURE 123.11. Osteochondritis dissecans of the medial femoral condyle. Crescentic lesion with radiolucent margin in a 14-year-old girl.

Early lesions may not be detected on plain films and alternate imaging modalities may improve overall sensitivity. When correlated with arthroscopic or surgical findings, MRI has been shown to have better ability to detect osteochondritis dissecans and accurately define the extent and stage of the lesion. MRI is increasingly used to guide therapy and monitor healing. Arthrograms have also been used to detect changes in the surface of the cartilage before bony changes occur, but these studies require intra-articular injection of dye and subject patients to additional radiation.

Management

The management of osteochondritis dissecans depends on the age and skeletal maturity of the patient, the location of the lesion, and the stage of the lesion. Conservative therapy consisting of restricted activity and relief of stress on the joint is the first-line treatment in children who have not reached skeletal maturity, and for those diagnosed at an early stage of the disease. Immobilization in a cast or non–weight bearing for lower extremity lesions is unnecessary. Patients should be followed closely by an orthopedic surgeon both for resolution of clinical symptoms and evidence of healing on serial radiographs or MRIs. Most stable lesions occurring in patients before physeal closure go on to heal; however, a few will progress to separation. Lesions occurring in adults generally do not heal without surgery.

Surgical intervention is generally recommended when lesions fail to improve clinically or radiographically after 6 months of rest. The presence of a loose or free body is also considered an indication for surgery. Many surgical procedures can now be performed arthroscopically. Fine drilling through the articular cartilage and subchondral bone into healthy bone appears to stimulate revascularization and promote healing. Loose fragments and larger free bodies may be reduced and fixed in place with the use of screws or Kirschner wires. Other free bodies need to be removed from the joint space.

Resulting defects may be repaired with the use of a bone graft or through stimulation of fibrocartilage or scar tissue formation to restore congruity to the articular surface.

Chondromalacia Patellae

Chondromalacia patellae is a pathologic diagnosis referring to damage of the articular cartilage of the patella. Specific changes include softening, fissures, and erosions. Patellofemoral pain syndrome, a term often used interchangeably with chondromalacia patellae, more accurately describes a constellation of symptoms, principally anterior knee pain arising from the patellofemoral joint. Whether the two conditions are actually related is the subject of debate. They share a number of symptoms and precipitating factors. Patellofemoral pain syndrome may represent the early end of the spectrum of injury, which ultimately may or may not progress to true pathologic changes within the cartilage.

Patellofemoral pain syndrome and chondromalacia patellae are first seen in early adolescents. The rise in incidence tends to parallel the growth spurt. A number of underlying causes or associated factors have been identified. Malalignment of the patella and an abnormal tracking of the patella over the femoral condyles appear to be the major contributors to patellofemoral disorders. The quadriceps or Q angle is the angle between a line from the center of the tibial tubercle to the center of the patella, and a second line from the center of the patella to the anterior superior iliac spine. A Q angle greater than 20 degrees has been found in a significant number of affected individuals and results in disproportionate lateral traction applied to the patella during extension. The wider pelvic bones in females result in a generally wider Q angle, which may account for the higher proportion of patellofemoral problems in females. Another contributing anatomic factor is a relative strength imbalance of the four muscles composing the quadriceps. A shallow femoral intra-condylar sulcus has also been associated with the disorder.

Chondromalacia patellae and patellofemoral pain syndrome are often classified as overuse syndromes because individuals exposed to repetitive trauma are at higher risk for these disorders. Runners are particularly predisposed to develop these conditions. Poor training regimens, rapid increases in duration or intensity of training, hard or uneven running surfaces, and inadequate shoes have been blamed.

Symptoms consist mainly of anterior knee pain often described as arising from beneath or on the sides of the patella. Pain is usually of gradual onset and is exacerbated by exercise. Activities that involve loading of the knee when it is in flexion, such as climbing steps, are particularly painful.

The physical examination is notable for tenderness along the patellar margins or the posterior surface, which is accessible when the patella is manually displaced medially or laterally. Pain, and occasionally crepitus, are elicited with flexion and extension of the knee, or tightening the quadriceps while compressing the patella against the femoral condyles. Range of motion is not limited and swelling is rare. The presence of an effusion is suggestive of significant cartilaginous damage. Provocative tests that reproduce the pain include climbing steps, squatting, or knee extension against resistance.

Radiographs are generally insensitive but may show changes to the patella in advanced cases. Radiographs may also be obtained to more accurately measure the intracondylar sulcus or Q angle, or to rule out alternative diagnoses. MRI, with a sensitivity greater than 80%, is considered the best noninvasive diagnostic modality for chondromalacia patellae. True confirmation of lesions requires arthroscopy.

Treatment is conservative. More than 90% of cases of patellofemoral pain syndrome resolve after instituting a program of rest, anti-inflammatory medications, and ice followed by physical therapy. Exercises that begin once the initial pain has resolved emphasize strengthening of the quadriceps muscles. Recommended exercise regimens include isometric contractions of the quadriceps with the knee in extension, straight leg raises, and knee extensions, first without and then with weights. Training routines for athletes may need modification and should emphasize soft, even running surfaces; proper biomechanics; and shoes with appropriate cushioning and support. Surgery is recommended only as a last resort in the most recalcitrant cases because results have been generally less than satisfactory. Surgery is directed at either correcting unequal tension applied to the patella or removing loose or nonviable cartilage from the posterior patellar surface.

Sever's Disease

Sever's disease is a calcaneal apophysitis occurring at the insertion of the Achilles' tendon at the posterior aspect of the calcaneus. It afflicts predominantly runners, jumpers, and soccer players. Sever's disease is often bilateral, is more common in males, and has its peak incidence between 10 and 12 years of age.

Localized tenderness occurs at the insertion of the Achilles' tendon on the calcaneus. A maneuver such as hanging the heels over the edge of a step, climbing steps, or hopping applies tension to the Achilles' tendon and exacerbates the pain. Patients are often found to have a tight gastrocnemius-soleus muscle complex and limited dorsiflexion of the foot. Radiographs of the site are usually normal and are unhelpful except to exclude bony injuries such as stress fractures.

Management includes rest, ice, and anti-inflammatory medications. Heel padding or lifts may be helpful in relieving tension in the area. Flexibility exercises should concentrate on both the hamstrings and the calf muscles. When therapy is initiated early in the disease, most patients are able to return to normal activity by 2 months.

Bursitis

Bursa sacs are both the shock absorbers and the ball bearings of the musculoskeletal system. They disperse forces from blows on bony prominences and reduce friction where tendons or ligaments are in frequent motion.

Trauma, either in a single blow or by repetitive forces, can inflame the bursa, which responds with increased production

of synovial fluid. The bursa sac subsequently swells and a cycle of swelling, irritation, and inflammation ensues. Bursitis is most commonly an overuse syndrome seen in adolescents and adults, and is less common in young children.

Injury or cellulitis of the skin overlying a bursa sac can predispose to infection. Aspiration and culture are necessary for definitive diagnosis. The organisms found in septic bursitis are the same as those in septic arthritis, with *S. aureus* accounting for more than 90% of cases. There is no consensus on the need for parenteral versus oral antibiotics. The prepatellar bursa and olecranon bursa are most commonly infected.

Bursae are located throughout the body, but bursitis occurs only in a few. Prepatellar bursitis, commonly called “housemaid's knee” results from frequent or prolonged kneeling. Pens anserinus bursitis occurs on the lateral aspect of the knee, where the tendons of the hamstring muscles overlie the tibia. Retrocalcaneal bursitis occurs between the calcaneus and Achilles' tendon and often is caused by direct pressure from ill-fitting footwear or high-heeled shoes. Olecranon bursitis most often results from a single direct blow to the elbow. Shoulder or subacromial bursitis is often associated with calcifications and produces severe pain with abduction. Other commonly affected bursae include the inferior calcaneal bursa and the trochanteric bursa.

An unusual form of bursitis is known as a popliteal or Baker's cyst. This occurs in the bursa, which cushions the tendons of the gastrocnemius and semimembranous muscles from the distal femur. The presence of this condition in adults is highly suggestive of intra-articular knee damage. In children with a Baker's cyst, there is often a congenitally wide opening joining the bursa sac with the knee joint itself. One-way flow of synovial fluid into the bursa produces swelling just below the popliteal fossa on the medial side. Patients with chronic inflammatory conditions of the knee, such as juvenile rheumatoid arthritis (JRA), are at increased risk of developing popliteal cysts. The swelling limits full flexion of the knee and produces the sensation of tension with extension. An arthrogram or bursagram may outline the cyst, document the articular connection, and detect ruptures of the cyst. Ultrasound is a useful noninvasive diagnostic modality. MRI is more accurate than ultrasound, but not as essential in children given the lower incidence of accompanying intra-articular injury.

Bursa inflammation produces swelling and localized pain with direct palpation. Any movement of the tendons overlying the site will reproduce the pain.

Conservative therapy consisting of restricted activity, frequent application of ice, and regular use of non-steroidal anti-inflammatory medications is successful in most cases. Resistant cases respond well to aspiration of synovial fluid and injection of corticosteroids. Often, recurrent cases may require surgical removal of the bursa sac. A new bursa will be generated.

COMPARTMENT SYNDROME

Compartment syndrome refers to vascular insufficiency caused by elevated tissue pressures. It usually occurs after an injury causes hemorrhage or edema within an enclosed fascial compartment. Tight circumferential bandages or casts can also limit expansion of swollen tissues and result in elevation of tissue pressures. Fluid extravasation from intravenous or intraosseous lines, especially pressure-driven extravasation, may significantly elevate compartment pressures. Direct injury to a vessel is less common as the cause of vascular insufficiency after injury. When compartment pressures approach the perfusion pressure of muscle, which is approximately 30 mm Hg, arterial inflow is reduced and veins and capillaries are collapsed. Ischemia of muscle leads to further swelling, and an ischemia–edema cycle can lead to complete cessation of tissue perfusion. Muscle necrosis is irreversible after 6 to 8 hours of tissue anoxia. Fibrosis develops and ischemic contracture results in permanent disability. The emergency physician must identify patients at risk for compartment syndromes and consult with an orthopedist, who can monitor tissue pressures and treat compartment syndromes before irreversible injuries occur.

Knowledge of the common pediatric injuries that are associated with compartment syndromes can raise the clinician's index of suspicion appropriately (Table 123.3). Displaced supracondylar fractures may lead to Volkmann's contracture, which involves the distribution of the anterior interosseous artery and the flexor compartment of the forearm. Forearm fractures may also cause compartment syndromes, affecting either the flexor or extensor musculature. Fractures of the tibia and/or fibula can lead to compartment syndrome of the lower leg. Compartment syndromes may occur from crush injuries and other soft-tissue trauma that does not necessarily involve a fracture.

Fractures	Ischemic Compartments
Supracondylar fractures of the humerus (displaced)	Deep flexor (anterior) compartment
Radius/Ulna fractures (diaphyseal)	Anterior tibial and peroneal compartments
Tibia/fibula fractures (diaphyseal)	
Femur fractures	

Table 123.3. Compartment Syndromes

Compartment syndromes are diagnosed clinically by assessing the “Five Ps”: pain, paresthesia, pallor, paralysis, and

pulselessness. All five need not be present for a compartment syndrome to exist.

Pain is the hallmark of compartment syndromes, but it can be difficult to distinguish the pain of the injury itself from that related to the vascular insufficiency. Pain that increases over time after the injury suggests muscle ischemia. Compartment syndrome must be suspected when the pain seems out of proportion to the injury itself. Stretching ischemic muscles exacerbates the pain. Increased pain with extension of the fingers or toes is an important clinical finding in patients with compartment syndromes.

Paresthesia may be noted in the distribution of the nerves that traverse the ischemic compartment. When the flexor compartment of the forearm is involved, the median nerve is usually affected. Over time, paresthesias may progress to complete anesthesia, and pain may decrease.

Pallor from decreased perfusion may be noted distally. Sluggish circulation may cause cyanosis. Paralysis is a late finding and is probably the least sensitive marker for compartment syndrome. Pulselessness is a useful finding if present, but some physicians are falsely reassured when distal pulses are palpable. The ischemia in compartment syndromes results from vascular occlusion of small vessels. Pressures seldom exceed systolic blood pressure. Larger arteries may not be occluded, and pulses often remain intact.

Treatment of a compartment syndrome should begin when it is suspected. All circumferential bandages should be removed. If symptoms persist, orthopedic consultation should be obtained for measurement of compartment pressures. Reduction of displaced fractures can improve blood flow to affected compartments. Fasciotomy in the operating room is indicated if compartment pressures remain high.

REFLEX SYMPATHETIC DYSTROPHY

Reflex sympathetic dystrophy (RSD) is a poorly understood disorder characterized by pain, abnormal sensation, and circulatory irregularities. Over time, atrophic changes of the extremity develop. *Causalgia, algodystrophy,* and *Sudeck's atrophy* are also terms that have been used for this mysterious disorder, first reported in gunshot victims during the American Civil War. The average time from onset to diagnosis of RSD in children is 1 year. Emergency physicians play an important role in the early diagnosis and treatment of RSD, which may prevent prolonged disability.

RSD is well known in adults, but children with RSD as young as 3 years have been described. The average age of children with RSD is approximately 12 years, girls outnumbering boys by as much as 6:1. Most cases in children involve the lower extremity. RSD usually follows minor trauma, but some cases develop without an identified precipitant.

The pathophysiology of RSD is not understood. Early theories suggested abnormal synapses develop between sensory afferent nerves and sympathetic efferents after an injury. “Sympathetic” dystrophy may be a misnomer, however, because local epinephrine and norepinephrine levels are lower, not higher than normal, and vasodilation, not sympathetic vasoconstriction, may predominate. Theories of sympathetic receptor hypersensitivity or central, self-exciting pathways in the substantia nigra remain unproven.

Pain is usually the presenting complaint with RSD. The pain is continuous, often burning in quality, with exacerbations but no complete remissions. Abnormal sensitivity is distinctive, with severe pain provoked by normally non-tender touching (allodynia). The extremity is usually swollen and cool to the touch, although warmth has also been reported. Dusky discoloration of the skin with hyperhidrosis or anhydrosis may be present. The arm or leg is not used, and atrophic muscle, skin, and bony changes develop in some patients over time. There is some evidence that demineralization of bone occurs more rapidly than would be expected from disuse alone.

Psychiatric and personality problems have been suspected in many patients with RSD, but controlled prospective studies are lacking. Factitious illness or conversion reactions may be considered, given that symptoms are out of proportion to the inciting injury.

The characteristic history and physical examination, including pain, loss of function, and evidence of autonomic dysfunction, allow for a clinical diagnosis of RSD in most cases. Radiographs in children may not demonstrate the osteoporosis described in adults, especially early after the onset of symptoms. Radionuclide bone scans generally show increased blood flow and periarticular uptake in adults, but in children with RSD the blood flow and osseous uptake is more often reduced. Thermography may document decreased temperature in the affected extremity. Treatment of RSD focuses on early mobilization of the extremity through physical therapy to avoid atrophic changes. Physiotherapy may initially exacerbate symptoms, but experienced clinicians believe it both prevents atrophy and decreases the duration of pain. The knee-jerk response to splint for comfort may be counterproductive with RSD. Referral to a pediatric pain program is advisable should symptoms persist. Intravenous regional block with guanethidine, sympathetic block, transcutaneous nerve stimulation, and, with intractable cases, sympathectomy have all been performed, reportedly with some success.

Suggested Readings

OSTEOMYELITIS

Dich VQ, Nelson JD, Haltalin KC. Osteomyelitis in infants and children. Am J Dis Child 1975;129:1273–1278.

Faden H, Grossi M. Acute osteomyelitis in children. Am J Dis Child 1991;145:65–69.

Fox L, Sprint K. Neonatal osteomyelitis. Pediatrics 1978;62(4):535–542.

Gold R. Diagnosis of osteomyelitis. Pediatr Rev 1991;12:292–297.

Howard CB, Einhorn M, Dagan R, et al. Fine-needle bone biopsy to diagnose osteomyelitis. J Bone Joint Surg 1994;76-B:311–314.

LaMont RL, Anderson PA, Dajani AS, et al. Acute hematogenous osteomyelitis in children. J Pediatr Orthop 1987;7(5):579–583.

Rosovsky M, FitzPatrick M, Goldfarb CR, et al. Bilateral osteomyelitis due to intraosseous infusion: case report and review of the English-language literature. Pediatr Radiol 1994;24:72–73.

Sonnen GM, Henry NK. Pediatric bone and joint infections. Pediatr Clin North Am 1996;43(4):933–947.

Tuson CE, Hoffman EB, Mann MD. Isotope bone scanning for acute osteomyelitis and septic arthritis in children. J Bone Joint Surg 1994;76-B:306–310.

Unkila-Kallio L, Kallio MJT, Eskola J, et al. Serum C-reactive protein, erythrocyte sedimentation rate, and white blood cell count in acute hematogenous osteomyelitis of children. Pediatrics 1994;93:59–62.

Weissberg ED, Smith AL, Smith DH. Clinical features of neonatal osteomyelitis. Pediatrics 1974;53:505–510.

SEPTIC ARTHRITIS

Adams WG, Deaver KA, Cochi SL, et al. Decline of childhood *Haemophilus influenzae* type b (Hib) disease in the Hib vaccine era. JAMA 1993;269(2):221–226.

Barton LL, Dunkle LM, Habib FH. Septic arthritis in childhood. Am J Dis Child 1987;141:898–900.

Canale ST, Harkness RM, Thomas PA, et al. Does aspiration of bones and joints affect results of later bone scanning? J Pediatr Orthop 1985;5(1):23–26.

DelBeccaro MA, Champoux AN, Bockers T, et al. Septic arthritis versus transient synovitis of the hip: the value of screening laboratory tests. Ann Emerg Med 1992;21:1418–1422.

Fisher MC, Goldsmith JF, Gilligan PH. Sneakers as a source of *Pseudomonas aeruginosa* in children with osteomyelitis following puncture wounds. J Pediatr 1985;106(4):607–609.

Jacobs RF, McCarthy RE, Elser JM. *Pseudomonas* osteochondritis complicating puncture wounds of the foot in children: a 10-year evaluation. J Infect Dis 1989;160(4):657–661.

SEPTIC ARTHRITIS

Kallio MJT, Unkila-Kallio L, Aalto K, et al. Serum C-reactive protein, erythrocyte sedimentation rate and white blood cell count in septic arthritis of children. Pediatr Infect Dis J 1997;16(4):411–413.

Kunnamo I, Kallio P, Pelkonen P, et al. Clinical signs and laboratory tests in the differential diagnosis of arthritis in children. Am J Dis Child 1987;141:34–40.

Mandell GA. Imaging in the diagnosis of musculoskeletal infections in children. Curr Probl Pediatr 1996;26:218–237.

McKillop JH, Maharaj D, Boyce BF, et al. Bone scan appearances following biopsy of bone and bone marrow. Radiology 1984;153:241–242.

Miralles M, Gonzalez G, Pulpiero JR, et al. Sonography of the painful hip in children: 500 consecutive cases. Am J Rheumatol 1989;152:579–582.

Nelson JD, Koontz WC. Septic arthritis in infants and children: a review of 117 cases. Pediatrics 1966;38(6):966–971.

Sundberg SB, Savage JP, Foster BK. Technetium phosphate bone scan in the diagnosis of septic arthritis in childhood. J Pediatr Orthop 1989; 9:579–585.

Welkon CJ, Long SS, Fisher MC, et al. Pyogenic arthritis in infants and children: a review of 95 cases. Pediatr Infect Dis 1986;5:669–676.

Yagupsky P, Bar-Ziv Y, Howard CB, et al. Epidemiology, etiology, and clinical features of septic arthritis in children younger than 24 months. Arch Pediatr Adolesc Med 1995;149:537.

Zawin JK, Hoffer FA, Rand FF, et al. Joint effusion in children with an irritable hip: US diagnosis and aspiration. Radiology 1993;187:459–463.

TOXIC SYNOVITIS

Del Becarro MA, Champoux AN, Bockers T, et al. Septic arthritis versus transient synovitis of the hip: the value of screening laboratory tests. Ann Emerg Med 1992;21:1418–1422.

Fink AM, Berman L, Edwards D, et al. The irritable hip: immediate ultrasound guided aspiration and prevention of hospital admission. Arch Dis Child 1995;72:110–114.

Kunnamo I, Kallio P, Pelkonen P, et al. Clinical signs and laboratory tests in the differential diagnosis of arthritis in children. Am J Dis Child 1987;141:34–40.

Marchal GJ, Van Holsbeeck MT, Raes M, et al. Transient synovitis of the hip in children: role of US. Radiology 1987;162:825–828.

Miralles M, Gonzalez G, Pulpeiro JR, et al. Sonography of the painful hip in children: 500 consecutive cases. Am J Rheumatol 1989;152:579–582.

Mukamel M, Litmanovitch M, Yosipovich Z, et al. Legg-Calve-Perthes disease following transient synovitis. Clin Pediatr 1985;24:629–631.

Taylor GR, Clarke NM. Management of irritable hip: a review of hospital admission policy. Arch Dis Child 1994;71:59–63.

Zawin JK, Hoffer FA, Rand FF, et al. Joint effusion in children with an irritable hip: US diagnosis and aspiration. Radiology 1993;187:459–463.

PENETRATING INTRA-ARTICULAR INJURIES

Collins DN, Temple SD. Open joint injuries: classification and treatment. Clin Orthop 1988;243:48–56.

Goodnough CP, Frymoyer JW. Synovitis secondary to non-metallic foreign bodies. J Trauma 1975;15(11):960–965.

Patzakis MJ, Dorr LD, Ivler D, et al. The early management of open joint injuries. J Bone Joint Surg 1975;57-A(8):1065–1071.

Voit GA, Irvine G, Beals RK. Saline load test for penetration of periarticular lacerations. J Bone Joint Surg 1996;78-B:732–733.

RADIAL HEAD SUBLUXATION

Illingsworth CM. Pulled elbow: a study of 100 patients. Br Med J 1975; 2:672–674.

Quan L, Marcuse EK. The epidemiology and treatment of radial head subluxation. Am J Dis Child 1985;139:1194.

Schunk JE. Radial head subluxation. Epidemiology and treatment of episodes. Ann Emerg Med 1990;19:1019.

SLIPPED CAPITAL FEMORAL EPIPHYSIS

Aronsson DD, Loder RT. Treatment of the unstable (acute) slipped capital femoral epiphysis. Clin Orthopaed Related Research 1996;(322):99–110.

Causey AL, Smith ER, Donaldson JJ, et al. Missed slipped capital femoral epiphysis: illustrative cases and a review. J Emerg Med 1995;13(2):175–189.

Hagglund G. The contralateral hip in slipped capital femoral epiphysis. J Pediatr Orthopaed 1996;5(3):158–161.

Koop S, Quanbeck D. Three common causes of childhood hip pain. Pediatr Clin North Am 1996;43(5):1053–1066.

Loder RT, Hensinger RN. Slipped capital femoral epiphysis associated with renal failure osteodystrophy. J Pediatr Orthoped 1997;17(2):205–211.

Loder RT. Slipped capital femoral epiphysis in children. Curr Opin Pediatr 1995;7(1):95–97.

LEGG-CALVE-PERTHES DISEASE

Skaggs DL, Tolo VT. Legg-Calve-Perthes disease. J Am Acad Orthop Surg 1996;4:9–16.

Stulberg SD, Cooperman DR, Wallensten R. The natural history of Legg-Calve-Perthes' disease. J Bone Joint Surg 1981;63-A:1095–1108.

DISCITIS

King HA. Back pain in children. Pediatr Clin North Am 1984;31(5): 1083–1095.

Lascari AD, Graham MH, MacQueen JC. Intervertebral disk infection in children. J Pediatr 1967;70(5):751–757.

Moskal MJ, Villar LA. Childhood diskitis: report of 2 cases and review of the literature. J Am Orthoped Assoc 1986;86(3):169–174.

Rocco HD, Eyring EJ. Intervertebral disk infections in children. Am J Dis Child 1972;123:448–451.

SPONDYLOLYSIS AND SPONDYLOLISTHESIS

Friberg O. Instability in spondylolisthesis. Orthopedics 1991;14(4):463–465.

Stinson JT. Spondylolysis and spondylolisthesis in the athlete. Clin Sports Med 1993;12(3):517–528.

OVERUSE SYNDROMES

Clain MR, Hershman EB. Overuse injuries in children and adolescents. Phys Sports Med 1989;17(9):111–123.

Dyment PG, ed. Sports Medicine. Health Care for Young Athletes. Elk Grove Village, IL: American Academy of Pediatrics, 1991.

Gill TJ, Micheli LJ. The immature athlete: common injuries and overuse syndromes of the elbow and wrist. Clin Sports Med 1996;15(2):401–423.

Micheli LJ. Overuse injuries in children's sports: the growth factor. Orthoped Clin North Am 1983;14(2):337–360.

Micheli LJ, Ireland ML. Prevention and management of calcaneal apophysitis in children: an overuse syndrome. J Pediatr Orthoped 1987;7:34–38.

Ogden JA, Tross RB, Murphy MJ. Fractures of the tibial tuberosity in adolescents. J Bone Joint Surg 1980;62-A(2):205–215.

Ogden JA, Southwick WA. Osgood-Schlatter disease and tibial tuberosity development. Clin Orthoped Rel Res 1976;116:180–189.

Saperstein AL, Nicholas SJ. Pediatric and adolescent sports medicine. Pediatr Clin North Am 1996;43:1013–1033.

OSTEOCHONDRITIS DISSECANS

Bradley J, Dandy DJ. Osteochondritis dissecans and other lesions of the femoral condyles. J Bone Joint Surg 1989;71-B:518–22.

DeSmet AA, Fisher Dr, Graf BK, et al. Osteochondritis dissecans of the knee: value of MR imaging in determining lesion stability and the presence of articular cartilage defects. Am J Rheumatol 1990;155:549–553.

Schenck RC Jr, Goodnight JM. Osteochondritis dissecans. J Bone Joint Surg 1996;78-A:439–456.

Twyman RS, Desai K, Aichroth PM. Osteochondritis of the knee: a long-term study. J Bone Joint Surg 1991;73-B:461–464.

Wood JB, Klassen RA, Peterson HA. Osteochondritis dissecans of the femoral head in children and adolescents: a report of 17 cases. J Pediatr Orthop 1995;15:313–316.

CHONDROMALACIA PATELLAE

Davidson K: Patellofemoral pain syndrome. Am Fam Phys 1993;48(7): 1254–1262.

Dyment PG, ed. Sports Medicine. Health Care for Young Athletes. Elk Grove Village, IL: American Academy of Pediatrics, 1991.

Insall J, Falvo KA, Wise DW. Chondromalacia patellae: a prospective study. J Bone Joint Surg 1976;58A(1):1–8.

Tria AJ, Palumbo RC, Alicea JA. Conservative care for patellofemoral pain. Orthoped Clin North Am 1992;23(4):522–526.

BURSITIS

Lang IM. MRI Appearance of Popliteal Cysts in childhood. Pediatr Radiol 1997;27(2):130–132.

Paisley JW. Septic bursitis in childhood. J Pediatr Orthoped 1982;2(1):57–61.

Raddatz DA, Hoffman GS, Franck WA. Septic bursitis: presentation, treatment, and prognosis. J Rheumatol 1987;14(6):1160–1163.

Southmayd W, Hoffman M. Sports Health: The Complete Book of Athletic Injuries. New York: Quick Fox, 1981.

Szer IS, Klein-Gitelman M, DeNardo BA, et al. Ultrasonography in the study of prevalence and clinical evolution of popliteal cysts in children with knee effusions. J Rheumatol 1992;19(3):458–462.

COMPARTMENT SYNDROME

Paletta CE, Dehghan K. Compartment syndrome in children. Ann Plast Surg 1994;32(2):141–144.

Sharrard WJW. Paediatric Orthopaedics and Fractures. London: Blackwell Scientific, 1993.

Tachdjian MO. Pediatric Orthopedics. Philadelphia: WB Saunders, 1990.

REFLEX SYMPATHETIC DYSTROPHY

Lloyd-Thomas AR, Lauder G. Reflex sympathetic dystrophy in children. Br Med J 1995;310:1648–1649.

Silber TJ, Majd M. Reflex sympathetic dystrophy syndrome in children and adolescents. Am J Dis Child 1988;142:1325–1330.

Stanton RP, Malcolm JR, Wesdock KA, et al. Reflex sympathetic dystrophy in children: an orthopedic perspective. Orthopedics 1993;16(7):773–780.

Wilder RT, Berde CB, Wolohan M, et al. Reflex sympathetic dystrophy in children. Clinical characteristics and follow-up of seventy patients. J Bone Joint Surg 1992;74(6):910–919.

CHAPTER 124

Dental Emergencies

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Nontraumatic orofacial emergencies can appear suddenly and can be frightening for pediatric patients and their families. The major task in evaluating a child with a nontraumatic orofacial emergency is to identify the cause of the problem. In cases of facial swellings, the first step in treatment is determining that a tooth is the causative agent. In cases of postextraction complications, historical information suggesting a preextraction infection, fractured tooth, or overlying chronic systemic problem may be elicited. Therefore, initial assessments must be performed in the same manner as traumatic orofacial emergencies (see Chapter 113).

POSTEXTRACTION COMPLICATIONS

Hemorrhage

It is expected that any extraction site may ooze for 8 to 12 hours and perhaps longer for a permanent site. However, it is important to check the history for any prior bleeding episodes to rule out a systemic hematologic abnormality. A complete blood count and coagulation profile would be indicated.

Emergency treatment may include the following steps:

1. Apply pressure, using folded gauze sponges that are placed over the socket with biting pressure applied for 30 minutes. If unsuccessful, proceed to the second step.
2. Physically close the socket by suturing. Administer local anesthesia (2% Xylocaine with 1:100,000 epinephrine infiltration), approximate the extraction site with the appropriate sutures. Alternatively, the socket may be packed with Gelfoam.

A possible home remedy before coming to the emergency department (ED) might include the use of a tea bag. A tea bag is dipped in hot water and allowed to cool, then placed over the socket with pressure. The tannic acid in the tea bag may accelerate or initiate coagulation.

Infection

Postextraction infection is rare in children. If it occurs, it may present as localized swelling or edema surrounded by an erythematous zone. A purulent exudate may be evident from the socket. Emergency treatment includes the application of moist heat, oral saline rinses (if the age is appropriate), and antibiotic therapy. Penicillin remains the drug of choice. (See the section on “Dentoalveolar Abscess” for dose and duration.)

Alveolar Osteitis

Alveolar osteitis, or “dry socket,” is a painful postoperative condition produced by a disintegration of the clot in the tooth socket. This condition usually is seen in adults and only rarely in children less than 12 years of age. It usually follows

(approximately 72 hours) mandibular extractions and is painful. Emergency dental treatment is variable, but the immediate goal is relief of pain. Under local anesthesia, the socket may be debrided and then packed with ¼-inch iodoform gauze or Bipp's paste (bismuth, iodoform, benzocaine, and petrolatum). Oral analgesic medication should be prescribed along with antibiotics.

ORAL INFECTIONS

In a retrospective analysis of pediatric dental patients presenting to the ED and dental clinic at The Children's Hospital, Boston, from 1989 to 1990, toothaches, pain, and facial swellings accounted for 44% of the chief complaints. This is consistent with other studies. It is important to remember that the infant or small child who may be in pain often cannot localize the discomfort. It may be the first opportunity for many children to receive dental care. A complete history from the parents and a thorough oral examination are mandatory. Figure 124.1 is a diagram of the normal tooth.

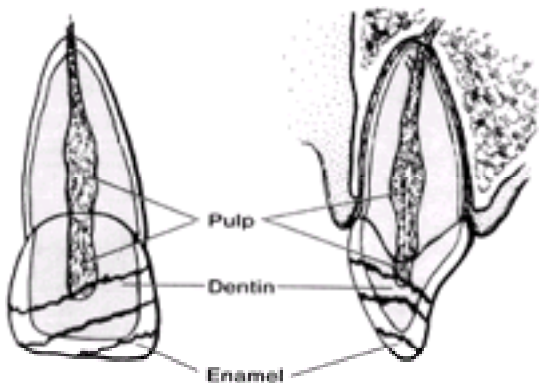


FIGURE 124.1. The anatomy of a tooth that should be considered during a traumatic injury. Enamel fracture, no emergency treatment; dentin fracture, emergency treatment as soon as convenient; and pulpal fracture, emergency treatment as soon as possible.

Odontalgia—Simple Toothache

The child with a simple toothache often complains of diffuse mouth pain and may not be able to point to a specific tooth. The emergency physician may note a grossly carious tooth or large restoration. Swelling or inflammation in the surrounding soft tissue may be present. The tooth may be sensitive to percussion and may exhibit excessive mobility. A dental consultation is necessary, especially if swelling is noted. In the case of swelling, the tooth may be opened for drainage to relieve the pressure, in a manner similar to the management of any abscess.

Dentoalveolar Abscess

Dental abscesses are common in children because of the morphologic characteristic of the primary tooth and immature permanent tooth. In the dentoalveolar abscess, the causative factors are gross or recurrent decay, trauma, or perhaps, chronic irritation from a large restoration. Suppuration is usually confined to the bone around the tooth. If the infection is long-standing, it can perforate the thin buccal bony plate adjacent to the root of the involved tooth and spread into the subperiosteal area and then to the surrounding soft tissues. In a child, the dentoalveolar abscess usually perforates the buccal plate of bone because of the position of the tooth and the thinness of the overlying bone. If it does not drain intraorally, the infection can spread rapidly through the fascial planes of the face or neck.

The following are clinical manifestations of a dentoalveolar abscess in a child:

1. Pain: The tooth may be painful to percussion, or may exhibit spontaneous painful episodes.
2. Mobility: The tooth may have a greater than normal degree of movement in the socket when palpated.
3. Swelling: The soft tissues surrounding the tooth may be edematous and erythematous.
4. Temperature elevation: The child may be febrile (greater than 37.5°C [99.5°F]), have general malaise, and a decrease in appetite.
5. Fistulous tracts: These appear clinically as a pustule-like lesion on the gingiva (rarely on the face) when the infection has been present for a long time.
6. Extrusion: The tooth may become extruded because of the presence of fluid in the periradicular space.
7. Lymphadenopathy: Major lymph node enlargement can occur at any time during the infective process.

The first step in treating a perioral abscess is to determine whether a tooth is the causative agent. This can be accomplished by clinical examination and available radiographs. Corroboration of dental origin can be established by reviewing intraoral radiographs. The location and extent of any swelling and/or fistulous tracts, both intraorally or extraorally, should be noted.

It is important that the treatment of choice for a localized dentoalveolar abscess is local in its focus (e.g., drainage, moist heat). In cases of facial cellulitis with lymphadenopathy caused by acute dentoalveolar abscess, the antibiotic of choice is penicillin (or erythromycin, if there is a known allergy to penicillin). The initial dose for children who weigh more than 60 pounds (27 kg) is 1 g orally, followed by 500 mg every 6 hours until the patient can be seen by a dentist. For children who weigh less than 60 pounds (27 kg), the initial penicillin dose is 500 mg, followed by 250 mg every 6 hours.

Penicillin-sensitive streptococci and anaerobic organisms predominate as the cause of acute dentoalveolar abscesses. If

there is a facial cellulitis over the maxilla extending toward the inferior border of the orbital rim or if there is a mandibular cellulitis, which might be a potential cause of airway compromise, the child should be admitted to the hospital where intravenous antibiotic therapy can be managed. Treatment for facial cellulitis is covered in Chapter 84.

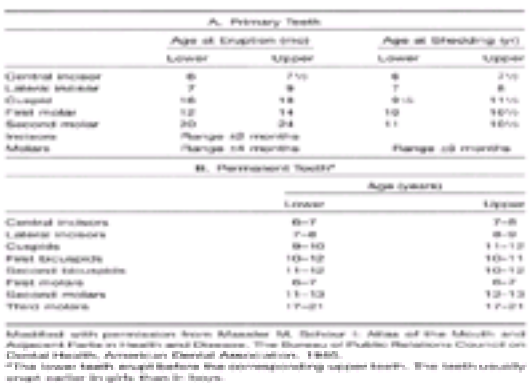
Other factors to consider in determining the need for hospital admission include the child's ability to take fluids and the likelihood of the parent's cooperation for follow-up dental care. Obviously, if the child is toxic, a hospital admission is indicated. In addition to antibiotics, warm oral saline rinses should be used and heat should be applied extraorally. There is some feeling that extraoral heat will cause the abscess to point extraorally and thus produce an exterior fistula. This has not proven true in our experience. Mild analgesic therapy such as acetaminophen is usually sufficient. Dental consultation should be obtained in order to vent the offending tooth, to establish drainage, to incise a fluctuant mass, or to remove the tooth.

As with infection elsewhere in the body, the basic surgical principles of treatment must be used: 1) establish drainage and 2) remove the cause. An abscessed primary tooth must be vigorously treated because such infections can affect the developing unerupted permanent tooth bud. A facial cellulitis can have severe systemic consequences, including cavernous sinus thrombosis, airway obstruction, brain abscess, and septicemia.

In some cases, there may be a need for additional consultation with infectious disease experts, especially in a situation in which systemic disorders render the child more susceptible to infection.

Pericoronitis

Pericoronitis is a localized infection surrounding an erupting tooth. It is usually associated with erupting molars in the adolescent patient, although a mild form may be associated with the eruption of the first permanent molar at age 6 (Table 124.1). Symptoms usually include pain distal to the last erupted tooth in the dental arch, along with erythema and edema localized to the gingiva in the retromolar area. Lymphadenopathy, trismus, and dysphagia may accompany these symptoms. An elevated body temperature is an occasional finding. It is not unusual to see or palpate the cusps of the erupting tooth. The patient may complain of an inability to completely close his or her mouth, as a result of the edematous gingiva. Otalgia is an uncommon complaint.



A. Primary Teeth

	Age at Eruption (mo)		Age at Shedding (yr)	
	Lower	Upper	Lower	Upper
Central incisor	6	7½	6	7½
Lateral incisor	7	9	7	8
Cuspid	16	18	9½	11½
First molar	12	14	10	10½
Second molar	20	24	11	10½
Incisors	Range 6–9 months			
Molars	Range 12 months		Range 2 months	

B. Permanent Teeth*

	Age (years)	
	Lower	Upper
Central incisors	6–7	7–8
Lateral incisors	7–8	8–9
Cuspids	9–10	11–12
First bicuspids	10–12	10–11
Second bicuspids	11–12	10–12
First molars	6–7	6–7
Second molars	11–13	12–13
Third molars	17–21	17–21

Modified with permission from Wheeler M, Schour I: Atlas of the Mouth and Adjacent Parts in Health and Disease. The Bureau of Public Relations Council on Dental Health, American Dental Association, 1960.

*The lower teeth erupt before the corresponding upper teeth. The teeth usually erupt earlier in girls than in boys.

Table 124.1. Eruption Schedule for Specific Teeth

Emergency treatment includes local curettage, oral rinses, heat, and scrupulous oral hygiene. Penicillin may be necessary (for dose, see “Dentoalveolar Abscess”) when there are systemic symptoms or facial swelling. Antibiotics should be continued until the tooth has erupted or treatment is completed.

Primary Herpetic Gingivostomatitis or Herpes Simplex Virus Type 1

Primary herpetic gingivostomatitis, or herpes simplex virus type 1 is a communicable childhood disease that is not a true dental emergency, but is a common cause of ED visits. The child is usually an infant or toddler who stops eating, drinking, or talking and is extremely irritable. The child usually has had an elevated temperature for 3 to 5 days before any clinical oral findings. A higher incidence of primary herpes has been noted after other viral illnesses. Older children may complain of headaches, malaise, nausea, regional lymphadenopathy, and/or bleeding gums. The physical examination reveals fiery red marginal gingiva with areas of spontaneous hemorrhage. Within 1 or 2 days, yellowish, fluid-filled vesicles develop on the mucosa, palate, or tongue and coalesce. The vesicles rupture spontaneously, leaving extremely painful ulcers, covered by a yellow or gray membrane and surrounded by an erythematous zone. Ulcers, especially on the lips, may become encrusted, as seen in Figure 124.2.

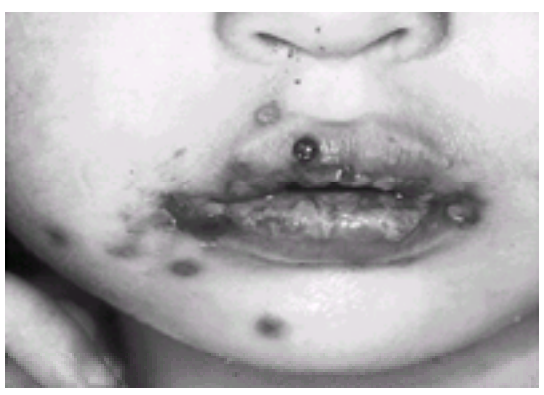


FIGURE 124.2. A child with typical crusted extraoral lesions of late primary gingivostomatitis.

If necessary, a definitive diagnosis can be made by isolation of herpes simplex virus in tissue culture (although this is rarely indicated). Emergency treatment includes reassuring the parent and rehydrating the patient. The disease, like recurrent herpes labialis, is self-limiting, with a duration of 7 to 14 days. Dehydration and weight loss are the major concerns; therefore, milk shakes, ice cream, and liberal quantities of clear fluids should be encouraged. The young child with extensive lesions may require hospitalization for intravenous hydration.

Viscous Xylocaine rinses and “magic mouthwash,” Kaopectate and Benadryl, may be unrealistic for children in this age range. The unpleasant taste sometimes negates any benefit that the topical anesthetic gives, and makes administration difficult.

Secondary infection, although rare, is of concern for those children who may be immunosuppressed and, in those cases, antibiotic therapy may be indicated.

Acute Necrotizing Ulcerative Gingivitis, Vincent's Disease, Trench Mouth

Acute necrotizing ulcerative gingivitis (ANUG), Vincent's disease, or trench mouth is characterized by increases in the fusiform bacillus and *Borrelia vincentii,* a spirochete, which usually coexist in a symbiotic relationship with other oral flora. Adolescents complain of soreness and point-tenderness at the gingiva and often tell the physician that they feel as if they “cannot remove a piece of food that is painfully stuck between their teeth” (a wedging sensation). They may also complain of a metallic taste in their mouth and of bleeding gums. Upon examination, the breath has an obvious fetid odor. The gingivae are hyperemic and the usually triangular gingiva between the teeth is missing, or “punched out” (Fig. 124.3). Intense pain is produced with probing, and a gray, necrotic pseudomembrane may cover some areas of gingiva.

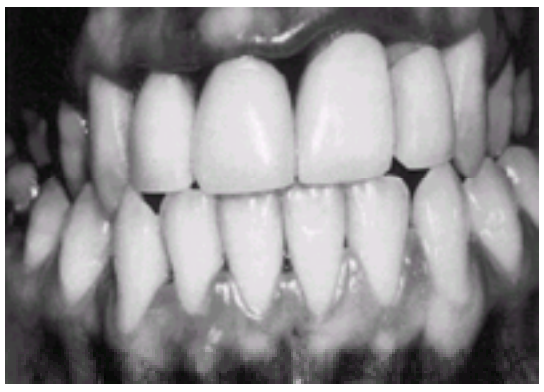


FIGURE 124.3. A child with typical “punched out” gingiva—pathognomonic for acute necrotizing ulcerative gingivitis. (Courtesy of Dr. Mark Snyder.)

It is extremely rare to find ANUG in a young child, but a mistaken diagnosis is often made by physicians, confusing this disease with primary herpetic gingivostomatitis. Primary herpes is usually seen in infants and toddlers, and ANUG is characteristically seen in adolescents and young adults (ages 15 to 35). Emotional stress has been linked to the onset of the disease process. The adolescent should be advised to maintain better oral hygiene and to use frequent hydrogen peroxide mouth rinses. Diluted 1:1 with warm water, the hydrogen peroxide is vigorously swished and forced between the teeth as often as possible throughout the acute phase. Because of the rapidity of tissue destruction and sensitivity of the organisms, as well as risk of secondary infection, penicillin should be prescribed for the first week. When the acute phase is over, the patient should be sent to the dentist for a thorough debridement of the area.

ORAL AND PERIORAL PATHOLOGY PRESENTING AS DENTAL EMERGENCIES

Erythema Multiforme

Erythema multiforme is primarily a dermatologic disease characterized by macular, papular, vesicular, or bullous lesions on the skin or oral mucosa (Fig. 124.4). The lips may appear crusted, as in primary herpes. Lesions arise from an erythematous area that enlarges and develops a central vesicle or “target lesion.” Oral lesions arise at about the same time as skin lesions and are also variable in their clinical appearance, producing painful, bleeding, crusting erosions. These symptoms may occur as an acute drug reaction, but can be precipitated by herpes simplex. Stevens-Johnson syndrome is a more disseminated form of erythema multiforme in which conjunctival and genital lesions are seen concomitant with the oral and cutaneous lesions (see Chapter 99). Identifying and eliminating the precipitating drug is the first step in treatment. Immediate care may include caloric and fluid support. Steroids may be of help in severe cases.

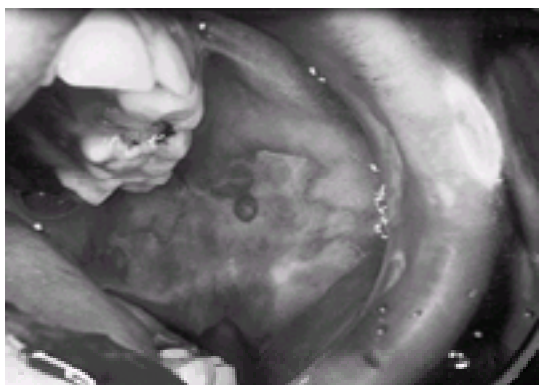


FIGURE 124.4. Intraoral view of erythema multiforme.

Epidermolysis Bullosa

Epidermolysis bullosa is a hereditary vesiculobullous condition affecting the skin, mucous membranes, and teeth. There are several forms of the disease. In the dominant form, oral bullae have been documented. In the recessive dystrophic form, the teeth have hypoplastic enamel, have an increased susceptibility to dental caries, and delayed eruption. Oral mucosal involvement appears soon after birth with vesicles from the negative pressure of the sucking reflex. The labial mucosa and lips can appear scarred and taut. Even routine dental management such as toothbrushing may cause the eruption of bullae on the mucosa and lips. Emergency visits may result from pain or bleeding from oral lesions. Treatment should palliate pain and support nutritional requirements.

Pyogenic Granuloma

Pyogenic granulomas develop as granulation tissue in response to an irritant or trauma. Clinically, they are red, elevated, and usually ulcerated. Initial growth is rapid. Pyogenic granulomas are most common on the gingivae and may remain static for a time before becoming fibrotic. Treatment consists of simple excision, but recurrence is common unless the causative agent (calculus or foreign body) is removed.

Epstein's Pearls

Epstein's pearls are keratin-filled cystic lesions located along the midpalatine raphe in the newborn. Often, only a few can be visualized, but sometimes there are too many to count. They are thought to arise from embryologically trapped epithelium. They are present in about 80% of neonates and should be considered a variation of normal. No treatment is necessary because they disappear within several weeks.

Bohn's Nodules

Bohn's nodules are remnants of the dental lamina that appear as cysts on the buccal or lingual aspect of the maxillary and mandibular dental ridges in the newborn. They may appear in the palate but are far removed from the midpalatine raphe and should not be confused with Epstein's pearls. No treatment is necessary because they disappear in several weeks.

Dental Lamina Cysts

Dental lamina cysts are multiple, or occasionally solitary, nodules on the alveolar ridge of newborn (Fig. 124.5) or young infants. They represent trapped remnants of the dental lamina. They are soft and spongy, asymptomatic, and tend to disappear with time or the eruption of teeth.

FIGURE 124.5. Dental lamina cyst in a neonate.

Eruption Cysts

Eruption cysts arise from the pre-eruptive dental sac and appear as a swelling of the alveolar ridge. They are associated with the eruption of primary (Fig. 124.6) and permanent teeth. Occasionally, they fill with blood and may be termed

eruption hematomas (Fig. 124.7). Treatment is unnecessary because the erupting tooth usually emerges within several days. If treatment is necessary because of the size of the lesion, excision of the overlying soft tissue to expose the erupting tooth eliminates the problem.

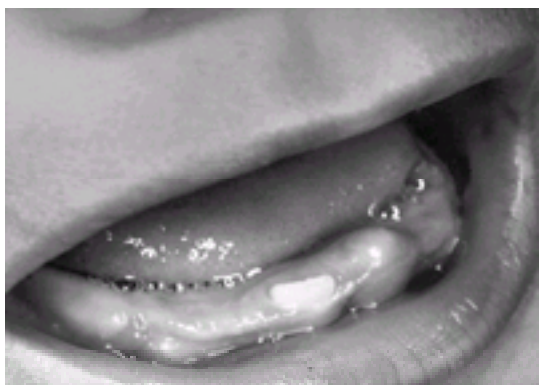


FIGURE 124.6. An eruption cyst associated with an erupting primary central incisor.

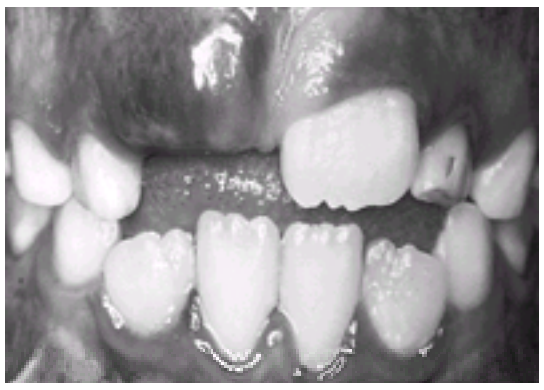


FIGURE 124.7. Erupting hematoma over erupting maxillary permanent central incisor.

Riga-Fede Disease

Riga-Fede disease is a condition observed in infants with natal or neonatal teeth. It is characterized by ulcerations on the ventral surface of the tongue from irritation caused by the incisal edges of lower incisors during nursing or suckling. Treatment should be avoided, but extraction may be necessary if they interfere with feeding.

Orofacial Neoplasms

Orofacial neoplasms in children are rare, but may be frightening to the patient and their family. Common benign and malignant neoplasms may result in emergency visits and, therefore, are included. Identification is central to the triage process.

The oral papilloma is a benign epithelial neoplasm that is an exophytic elevation of the surface epithelium with small fingerlike projections from its surface. These lesions, which rarely become malignant, constitute about 8% of all oral neoplasms in children. Slightly more than a third of the lesions occur on the tongue and (in decreasing order of frequency) palate, buccal mucosa, gingiva, and lip. If spontaneous involution does not occur, the usual treatment is surgical removal.

The fibroma is a common smooth-surfaced lesion with a sessile base. Its consistency varies from soft to firm, and its size ranges from a few millimeters to a centimeter or more in diameter. It may become whitened secondary to the overlying hyperkeratosis caused by trauma. Fibromas occur during the first and second decades of life and are usually found on the palate, tongue, cheek, and lip. Surgical removal is sometimes indicated and recurrence is rare if the source of the irritation is removed.

The mucocele appears as a soft, raised, fluid-filled and well-delineated nodule, most commonly on the lower lip (Fig. 124.8). Superficial lesions appear translucent and are bluish, whereas deep-seated lesions have a normal color. A mucocele in the floor of the mouth is termed a ranula and is seen as a dome-shaped, fluid-filled lesion. Mucoceles are thought to result from severance or obstruction of a salivary gland duct, with pooling of mucin in the lamina propria. Complete excision of the mucocele or marsupialization of the ranula is indicated.

FIGURE 124.8. Mucocele associated with minor salivary gland of the lower lip.

Suggested Readings

Kaban L. Pediatric Oral and Maxillofacial Surgery. Philadelphia: WB Saunders, 1990.

Shusterman S. Pediatric dental update. Pediatr Rev 1994;15:311–318.

CHAPTER 125

Neurosurgical Emergencies, Nontraumatic

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Patients with non-traumatic acute neurosurgical problems come to the emergency department (ED) with a variety of non-specific signs and symptoms, including headache, vomiting, seizures, changes in mental status, weakness, and coma. Because headache and vomiting are common, a high index of suspicion is required. This point is amplified by a case series from a large center that reports seven children who died unexpectedly from acute hydrocephalus associated with a previously undiagnosed intracranial tumor, most of whom had been managed as presumed acute gastroenteritis and treated with intravenous hydration.

Because most of the acute problems that result in an ED visit for non-traumatic neurosurgical problems are related to the onset of increased intracranial pressure (ICP), this chapter opens with a general discussion of ICP. A detailed description of the specific congenital, infectious, vascular, and neoplastic entities that may result in intracranial hypertension follows.

INCREASED INTRACRANIAL PRESSURE

Pathophysiology

The functions of the buffering systems that maintain ICP at normal levels have been detailed in Chapter 105. The relatively rigid cranium contains three components: 1) brain tissue predominantly, 2) cerebrospinal fluid (CSF), and 3) blood. An abnormal increase in volume of any of these components (by means of edema or mass lesion of the brain tissue, increased production or diminished absorption of CSF, or increased cerebral perfusion pressure related to increased blood flow) may result in elevated ICP. This closed space has limited capacity to compensate for increased volume from hydrocephalus, cerebral edema, hemorrhage, mass lesions, or pus.

ICP levels normally rise and fall over the course of the day. ICP is at its peak during sleep, because the horizontal posture and the relative hypoventilation that increases arterial P CO_2 result in increased cerebral blood flow. This condition is especially true during rapid eye movement (REM) sleep, which usually occurs just before awakening. Headaches that awaken a child during the night or that occur on awakening are always significant, because they may be caused by ICP waves elevated beyond the normal nighttime peaks. With the improved ventilation and the assumption of upright posture on arising, P CO_2 is lowered and cerebral vasodilation is lessened. If vomiting occurs, the accompanying hyperventilation also decreases cerebral blood flow. As a result, the severity of the headache may then lessen significantly in the awakened child.

Clinical Manifestations

A careful history must be taken with respect to the timing and severity of headaches, vomiting, changes in behavior, visual changes, and episodic decreases in level of consciousness (Table 125.1). Nighttime and morning headaches that improve on arising are always ominous suggestions of elevated ICP, as is recurrent vomiting without fever, abdominal pain, or diarrhea.

Symptoms	Signs
Headache	Papilledema
Nocturnal, episodic severe	Cranial nerve palsies
Vomiting	Meningismus
Stiff neck	Head tilt
Double vision	Retinal hemorrhage
Transient visual loss	Macewen's (cracked pot) signs
Gait difficulties	Decorticate/decerebrate posturing
Dulled intellect	Coma
Irritability	Progressive hemiparesis
	Bradycardia

Modified with permission from: Bruce DA. Neurosurgical emergencies. In: Fleisher GR and Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 3rd ed. Baltimore: Williams & Wilkins, 1993:1410.

Table 125.1. Signs and Symptoms of Elevated Intracranial Pressure

The clinical examination can help confirm the presence of intracranial hypertension, but a normal examination cannot reliably exclude it. Funduscopic examination should be performed to look for papilledema or optic atrophy. Visual fields and visual acuity should be checked. Cranial sutures may split in infants and young children with chronic elevation of ICP, resulting in a hyperresonant note when the skull is percussed, a “cracked pot” sound known as Macewen's sign. Cranial nerve palsy may occur, usually affecting the third and sixth nerves, resulting in dilated pupil, diplopia, and strabismus. When the fourth nerve is affected, the child may exhibit a “cock robin” head tilt. Cerebellar herniation may also cause head tilt; if herniation is bilateral, the neck may be held in an extended position.

The possibility of cerebral herniation should be considered and aggressively treated in the presence of an evolving pattern of diminishing level of consciousness, unexplained bradycardia, pupillary changes, or abnormal asymmetric motor responses to stimuli. Abnormal motor posturing in this setting may be confused with seizure activity. Cushing triad (hypertension, bradycardia, and irregular respirations) is a near terminal event, with bradycardia usually the first and most sensitive indicator.

Other diagnoses must be considered, but some can be ruled out by the history and examination. Migraine headaches are rarely present on waking and are usually relieved by rest. Children up to at least 7 years of age do not have frontal sinuses, ruling out the possibility of frontal headache caused by sinusitis in this younger age group. However, frontal sinusitis in adolescents has a significant rate of intracranial complications, warranting meticulous evaluation for this possible cause. The headaches of school phobia do not usually occur during sleep nor do they lessen on arising.

Management

The emergency treatment of increased ICP depends on the patient's clinical state and the cause of the intracranial hypertension (Table 125.2). However, the first priority in all patients is to follow the ABCs (airway, breathing, and circulation) of resuscitation and to prevent hypoxemia, hypercarbia, and systemic hypotension with oxygenation, ventilation, and appropriate fluid therapy. Seizures should be prevented if possible and treated aggressively when they occur, because the ICP spikes during seizures aggravate intracranial hypertension. Phenytoin (15 to 20 mg/kg) and fosphenytoin are the preferred drugs.

Prevent hypoxia and hypercarbia
Tracheal intubation/controlled ventilation
Seizure treatment and prophylaxis
Maintain adequate cerebral perfusion pressure and cerebral perfusion
Treatment of shock
Limitation of excessive hyperventilation
Decrease cerebral blood volume
Acute hyperventilation
Decrease brain tissue volume
Mannitol
Dexamethasone for vasogenic edema
Decrease cerebrospinal fluid (CSF) volume
CSF drainage
Acetazolamide
Removal of mass lesion
Surgical removal/decompression

Table 125.2. Treatment of Increased Intracranial Pressure

When the clinical picture suggests intracranial hypertension, computed tomography (CT) of the head should be performed without contrast material as soon as the patient is stable. Lumbar puncture should be withheld until the scan has been read for fear of precipitating herniation. The temptation to give sedative agents to the agitated patient to accomplish head CT or to facilitate transport should be avoided. Sedative agents given without controlled ventilation may result in hypercarbia, causing an increase in cerebral blood volume and, therefore, in ICP. Sedatives may also block protective airway reflexes, increasing the risk of aspiration. Therefore, inserting an endotracheal tube before CT scan or before transport is often preferable.

However, intubation should not be attempted without appropriate expertise and use of proper medications to blunt the increases in ICP associated with the procedure. Rapid sequence intubation (Chapter 5 and Chapter 105) should be accomplished after pre-oxygenation by giving a sequence of atropine (0.02 mg/kg), lidocaine (1.5 mg/kg), and thiopental (2 to 5 mg/kg), followed by a neuromuscular blocker. Succinylcholine (1 to 1.5 mg/kg) has rapid onset and short duration, but the fasciculations related to its depolarizing effects may cause transient increases in ICP. Rocuronium (0.6 to 1.2

mg/kg) and vecuronium (0.1 to 0.2 mg/kg) are non-depolarizing blockers that provide adequate alternatives.

In the absence of signs of impending herniation, P_{CO_2} should be controlled by mild hyperventilation in the range of 30 to 35 mm Hg. Prolonged excessive hyperventilation may cause cerebral ischemia. Continuous, portable capnometry may be useful as a monitor to avoid excessive hyperventilation during transport or diagnostic imaging.

Acute hyperventilation (hand ventilation) is used as an attempt to reverse signs of acute herniation. The head of the bed should be elevated to 30 degrees and the head maintained in a neutral position to promote venous drainage. Also, when the child exhibits acute herniation, drainage of CSF, either from a shunt reservoir or by a ventricular tap via an open fontanel, a split suture, or a burr-hole, allows controlled reduction of CSF volume. A ventriculostomy catheter may also be used to directly measure ICP, to direct medical therapy, and to allow drainage of CSF. Mannitol (0.25 to 2.0 g/kg) is the most useful drug to acutely decrease ICP in the deteriorating patient. Acetazolamide and furosemide have a limited role in acute management. Dexamethasone (1 mg/kg) is of controversial benefit, has slow onset, and appears to be most useful in treating vasogenic brain edema associated with tumors and brain abscess and non-traumatic hemorrhage. Surgical removal of collections of blood or pus may be indicated to lower ICP, as discussed later in this chapter.

HYDROCEPHALUS

Hydrocephalus is characterized by dilated cerebral ventricles that contain an excessive amount of CSF, resulting from imbalance between production and absorption. Production, which is accomplished in the choroid plexus, almost always remains stable and is only rarely excessive. In non-communicating hydrocephalus, CSF in the ventricular systems is blocked from communicating with CSF in the subarachnoid spaces and basal cisterns by a congenital or acquired defect. In communicating hydrocephalus, the block in absorption is on the meningeal surfaces, outside the ventricular system. Congenital hydrocephalus may result from aqueductal stenosis or in association with Dandy-Walker or Arnold-Chiari malformations. Acquired hydrocephalus may follow bacterial meningitis, be secondary to tumor (particularly in the posterior fossa), or result from the inflammatory response to subarachnoid or intracranial hemorrhage.

Previously Undiagnosed Hydrocephalus

Children with undiagnosed hydrocephalus rarely present first to the ED, but hydrocephalus must be considered in all children with symptoms suggestive of increased ICP. A careful history should be taken of all previous illnesses and traumas. In a child with unexplained headache, chronic vomiting, and irritability, head circumference should be recorded and the fontanel, if open, evaluated. The skull should be transilluminated and dilated scalp veins should be noted. A “cracked pot” sound may be noted on percussion if the sutures are split. The pupils and extraocular movements should be examined. Difficulty with upward gaze (“sunset” sign) may be seen. Muscular spasticity, particularly in the lower extremities, may develop as cortical motor fibers are stretched by the ventricular dilation.

Management

Non-contrast head CT demonstrates enlarged ventricles. The urgency with which either insertion of a ventricular shunt or ventricular drainage needs to be performed depends on the child's condition. Ventricular puncture through an open fontanel or coronal suture may be lifesaving in a child with evidence for impending cerebral herniation who is unresponsive to hyperventilation and mannitol.

Previously Shunted Hydrocephalus

The placement of CSF shunts is currently the most commonly performed neurosurgical procedure. These shunts allow diversion of CSF into another area of the body outside the brain, most commonly the peritoneal cavity, thereby relieving pressure on the brain. Unfortunately, placement of shunts is accompanied by complications, including malfunction, obstruction, infection, malposition, and migration. Children who have shunts represent a heterogeneous group with multiple causes for hydrocephalus, including: congenital defects, intraventricular hemorrhage, myelodysplasia, central nervous system (CNS) infection, and brain tumor.

Pathophysiology

Many different types of shunts are in use; therefore, emergency physicians must become familiar with those commonly used in their area. All shunts share three common features 1) a radiopaque ventricular catheter; 2) a one-way valve; and 3) a long distal tubing that is palpable subcutaneously and that enters, most commonly into the peritoneal cavity, but rarely into the right atrium or pleural cavity. Many shunts also include a pumping mechanism and a reservoir, which enables percutaneous sampling of CSF without damage to the shunt.

Shunt malfunctions are the most common complication of CSF shunts. The risk for shunt failure is greatest in the first months after placement, with a mean survival time of 5 years. Approximately 80% of patients require a revision by 10 years. Obstructions occur most commonly (approximately 80%) at the proximal (ventricular) end as a result of occlusion by tissue or migration of the shunt tip into the brain parenchyma. The valve may occasionally become blocked. Distal obstruction also may result from disconnection of the shunt tubing, migration of the catheter outside the peritoneum, perforation of the bowel, or pseudocyst formation. Shunt infections eventually result in obstruction.

Shunt Malfunction

Clinical Manifestations

Patients with shunt malfunction commonly present with manifestations of increased ICP. Children may complain of headache (often worse in the morning), screaming episodes, lethargy, and other behavioral changes and/or visual

symptoms. Vomiting is common. Parents, particularly those who have witnessed prior episodes of shunt malfunction, are perceptive to subtle, and often intermittent symptoms that herald shunt malfunction. On physical examination, unilateral or bilateral cranial nerve palsies, especially a non-localizing sixth nerve palsy, may be present. Intermittent downward gaze (sunset sign) may be reported or observed. Swelling from CSF tracking along the shunt tract is indicative of obstruction. The fontanel may be full and tense, even when the infant is upright. Rapid enlargement in head circumference or increase in the prominence of scalp veins may occur. Infants and young children with split sutures have a characteristic hyperresonant sound when the skull is percussed (“cracked-pot” sound) known as a positive Macewen's sign. Papilledema is uncommon in acute shunt malfunction. Head tilt may be seen as a result of fourth cranial nerve palsy or cerebellar tonsillar herniation.

Of particular concern are waves of severe headache with or without visual changes, loss of consciousness, decerebrate posturing, or new third nerve palsy. Although seizures are common in patients with CSF shunts, one reported series revealed that only 2.9% of ED visits for seizures in patients with shunts culminated in shunt revision. Thus, a seizure alone is seldom an indication of shunt malfunction. Most shunts have a pumping mechanism, but pumping the shunt correlates poorly with shunt obstruction. In one series, shunt pumping in patients with suspected shunt malfunction had a sensitivity of 18% with a positive predictive value of only 17%. The shunt should not be routinely pumped because the negative pressure generated in a small ventricle may occasionally result in obstruction. However, if the ventricular catheter is shown on CT scan to be in the center of a dilated ventricle and the shunt umbilicates on depression with slow refill, shunt obstruction is likely.

Management

If the history or physical examination suggests shunt malfunction, early neurosurgical consultation is strongly recommended. A plain radiographic “shunt series” should be done, consisting of anteroposterior and lateral views of the skull, neck, thorax, and abdomen. These radiographs allow the type, location, connections, and intactness of the system to be evaluated. Occasionally, split sutures on the skull film suggest increased ICP. Noncontrast head CT should also be taken and compared, if possible, with previous scans taken when the shunt was functioning. In most cases (approximately 80%), these studies identify shunt malfunction.

Shunt tap may also be useful in patients in whom the function of the shunt is questionable. This procedure should be completed under sterile conditions with a knowledge of the anatomy of the patient's shunt. A small, but significant risk of causing a shunt infection by performing a shunt tap exists. Therefore, diagnostic shunt taps are usually performed selectively by the neurosurgical consultant. Three pieces of information can be obtained. First, assuming the ventricular end is patent, the pressure in the system can be estimated by the level to which CSF rises when the butterfly tubing is held erect. A falsely low reading results if the ventricular end is obstructed. If the pressure is high, CSF may be withdrawn until a pressure of approximately 10 cm of water is reached. Second, downstream drainage is checked by permitting CSF to flow distally, observing the height at which the CSF column ceases draining. Third, the fluid is sent to the laboratory to evaluate for CSF infection.

The urgency of shunt revision depends on the patient's status. Patients with evidence of obliteration of the perimesencephalic cistern appear to be at particularly high risk for sudden deterioration. Patients with proximal obstructions may worsen quickly, and if the child suddenly deteriorates, CSF cannot be quickly withdrawn from the shunt reservoir to relieve pressure. Because fluid cannot be drawn from the shunt, the ventricle must be tapped through the fontanel, if open, through the sutures if they are split, or through the shunt burr hole. This latter maneuver usually damages the shunt and is therefore simply a temporizing measure to lower the ICP before operative shunt revision.

When the distal end of the shunt is blocked, the ICP can be immediately lowered by removing CSF through a shunt tap. The need for emergency shunt revision is less in this setting because the ICP can be easily controlled by tapping the shunt. Acetazolamide and dexamethasone may be used as temporizing measures if shunt revision is to be delayed, but may not be effective.

In some patients, ventriculomegaly may persist despite a functioning shunt. In these patients, prior CT scans are particularly helpful. Conversely, some patients have symptoms and signs of increased ICP despite small or unchanged ventricules, which is called the “slit ventricle” syndrome. This condition may be the result of intermittent proximal obstruction, poor ventricular compliance, or overdrainage of CSF. A history of onset or worsening of symptoms with upright posture suggests overdrainage. These children require careful further evaluation, which may include ICP monitoring, to determine the functions of their symptoms.

Shunt Infection

About 70% of all shunt infections occur within 2 months of surgery.

Pathophysiology

Infections are caused by low virulence organisms found in skin flora. *Staphylococcus epidermidis* accounts for approximately 75% of shunt infections, followed by Gram-negative organisms and *Staphylococcus aureus. S. epidermidis* recovered from CSF fluid shunt devices secrete an extracellular polysaccharide “slime” substance that coats the shunt, making the enclosed colonies of organism highly resistant to phagocytosis and systemic antibiotics.

Clinical Manifestations

In the post-operative period, erythema and warmth along the course of the shunt are highly predictive of early wound infection. Later, signs of indolent infection are often variable and nonspecific. Signs of shunt malfunction occur commonly. The adage, “an infected shunt is an obstructed shunt” is well remembered. Although fever raises concern for shunt infection, documented shunt infections were associated with fever in only 42% of patients in one series. Meningeal

signs have been reported in only about 33% of patients. Abdominal symptoms from an associated peritonitis may predominate in cases of pseudocyst formation with distal obstruction.

Management

A definite diagnosis of shunt infection is made by tapping the shunt and obtaining a CSF specimen. A definite, although small, rate of infection occurs as a result of a tap, and in someone with the potential for bacteremia, the blood carried into the shunt reservoir on the tip of the needle may be contaminated and produce a shunt infection. Thus, shunt tap is not indicated in all children with a shunt who present with fever. A diligent search for alternative explanations of fever should be undertaken. Urinary tract infections are a particularly common cause of fever in children with myelomeningocele. Debilitated patients with severe developmental delay are at risk for pneumonia. These potentially occult sources should be excluded. Fever without localizing signs in those patients in whom the current shunt was placed or revised many months to years ago and who lack signs and symptoms of shunt malfunction may be appropriately managed with close follow-up and observation without shunt tap.

In patients with noncommunicating hydrocephalus, if the shunt tap is negative but a CNS infection is a likely clinical diagnosis, a lumbar puncture is required because ventriculitis may not accompany meningitis. CSF obtained from the shunt tap is sent for Gram stain, cell count, glucose and protein, and culture. If the CSF contains more than 100 cells/mm^3, cultures are positive in about 90% of patients. However, most infected shunts have only modest pleocytosis (less than 200 white blood cells/mm^3), slight decrease in glucose, and modest protein elevation. The Gram stain is positive for Gram-positive cocci in clusters in approximately 50% of infections caused by *S. epidermidis.* Blood cultures are positive in only a minority of patients. The peripheral white blood count is of little use in predicting patients with infected shunts.

In patients with abdominal symptoms, neurosurgical consultation should be obtained before exploratory laparotomy for suspected appendicitis. Abdominal ultrasonography is especially useful for diagnosing abdominal pseudocyst, which is always secondary to infection. If shunt infection is diagnosed and appropriate cultures are obtained (including CSF via shunt tap, blood culture, and possibly urine and soft-tissue aspirate cultures from areas of local inflammation), treatment with intravenous vancomycin (15 mg/kg) may be started, pending growth on the cultures. Patients require complete removal of the entire shunt and a period of external drainage, followed by placement of a new shunt into a different anatomic location.

STROKE

Stroke denotes a sudden onset of a persistent focal neurologic deficit, resulting from interruption of blood flow to a localized area of the brain. Pediatric stroke has a wide range of causes and risk factors distinct from those in adults, thus limiting comparison to stroke in adults.

Hemorrhagic Stroke

In children and adolescents, rupture of an arteriovenous malformation is the most common cause of spontaneous intracranial hemorrhage. (General causes of hemorrhagic stroke are listed in Table 125.3.) These lesions are within the cerebral parenchyma; therefore, when they bleed, the hematoma is intracerebral and the bleeding is arterial. Arterial intraparenchymal bleeding results in progressive surrounding edema and focal mass effect. Congenital or acquired coagulation disorders, such as severe factor VIII deficiency or severe thrombocytopenia, may result in spontaneous intracranial bleeding with minimal or no preceding head trauma.

Secondary hemorrhage into ischemic brain
Vascular malformations
Arterio-venous malformations
Sickle cell disease
Saccular (berry) aneurysms
Hemorrhage into intracranial tumor
Coagulopathy
Hemorrhagic disease of the newborn (vitamin K deficiency)
Clotting factor deficiency (VIII, IX, XI)
Thrombocytopenia
Arterial hypertension
Renal vascular or parenchymal disease
Coarctation of the aorta
Pheochromocytoma
Illicit drugs with sympathomimetic effect
Amphetamines, cocaine

Table 125.3. Causes of Hemorrhagic Stroke

Ruptured aneurysms are rare, accounting for only 10% of intracranial hemorrhage in children. Congenital ruptured aneurysms rarely may be seen as early as the first week of life. The bleeding occurs from an aneurysm located at branching points of the major arteries coursing through the subarachnoid space at the base of the brain. The incidence of aneurysm is increased in several inherited conditions, including autosomal-dominant polycystic kidney disease, Ehlers-Danlos type IV, neurofibromatosis type 1, and Marfan's syndrome.

Other vascular abnormalities associated with intracranial bleeding include cavernous angiomas and hemangioblastoma associated with von Hippel- Lindau syndrome. Cavernous angiomas are low-flow lesions that can occur anywhere in the cerebrum, brainstem, cerebellum, or spinal cord. Because they lack large arterial feeders, onset of symptoms is usually subacute. The greatest danger is of acute hydrocephalus caused by occlusion of the fourth ventricle, resulting from

posterior fossa hemorrhage and/or swelling.

Ischemic Stroke

Ischemic injury to the brain occurs as a result of embolism from the heart or proximal arterial circulation, or from thrombosis in the arterial or sinovenous system. The most common risk factor (in about 25% of patients) is congenital heart disease.

Although it is difficult to separate risk factors from causes, several conditions have been associated with embolic or thrombotic stroke (Table 125.4). Embolic sources are primarily from the heart (dilated or abnormal chambers), from abnormal or infected heart valves, or from “paradoxical” emboli via lesions associated with right-to-left cardiac shunts. Other risk factors include congenital or acquired vascular disorders and factors that result in hypercoagulability, such as 1) oral contraceptive use, 2) anticardiolipin antibodies, and 3) deficiencies of protein S, protein C, or antithrombin III. Stroke occurs in 6 to 9% of patients with sickle cell disease (SCD). In children with SCD, most strokes are ischemic, resulting from occlusion of intracranial carotid and middle cerebral arteries. An important cause of stroke is arterial dissection, which may be spontaneous or associated with minimal trauma. This condition often results in acute thrombosis and has been treated with anticoagulation and endovascular thrombolytic treatment.

Cardioembolic
Cyanotic congenital heart disease
Right-to-left shunts (e.g., patent foramen ovale)
Congenital or acquired valvular defects
Contractile dysfunction
Rhythm disturbance
Vascular disease
Sickle cell disease
Arterial dissection
Homocystinuria
Vasculitis
Moyamoya
Migraine
Thrombotic (arterial and sinovenous)
Hypercoagulable state, *congenital or acquired*
Hyperviscosity (polycythemia, dehydration)
Genetic/metabolic

Table 125.4. Causes of Ischemic Stroke

Clinical Manifestations

Diagnosis of stroke syndrome in children is often delayed by the failure to consider it. Focal weakness in association with headache or after a seizure should not be dismissed as hemiplegic migraine or post-ictal Todd's paresis. Presenting symptoms are non-specific as to the cause, but several patterns exist. Depending on the location and nature of the intraparenchymal lesion, stroke in children may present with sudden onset of hemiplegia or hemiparesis, aphasia, and sensory symptoms. These focal signs are frequently accompanied by seizures, fever, acute change in mental status, and signs and symptoms of increased ICP. Arterial dissection commonly presents as stroke with the patient complaining of sudden ipsilateral pain in the head, neck, or eye. It may be associated with Horner's syndrome if cervical sympathetic chain is involved, and a bruit may be heard over the involved carotid. The posterior circulation can also be involved, resulting in vertebro-basilar insufficiency resulting in cranial neuropathies, difficulties with balance and coordination, and tremor. Subarachnoid hemorrhage causes sudden severe headache and meningismus caused by the breakdown of blood products in the subarachnoid space, leading to meningeal irritation. As CSF circulates, symptoms of lower back pain and radicular leg pain may subsequently predominate.

Management

After initiation of supportive care to prevent secondary hypoxic ischemic injury and to ameliorate increased ICP, the next priority is to exclude an acute intraparenchymal or subarachnoid hemorrhage. Non-contrast head CT is sensitive for acute bleeding and should be obtained emergently. Noncontrast CT is a good test to exclude hemorrhagic causes of stroke, but it may be normal or near normal soon after the onset of symptoms of ischemic stroke. If subarachnoid hemorrhage is thought likely but not documented by CT, a lumbar puncture may be performed to enumerate red blood cells in the CSF. Xanthochromia from breakdown of blood products may also be seen, particularly if lumbar puncture is performed after the acute episode. Lumbar puncture has been associated with herniation in the setting of increased ICP and should be deferred until after a mass lesion or noncommunicating hydrocephalus has been excluded.

If intraparenchymal hemorrhage is found, most children with bleeding from suspected rupture of an arteriovenous malformation require early angiography to localize the bleeding and the arterial feeders. Medical therapy of associated edema and increased ICP may include dexamethasone (0.5 to 1.0 mg/kg; maximum 16 mg/day) and mannitol (0.25 to 2.0 g/kg). Seizures should be treated aggressively, and prophylactic administration of phenytoin (15 to 20 mg/kg) or fosphenytoin may play a role in this treatment. Coagulation defects should be corrected as appropriate (see coagulation emergencies, Chapter 87). Emergent splenectomy is indicated for intraparenchymal bleeding associated with idiopathic thrombocytopenic purpura (ITP).

Hemorrhagic stroke or secondary gross hemorrhage into an area of ischemic infarction may produce a rapidly expanding intracranial mass. Depending on the site of the hemorrhage, emergency surgical evacuation of the hematoma may be indicated to reverse cerebral herniation and lower ICP.

The management of ischemic stroke remains largely supportive. Acute carotid occlusion may result in significant hemispheric swelling sufficient to produce elevated ICP. This condition may require ICP monitoring and intensive medical

and perhaps surgical therapy to prevent or reverse transtentorial herniation. Invasive angiography and possibly endovascular thrombolytic therapy may be needed for definitive diagnosis and should be considered early in the diagnostic workup of ischemic stroke. Magnetic resonance imaging (MRI) permits visualization of brain infarction. Magnetic resonance angiography (MRA) yields further information about blood flow and the structure of cervical and intracranial vessels. Ultrasound may be used to evaluate the extracranial carotid circulation. Cardiac lesions are often found and may be evaluated with transthoracic or trans-esophageal echocardiography.

The decision to use anticoagulation must balance the likelihood of either extension of infarction or a second embolus with the risk of inducing hemorrhage. Anticoagulation is often used in children with arterial dissection, dural sinus thrombosis, or coagulation disorders, in those at high risk of embolism; or in response to progressive deterioration during the initial evaluation of a new cerebral infarction. The loading dose of heparin is 75 units/kg intravenously, followed by 20 units/kg per hour for children older than 1 year of age (or 28 units/kg per hour for children less than 1 year of age) titrated to a target activated partial thromboplastin time (aPTT) of 60 to 85 seconds. Alternately, low-molecular-weight heparin (Enoxaparin 1 mg/kg subcutaneously twice daily) has also been used.

Stroke in a patient with SSD is treated with simple or partial exchange transfusion to achieve a hemoglobin SS fraction of less than 30% and a hemoglobin level not greater than 10 g/dL to avoid problems of hyperviscosity.

The use of thrombolytic agents for ischemic stroke, administered intravenously, or locally utilizing angiographic catheters, is currently receiving significant attention. A single controlled trial of intravenous recombinant tissue plasminogen activator (t-PA) resulted in overall better outcomes in carefully selected adults treated within 3 hours after onset of symptoms. This therapeutic success has led to the concept of stroke as a “brain attack” analogous to a heart attack. In adults, placebo-controlled studies of intra-arterial thrombolysis are under way. However, as yet, no clinical efficacy trials of intra-arterial thrombolysis, nor a direct comparison between intravenous and intra-arterial thrombolysis, has been reported. The relative infrequency of childhood stroke and the heterogeneous patient population has hindered clinical trials of the sort possible in adults. There are anecdotal reports of successful endovascular thrombolysis in children. Intravenous thrombolytic therapy in children using t-PA (0.5 mg/kg) mostly for non-cerebral thrombotic complications has resulted in successful clot lysis, but at the expense of serious bleeding complications. If the diagnosis is delayed beyond 3 to 6 hours, risk of hemorrhage into ischemic infarcts clearly precludes any attempt at clot lysis. Some pediatric patients with acute ischemic stroke may benefit from thrombolytic therapy, when administered with caution, in a highly individualized manner using guidelines defined by ongoing randomized studies in adults.

CENTRAL NERVOUS SYSTEM INFECTIONS (SEE CHAPTER 84)

Infection of the CNS, including subdural empyema, brain abscess, bacterial and viral meningitis, and viral encephalitis, are often associated with some degree of increased ICP. Acute hydrocephalus may complicate tuberculous, fungal, amebic, and rarely bacterial meningitis. Subdural empyema occurs as a complication of parameningeal infections, such as sinusitis, orbital cellulitis, or mastoiditis, and is a true neurosurgical emergency, often progressing to death if not recognized early. Children with CNS infectious processes may present with fever, evidence of meningeal irritation, seizures, and increased ICP, which may progress to herniation and focal neurologic findings.

Management

Supportive care and antibiotics are the first priority in treatment of meningitis. Lumbar puncture should be deferred in the patient with cardiorespiratory instability. When an increase in ICP or mass lesion is suspected, the clinician must use extreme caution, and should not perform a lumbar puncture before excluding these diagnoses with an appropriate imaging study. A contrast-enhanced CT is necessary if a subdural empyema or brain abscess is suspected. Subdural empyemas require immediate neurosurgical drainage. Brain abscess is treated with dexamethasone, antibiotics, general management of intracranial hypertension, and often surgical drainage. An ICP monitor may assist in the medical management of intracranial hypertension associated with meningitis or encephalitis.

BRAIN TUMORS

Brain tumors in children usually present with a chronic history, but these signs and symptoms may have been attributed to benign causes. Neurologic abnormalities may be subtle. Children with brain tumors may develop the following four major symptom complexes: 1) cerebellar ataxia, 2) acute deterioration in the level of consciousness, 3) acute onset of cranial nerve palsies, and 4) severe recurrent headaches or vomiting. Headaches on awakening may be ominous, but many of the headaches associated with intracranial mass lesions are nonspecific.

Physical examination should include funduscopy to look for papilledema, cranial nerve examination for palsy, and search for focal motor deficits and ataxia. Acute symptoms may result from seizure, acute intracranial hypertension with pressure waves, or acute obstructive hydrocephalus or hemorrhage into tumor.

Management

Most children with dramatic acute symptoms requiring emergent management (obstructive hydrocephalus, mass effect, hemorrhage into tumor) are recognized on non-contrast CT, which is readily available to most EDs. Subsequent MRI with contrast is required for definitive management and evaluation of subtle abnormalities. The probability of elevated ICP should always be considered. Children with headache, papilledema, and altered levels of consciousness, which may suggest herniation, are at risk for sudden deterioration; therefore, early neurosurgical consultation should be obtained. Dexamethasone is useful in treating increased ICP before surgery. Patients are best managed by a multidisciplinary team that incorporates pediatric neurosurgery, neurology, and oncology.

SPINAL CORD COMPRESSION

Nontraumatic acute spinal cord dysfunction occurs in 4% of children undergoing treatment for cancer, usually because of spinal cord compression. Many children undergoing treatment for cancer develop back pain, which should raise concern for cord compression until proven otherwise.

Pathophysiology

Spinal cord compression may be the presenting sign of neuroblastoma, lymphoma, or sarcoma. Other causes of acute spinal cord symptoms include spinal epidural and, more rarely, subdural abscess, epidural hematoma, and congenital tethered cord.

Clinical Manifestations

Back pain in children commonly signals an important diagnosis. A history of localized or radicular back pain or refusal to walk mandates a careful evaluation. A history of change in gait or difficulty with bowel or bladder control should be sought. Localized tenderness to palpation is commonly found, and the level of maximal spinal tenderness is usually the site of pathology. A detailed neurologic examination should be documented with attention to extremity strength, reflexes, anal tone, and evaluation of sensory level. Compression of the spinal cord above the conus may be associated with increased or absent deep tendon reflexes, an extensor Babinski's reflex with symmetric (and profound) weakness, and a symmetric sensory level. Sphincter tone is spared until late, and progression is characteristically rapid. Compression of the conus medullaris results in increased knee and decreased ankle reflexes, extensor Babinski's reflex with a symmetric saddle distribution of weakness, and early sphincter involvement. Compression of the cauda equina typically results in asymmetric, often mild weakness, and asymmetric and radicular sensory distribution. Deep tendon reflexes are decreased with a plantar Babinski's response.

Management

Evidence for progressive cord dysfunction in the presence of significant neurologic deficits mandates immediate high-dose corticosteroid therapy with methylprednisolone 30 mg/kg or dexamethasone 2 mg/kg. Plain radiographs of the spine may be helpful, but emergency MRI is necessary to view the anatomic cause and degree of spinal compression. A previously unknown primary tumor is a clear indication for immediate decompressive surgery because it offers the benefit of decompression plus identification of the tumor type. Radiation therapy or chemotherapy may be indicated if the tumor type is known. Abscess formation, subdural or epidural hemorrhage, and symptoms related to cord tethering are usually indications for surgical intervention.

Suggested Readings

Burton LJ, Quinn B, Pratt-Cheney JL, Pourani M. Headache etiology in a pediatric emergency department. Pediatr Emerg Care 1997;13:1–4.

Shemie S, Jay V, Rutka J, Armstrong D. Acute obstructive hydrocephalus and sudden death in children. Ann Emerg Med 1997;29:524–528.

INCREASED INTRACRANIAL PRESSURE

Fenichel GM. Increased intracranial pressure. In: Clinical Pediatric Neurology: A Signs and Symptoms Approach. 3rd ed. Philadelphia: WB Saunders, 1977:91–105.

Gerardi MJ, Sacchetti AD, Cantor RM, et al. Rapid-sequence intubation of the pediatric patient. Ann Emerg Med 1996;28:55–74.

PREVIOUSLY UNDIAGNOSED HYDROCEPHALUS

Scott RM. Hydrocephalus. Baltimore: Williams & Wilkins, 1990.

Shallat RF, Pawl RP, Jerva MJ. Significance of upward gaze palsy (Parinaud's syndrome) in hydrocephalus due to shunt malfunction. J Neurosurg 1973;38(6):717–721.

Wiley JF, Duhaime A. Ventricular puncture. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1986:573.

PREVIOUSLY SHUNTED HYDROCEPHALUS

Duhaime A, Wiley JF. Ventricular shunt and burr hole puncture. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1986:559–565.

Fouyas IP, Casey AT, Thompson D, et al. Use of intracranial pressure monitoring in the management of childhood hydrocephalus and shunt-related problems. Neurosurgery 1996;38(4):726–731.

Hack CH, Benedicta GE, Donat JF, et al. Seizures in relation to shunt dysfunction in children with meningomyelocele. J Pediatr 1990;116:57–60.

Iskandar BJ, McLaughlin C, Mapstone TB, et al. Pitfalls in the diagnosis of ventricular shunt dysfunction: radiology reports and ventricular size. Pediatrics 1998;101:1031–1036.

Johnson DL, Conry J, O'Donnell R. Epileptic seizure as a sign of cerebrospinal fluid shunt malfunction. Pediatr Neurosurg 1996;24:223–227; discussion 227–228.

Johnson DL, Fitz C, McCullough DC, Schwarz S. Perimesencephalic cistern obliteration: a CT sign of life-threatening shunt failure. J Neurosurg 1986;64(3):386–389.

Key CB, Rothrock SG, Falk JL. Cerebrospinal fluid shunt complications: an emergency medicine perspective. Pediatr Emerg Care 1995;11:265–273.

Madikians A, Conway EE. Cerebrospinal fluid shunt problems in pediatric patients. Pediatr Ann 1997;26:613–620.

Piatt JH. Physical examination of patients with CSF shunts: is there useful information in purging the shunt? Pediatrics 1992;89:470–473.

Rekate HL. Classification of slit-ventricle syndromes using intracranial pressure monitoring. Pediatr Neurosurg 1993;19(1):15–20.

Sood S, Kim S, Ham SD, et al. Useful components of the shunt tap test for evaluation of shunt malfunction. Childs Nerv Syst 1993;9(3):157–161.

Watkins L, Hayward R, Andar U, Harkness W. The diagnosis of blocked cerebrospinal fluid shunts: a prospective study of referral to a paediatric neurosurgical unit. Childs Nerv Syst 1994;10(2):87–90.

Woodward GA, Carey CM. Evaluation of ventricular shunt function. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1986:553–558.

SHUNT INFECTION

Ersahin Y, Mutluer S, Tekeli G. Abdominal cerebrospinal fluid pseudocysts. Childs Nerv Syst 1996;12(12):755–758.

Ronan A, Hogg GG, Klug GL. Cerebrospinal fluid infections in children. Pediatr Infect Dis J 1995;14(9):782–786.

HEMORRHAGIC AND ISCHEMIC STROKE

American Heart Association Science Advisory and Coordinating Committee. Guidelines for thrombolytic therapy for acute stroke: a supplement to the guidelines for the management of patients with acute ischemic stroke. Circulation 1996;94:1167–1174.

Ferrera PC, Curran CB, Swanson H. Etiology of pediatric ischemic stroke. Am J Emerg Med 1997;15:671–679.

Fisher M, Bogousslavsky J. Further evolution toward effective therapy for acute ischemic stroke. JAMA 1998;279:1298–1303.

Ganesan V, Kirkham FJ. Lesson of the week: carotid dissection causing stroke in a child with migraine. Br Med J 1997;314:291–292.

Harisson MA, Witte M, Dean M. Cerebrovascular disease and vascular anomalies. In: Rogers MC, ed. Textbook of Pediatric Intensive Care. Baltimore: Williams & Wilkins, 1996:859–874.

Patel H, Smith RR, Garg BP. Spontaneous extracranial carotid artery dissection in children. Pediatr Neurol 1995;13(1):55–60.

Roach ES, de Veber G, Riela AR, Wiznitzer M. Recognition and treatment of stroke in children. Child Neurology Society Ad Hoc Committee on Stroke in Children. Proceedings of a National Symposium on Rapid Identification and Treatment of Acute Stroke. National Institute of Neurological Disorders and Stroke. December 12–13, 1996.

CENTRAL NERVOUS SYSTEM INFECTIONS

Ashwal S. Neurologic evaluation of the patient with acute bacterial meningitis. Neurol Clin 1995;13(3):549–577.

Yogov R. Focal Supportive Infections of the Central Nervous System. In: Long SS, Pickering LK, Prober CG, eds. Principles and Practice of Pediatric Infectious Diseases. New York: Churchill Livingstone, 1997:349–358.

BRAIN TUMORS

Edgeworth J, Bullock P, Bailey A, et al. Why are brain tumours still being missed? Arch Dis Child 1996;74:148–151.

Honig P, Charney E. Children with brain tumor headaches: distinguishing features. Am J Dis Child 1982;136:121–124.

Sjors K, Blennow G, Lantz G. Seizures as the presenting symptom of brain tumors in children. Acta Paediatr 1993;82(1):66–70.

Squires RH Jr. Intracranial tumors. Vomiting as a presenting sign. A gastroenterologist's perspective. Clin Pediatr (Phila) 1989;28(8):351.

SPINAL CORD COMPRESSION

Hesketh E, Eden OB, Gattamaneni HR, et al. Spinal cord compression: do we miss it? Acta Paediatr 1998;87:452–454.

Jacobsen FS, Sullivan B. Spinal epidural abscesses in children. Orthopedics 1994;17:1131–1138.

Kelly KM, Lange B. Oncologic emergencies. Pediatr Clin North Am 1997;44(4):809–831.

Sinha AK, Seki JT, Moreau G, et al. The management of spinal metastasis in children. Can J Surg 1997;40:218–226.

CHAPTER 126

Transplantation Emergencies

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Solid organ transplantation provides many children with a second chance at life, whether that means freedom from the disabling effects of end-stage liver, heart, or lung disease, or from the burden of chronic dialysis. However, transplant recipients differ significantly from other children, because of their susceptibility to infection, risk for organ rejection, and risk for early and late postsurgical complications. Most post-transplant problems encountered follow a pattern in terms of their timing and presentation, but others are dangerously subtle and insidious. Aggressive workup and compulsive examination are the rule when complications and emergencies arise in a child who has had a transplant. A thoughtful and focused approach is necessary for proper care. Although these children are naturally susceptible to the usual illnesses and injuries of childhood, their abilities to handle even minor infections or wounds may be compromised. Many illnesses and emergencies are unique to this patient group. The goal of this chapter is to provide the necessary background information and tools for the management of the transplant recipient presenting to the emergency department (ED). The focus is on recipients of kidney and liver grafts, but many of the problems discussed are germane to all transplant recipients.

OVERVIEW OF RENAL TRANSPLANTATION

End-stage renal disease is uncommon in children, with an incidence of 11 per 1 million population, less than 20 years of age. Renal transplantation is the preferred treatment for most children with end-stage renal disease because it restores the potential for optimal growth and development. Congenital structural abnormalities are the most common cause of renal failure in pediatric renal transplant recipients (Table 126.1). Renal transplantation is indicated when there are significant complications of chronic renal failure, including symptoms of uremia unresponsive to medical management, failure to thrive because of inadequate dietary intake, hypervolemia, metabolic bone disease, hyperkalemia, or delayed development. The absolute contraindications to renal transplantation include active malignancy, chronic human immunodeficiency virus (HIV) infection, chronic active infection with hepatitis B, and severe multi-organ failure. The relative contraindications are uncommon and would be decided on a case-by-case basis. These conditions similar to those listed for the liver transplant patient in Table 126.2.

Cause	Percentage
Obstructive uropathy	16%
Renal dysplasia/hypoplasia	16%
Focal segmental glomerulosclerosis	12%
Chronic glomerulonephritis	10%
Reflux nephropathy	6%
Systemic immunologic disease	5%

Table 126.1. Most Common Causes of Renal Failure in Children Undergoing Renal Transplantation

Indications	Contraindications
Poor or arrested growth	Untreated bacterial or fungal infection outside of the liver
Intractible coagulopathy	Malignant disease outside of the liver
Progressive hypoalbuminemia (<2.5 g/dL)	Irreversible brain damage
Intractible hepatic encephalopathy	Other irreversible organ failure
Refractory ascites	Poor or no psychosocial support
Recurrent esophageal variceal bleeding	Chronic human immunodeficiency virus infection
Spontaneous bacterial peritonitis	
Others: Intractable pruritus, pathologic fractures	

Table 126.2. Indications and Contraindications for Liver Transplantation in Pediatric Liver Disease

Approximately half of pediatric renal transplant recipients receive a kidney from a living donor, most often a parent, and the other half receive a kidney from cadaver donors. The transplant procedure is the same for either donor source. In larger children, the kidney is placed extraperitoneally in the iliac fossa with vascular anastomoses to the iliac vessels. In smaller children (less than 20 kg), the kidney is placed intraperitoneally with vascular anastomoses to the aorta and vena cava. Because of these vascular anastomoses, many centers recommend antibiotic prophylaxis after transplantation for dental procedures and other procedures that are likely to produce bacteremia, such as colonoscopy, to decrease the risk of arteritis, following the current recommendations for endocarditis prophylaxis. The ureter is implanted into the bladder in most children. An intestinal conduit is used in uncommon circumstances when no other options for urinary drainage are available (Fig. 126.1). The native kidneys remain in situ in 75% of patients. Indications for native nephrectomy include severe hypertension, chronic or recurrent pyelonephritis, risk of malignancy (Wilms' tumor in Denys Drash syndrome), or young recipient age. Because the native kidneys remain in place, the possibility of urine production by these kidneys must be kept in mind when assessing renal transplant recipients.

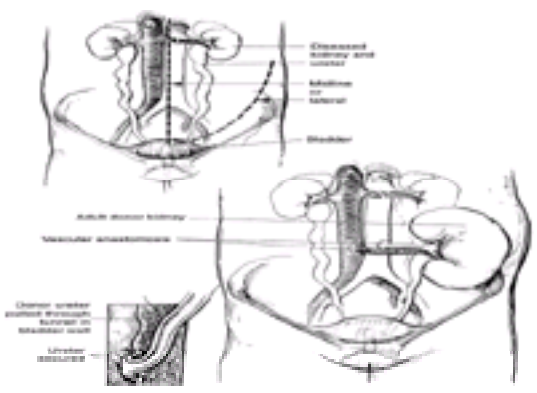

FIGURE 126.1. Anastomoses in renal transplantation

OVERVIEW OF LIVER TRANSPLANTATION

Human orthotopic liver transplantation is a reliable life-saving technique for patients with acute or chronic liver failure. Since 1963, when Starzl performed the first liver transplant, the technique and success of transplantation has grown exponentially. Presently, most transplant centers report 5 year survival rates of at least 80 to 90% in children after liver transplant. The most common indication for liver transplantation in the pediatric age group is biliary atresia with chronic liver failure after a failed hepatoportoenterostomy (Kasai procedure). The second most common indication is the metabolic disorder-a-antitrypsin deficiency. Smaller percentages of children receive transplants to treat other metabolic diseases, chronic liver disease secondary to hepatitis, and acute liver failure resulting from viral or toxic causes. The main indication for transplantation is end-stage organ dysfunction and failure (Table 126.2). The timing of the transplant procedure is crucial to the outcome and is based on the severity of the disease. The risks of the procedure make it inappropriate to operate on a child who has a good quality of life, but it is essential not to delay until end-stage liver failure with associated multi-organ involvement occurs. The aim of the pretransplant evaluation of the potential transplant recipient is to detect any likely surgical problems, assess the extent of any extrahepatic problems, and prepare the child and the family for the operation. Patients are listed with The United Network for Organ Sharing (UNOS), which administers the national organ procurement computer-matching network according to blood group, weight range of the donor, and medical urgency of the recipient.

Pediatric patients may receive transplants from reduced-size adult cadaver livers, whole cadaver organs from a donor of similar size, or donated left-lateral segments from a living donor. The transplantation operation in children differs from the procedure in adults based on the accommodations made for small sizes of the vessels and biliary ducts, which need to be anastomosed between the donor and recipient. A Roux-en-Y jejunal limb is usually used in most children for the biliary anastomosis in children, which changes the dynamics of biliary drainage. The operation involves the anastomosis of the donor and recipient portal veins, hepatic arteries, hepatic veins and bile duct–to–bile duct or biliary–enteric (Roux-en-Y) (Fig. 126.2A) anastomosis. After the transplant, any of these anastomotic sites may be a source of complications or emergencies. Despite these potential complications, liver transplant recipients generally have an excellent quality of life and return to a normal lifestyle shortly after receiving their graft.

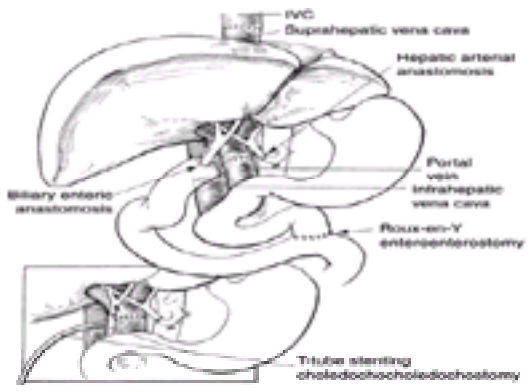

FIGURE 126.2. Anastomoses in liver transplantation.

IMMUNOSUPPRESSIVE AGENTS

The outcome of transplantation in children has improved progressively with the introduction of new immunosuppressive agents. The goal of immunosuppressive therapy is to down-regulate the immune response. Failure to attain an adequate degree of immunosuppression is associated with a high incidence of graft rejection. Acute and chronic allograft rejection remains a significant cause of morbidity and a contributing factor in mortality after solid organ transplantation. Post-transplant immunosuppression varies among transplant centers, organs transplanted, and individual patients. In all cases, however, a combination of agents is used to optimize immunosuppressive effects while minimizing adverse effects, such as opportunistic infection and renal toxicity. Initial immunosuppression for some transplant recipients includes induction with monoclonal or polyclonal anti-lymphocyte antibodies. Most long-term immunosuppression regimens consist of varying combinations of cyclosporine or tacrolimus, azathioprine or mycophenolate mofetil, and corticosteroid. The immunosuppressive agents are generally administered at high dosages in the early post-transplant period and are then weaned progressively if the patient is doing well. Many post-transplant complications observed are related to adverse effects of these medications. Immunosuppression remains complicated by the resultant increased incidence of major infections and malignancy in transplanted patients (Fig. 126.3).

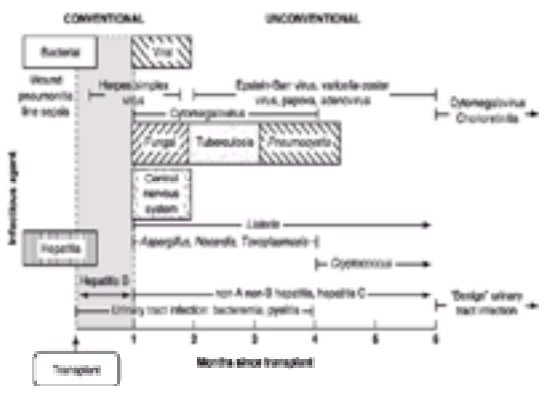

FIGURE 126.3. Timing of posttransplantation infections.

OKT3/Antilymphocyte Globulin (ALG)/Antithymocyte Globulin (ATGAM)

Polyclonal (ALG, ATGAM) and monoclonal (OKT3) antibodies against lymphocytes generally or effector T cells specifically are often effective in reversing established rejection refractory to high-dose steroid pulse therapy. These antibodies may also be used as part of initial immunosuppressive regimens at some centers and are particularly useful in the early postoperative management of the anuric kidney transplant or the liver recipient with poor renal function. Polyclonal antibodies are used in 37% of living donor kidney recipients and 47% of cadaver donor recipients. OKT3, a monoclonal mouse anti-human antibody to the CD3 epitope of lymphocytes, and a potent T-cell inhibitor, is used in 12% of living donor kidney recipients and 19% of cadaver donor recipients. These preparations are administered intravenously on an inpatient basis. The use of these agents increases graft survival in some situations but also increases the risk of infection (e.g., cytomegalovirus [CMV]) and malignancy (e.g., Epstein-Barr virus [EBV]-related lymphoproliferative disease). A patient who has received a recent course of OKT3 or ALG may be profoundly immunosuppressed for several weeks after administration and is at increased risk for opportunistic infections.

Cyclosporine (Neoral®/CyA)

Cyclosporine is the backbone of many immunosuppression regimens. Cyclosporine primarily affects the T-cell immune response by blocking the production of interleukin-2 (IL-2), thereby inhibiting T-cell activation. Cyclosporine also inhibits gene transcription for interferon-g, IL-3, IL-4, and other genes required for the differentiation and proliferation of T and B lymphocytes.

Empiric initial dosing of cyclosporine is modified on the basis of trough cyclosporine levels drawn just before the next dose or analysis of area-under-the-curve by formal pharmacokinetic studies. Blood or plasma levels may be measured by high-performance liquid chromatography (HPLC) or radioimmunoassay techniques. The target level varies by organ, assay method, and the time interval since transplant. For liver transplant recipients, cyclosporine trough levels of 250 to 400 ng/mL (HPLC of the parent compound) are recommended with slow reduction to half that level over time. The dosage

required to achieve these levels may range from 2 mg/kg to 30 to 40 mg/kg per day in young infants with decreased drug absorption. This variation has decreased with the advent of the microemulsion preparation of cyclosporine (Neoral ®), which has more predictable dosing and absorption/metabolism kinetics. Cyclosporine is highly lipid soluble and is only poorly soluble in water. Its absorption from the gastrointestinal (GI) tract is variable and depends in part on good bile flow for solubilization by bile salts and in part on the small intestinal length available for absorption. Therefore, any factor that impairs bile flow (i.e., bile duct stricture, external T-tube drainage of bile, allograft rejection, or cholestasis) may decrease absorption of cyclosporine. Similarly, any factor that decreases the small intestinal length available for absorption of the drug (i.e., long Roux-en-Y jejunal limb) or that increases transit time through the small intestine (as in diarrhea) may impair the absorption of cyclosporine. Higher dosages of the drug may be required in such cases to achieve therapeutic levels. The absorption of the Neoral ® preparation that is currently used is more consistent than that of the earlier non-microemulsified preparation. Cyclosporine is metabolized by the cytochrome P-450 liver microsomal system into multiple metabolites. Most metabolites are excreted in bile and undergo enterohepatic circulation. With severe liver dysfunction, in which the liver is unable to metabolize the cyclosporine or excrete the metabolites, the potential for toxicity of the drug is increased. Many drugs can alter the metabolism of cyclosporine by inhibition or stimulation of the P-450 system (Table 126.3).

Agents That Decrease Cyclosporine/Tacrolimus Metabolism (Increase Levels)
Erythromycin
Chloramphenicol
Ketoconazole
Fluconazole
Diltiazem
Verapamil
Nicardipine
Metoclopramide
Agents That Increase Cyclosporine/Tacrolimus Metabolism (Decrease Levels)
Phenytoin
Phenobarbital
Carbamazepine
Rifampin
Isoniazid
Agents That Cause Synergistic Nephrotoxicity
Aminoglycosides
Amphotericin B
Sulfonamides

Table 126.3. Medications That Alter Cyclosporine and Tacrolimus Metabolism

The major short-term and long-term side effect of cyclosporine is nephrotoxicity, which can cause particular confusion to the physician caring for the renal transplant patient. Some degree of renal dysfunction is present in 25 to 40% of transplant recipients. Acute, reversible renal dysfunction caused by intrarenal vasoconstriction secondary to cyclosporine may be manifested by hypertension, oliguria, azotemia, and hyperkalemia. These effects may be reversed with a decrease in the drug dosage. Chronic cyclosporine nephrotoxicity can result from interstitial fibrosis. Some drugs may act synergistically to amplify the nephrotoxic effect (Table 126.3). Other common side effects of cyclosporine include gingival hypertrophy, hirsutism, tremor, seizure, headache, hepatotoxicity, and hyperuricemia with occasional clinical gout (Table 126.4). Over time, the risk of de novo malignancy, such as lymphoma, is also increased. Seizures associated with cyclosporine occur in more than 20% of children, whether from high levels of the drug or in association with drug-induced hypomagnesemia caused by excessive magnesium wasting in the urine. To some degree, these side effects can be lessened by decreases in dosage, but their occurrence in any given patient is unpredictable and often idiosyncratic.

Adverse Effect	Cyclosporine	Tacrolimus	Azathioprine	Corticosteroids	Sirolimus
Systemic increased risk of infection	+	+	–	+	–
Increased risk of malignancy	+	+	–	+	–
Hyperglycemia	+	+	–	+	+
Bone marrow suppression	+	+	+	+	–
Nephrotoxicity	+	+	–	+	–
Hyperkalemia, hypomagnesemia	+	+	–	–	–
Hypertension	+	+	–	–	+
Dermatologic					
Alopecia	+	–	–	+	+/–
Rash	+	+	+	+	+
Gastrointestinal					
Gingival hypertrophy	+	+	–	–	–
Abdominal pain	+	–	–	+	+
Diarrhea, nausea, emesis	+	+	+	+	+
Hepatotoxicity	+	+	+	+	+
Neurologic					
Headache	+	+	–	+	+
Tremor	+	+	–	–	–
Seizures	+	+	–	–	–

*Does not include adverse effects rarely encountered with a medication.

Table 126.4. Adverse Effects of Immunosuppressive Agents[a]

Tacrolimus (Prograf®/FK506)

Tacrolimus is a potent immunosuppressive agent that, in the last 5 years, has been established as an effective drug for both primary immunosuppression after pediatric liver transplantation and as rescue therapy for the management of refractory and chronic rejection. An analysis of graft survival in pediatric liver transplant recipients showed 4-year graft survival of 86% in those patients receiving tacrolimus-based immunosuppression compared with 66% in those patients receiving cyclosporine. Tacrolimus in combination with prednisone alone has replaced cyclosporine in many liver transplant immunosuppressive regimens. Presently, tacrolimus may be used for induction and maintenance immunosuppression, usually in combination with steroids, which are often subsequently successfully withdrawn. Tacrolimus was developed as a macrolide antibiotic but functions in immunosuppression similarly to cyclosporine. It inhibits IL-2, interferon-g, and IL-3 production, transferrin and IL-2 receptor expression, mixed lymphocyte reactions, and cytotoxic T-cell generation.

Tacrolimus is administered at initial oral dosages of 0.1 to 0.3 mg/kg per day divided in two doses, with the dosage adjusted to maintain target whole blood trough levels. The target level varies from 5 to 20 ng/mL, depending on the organ transplanted, the transplant center, and the time since the transplant. Higher levels are used in the early post-transplant period and to treat acute rejection. Unlike cyclosporine, the oral absorption of tacrolimus is not dependent on solubilization by bile acids. The absorption of tacrolimus is enhanced on an empty stomach. Like cyclosporine, tacrolimus is primarily metabolized in the liver through the cytochrome P-450 system and its metabolism can be altered by concurrent administration of a number of medications (Table 126.3). Therefore, in the setting of liver dysfunction, decreased excretion may occur, which may lead to toxic levels of the drug.

The adverse effects of tacrolimus are similar to those associated with cyclosporine and include nephrotoxicity, hypertension, renal tubular acidosis, neurotoxicity, and hyperglycemia (Table 126.4). Acute nephrotoxicity and neurotoxicity (in the form of headaches, tremors, and seizures) are as frequent and severe as those seen with cyclosporine and are generally controllable with dosage reduction. Hyperglycemia may require therapy with insulin and does not appear to be dose dependent. Unlike cyclosporine, hirsutism and gingival hypertrophy are not a problem with this agent.

Corticosteroids

Corticosteroids were the first immunosuppressive agents used in solid organ transplantation. They are potent immunosuppressive and anti-inflammatory agents. Low-dose prednisone is a key component of mainstay post-transplant immunotherapy, whereas larger doses of both prednisone and methylprednisolone are often used as the first-line treatment for acute allograft rejection. Corticosteroids decrease inflammatory response by preventing inflammatory cell chemotaxis and recruitment and inhibition of extravascular migration of cells and plasma protein via microvascular constriction. In addition, steroids inhibit macrophage secretion of IL-1. At high dosages, steroids are lympholytic. Steroid activity is primarily inactivated by liver metabolism through reduction and conjugation with urinary excretion. Thus, changes in the liver function may significantly influence activation, metabolism, and elimination of the steroids.

The dosages of prednisone vary from 0.1 to 2 mg/kg per day, with higher dosages used in the early post-transplant period, in younger children, and in recipients of renal transplants. Some patients are treated eventually on an alternate day schedule or have steroids withdrawn entirely.

The adverse effects of corticosteroids are diverse and include both short-term and long-term toxicities. The most common short-term effects include sodium and fluid retention, hypertension, hyperglycemia, poor wound healing, gastritis, and central nervous system (CNS) changes (depression, euphoria, or rarely psychosis). The adverse effects of chronic steroid treatment include Cushing's features with truncal obesity, acne, striae, dermal atrophy, and hirsutism. Additional consequences include osteoporosis and aseptic necrosis of bone (particularly of the vertebral bodies and the femoral heads), glucose intolerance, cataracts, and growth retardation. The effect of steroid therapy on the immune system may render the transplant recipient susceptible to lethal bacterial, viral, or fungal infections.

Azathioprine (Imuran®/AZA)

Azathioprine (AZA) has been a key component of antirejection immunotherapy for more than 30 years. In many programs, it is used in a triple drug regimen with cyclosporine and prednisone. AZA is an antimetabolite that inhibits purine nucleotide synthesis and metabolism. The alteration in RNA synthesis caused by the drug prevents mitosis and proliferation in rapidly dividing cells such as B and T lymphocytes. AZA is administered intravenously or orally at dosages of 1 to 2 mg/kg per day as a single daily dose. The metabolites of AZA are primarily excreted by the kidneys, necessitating a dosage reduction patients with in renal failure to avoid increased toxicity.

The most common adverse effect of AZA therapy is myelosuppression, especially granulocytopenia with an increase risk of infection. The myelosuppression is dose-dependent and generally occurs within 1 to 2 weeks after the initiation of therapy. Monitoring of the peripheral blood counts, particularly during the early stages of therapy, is critical. Idiosyncratic reactions, such as fever, hepatotoxicity, and pancreatitis, occur but are relatively rare. Toxicity may occur during periods of renal insufficiency from diminished metabolite elimination and the increased sensitivity noted in uremic patients. AZA may also contribute to the risks of infection and late neoplasia in transplant recipients.

Mycophenolate Mofetil (MMF/Cellcept®)

The active metabolite of mycophenolate mofetil (MMF), mycophenolic acid, is a potent inhibitor of lymphocyte proliferation. It may also interfere with the binding of lymphocytes to activated endothelial cells, decreasing the migration of lymphocytes into the allograft. MMF may also inhibit arterial smooth muscle proliferation and thereby decrease the arteriopathy associated with chronic rejection. MMF is typically used in combination with cyclosporine and prednisone, or less commonly added to tacrolimus and prednisone as rescue therapy for rejection. The usual pediatric dosage is 600 mg/m^2 body surface area (BSA) per day administered in two divided doses or 20 mg/kg per day in two divided doses up to an adult dose of 1 g twice daily. The absorption is decreased by concurrent administration with antacids, and the blood level may be decreased if cholestyramine is also taken. As is the case for other immunosuppressive agents, MMF treatment increases the risk of serious infections and malignancy. GI complaints, including esophagitis, gastritis, diarrhea, and emesis, are common. These symptoms may be ameliorated with treatment with an H_2 blocker or proton-pump inhibitor (e.g., omeprazole). Bone marrow suppression, including leukopenia, anemia, and thrombocytopenia, is dose related and may necessitate a decrease in dosage. Isolated anemia associated with MMF treatment is more common in patients with decreased renal function. Adverse fetal effects have been reported in animal studies, but no studies have been conducted in pregnant humans. Therefore, MMF should be used with caution during pregnancy.

INFECTION PROPHYLAXIS

The immunosuppressed transplant recipient is susceptible to a wide variety of infections (Fig. 126.3). Therefore, antimicrobial prophylaxis is used to decrease the risk of serious infection with the most common pathogens (e.g., *Pneumocystis, Candida,* CMV, herpes simplex virus [HSV], and urinary pathogens). The agents used and the dosing schedules vary with the organ transplanted, the transplant center, and the infection risk of the individual patient.

Most programs consider *Pneumocystis carinii* to be an important pathogen. Prophylactic regimens may include trimethoprim–sulfamethoxazole as a single dose (at half the therapeutic dose) daily or full dose (10 mg/kg per day trimethoprim) in two divided doses three times per week. Other regimens include monthly intravenous (IV) pentamidine and monthly aerosolized pentamidine. Trimethioprim–sulfamethoxisole may also serve as a urinary tract infection suppressant in renal transplant recipients. These patients are also at increased risk for oral and esophageal candidiasis as a result of steroid and antibiotic treatment. Therefore, oral nystatin is given for 6 to 12 months after the transplant.

Both primary and reactivated CMV infections can be serious threats to transplant recipients. The risk is particularly high in CMV-negative recipients who have received CMV-positive allografts. CMV infections may be asymptomatic or may present with symptoms such as fever, leukopenia, pulmonary disease, hepatic dysfunction, intestinal bleeding, or diarrhea. Prophylactic therapy for CMV infections may include CMV hyperimmunoglobulin, oral acyclovir, and IV or oral ganciclovir for the first 4 to 6 months after the transplant and during treatment with monoclonal or polyclonal antibodies. Some programs monitor the presence of CMV antigen in the blood and treat intermittently with IV ganciclovir.

ANTIHYPERTENSIVE MEDICATIONS

Hypertension is a common problem in pediatric transplant recipients, with an incidence of at least 50% in liver transplant recipients and 85% in renal transplant recipients in the early post-transplant period. A number of factors may contribute to post-transplant hypertension, including medications (cyclosporine, tacrolimus, and corticosteroids), acute or chronic renal allograft rejection, renal artery stenosis, high renin production by native kidneys, and recurrence of primary renal disease.

Several agents are effective in treating hypertension in this group of patients. Long-acting agents that can be dosed once or twice a day are optimal. Calcium channel antagonists may be optimal because they can counteract some of the intrarenal vasoconstriction caused by cyclosporine or tacrolimus and ameliorate their nephrotoxicity. However, they may also increase gingival hypertrophy. Long-acting preparations, such as amlodipine or nifedipine, provide the most consistent blood pressure control and are dosed daily or twice a day. The amlodipine tablet, which maintains its pharmacokinetic profile when dissolved in water, may be administered to children more than 1 year of age. The total oral daily dose of the two formulations is the same (0.25 to 1.0 mg/kg). Other vasodilators, such as minoxidil, may also be used. Diuretic treatment is usually necessary in concert with the vasodilator. The most commonly used agent is furosemide (1 to 5 mg/kg per day in one to two divided doses). b-Adrenergic antagonists are useful second-line agents. They are also effective when added to a vasodilator. Several selective b-blocker drugs have a greater effect on cardiac b-receptors than on bronchial b_2-receptors. One such agent is atenolol, which is administered daily or twice a day (1 to 2 mg/kg per day).

Angiotensin-converting enzyme inhibitors are used in a number of programs. In renal transplant recipients, they may be helpful if renin-mediated hypertension caused by high levels of renin secretion by the native kidneys is present. However, these agents should be used with caution in renal transplant recipients because they can cause profound graft dysfunction if renal artery stenosis is present and can exacerbate graft dysfunction in the presence of cyclosporine or tacrolimus therapy and volume depletion. The available agents vary in half-life; therefore, the dosing interval is shortest with captopril (0.3 to 5 mg/kg per day divided into two to three doses per day) and longer with enalapril (starting dosage 0.08 mg/kg per dose once or twice a day) or ramipril (once a day).

Severe hypertension is most commonly a complication of renal transplantation. When a child presents to the ED with severe hypertension, it may be important to rapidly reduce the blood pressure in a symptomatic patient to avoid the sequelae of hypertensive encephalopathy. In an emergency setting, sublingual nifedipine (0.25 to 0.5 mg/kg) is often effective. It can be repeated after 20 minutes if blood pressure is not adequately reduced. The dose is difficult to accurately administer to small children, but the capsule can be punctured and the liquid aspirated into a tuberculin syringe and the appropriate amount (0.035 mL corresponds to about 1 mg) administered. After the nifedipine is administered, IV access should be obtained for further treatment. Sublingual nifedipine should be used with caution to avoid too rapid a drop in blood pressure or frank hypotension. Hydralazine can be an effective agent administered intravenously or intramuscularly (0.1 to 0.2 mg/kg). The dose can be doubled and repeated every 20 minutes. A more effective agent is IV labetalol, which can be administered as a bolus (0.25 mg/kg) and repeated in escalating doses (0.5 to 1.0 mg/kg). Blood pressure control may be maintained by the constant infusion of labetalol at 1 to 3 mg/kg per hour. If this agent is inadequate, the blood pressure can be controlled with infusion of nitroprusside (0.5 to 3.0 µg/kg per minute). A child who requires an infusion is typically cared for in the intensive care unit.

IMMUNIZATIONS

When possible, children should be fully immunized before transplant with all age-appropriate vaccines, including vaccines for varicella, pneumococci, and hepatitis B. In general, live virus vaccines are contraindicated after transplant, and oral polio, measles, mumps, and rubella vaccines should be avoided. Limited experience exists with the use of the varicella vaccine after transplant. Immune response to vaccines administered after transplant may not be as complete as they might be in a non-immunosuppressed (non-transplant) patient.

GENERAL PRINCIPLES OF MANAGEMENT

Hospitalizations in the first 6 months after transplant are common. As an example, 60% of recipients of cadaver donor renal allografts and 50% of living donor recipients are hospitalized in the first 6 months after transplant. The most common causes for hospitalization include treatment of acute rejection, viral and bacterial infections, and treatment of hypertension. These reasons are also the most likely for presentation to the ED. The encounter in the ED with the renal or liver transplant recipient need not provoke a sense of uneasiness in the nontransplant physician if the following pertinent principles of evaluation and treatment are applied:

1. The transplant center should be regarded as a valuable source of information and assistance, and dialog with the center should be recognized as critically important to patient care. The transplant center should be contacted during the ED visit and should be contacted immediately if the patient has abnormal laboratory tests. No changes in immunosuppressive medication should be undertaken independent of input from the transplant center. A copy of the ED record should also be forwarded to the transplant center.
2. If the transplant patient is critically ill or hemodynamically unstable, the patient should be stabilized in the usual manner with attention to the ABCs of resuscitation. The primary focus should be on the patient and not on graft survival. The same life-saving interventions used for any critically ill individual should be used in these acute situations, and specific considerations regarding the graft should be secondary. A critically ill transplant patient should be stabilized and then transported to a transplant center.
3. All symptoms require thorough evaluation because a patient receiving immunosuppressive medications may have blunted or atypical presentations of severe disease, particularly when infection is involved.
4. Conditions that impair the patient's ability to take or absorb medications (e.g., vomiting, diarrhea), necessitate hospital admission for parenteral administration of the immunosuppressive medications and careful monitoring. Simple conditions, such as gastritis with vomiting or gastroenteritis with diarrhea, may lead to significantly diminished cyclosporine levels and the potential for rejection and graft loss.
5. Children and adolescents with renal transplants can become dehydrated more readily than healthy children with what appear to be trivial illnesses. The volume of the urine output cannot be used as an indication of adequate hydration in renal transplant recipients. Maximal urinary concentrating ability may not be present; therefore, the patient more readily becomes dehydrated. Volume depletion increases the acute nephrotoxicity potential of cyclosporine and tacrolimus, resulting in an increased serum creatinine. Therefore, renal transplant recipients often require IV fluids to maintain hydration. These patients also require IV hydration if the serum creatinine is increased above baseline levels and if ability to maintain oral intake is in question. Home trials of oral rehydration therapy are not appropriate for these children.
6. Fever in the immunocompromised patient requires aggressive investigation because it could be a manifestation of a wide spectrum of disease from severe opportunistic infection to acute graft rejection. If the patient is obviously septic, blood cultures should be drawn and broad-spectrum antibiotics administered expeditiously. Headache, seizures, or neurologic changes in the setting of a fever are indications for a lumbar puncture with cerebrospinal fluid cell count, and comprehensive stains and culture for bacteria, viruses, fungi, and acid-fast organisms, to be performed as a part of the primary evaluation. The transplant center should be notified of the patient's condition as soon as possible.
7. Abdominal pain may represent a surgical emergency even when mild in nature. Steroid therapy may blunt the inflammatory response in a transplant patient such that visceral perforation, urine or bile leak, or infectious peritonitis may not be accompanied by classic peritoneal signs. All complaints therefore warrant full investigation.
8. Renal or liver allograft dysfunction may be secondary to causes other than rejection. Problems such as bile duct or ureter obstruction, infection, drug toxicity, and rejection may mimic each other but require different therapy. In addition, volume depletion can significantly increase the serum creatinine level in renal transplant recipients. Therefore, these patients must be fully evaluated in a transplant center where they may have prompt immunosuppressive drug levels, graft-specific imaging studies, and possibly graft biopsy before antirejection therapy is initiated.
9. Cyclosporine and tacrolimus have several drug interactions (listed in Table 126.3) that alter patients' metabolism and may lead to dangerously high or low levels. A number of medications can act synergistically with these agents to increase their nephrotoxic effects. The effects of these medications on the levels of cyclosporine and tacrolimus are not absolute contraindications to their use, but an alternative medication should be used if possible. Levels of cyclosporine or tacrolimus should be carefully monitored.
10. Upper extremity cut-down IV access should be avoided in the renal transplant patient because these veins may be required for future hemodialysis access. Established arteriovenous fistulas should not be used for IV access, blood pressure monitoring, or blood drawing.
11. Non-steroidal anti-inflammatory agents should be avoided in renal and liver transplant recipients, if possible. These agents can increase the nephrotoxicity of cyclosporine and tacrolimus.

INFECTIONS

In general, the onset and type of infection and the responsible agent is related to the intensity and type of immunosuppression, and the time since the transplant procedure (Fig. 126.3). In the first few weeks after transplant, the most common infections are urinary tract infection and wound infection with bacterial pathogens. After this period, the incidence of opportunistic infections increases.

Pneumocystis carinii *Pneumonitis*

P. carinii pneumonia (PCP) is becoming increasingly uncommon in transplant patients as a result of routine prophylaxis. However, although most common within the second to sixth months after transplant, infection may occur at any time. In most cases, a patient with PCP presents with dry cough, dyspnea, diffuse interstitial infiltrates, and hypoxemia. Suspicion of infection with this pathogen necessitates a diagnostic intervention such as bronchoscopy with a bronchoalveolar lavage. The isolation of pneumocysts on the silver stain of the lavage fluid confirms the diagnosis. Infection is treated with IV trimethoprim–sulfamethoxazole (15 to 20 mg/kg per day of trimethoprim intravenously in three to four divided

doses) or pentamidine (4 mg/kg intravenously in a single daily dose).

Cytomegalovirus Infection

CMV infection is the single most important infection in the transplant recipient. CMV disease may represent 1) a primary infection in a CMV non-immune recipient who acquires the infection from the allograft or transfusion of CMV-positive blood products; 2) reactivation of latent virus as a consequence of immunosuppression (most recipients in this situation show some evidence of CMV infection, although only 20% become symptomatic); and 3) superinfection of a CMV-immune patient with a strain of CMV of donor or environmental origin. The course of CMV infection is influenced by the type and intensity of immunosuppression the patient has received. Steroids alone do not put the patient at great risk of reactivation of CMV disease. However, AZA and particularly OKT3 and ALG have the highest risk of reactivating CMV disease. CMV infection in an immunocompetent host is usually asymptomatic, whereas in a transplant patient, it may be devastating. CMV causes a syndrome characterized by anorexia, malaise, and myalgias and arthralgias usually heralded by the initial presentation of fever. These symptoms may be accompanied by an atypical leukocytosis and usually occur within 2 to 3 months of transplant but may occur at any time. In approximately one-third of febrile patients with CMV infection, pulmonary disease develops and may rapidly progress to adult respiratory distress syndrome (ARDS) and death. CMV may also cause localized disease, which in transplant patients may localize to the renal or hepatic graft (causing a picture of hepatitis) and mimic rejection. Another specific site of CMV involvement is the mucosal lining of the GI tract where the virus may cause ulceration and massive GI tract hemorrhage. The effects of the organ-specific infection are amplified by several of the systemic effects of the virus, including thrombocytopenia and leukopenia caused by bone marrow suppression, and impairment in alveolar macrophage function and cell-mediated immunity, which predispose the patient to further opportunistic infections by other viral agents or *Pneumocystis.* Diagnosis of CMV is made by culture, serology, or antigen detection. Treatment of active CMV includes lowering of the immunosuppression and administering IV ganciclovir (5 to 10 mg/kg per 24 hours in two divided doses).

Epstein-Barr Virus and Posttransplant Lymphoproliferative Disease

EBV is a ubiquitous virus and, similar to CMV, is associated with hepatitis as well as a more systemic illness. Acute EBV infection presents as a mononucleosis-like syndrome with diffuse B-cell hyperplasia. However, of particular concern is the risk of post-transplant lymphoproliferative disorder (LPD). This disorder is most common in children who are not immune to EBV at the time of transplant and develop EBV infection. The overall incidence in pediatric renal transplant recipients is less than 1%, but the risk increases with increasing exposure to immunosuppressive medications such as OKT3 and tacrolimus-based immunosuppression. LPD can present as early as 1 month after transplant or many years after transplantation. A high index of suspicion must be maintained because LPD can present at any lymph tissue site (e.g., tonsils, GI tract, cervical nodes, CNS). Patients may present with fever, upper airway obstruction, GI bleeding, or intestinal obstruction or perforation. In all cases, the first line of treatment is to discontinue immunosuppressive agents (prednisone may be maintained) even at the risk of possible rejection. Additional treatment with acyclovir or ganciclovir is added at most centers. In many cases, the LPD responds to discontinuation of immunosuppressive medications without ensuing allograft rejection. In other cases, cytotoxic chemotherapeutic agents, such as cyclophosphamide, are added.

Herpes Simplex Virus

Reactivation of latent HSV infection is common in transplant recipients. Patients presenting with an oral or genital lesion resembling herpes should have a scraping for immunofluorescence performed on the lesion. If herpes simplex is present, oral acyclovir is used for minor lesions without systemic symptoms. Extensive lesions, fever, or other systemic symptoms require IV acyclovir treatment (750 to 1500 mg/m^2 per 24 hours intravenously in three divided doses) to prevent disseminated herpes infection.

Varicella

Varicella is a highly contagious pathogen that is common among school children. In a transplant patient who is immunocompromised, it may become disseminated disease spreading to the liver, lungs, and CNS. If a patient has been exposed only to varicella (household contact or played in the same room with an infected individual), the patient should receive varicella-zoster immunoglobulin (VZIG). VZIG (125 units or one vial per 10 mg) should be given within 96 hours of exposure, but the sooner it is administered, the more efficacious it will be. If the transplant patient is diagnosed with varicella, he or she should be admitted to the hospital for IV acyclovir therapy (1500 mg/m^2 per 24 hours in three divided doses) and a sharp reduction in steroid dosage. Herpes zoster may occur in as many as 5 to 10% of adult transplant patients, representing reactivation of old varicella infection, although it rarely disseminates. Acyclovir may hasten resolution of lesions, but no change in immunosuppressive regimen is usually needed.

Adenovirus

This viral infection occurs in up to 7% of pediatric transplant patients and should be considered when the patient presents with high fever and liver and/or pulmonary dysfunction with or without diarrhea. A nasopharyngeal swab and stool for viral culture should be sent to screen for the virus in the febrile posttransplant patient.

Fungal and Nocardial Infections

Fungal infections in the posttransplant patient can have a variety of clinical presentations. They may present as a subacute respiratory illness with local or disseminated findings on chest radiograph. Alternatively, the patient may have a systemic illness with nonspecific symptoms of malaise and fever that may be acute or chronic. Fungal infections may also present with metastatic disease. Examples are the 20 to 30% of patients with cryptococcal infection who demonstrate skin lesions weeks or months before the development of CNS lesions and the 10 to 15% of patients with disseminated *Candida* infection who have skin lesions early on in its course. Similarly, *Nocardia* and *Mucor* species may show early skin lesions before evidence of more serious deep-seated infection presents itself. Another common fungal infection is

Candida esophagitis, which may present with dysphagia or odynophagia. CMV and HSV infection of the esophagus may also occur with similar symptoms.

Fungal infections generally do not occur in the first month after transplant but rather in the subsequent months. Between the first and sixth months, *Candida* species are the major fungal pathogens. Infections with *Aspergillus* are less common but are associated with high mortality.

Fungal infection of the CNS may be difficult to assess because the classic signs of CNS infection, such as meningismus, are often absent in immunosuppressed patients. The common presentation of headache, often without fever, may be the only indication that a CNS infection exists and warrants thorough neurological evaluation, lumbar puncture with fungal stains and cultures, and possibly an imaging study of the brain, preferably magnetic resonance imaging. Acute or subacute meningitis is most commonly caused by *Listeria monocytogenes,* whereas chronic meningitis is most often caused by *Cryptococcus neoformans* and focal brain abscess is often indicative of *Aspergillus* infection.

APPROACH TO FEVER

Fever is the most common reason for the pediatric transplant recipient to seek evaluation in an ED. A fever in a transplant patient may be a manifestation of any number of infections or processes from acute graft rejection to systemic sepsis. Assessing these patients is challenging because their immunosuppression may mask many of the typical physical findings associated with their disease process (e.g., peritoneal signs). All fevers and new symptoms in these patients require investigation. If the patient appears well, has normal laboratory evaluation and chest radiograph (if presenting with respiratory symptoms), and has an obvious minor source of infection, such as otitis media or an upper respiratory tract infection, the patient may be sent home with appropriate therapy. As previously mentioned, the transplant center should always be notified at the time of the visit that the patient had been seen and what the diagnosis was. Close outpatient follow-up is mandatory within 48 hours.

Patient assessment should include 1) a careful physical examination that includes examination of all wounds; 2) pulse oximetry, chest radiograph, and arterial blood gases if the patient has cough, dyspnea, tachypnea, or other signs of hypoxia; 3) a screen of graft function (liver function tests and creatinine) in addition to other routine laboratory tests, such as complete blood count and differential, disseminated intravascular coagulation panel, and prothrombin time/partial thromboplastin time, if the patient appears septic; and 4) a blood culture for bacterial and viral pathogens, blood buffy coat CMV antigen assay by rapid assay technique, urine for urinalysis, rapid CMV assay, and viral culture of the urine and a standard urine culture if the child has a fever or appears ill. Because some patients may have a central venous catheter, line sepsis should always be considered in the differential diagnosis.

In the liver transplant patient, elevated liver aminotransferases may be a sign of rejection, arterial or venous thrombosis of the graft, or even biliary stricture and obstruction with resultant cholangitis. The initial study required in this case is an ultrasound examination with Doppler flow study to view arterial and venous blood flow to the graft and to assess the biliary tree for evidence of dilation, which suggests obstruction. If obstruction is suspected from the ultrasound, percutaneous transhepatic cholangiography is usually necessary to image the biliary tree and biliary–enteric anastomosis. Recent progress has been made with magnetic resonance cholangiography to obtain clear non-invasive imaging of the biliary tree. The patient is given broad-spectrum antibiotic coverage for the common biliary pathogens (e.g., Gram-negative enteric organisms). Ampicillin (200 mg/kg per day) and cefotaxime (100 mg/kg per day) are usually adequate. If the ultrasound is normal and no source for the fever or increased liver function tests is found, the patient requires admission and liver biopsy to rule out rejection or viral infection. If the ultrasound is abnormal, the situation could require surgical revision of the biliary anastomosis or biliary stent placement (Fig. 126.2), either by the interventional radiologists or by open procedure by the transplant surgeons.

In the renal transplant recipient, particular attention should be paid to the graft site because fever, tenderness over the transplanted kidney, poorly controlled hypertension, diminished urinary output, and recent weight gain may all be signs of rejection. A rise in the blood urea nitrogen and creatinine levels also suggests rejection. However, ascending urinary tract infection, infected perinephric collections (lymphocele, seroma, and urinoma), ureteral stenosis or obstruction, and renal vascular thrombosis may also present with similar findings. These emergencies require in-hospital workup, beginning with urine culture, blood culture, Doppler and conventional ultrasound, nuclear renal scan, antegrade or retrograde pyelogram when indicated, and possible renal biopsy.

GASTROINTESTINAL EMERGENCIES

Transplanted patients may develop GI complications that are secondary to many different causes, ranging from infection to post-surgical complications. Perforation and bleeding may occur secondary to necrotic lymph nodes from LPD or because of small bowel ulcerations from CMV infection. Intussusception or luminal obstruction may occur secondary to enlarged lymph nodes from post-transplant lymphoproliferative disease (PTLD), and peritonitis may be caused by any of the aforementioned conditions, including bile leak from an anastomotic breakdown or bile duct ischemia, particularly in the early post-transplant period. Severe variceal bleeding is also seen in the liver transplant patient with portal venous thrombosis and consequent pre-hepatic portal hypertension. Pancreatitis may be seen as a complication of taking AZA.

The approach to these patients in the ED is the same as for the non-immunocompromised patient: 1) fluid resuscitation; 2) nasogastric intubation 3) blood products as needed; and 4) frequent monitoring of vital signs while transfer to the transplant center is arranged. In the evaluation of abdominal pain, a number of processes should be considered. Tacrolimus toxicity can produce complaints of severe epigastric pain or diffuse abdominal pain. MMF can cause gastritis, esophagitis, and diarrhea. In addition to viral involvement of the GI tract, infectious processes that are most common in transplant recipients include *Clostridium difficile* colitis and candidal esophagitis.

MISCELLANEOUS EMERGENCIES

Transplant recipients may also experience many of the usual childhood emergencies, including hydroceles and herniae. Scrotal edema on the side of the renal transplant is a common finding and is generally self-limiting and not a problem. Other possible complications include deep venous thrombosis on the side of the transplanted kidney, which is confirmed by noninvasive venous imaging and managed with anticoagulant therapy. Less common complications are suture-line disruption with subsequent “blow-out” or aneurysmal rupture at the anastomotic site in both liver and renal transplants.

Careful attention should be paid to a transplant recipient with a headache. In addition to usual causes of headaches, meningitis, pseudotumor cerebri, malignancy, and malignant hypertension must be considered. In a patient with papilledema, the most likely cause is pseudotumor cerebri, which is a complication of steroid treatment and may be related to changes in steroid dose.

CONCLUSION

Emergencies in the pediatric transplant patient are common, consequent to the complex nature of the procedures and the depressed state of the immune system. With the ever-increasing number of organ transplants being performed each year in children, more of these patients are likely to be seen in the ED. Therefore, a working knowledge of the nature of these procedures and the common emergency situations that may arise in this patient group is essential to proper assessment and management of these patients' problems.

Suggested Readings

Allen KA, Markin RS, Rennard SI, et al. Bronchoalveolar lavage in liver transplant patients. Acta Cytol 1989;33:539–543.

Bock GH, Sullivan EK, Miller D. Cytomegalovirus infections following renal transplantation--effects of antiviral prophylaxis: a report of the North American Pediatric Renal Transplant Cooperative Study (NAPRTCS). Pediatr Nephrol 1997;11:665–671.

Breinig MK, Zitelli B, Starzl TE, Ho M. Epstein-Barr virus, cytomegalovirus, and other viral infections in children after transplantation. J Infect Dis 1987;156:273–279.

Busuttil RW, Seu P, Millis JM, et al. Liver transplantation in children. Ann Surg 1991;213:48–57.

Cacciarelli TV, Esquivel CD, Cox KL, et al. Oral tacrolimus (FK506) induction therapy in pediatric orthotopic liver transplantation. Transplantation 1996;61:1188–1192.

Castelas AM, Sabate I, Grino JM, et al. Cyclosporine-drug interactions. Transplant Proc 1988;20(Suppl 6):66–69.

Chou S. Newer methods for diagnosis of cytomegalovirus infection. Rev Infect Dis 1990;12(Suppl 7):S727–736.

Davis ID, Bunchman TE, Grimm PC, et al. Pediatric renal transplantation--indications and special considerations. A position paper from the Pediatric Committee of the American Society of Transplant Physicians. Pediatr Transplant 1998;2:117–129.

Dreno B, Mansat E, Legoux B, Litoux P. Skin cancers in transplant patients. Adv Nephrol Necker Hosp 1997;27:377–389.

Drummer JS. Pneumocystis carinii infections in transplant recipients. Semin Respir Infect 1990;5:50–57.

Dunn DL, Mayoral JL, Gillingham KJ, et al. Treatment of invasive cytomegalovirus disease in solid organ transplant patients with ganciclovir. Transplantation 1991;51:98–106.

Feld LG, Stablein D, Fivush B, et al. Renal transplantation in children from 1987–1996: The 1996 Annual Report of the North American Pediatric Renal Transplant Cooperative Study. Pediatr Transplantation 1997; 1:146–162.

Fine RN, Salusky IB, Ettenger RB. The therapeutic approach to the infant, child, and adolescent with end-stage renal disease. Pediatr Clin North Am 1987;34:789–801.

Gershon AA. Immunizations for pediatric transplant patients. Kidney Int (Suppl) 1993;43:S87–90.

Giacchino R, Marcellini M, Timatilli A, et al. Varicella vaccine in children requiring renal or hepatic transplantation. Transplantation 1995;60:1055–1056.

Goldstein G, Kremer AB, Barnes L, Hirsch RL. OKT3 monoclonal antibody reversal of renal and hepatic rejection in pediatric patients. J Pediatr 1987;111:1046–1050.

Hecht DW, Snydman DR, Crumpacker CS, et al. Ganciclovir for treatment of renal transplant-associated primary cytomegalovirus pneumonia. J Infect Dis 1988;157:187–190.

Ho M, Jaffe R, Miller G, et al. The frequency of Epstein-Barr virus infection and associated lymphoproliferative syndrome after transplantation and its manifestations in children. Transplantation 1988;45:719–727.

Hughes WT, Rivera GK, Schell MJ, et al. Successful intermittent chemoprophylaxis for *Pneumocystis carinii* pneumonitis. N Engl J Med 1987;316:1627–1632.

Johnson PC, Hogg KM, Sarosi GA, et al. The rapid diagnosis of pulmonary infections in solid organ transplant recipients. Semin Respir Infect 1990;5:2–9.

June CH, Thompson CB, Kennedy MS, et al. Profound hypomagnesemia and renal magnesium wasting associated with the use of cyclosporine for marrow transplantation. Transplantation 1985;39:620–624.

Kahan BD. Cyclosporine. N Engl J Med 1989;321:1725–1738.

Kahan BD. Pharmacokinetics and pharmacodynamics of cyclosporine. Transplant Proc 1989;21(Suppl 1):9–15.

King SM, Petric M, Superina R, et al. Cytomegalovirus infections in pediatric liver transplantation. Am J Dis Child 1990;144:1307–1310.

Lautenschlager I, Suni J, Ahonen J, et al. Rapid diagnosis of cytomegalovirus infection in renal transplant recipients. Transplant Proc 1988;

20:408–409.

Lee H, Vacanti JP. Liver transplantation and its long-term management in children. Pediatr Clin North Am 1996;43:99–124.

Lerut J, Gordon RD, Iwatsuki S, et al. Biliary tract complications in human orthotopic liver transplantation. Transplantation 1987;43:47–51.

Lynfield R, Herrin JT, Rubin RH. Varicella in pediatric renal transplant recipients. Pediatrics 1992;90:216–220.

McDiarmid SV. Renal function in pediatric liver transplant patients. Kidney Int Suppl 1996;53:S77–84.

McDiarmid SV, Busuttil RW, Ascher NL, et al. FK506 (tacrolimus) compared with cyclosporine for primary immunosuppression after pediatric liver transplantation. Results from the U.S. Multicenter Trial. Transplantation 1995;59:530–536.

McDiarmid SV, Millis MJ, Olthoff KM, So SK. Indications for pediatric liver transplantation. Pediatr Transplantation 1998;2:106–116.

McGregor RS, Zitelli BJ, Urbach AH, et al. Varicella in pediatric orthotopic liver transplant recipients. Pediatrics 1989;83:256–261.

McKee M, Mattei P, Schwarz K, et al. Steroid withdrawal in tacrolimus (FK506) treated pediatric liver transplant recipients. J Pediatr Surg 1997;32:973–975.

Megison SM, Andrews WS. Combination therapy with ganciclovir and IV IgG for cytomegalovirus infections in pediatric liver transplant recipients. Transplantation 1991;52:151–154.

Michaels MG, Green M, Wald ER, et al. Adenovirus infection in pediatric liver transplant recipients. J Infect Dis 1992;165:170–174.

Myers BD, Ross J, Newton L, et al. Cyclosporine-associated chronic nephropathy. N Engl J Med 1984;311:699–705.

Patel R, Snydman DR, Rubin RH, et al. Cytomegalovirus prophylaxis in solid organ transplant recipients. Transplantation 1996;61:1279–1289.

Paya CV. Fungal infections in solid organ transplantation. Clin Infect Dis 1993;16:677–688.

Paya CV, Hermans PE, Washington JA II, et al. Incidence, distribution, and outcome of episodes of infection in 100 orthotopic liver transplantations. Mayo Clin Proc 1989;64:555–564.

Penn I. Tumors after renal and cardiac transplantation. Hematol Oncol Clin North Am 1993;7:431–445.

Rubin RH. Infectious disease complications of renal transplantation. Kidney Int 1993;44:221–236.

Sakr M, Hassanein T, Gavaler J, et al. Cytomegalovirus infection of the upper gastrointestinal tract following liver transplantation: incidence, location, and severity in cyclosporine- and FK506-treated patients. Transplantation 1992;53:786–791.

Salibe F, Gugenheim J, Samuel D, et al. Incidence of cytomegalovirus infection and effects of cytomegalovirus immune globulin prophylaxis after orthotopic liver transplantation. Transplant Proc 1987;19: 4081–4082.

Salvatierra O, Tanney D, Mak R, et al. Pediatric renal transplantation and its challenges. Transpl Rev 1997;11:51–69.

Sokal EM. Quality of life after orthotopic liver transplantation in children. An overview of physical, psychological and social outcome. Eur J Pediatr 1995;154:171–175.

Tzakis AG, Gordon RD, Shaw BW, et al. Clinical presentation of hepatic artery thrombosis after liver transplantation in the cyclosporine era. Transplantation 1985;40:667–671.

Whitington PF, Kehrer BH, Whitington SH, et al. The effect of biliary enteroenterostomy on the pharmacokinetics of enterally administered cyclosporine in rats. Hepatology 1989;9:393–397.

Wozney P, Zaiko AB, Bron KM, et al. Vascular complications after liver transplantation: a 5-year experience. Am J Roentgenol 1986;147: 657–663.

Zitelli BJ, Gartner JC, Malatack JJ, et al. Pediatric liver transplantation: patient evaluation and selection, infectious complications, and life-style after transplantation. Transplant Proc 1987;19:3309–3316.

CHAPTER 127

Approach to the Care of the Technology-Dependent Child

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Up to almost one-quarter of the visits to a pediatric emergency department (ED) are for complaints associated with chronic illness. Many children with chronic illness have indwelling hardware, such as cerebrospinal fluid (CSF) shunts, venous catheters, and gastrostomy tubes. Medical technology has enabled these children, who in the past would have required specialized inpatient or intensive care, to thrive at home. Emergency physicians must be able to diagnose and treat the common problems associated with these new technologies and recognize when it is appropriate to consult other specialists familiar with these children.

Devices most commonly found in the pediatric population include CSF shunts, tracheostomy tubes, venous catheters, and percutaneous gastrointestinal (GI) and urologic catheters. The goal of this chapter is to familiarize the emergency clinician with the equipment and with the clinical manifestations and management of the problems related to these apparatuses. In addition, the clinician can advocate more effectively for the patients if he or she is aware of the emotional and social issues accompanying these patients and families.

APPROACH TO THE CARE OF THE TECHNOLOGY-DEPENDENT CHILD

The technology-dependent child who visits the ED may pose a challenge for the practitioner. Because of several factors, the evaluation of these children may, at times, seem overwhelming. These children often present with several pieces of equipment, the history can be difficult to obtain because of its inherent complexity, and a thorough physical examination may be impeded by the technology. When a common illness is superimposed on a chronic condition, the illness may appear more complex, misleading the examiner. In addition, the ED visit may have been necessitated by multiple reasons. The more involved the equipment and problems, the more challenging the situation becomes.

When a technology-dependent child arrives in the ED, contacting the primary care provider early on during the evaluation of the patient may be helpful. The primary care provider may be able to offer suggestions about the management of the child, potentially avoiding unnecessary tests and admission. In many situations, a home health nurse may accompany the patient and family to the ED and can be a valuable source of information.

In recent years, developments have led to the facilitation of medical history gathering in the ED. For example, “Pedistat” is a computerized program used in Rhode Island that allows families and physicians of children with special needs to provide information about their child, which is entered into the database in the state's children's hospital ED. The American College of Emergency Physicians (ACEP) now provides a data set for children with special health care needs that can be accessed at the time of the ED visit. When the aforementioned approaches are not immediately available, checking for a Medi-Alert bracelet, which provides a patient identification number that can be used to obtain information about the patient, is always prudent. By accessing the Medi-Alert hot-line, relevant medical information about the patient can be faxed rapidly to the ED for immediate use. Deriving an accurate history in these scenarios is imperative and greatly improves the quality of care administered.

When caring for the technology-dependent child in the ED, several important points emerge and should be used in approaching these children in the acute care setting (Table 127.1).

Common pediatric illnesses can afflict chronically ill children.
The presence of foreign bodies or hardware predisposes the patient to infection.
Families are the experts in their children's problems—rely on them for important information.
Consider altering the usual criteria for admission.

Table 127.1. Approach to the Technology-Dependent Child in the Emergency Department

First, *common things are common;* common pediatric illnesses may afflict these children as they do others. This point is always important to remember when evaluating a seemingly complicated child who presents with the routine signs and symptoms characteristic of typical childhood diseases. For example, a child with a CSF shunt may have vomiting caused by gastroenteritis.

Second, the presence of indwelling devices *predisposes the patient to infection.* When a child presents with symptoms associated with a specific piece of equipment, the clinician must be suspicious of infection of that equipment. For example, if a child with a tracheostomy presents with fever, cough, and increasing secretions, it is crucial to evaluate for the possibility of tracheitis. At the same time, the equipment has a tendency to become colonized with commensal organisms. Therefore, all bacterial growth does not indicate acute infection, and other sources of infection should be considered.

Above all, *families should be relied on* for important information because the parents or caretaker of technology-dependent children have become sophisticated in their knowledge of specific illnesses and equipment. This information becomes crucial when an acutely ill patient presents to the ED with several forms of technology and an involved medical history. Parents are sensitive to subtle changes in their children because they provide most of the home medical care. *Families are experts* and should play an integral role in the evaluation, management, and ultimate disposition of their child in the ED setting.

Children with chronic illnesses have a higher likelihood of being admitted to the hospital, resulting in longer lengths of stay in the ED. The practitioner should realize that the families of technology-dependent children often have sufficient equipment and trained personnel available in the home setting to care for an exacerbation of a chronic problem or an unrelated acute problem. For example, a family whose child has a chronic respiratory illness often has supplemental oxygen in the home and is facile with its use. Knowing that families of technology-dependent children are compliant and likely to return to the ED if their child's degree of illness exceeds the capabilities of the home care is reassuring. Thus, the practitioner should consider *altering the usual criteria for admission* in this specific population.

Having a technology-dependent child in the home creates a stressful situation for family members and other caretakers. A visit to the ED for an acute problem exacerbates this level of stress. These families may be more likely to question the diagnostic tests and therapies offered during the evaluation of their child because of their level of medical knowledge as well as the constant illness-related anxiety that intrudes upon their lives. The ED visit is more effective if the practitioner recognizes the psychosocial issues associated with this population of patients.

Tracheostomy Care

Background

Advances in neonatology and pediatric critical care medicine have enabled children to survive the complications of premature birth, congenital anomalies, and severe life-threatening illnesses. Yet a significant number of children are unable to be weaned immediately from respiratory support. As home care has become more widely recognized as an alternative to prolonged and costly hospitalization, the number of children managed at home with tracheostomies and mechanical ventilation has increased dramatically. Consequently, these children seek care more often in the ED when acute problems arise. To approach these situations calmly and systematically, the emergency physician should 1) appreciate the physiologic differences in a patient with chronic respiratory insufficiency (CRI); 2) be familiar with the equipment used in the care; and 3) understand the commonly encountered complications and their management.

Pathophysiology

In healthy people, respiration is maintained via a complex mechanism involving the alveolocapillary network, the diaphragm and intercostal musculature, and the central centers in the brainstem. Respiratory compromise results when one or more components of this mechanism are affected by disease. Approximately 65% of patients with CRI have bronchopulmonary dysplasia and congenital airway anomalies. Another 30% have neuromuscular disorders such as muscular dystrophy or spinal cord injuries. The remaining 5% require mechanical ventilation to overcome central disorders such as a brain tumor or Chiari malformation. Many children are successfully weaned from ventilator-assisted to independent breathing. Survival and decannulation rates depend on the nature and severity of the underlying disease.

Equipment

The complexity of the many tubes and attachments extending from the patient's airway can be overwhelming, especially

in the emergent situation. Familiarity with the equipment used in caring for a patient with a tracheostomy ensures the emergency physician's adept management of these situations. Starting from the patient's neck, each piece of equipment can be easily identified (Fig. 127.1).

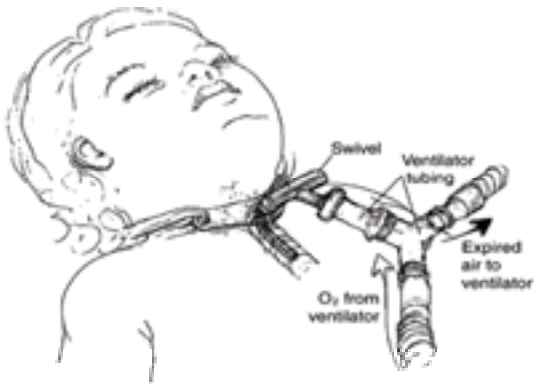

FIGURE 127.1. Tracheostomy parts.

Tracheostomy Tubes

Modern tracheostomy tubes are made of polyvinylchloride, a soft substance that conforms to the shape of the trachea, but is rigid enough to avoid collapse. Unlike their metal predecessors, they have little tissue reactivity, causing less tracheal wall irritation. Tracheostomy tubes are packaged by several manufacturers under sterile conditions for one-time use. Intensivists directing the long-term airway management of their patients may prefer one manufacturer over another, but the emergency physician does not need to know the minor differences among the products. The emergency physician should, however, know what types of tracheostomy tubes are stocked by the ED's facility and how to convert from the patient's brand and size to an available tube with suitable dimensions.

The size of a tracheostomy tube is determined by three dimensions: the inner diameter, the outer diameter, and the length. The inner diameter refers to the same measurement used in describing the size of an endotracheal tube, ranging from 2.5 to 10 mm. This measurement is generally imprinted on the flanges of the tracheostomy tube and is standardized among manufacturers. The outer diameter and length often are not identified on the tube and can vary considerably among manufacturers. The chart in Table 127.2 lists the dimensions of various tubes. When a tracheostomy tube change is indicated and an identical replacement is not available, this chart can be used in selecting the appropriate size tube of an available type.

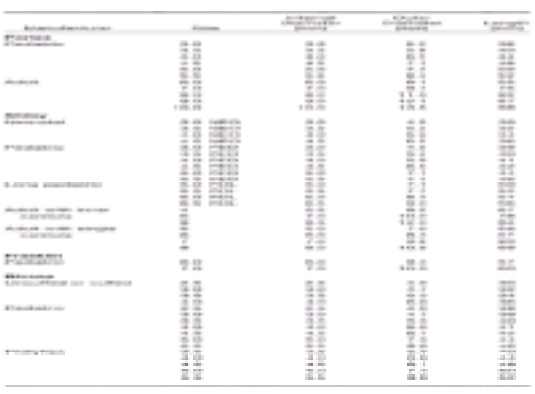

Table 127.2. Tracheostomy Tube Dimensions

A tracheostomy tube may be cuffed or uncuffed. Contrary to the general guideline that cuffed endotracheal tubes are used only for older children, an infant or young child may have a cuffed tracheostomy tube, especially if he or she has an airway anomaly or has developed tracheomegaly. Checking for the presence of a cuff in all patients, and deflating it before removing the tube is important.

Some tracheostomy tubes are fenestrated. The hole in the posterior aspect of the tube facilitates retrograde movement of air through the larynx, allowing vocalization. In addition, some tracheostomy tubes have an inner cannula. In patients with this type of tube, the inner cannula can be removed for cleaning while maintaining the airway with the outer cannula. Importantly, the proximal portion of the inner cannula is required to connect the tracheostomy to the manual resuscitator bag; therefore, the inner cannula must be in place when bag-valve ventilating.

Swivel

A swivel is often attached to the end of the tracheostomy tube. Some unique characteristics of children make the swivel particularly useful. First, children have a natural inclination to move and explore. The swivel device accommodates movement in the ventilator-assisted child so that traction is not placed on the ventilator tubing or on the tracheostomy tube. Second, the short neck and bulky soft tissues of young children can obstruct the tracheostomy tube opening. The swivel provides additional length so the tube extends beyond the soft tissues of the neck.

Heat–Moisture Exchanger

Air inspired directly into the trachea through a tracheostomy tube bypasses the important warming and humidification mechanisms provided by the natural upper airway. Therefore, a humidification system is an important component of the equipment used in a patient with a tracheostomy. The stationary humidification system in a home ventilator setup is used when the child is connected to the circuit. Similarly, a heat–moisture exchanger is attached to the end of the tracheostomy tube in patients who do not require the ventilator. The device is composed of a hydrophilic material that captures the patient's own heat and humidity on exhalation so that it can be inspired on inhalation. It should be placed between the tracheostomy tube and the manual resuscitator when prolonged bag-valve ventilation is performed.

Clinical Findings/Management

The approach to the ill patient with an artificial airway is the same as that for any patient who comes to the ED. The initial evaluation consists of a review of the patient's ABCDs (airway, breathing, circulation, and disability). Certainly, particular attention must be paid to the airway and breathing. An emergency physician who knows how to anticipate common problems and to recognize them early is able to institute appropriate therapy without delay.

Obstruction and Decannulation

The most life-threatening complication in a patient with an artificial airway is cannula obstruction or dislodgment. Younger children are more likely to experience accidental decannulation because of the short length of the trachea and tracheostomy tube. Some infant tubes are as short as 3 to 4 cm. In addition, the small lumen is more easily occluded by a mucous plug or by an accumulation of secretions. Infants with less developed intercostal muscles and children with neuromuscular disorders may be unable to generate an adequate cough to keep the airway clear of debris.

The presentation is similar to that of other children with respiratory compromise. The child may appear distressed with tachypnea, cyanosis, accessory muscle use, and nasal flaring. Alternatively, the child may be lethargic or obtunded as a result of prolonged respiratory effort or an elevated carbon dioxide level.

Any child with an artificial airway and respiratory distress is assumed to have an obstruction. The patient should be placed immediately on high-flow humidified oxygen. The physician should determine whether the tracheostomy tube appears to be in place, recognizing that a tube in the stoma does not necessarily indicate a tube in the trachea. If a cannula change was attempted before the child's arrival in the ED, a false passage into the paratracheal soft tissues may have occurred. Auscultation for the presence and symmetry of bilateral breath sounds should be performed and the quality of the patient's respiratory effort should be assessed. Immediate suctioning is appropriate in an attempt to assess tube patency and to clear the airway of secretions.

The physician should not hesitate to change the cannula. All the necessary equipment for the change should be present, including a replacement tracheostomy tube, an endotracheal tube one-half size smaller, a bag-valve-mask ventilation circuit with oxygen flow, scissors, and tracheostomy ties. The change is best accomplished with the participation of two people; one secures the patient and removes the old tube while the other inserts the new tube (see also Section VII).

Infection

Bacterial colonization of the trachea usually occurs in a child with a tracheostomy. Common colonizing organisms include Gram-positive cocci (*Staphylococcus aureus, Staphylococcus epidermidis, Streptococcus pneumoniae,* a- and b-hemolytic streptococcus), Gram-negative bacilli *(Klebsiella, Pseudomonas, Escherichia coli, Serratia marcescens, Haemophilus influenzae),* and anaerobes *(Peptostreptococcus, Bacteroides).* These same organisms can become pathogenic, causing tracheitis or pneumonia.

A peristomal cellulitis can result from infection with skin flora. Good tracheostomy care and regular cleaning with dilute hydrogen peroxide solution can prevent most peristomal infections. Similarly, inadequate padding of the neck area beneath the tracheostomy ties can result in a contact or monilial dermatitis.

Differentiating between bacterial colonization of the trachea and clinical infection can be difficult. The physician should elicit a history of any changes in the quantity, thickness, or odor of the tracheal secretions, and any systemic signs of infection or respiratory distress. Along with physical examination, there should be a determination of oxygenation by pulse oximetry. A Gram stain and bacterial culture, and a rapid viral detection assay of the tracheal secretions may be helpful in determining the presence and cause of an infection. Leukocytosis in the tracheal secretions and a predominant organism by Gram stain may be suggestive of bacterial tracheitis; radiographic evidence of a new infiltrate indicates pneumonia.

If the child appears well and follow-up can be ensured, outpatient antibiotic therapy may be appropriate. For children with increased oxygen or ventilatory requirements, hospitalization should be considered for intravenous (IV) antibiotic therapy, aggressive pulmonary toilet, and close monitoring.

Erythema of the peristomal skin is usually caused by irritation and should be managed by increasing the frequency of the tracheostomy care at home. The additional findings of warmth, tenderness, purulent drainage, or fever may suggest the presence of a peristomal cellulitis. Depending on its severity, this condition should be treated with oral or IV antibiotics.

The skin of the neck under the ties securing the tracheostomy tube also can become inflamed. Generally, this situation can be treated by increasing the amount of padding and by keeping the area dry. An erythematous rash with satellite

lesions classic for a monilial dermatitis should be treated with topical antifungal creams.

Asthma

The incidence of asthma in children with chronic lung disease has increased. Many children are maintained at home on inhaled b-agonists, inhaled steroids, and cromolyn sodium therapy. The usual viral and environmental triggers, such as dust, pets, and smoke, precipitate exacerbations of asthma in these children.

The presentation is similar to that of other asthmatic patients, with varying amounts of respiratory distress, wheezing, and hypoxemia. As previously mentioned, the physician must consider the possibility of cannula obstruction or dislodgment in all cases. Treatment with oxygen, bronchodilators, and steroids should be initiated promptly. Therapeutic options are the same as those used with other asthmatic patients, as described in Chapter 80 and Chapter 95. Emergency clinicians should recognize, however, that children with chronic lung disease have less pulmonary reserve. Chest radiography and arterial blood gas analysis should be performed as clinically indicated. Increased ventilatory support or continuous positive airway pressure (CPAP) may be required to overcome fatigue and atelectasis.

Bleeding and Granuloma

The tracheal mucosa located adjacent to the stoma, the cuff, and the distal tip of the tracheostomy tube are prone to bleeding or granuloma formation. The most common reason for bleeding is inadequate humidification causing drying and friability of the tracheal mucosa. Infection or granuloma formation can also result in small amounts of bleeding. Large amounts of blood coming from the tracheostomy tube opening can signify erosion of the tube into the innominate artery. This complication is rare, but life-threatening.

Small amounts of bleeding from the tracheal stoma usually resolve with increased humidification of the inspired air. The persistence of minor bleeding might indicate an intratracheal granuloma, which should be evaluated by direct visualization. This procedure is best performed by an otorhinolaryngologist.

A large amount of bleeding is a surgical emergency. IV access should be obtained immediately and volume replacement should be initiated. The tracheostomy tube should not be removed because it may be the best way to ensure an airway. Frequent suctioning aids in preventing aspiration. If the site of bleeding can be identified, direct pressure should be applied to the area. Overinflating the cuff may tamponade a bleeding vessel and provide a temporary treatment until it can be ligated.

Peristomal granulomas can usually be treated with topical antibiotics. In refractory cases, cauterization with silver nitrate is indicated.

Cerebrospinal Fluid Shunts

Background

CSF shunt placement is the most common neurosurgical procedure performed in children. CSF shunts are placed to divert CSF from the brain to another area of the body, most commonly the peritoneal cavity. The clinician evaluating a child with a CSF shunt should be aware of associated complications such as infection, obstruction, and overdrainage. Certain complications can be disastrous if unrecognized and untreated. Children with CSF shunts often may exhibit symptoms of their chronic illness that are unrelated to shunt placement.

Pathophysiology

CSF is an ultrafiltrate of plasma produced at a rate of 500 mL/day by the choroid plexus and various extrachoroidal sites within the brain. CSF travels from the lateral ventricles into the third ventricle through the foramen of Monro, and then again through the aqueduct of Sylvius to the fourth ventricle. The CSF then enters the subarachnoid space via the foramina of Magendie and Luschka, and travels through the brain and spinal canal. CSF is reabsorbed and enters the venous system through the “one-way valves” of arachnoid villi that penetrate the dura.

Hydrocephalus can result from oversecretion, impaired absorption, or blockage of CSF pathways. *Oversecretion* can occur in some choroid plexus tumors. *Impaired absorption* can occur as a result of increased CSF protein (e.g., subarachnoid hemorrhage, Guillain-Barré syndrome), severe congestive heart failure, or any other condition that raises venous pressure. Impaired absorption is the cause of communicating hydrocephalus, in which flow from the lateral ventricles to the foramina of Luschka and Magendie is not obstructed. *Blockage of CSF pathways* is the most common cause of hydrocephalus in children and is often located at the narrow aqueduct of Sylvius proximal to the fourth ventricle. Conditions that can cause obstruction are intraventricular bleeding or scarring, tumors, or congenital malformations. Dandy-Walker cysts cause obstruction of the foramina of Magendie and Luschka and therefore result in enlargement of all four ventricles.

Equipment

Different types of CSF shunts, which vary mostly by the location of the distal tubing and the type of reservoir or valve system, are available. The choice of CSF shunt depends on the individual patient's anatomy and cause of hydrocephalus, and the experiences and preferences of the neurosurgeon performing the procedure. Commonly, the patient or caretaker knows the location and type of shunt and is able to provide details regarding prior shunt placement and problems. Palpation of the hardware and plain radiographs may be used to acquire more information regarding the specific location of the shunt components. All CSF shunts have the following three components: 1) proximal shunt tubing,

2) reservoir system, and 3) distal shunt tubing (Fig. 127.2).

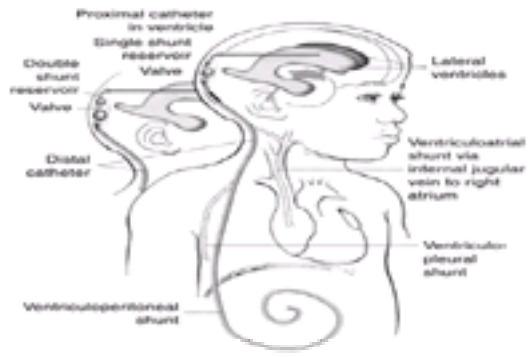

FIGURE 127.2. Diagram of typical ventriculoperitoneal shunt.

The *proximal shunt tubing* has a fenestrated tip that is usually located in the ventricle but may also be located inside a non-communicating cyst. This tip allows free passage of CSF into the shunt system. More than one proximal catheter may be present if multiple, noncommunicating areas of the brain require shunting. The *reservoir system* consists of one or two “domes” or “bubbles” and may contain a one-way internal valve unit. “Double-bubble” shunts provide selective access to (and assessment of) proximal or distal portions of the shunt system. Reservoirs may be placed directly over or slightly distal to the burr hole. This information is crucial when emergent access to the burr hole is needed. The *distal shunt tubing* leads from the reservoir unit to a part of the body that can accept the drained CSF, usually the peritoneum. The distal tubing may also be located in the vascular system or pleural cavity. Ventricular–atrial shunts are less commonly inserted because of the serious infectious complications that have occurred with these types of shunts. All modern shunt tubing is made of $^{1}/_{8}$-inch diameter Silastic, which causes less omental reaction and suffers less cracking than did prior materials.

CSF shunt systems contain a one-way valve to prevent backflow of CSF into the ventricles. These valves are designed to operate at high, medium, or low pressure, and may rarely incorporate an on–off control switch. An anti-siphon device may be inserted into the system to prevent overdrainage of CSF and concomitant low-pressure complications.

Clinical Findings/Management

Mechanical Malfunction

Malfunction of a CSF shunt can be caused by obstruction of the catheter lumen or disconnection of the various components. The proximal catheter lumen can be obstructed by fibrosis, debris, or choroid plexus; the distal catheter can be obstructed by the surrounding omentum or kinking of the catheter. Both proximal and distal portions can be occluded by the products of infection or by migration of the catheter tip into the brain parenchyma or intraabdominal structures. Particularly in neonates, poor absorption of excess fluid in the peritoneum can create the appearance of luminal obstruction. In addition, as the child grows, the tension on the shunt system can lead to disconnection of the distal tubing.

Almost 70% of patients with CSF shunts experience a shunt malfunction in their lifetime. Parental history is paramount in deciding whether a child is experiencing symptoms of shunt malfunction. The parent often notices that the child “just isn't acting right,” or is less active or thinking less clearly than usual. The statement, “This is exactly how he acted the last time his shunt was obstructed” is suggestive of another malfunction, regardless of the presence or absence of the symptoms listed in the following section.

Common signs and symptoms of mechanical shunt failure include headache, visual disturbances, vomiting, lethargy, and irritability (Table 127.3). The astute parent or clinician may note mild ataxia, increased head circumference or bulging fontanelle in an infant, poor cognition, or abnormal behaviors. Less subtle signs include paralysis of the fourth (sunsetting) or sixth cranial nerves (lateral gaze preference) and decreased level of consciousness. Increased tone, hyperreflexia, or Babinski's reflex represents stretching and disruption of the corticospinal fibers originating from the motor cortex and can suggest shunt malfunction in a patient with a previously normal examination. Patients with any component of Cushing's triad (hypertension, bradycardia, and abnormal respiratory pattern) require immediate maneuvers to decrease intracranial pressure (ICP) and guide them quickly toward operative repair of the shunt. Seizures are uncommon as the sole manifestation of CSF shunt malfunction. However, seizures can occur in children who have predisposing brain lesions, and many patients with CSF shunts have epilepsy. Shunt infection must be considered in the child with symptoms of shunt malfunction, especially if the child has a history of recent shunt revision. Ronan et al. reported that more than one-third of patients with shunt infection presented with symptoms of malfunction.

Symptoms
Fever
Headache
Altered mental status
Irritability
Lethargy/difficult arousal
Confusion
Vomiting
Visual disturbances
Seizures (rare to be only manifestation)
Signs
Papilledema
Bulging fontanelle/enlarged head
Engorged head veins
Macewen's sign (cracked pot sound during percussion)
Abnormal neurologic examination
Increased deep tendon reflexes (DTRs) or lower extremity tone
Positive Babinski's sign
Cranial nerve palsy—lateral (6th) or upward (4th) gaze (sunsetting)
Respiratory compromise

Table 127.3. Concerning Findings in Patients in Cerebrospinal Fluid Shunt Malfunction

If the history and physical examination of the ill child with a CSF shunt suggest a possible shunt malfunction, further evaluation includes a non-contrast computed tomography (CT) scan with comparison to the most recent prior study, if available. A plain radiograph of the skull, chest, and abdomen (“shunt series”) is helpful in assessing the integrity of the shunt connection and in identifying the components of the working system. The clinical suspicion of a shunt malfunction based on history and physical examination may outweigh the data obtained from radiographic studies.

Much discussion and controversy surround the clinician's ability to assess CSF shunt function by “pumping” the shunt reservoirs. In a single reservoir system, this procedure involves depressing the reservoir bubble. Resistance to depression suggests distal catheter malfunction. Poor filling suggests either proximal catheter malfunction or small ventricles. The maneuver in a double-bubble shunt requires the initial depression of the proximal bubble, depression of the distal bubble to check for resistance, and subsequent release of the proximal bubble to check for poor filling. Pumping the shunt to test for obstruction is not always reliable. Piatt found that this maneuver had a positive predictive value of 21% and a negative predictive value of 78% in patients for whom the diagnosis of shunt patency or malfunction was definite. In addition, frequent pumping of the shunt can cause entrapment of choroid plexus in the proximal shunt tubing and lead to proximal catheter obstruction where none previously existed.

If subsequent evaluation is still necessary to diagnose malfunction, a neurosurgeon should be consulted. It may be necessary to “tap” the shunt (Fig. 127.3). The patient's hair is either shaved or trimmed. The scalp is cleansed first with alcohol, then with three applications of Betadine that are allowed to dry after each application. The shunt tap is performed by inserting a 23- or 25-gauge butterfly obliquely into the reservoir and holding the butterfly tubing perpendicular to the floor. The height that the CSF rises into the butterfly tubing, measured in centimeters, is the ICP. Normal pressure is between 5 and 10 cm; pressure more than 20 cm is indicative of distal shunt malfunction requiring urgent revision. Slow or absent flow from the proximal reservoir (especially with occlusion of the distal reservoir of a double-bubble shunt) suggests proximal shunt obstruction. In this case, the physician may notice that the reservoir collapses when gentle suction is applied to the butterfly with a syringe.

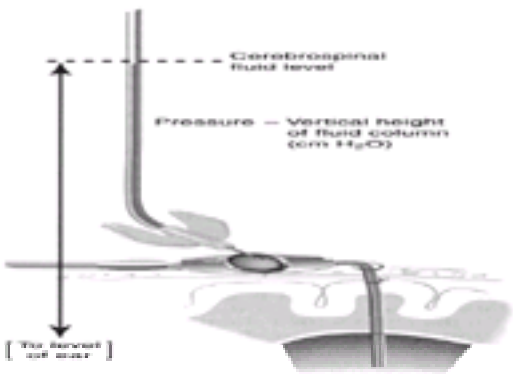

FIGURE 127.3. Tapping the cerebrospinal fluid shunt.

The shunt tap can be therapeutic as well as diagnostic. The child with a distal shunt obstruction or partial proximal obstruction may be eligible for urgent, rather than emergent, shunt revision if symptoms of increased ICP are alleviated after the tap. However, removal of too much fluid should be avoided because abrupt fluid shifts within the cranial vault can lead to disruption of subdural vessels. It is prudent to remove just enough fluid to decrease the ICP below 20 cm and repeat the procedure if symptoms return before definitive surgical management.

The child with complete obstruction of the proximal catheter does not obtain relief of symptoms after a shunt tap, because the obstruction prevents adequate aspiration of fluid from the ventricles. In most cases, these children may respond to medical management of increased ICP. This treatment includes the administration of acetazolamide (Diamox) 30 mg/kg per day and Decadron 1.0 mg/kg per day, and hyperventilation in the relatively unstable patient. If the child is experiencing life-threatening symptoms from proximal obstruction, is unable to undergo immediate surgical repair, and is unresponsive to medical management, a burr-hole puncture procedure may be performed (Fig. 127.4). Although the role of burr-hole puncture is clear in the patient who is in impending or existing neurologic failure, the procedure has many risks, including disruption of intraparenchymal vessels and tissue. By nature of the procedure itself, the proximal shunt catheter is torn and urgent revision is therefore mandatory. The burr hole is located either directly below or slightly proximal to the reservoir, depending on the type of shunt. For example, a Rickham reservoir is located directly over the

burr hole, whereas a double-bubble reservoir is located slightly distal to the burr hole. A 3½-inch spinal needle is inserted perpendicular to the skull through the burr hole to a depth of no more than 5 cm. After the stylet is removed, fluid should drain spontaneously and should be removed until flow slows down. The patient's condition should stabilize sufficiently for transport to an operating suite or tertiary care institution.

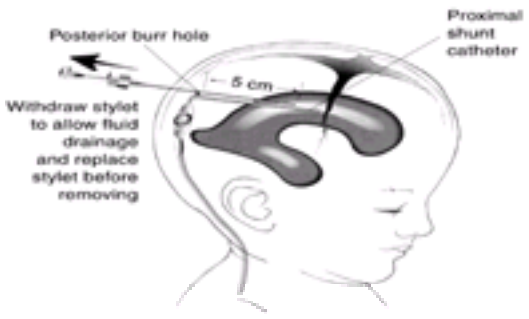

FIGURE 127.4. Burr-hole puncture.

Another method of temporarily relieving a lumen obstruction is to flush a small amount of sterile saline through the clogged tubing in an attempt to dislodge the obstruction. This method can be used for distal or proximal obstructions, with the caveat that instilling a few more milliliters into the ventricles may in fact worsen the patient's condition. In a double-bubble shunt, the reservoir that is not being used must be compressed to allow the fluid to go in only one direction.

In an infant with an open fontanelle, the physician can aspirate fluid through a direct ventricular puncture (Fig. 127.5). This procedure carries as great if not greater risk of parenchymal injury as the burr-hole puncture procedure, and likewise should be performed only when prompt surgery is impossible.

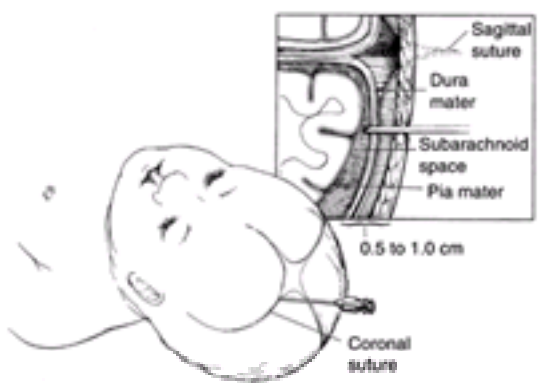

FIGURE 127.5. Ventricular tap through open fontanelle.

Infection

The reported incidence of CSF shunt infections ranges between 2 and 20% and depends on the center performing the study and the criteria used to define infection. The chance of infection is directly related to the timing of shunt placement. Infections generally occur within a few months of shunt placement, many of which occur within the first few weeks. The common organisms cultured from infected CSF shunts are Gram-positive (Table 127.4). Staphylococci adhere well to Silastic tubing, and these infections are often difficult to eradicate without removal of the catheter. Infection with staphylococci is common within the first 2 weeks after surgery. Gram-negative infections tend to occur later in the perioperative course. Fungi are rare pathogens seen occasionally in premature infants.

Table 127.4. Common Organisms Involved in Cerebrospinal Fluid Shunt Infections

External infection of skin and subcutaneous tissue overlying the shunt hardware can occur; however, these superficial infections may not lead to shunt infection if treated promptly. Necrosis of the area around the reservoir can occur as a result of the constant pressure in infants or non-ambulatory patients. Skin breakdown leading to visualization of the shunt mechanism is, by definition, a shunt infection, and must be treated accordingly.

The peritoneal portion of the shunt may become infected through the shunt mechanism or via a primary peritoneal

infection. Peritoneal infection can result in loculated, cystic pools of infection around the terminal portion of tubing (pseudocysts). These infections may be indolent in their presentation, and the shunt tap from the reservoir may not show evidence of infection.

Shunt nephritis is a rare but serious complication of ventricular–atrial shunts. Renal deposition of antigen–antibody complexes leads to complement activation, which damages the renal tissue.

Unfortunately, the child with an infected CSF shunt may present with nonspecific signs and symptoms (Table 127.5). Children commonly develop symptoms of shunt malfunction, such as lethargy or irritability. However, infection may also manifest as abdominal complaints, such as pain or vomiting. Fever is present in a little more than half of patients with shunt infections, and is rarely the only sign. As previously mentioned, infection is most common within a few months of the shunting procedure and is rare more than 1 year afterward. These rules are less applicable in patients with Gram-negative infections, which can occur later after shunt placement. Children with Gram-negative infections are often bacteremic, if not septic appearing.

Change in sensorium	Shunt malfunction
Fever	Vomiting
Irritability	Abdominal pain

Modified with permission from Odio C, McCracken GH, Nelson JD. Am J Dis Child 1984;138:1103-1108.

Table 127.5. Signs and Symptoms of Shunt Infection in Patients without Wound Infection

A wound infection overlying any portion of the shunt mechanism manifests as erythema and tenderness or swelling along the shunt tract or over the reservoir.

In the absence of overlying infection, aspiration of a small amount of CSF from the shunt system should be performed to identify the presence of a bacteriologic cause of shunt infection. This procedure is usually performed by a neurosurgeon, if possible. The white blood cell (WBC) count can range from 0 to 2600 if the shunt is infected, and patients without evidence of infection can have up to 500 WBC/mm^3. Many clinicians use greater than 50 WBC/mm^3 in the presence of clinical signs or symptoms of infection to secure the diagnosis. Gram stain of the fluid may be helpful in broadening antibiotic coverage if Gram-negative organisms are present. However, the Gram stain should not be used to narrow the usual antibiotic coverage until the culture and sensitivities of the causative organisms are obtained.

Various permutations of medical and surgical therapy have been suggested for treatment of CSF shunt infections. Medical therapy alone has been found to have a relatively low success rate compared with the surgical approach. Potential surgical interventions include immediate shunt replacement, or the insertion of an extraventricular drainage (EVD) catheter followed by delayed shunt revision. This latter method improves the bacteriologic cure rate significantly, although it must be performed in an institution that is facile in managing and preventing infection of EVD catheters. This procedure also requires the patient to visit the operating room twice.

Medical therapy provided in the ED for children with suspected CSF shunt infections is limited to the administration of broad-spectrum IV antibiotics. The antibiotics should be effective against the most common infecting organisms, as well as any organisms identified from previous infections. A reasonable choice of empiric therapy is vancomycin and ceftazidime. Therapy can be narrowed based on culture results of the shunt fluid. In patients with Gram-negative or fungal infections, intrathecal antibiotics may be used; however, this procedure is not considered appropriate in an ED.

Overdrainage

Occasionally, children with CSF shunts experience symptoms related to the system working too well, resulting in low ICP. One consequence is the slit ventricle syndrome, in which the ventricles collapse around the proximal catheter port and block further drainage. The best means of diagnosing this condition is the patient's history, rather than physical examination or radiographic analysis.

Until fluid increases to the point that the proximal catheter becomes patent, the child may exhibit symptoms of headache, nausea, vomiting, and lethargy. The drainage of CSF shunts increases when the patient is upright and decreases when supine. In contrast to the classic timing of symptoms related to increased ICP, patients with slit ventricle syndrome are often worse when in the standing position or after they are awake for several hours. Lying supine for a few hours tends to relieve symptoms of slit ventricle syndrome. Many patients with CSF shunts have CT scans that reveal small ventricles; however, only a small proportion of these patients have slit ventricle syndrome. Therefore, the CT scan is best used to differentiate between shunt malfunction and other causes of symptoms, rather than to diagnose an overdrainage problem.

Chronic or recurrent episodes of slit ventricle syndrome can be addressed surgically by upgrading the resistance of the valve or by insertion of an antisiphon device. Oral analgesics may be helpful in managing mild cases.

Other Complications

A number of other complications related to CSF shunts deserve mention. The most common of these complications is a benign postoperative leakage of CSF around the proximal shunt tubing into the subgaleal space around the reservoir. The resulting boggy mass, often called a “bumble,” resolves spontaneously, so drainage of this fluid should be avoided.

Patients with CSF shunts have an increased risk of seizures compared with the general population. These seizures often begin years after shunt placement and are caused by epileptogenic scars. They are more common in patients with other abnormalities correlated with seizures, such as porencephalic cyst or intracranial hemorrhage.

Overdrainage can lead to shrinkage of brain tissue and concomitant subdural accumulations (hematomas or effusions). Similarly, a decreased rate of head growth because of overdrainage can result in craniosynostosis in the infant.

Some important, albeit rare, complications are related to specific types of CSF shunts. The distal portions of a ventriculoperitoneal shunt can migrate and cause perforation of the colon or genital tract. This section of tubing can act as a fulcrum for intestinal volvulus. Ascites and abdominal cysts can form as a result of drainage of excess fluid into the peritoneum. Increased intra-abdominal pressure can precipitate the formation of an inguinal hernia through a patent processus vaginalis.

Ventricular–vascular shunts can be associated with an increased risk of bacteremia. Shunt nephritis can result from complement activation renal deposition of bacteria. Patients with ventriculo-atrial shunts can experience cardiac arrhythmias or atrial perforation, usually perioperatively. Bacterial endocarditis, cardiac foreign body, and mural thrombus are rare but notable complications of vascular shunts.

Indwelling Venous Access Devices

Background

Broviac designed the first Silastic indwelling central venous catheter in 1973. These devices provide children with relatively permanent and secure venous access during chemotherapy, total parenteral nutrition, or prolonged IV antibiotic therapy. Pediatricians, family practitioners, and emergency physicians have increasingly been called on to access and assess these catheters. Clinicians must be familiar with the procedures for establishing patency, drawing blood, dealing with catheter occlusion or breakage, and assessing for infection.

Pathophysiology

The distal tip of indwelling venous catheters is located in the right atrium or at the junction of the right atrium and the superior vena cava. The site of venous entry is usually the subclavian or internal jugular vein; however, access is occasionally obtained through the external jugular, cephalic, and brachiocephalic veins. The catheter is tunneled under the skin to a site in the chest away from the venous entry site, and then either externalized or connected to a subcutaneous reservoir.

Equipment

Partially implantable central venous catheters (CVCs) are also known as *externalized* CVCs (Fig. 127.6). They come in various types, such as Broviac, Hickman, Leonard, Raaf, Hermed, Groshong, and Corcath. All of these catheters are made of Silastic and are tunneled under the skin a few centimeters before externalization. A Dacron cuff located around the catheter at the skin line anchors the catheter after it fibroses. The most proximal portion of the catheter contains a female Luer lock tip, allowing a direct and solid connection to most syringes and IV tubing. A clamp is present just before this tip, under which a reinforced sleeve protects against catheter breakage. The catheters can vary in length, diameter, and numbers of lumen and access ports. The Groshong type catheter is valved at the distal tip to keep blood out of the lumen and therefore requires only saline flushes and does not require clamping. A kit is available that contains the equipment necessary to repair broken external catheters (Evermed, Inc.).

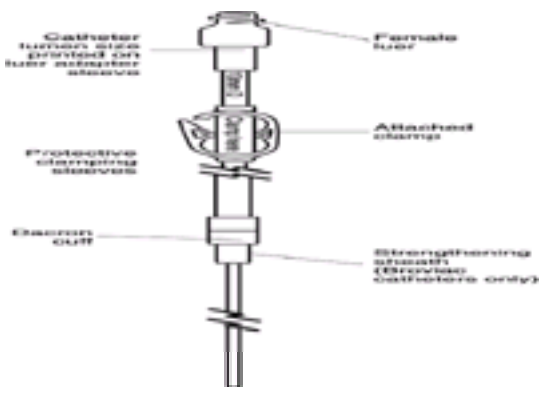

FIGURE 127.6. Partially implantable (externalized) venous catheter.

Implantable venous access devices (Infusaport, Port-A-Cath) are also called *internalized* CVCs (Fig. 127.7). Like the externalized CVCs, they use a Silastic catheter with the distal tip located in the right atrium. However, the proximal end is

tunneled and connected to a subcutaneous reservoir chamber. The reservoir has a self-sealing silicone septum and a hard back surface. The chamber is accessed using a tapered 20- or 22-gauge Huber noncoring needle (solid tip with side ports). The needle is angled at 90 degrees for ease of insertion and stabilization. If emergency access is required, a 19-gauge straight needle can be used.

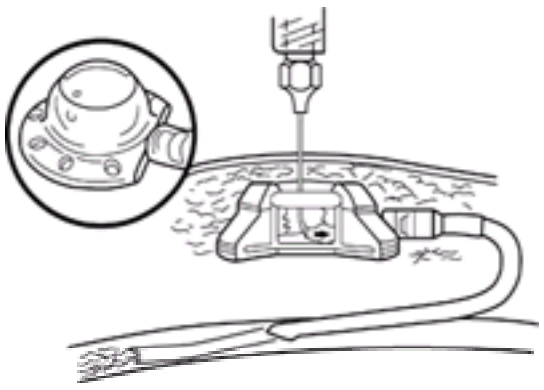

FIGURE 127.7. Implantable (internalized) venous access device.

Specific equipment is available for accessing externalized and internalized indwelling CVCs (Table 127.6). The medical office or ED that occasionally sees these children should have a prepared kit containing these items at hand.

Clamp or hemostat without teeth, possibly with rubber tips	Povidone-iodine ointment
T-extension tubing with clamp	Alcohol swabs
10-mL syringe of normal saline	Sterile drapes and gloves
10-mL syringe of heparin 100 U/mL	Dressing gauze and tape
(4) Injection caps	Tegaderm sterile dressing
Povidone–iodine solution with sterile gauze	(2) Tapered Huber non-coring needles, 20- and 22-gauge

Table 127.6. Equipment Needed to Access Central Venous Catheters

Procedures that can be accomplished by the generalist or ED personnel include establishing access, performing phlebotomy, and infusing fluids or medications. The general procedure for establishing access and patency is similar for both externalized and internalized devices (Table 127.7). Aseptic technique is mandatory. Because tincture of iodine solution can damage Silastic catheters, povidone–iodine solution is used. Clamps or hemostats with teeth can also damage the external portion of the catheter. In addition, smaller (less than 3 mL) syringes can generate too much pressure inside the catheter, causing catheter breakage. Therefore, 5- or 10-mL syringes are recommended to flush the system. Fluid or medications should never be infused until patency is established because the risk of administering these solutions into a non-vascular space is high. To prevent air emboli from occurring, all clamps must remain closed when any part of the circuit is open. For accurate blood test results, the amount of blood that needs to be withdrawn unused is 5 mL from an externalized CVC and 3 to 5 mL from an internalized CVC.

Aseptic technique
Do not use:
Clamps or hemostats with teeth
Tincture of iodine solution
Small (≤3-mL) syringes
Flush entire intravenous circuit before accessing system.
Always close clamps when any part of the circuit is open.
Do not infuse fluids or medications until patency is established.
Flush the catheter with 10 mL of saline between medications.
Flush cap or reservoir with heparin when procedure is complete.

Table 127.7. Tips for the Routine Use of Indwelling Venous Access Devices

When an externalized CVC is accessed, these steps should be followed:

1. Before accessing the system, flush the intended IV circuit, including T-connectors, with saline to remove air. Clamp the T-connector closed.
2. Clamp any external portions of tubing 3 cm from tip.

3. Connect to the system.
4. Flush the system with 3 to 5 mL of saline in a 5- to 10-mL syringe, then aspirate 3 to 5 mL of blood to check patency.
5. Draw off blood needed for laboratory analysis, and administer medications or fluids as needed.
6. When these steps are completed, clamp the line.
7. Inject 1 mL of 100 U/mL heparin solution into the cap after the procedure is completed. The recent advent of "needleless" systems have decreased the need for puncturing the caps of external catheters. If this type of system is used, remember to discard the old cap and replace it with a new one using sterile technique.

The procedure differs slightly when accessing an internalized CVC. Because intact skin is penetrated, the use of EMLA at least 60 minutes before access should be considered when feasible. If used, the EMLA should be wiped off and the skin should be cleansed using povidone–iodine, allowed to dry, and followed by an alcohol swab. Instead of connecting to an external catheter, a Huber needle should be inserted through the skin directly into the reservoir diaphragm but stopped if resistance is met at the back of the reservoir. The needle should be taped in place and patency should be established with aspiration and flushing. After use, the internalized device must be flushed using 1 mL of heparin diluted in 4 mL of saline or similar solution.

Complications resulting from accessing CVCs include occlusion, air embolus, catheter breakage or displacement, and infection. Although most of these complications can be avoided if care is taken to maintain aseptic technique, the clinician should be aware of their diagnosis and management.

Clinical Findings/Management

Catheter Occlusion

Difficulty in drawing blood or infusing fluid through a CVC can be the result of catheter malposition or occlusion. The catheter may be positioned against a vessel wall or fibrin or blood may be clotted in the lumen. In addition, various precipitates can occlude the lumen of the catheter. Waxy precipitates can result when parenteral nutrition solutions contain combinations of fat, protein, and carbohydrate, and particulate precipitates can result from the poor solubility of calcium and phosphorus. IV phenytoin (especially when administered in a glucose-containing solution) and diazepam can precipitate as well.

Children who require IV medications or fluids at home may present for acute management of catheter occlusions. The problem is often noted only when the acute-care nurse or physician cannot access the line during the evaluation of another problem.

Phlebotomy can be facilitated by increasing the venous pressure gradient along the catheter. These maneuvers include having the patient hold his or her arms above the head, cough or Valsalva, and placing the patient in reverse Trendelenburg position. If blood still cannot be drawn, 3 mL of saline should be used to gently irrigate the clot and aspirate it into the syringe. Two to three mL of fluid should be used in a back-and-forth motion to avoid forcing the clot into the venous system. A number of complications can result from this maneuver. The pressure can force the clot into the bloodstream or rupture the catheter, particularly if the practitioner uses too much force or too small a syringe. Care should be taken to observe the catheter for a balloon "aneurysm," a sign of impending rupture.

Internalized systems are much less likely to clot than are externalized catheters. This situation is fortunate because irrigating the clot is admittedly more difficult, if not impossible, to perform on an internalized system.

Specific agents may help dissolve precipitates or clots. For waxy precipitates, 70% ethanol should be used, and for particulate precipitates, 0.1 normal hydrochloric acid (HCl) should be used. Fibrinolytic agents such as urokinase (0.5–1 mL of a 5000 U/mL solution) may dissolve a blood clot, and similar to HCl may be used up to three times if necessary. Ethanol should only be used one time per episode. Urokinase infusions may be started at the suggestion of the surgical consultants, who should be involved in the treatment plan if initial attempts are unsuccessful.

Air Embolism

Failure to maintain a closed system during manipulation of indwelling venous catheters can result in embolism of air into the chambers of the heart. Passage of the embolus to the systemic or pulmonary circulation can result in severe and irreversible tissue damage.

Air embolus can cause a patient to experience sudden onset of tachypnea, tachycardia, hypotension, or loss of consciousness. Other diagnoses that should be considered in patients with these symptoms are pneumothorax, liberation of septic emboli, and direct cardiac insult. If an air embolus is suspected, the patient should be placed in the left-sided Trendelenburg position, and oxygen should be administered. In addition, the indwelling catheter should be clamped and remain unused as other peripheral access is obtained.

Catheter Breakage

The family members and physicians caring for the child with an externalized catheter may have considered the nightmare of catheter breakage and subsequent exsanguination. Although catheter breakage is a distinct possibility, most events occur during routine care rather than during playtime, and therefore the blood loss is easily apparent and correctable. An external catheter can acquire a small hole from inadvertent needle puncture or even ordinary wear and tear. Internalized catheters, on the other hand, are less susceptible to local events or wear and tear. However, trauma to the area can result in detachment of the proximal portion of the catheter from the implanted port.

Leakage of blood or fluid from the externalized portion of an indwelling catheter is easily noticed. Externalized catheters

must be immediately clamped proximal to the break, cleaned with povidone–iodine solution, and covered with sterile dressing until repair can be made. Repair kits are available for each catheter size (Fig. 127.8). These kits contain a new external catheter segment with a hollow male connector that fits into a cleanly sliced proximal end. The kits also contain a syringe and needle to apply the glue to the male connector. Optimally, a person familiar with the procedure will be available within a short time of clamping the catheter. If the externalized portion is too small to clamp, hemostasis may be achieved by putting pressure on the site of venous entry. A scar is usually apparent at this site. However, if the scar is not apparent, the catheter should be palpated from the exit site on the skin to the location at which it can no longer be palpated, and pressure should be applied at that site.

FIGURE 127.8. Repair kit for externalized catheters

If an implantable catheter leaks, fluid or blood that collects subcutaneously may cause a bulge or painful swelling at the site. A broken internalized catheter must undergo prompt surgical management.

Catheter Displacement

Occasionally, the patient or caretaker inadvertently pulls on the externalized portion of an indwelling catheter. The venous portion of the catheter may eventually be displaced from the venous system. Externalized catheters are at higher risk for dislodgment within a few weeks of insertion, because the cuff is not fully anchored by fibrosis. Exsanguination after catheter dislodgment is a rare event because of the advancement of the tip inside the vein and the natural tendency toward venous hemostasis. However, children with clotting disorders are at increased risk of life-threatening blood loss after catheter displacement. Internalized devices are at risk of dislodgment at both ends; however, few events aside from major thoracic trauma place enough tension on the catheter to dislodge it from the vein. Migration of the venous catheter tip is rare but can lead to cardiac arrhythmias, pneumothorax, cardiac tamponade, and superior vena cava syndrome.

Detecting catheter dislodgment is easier in patients with externalized catheters. If the Dacron cuff is noted outside of the skin surface, the catheter must be considered dislodged and should not be used. Failure to draw back free-flowing blood from the device increases the suspicion that the catheter is no longer in the central vein. In this situation, the catheter should be clamped and secured close to the skin, and immediate surgical consultation should be obtained. As previously mentioned, hemostasis is obtained by putting pressure on the site of venous entry. A dye study may be necessary to locate the catheter tip. For internalized devices, dislodgment of the catheter from the vein should be suspected if the device no longer functions after thoracic trauma. If the catheter is disconnected from the reservoir, fluid or blood may collect subcutaneously and cause a bulge or painful swelling at the site. Prompt surgical management is required.

Catheter migration should be considered in patients with indwelling venous catheters who experience respiratory distress or palpitations. Radiologic evaluation of catheter location should rapidly ensue, with subsequent surgical consultation if the catheter tip has migrated.

Infection

Fever in a patient with an indwelling venous catheter is usually caused by a routine viral or bacterial infection. However, certain clinical findings suggest a line infection. The presence of an indwelling venous catheter places a patient at higher risk for infection. Immunocompromised patients can exhibit rapid deterioration, and more commonly acquire fungal, Gram-negative, and polymicrobial infections. Patients receiving parenteral alimentation are also at higher risk for Gram-negative infections. Still, the most common pathogens in patients with indwelling catheters are Gram-positive organisms such as *S. epidermidis, S. aureus,* and *S. viridans.* Externalized catheters carry a higher overall risk of infection compared with internalized devices. Catheter infection can occur at the site of catheter exit or reservoir, at the subcutaneous tunnel, or at the site of venous entry. The signs of infection may be more subtle or absent in neutropenic patients. Catheter-related bacteremia can also occur without apparent skin manifestations.

The presence of erythema, tenderness, or purulent drainage at any skin site related to an indwelling catheter suggests a line infection. The entire dressing must be removed for all of these sites to be inspected. Fever is common in patients with catheter-related bacteremia or sepsis but may be absent in early localized infection.

Blood cultures should be obtained from the catheter, and in most cases from a peripheral vein as well. Fungal cultures are appropriate in immunocompromised patients or those who have had prior invasive fungal infections. Cultures of any purulent fluid are helpful. A complete blood count with differential is warranted, although a normal result should not dissuade the clinician from suspecting an invasive bacterial infection. Other blood tests, such as coagulation studies, should be considered if the patient is ill appearing.

Initial treatment consists of IV antibiotic therapy and supportive measures. Many catheter infections can be eradicated without catheter removal. Infection of the subcutaneous tunnel is a strong indication for catheter removal. Initial antibiotic therapy should include agents active against both Gram-positive and Gram-negative infections. Many centers use vancomycin and gentamicin as the first choice. If the patient is neutropenic, ceftazidime should be added for presumptive treatment of *Pseudomonas* infection. Local bacterial resistance patterns may alter these choices, such that centers may reserve the use of vancomycin for culture-positive resistant strains or use netilmicin in place of gentamicin. These issues should be discussed with the patient's personal physicians and the infectious disease consultants.

Other Complications

Other complications related to indwelling catheters can occur, albeit rarely. Direct injury to the exit site can be a result of erosion of tissue by the Dacron cuff of an externalized catheter, or of breakdown of the skin site from vigorous cleansing. This condition can lead to a localized infection. On physical examination, excoriation, erythema, tenderness, or purulent drainage is present at the exit site of the catheter. Select patients with a localized site infection who are afebrile and well appearing, have a normal leukocyte count, and have pre-arranged follow-up may be managed as outpatients with oral antibiotic therapy.

As previously mentioned, phenytoin and diazepam can interact with the silicone lining of the catheters, and the administration of these medications through silastic catheters should be avoided if possible. In addition, a large volume of saline flush should be administered between medications that are incompatible with each other, such as calcium and bicarbonate.

Enteral Feeding Tubes

Background

A stoma, derived from the Latin word for “mouth,” is an opening from the GI or urinary tract to the outside of the body. A gastrostomy is a surgically created stoma that brings the stomach to the level of the skin. A jejunostomy is a surgically created stoma that brings the jejunum to the skin surface. Gastrostomy is performed most typically in children who are predicted to be unable to take adequate oral nourishment for a prolonged period. The inability to tolerate sufficient oral feedings can be related to several conditions, including esophageal atresia, chronic malabsorptive syndromes, significant craniofacial abnormalities, neurologic impairment, severe gastroesophageal reflux, and esophageal burns. Jejunostomy feedings are used when post-pyloric feeding is required, such as patients with delayed gastric emptying, recurrent aspiration pneumonia, and severe gastroesophageal reflux. Gastric feedings are much more common than jejunal feedings.

Enteral feeding via gastrostomy and jejunostomy tubes has become more common in recent years. Therefore, ED physicians should become comfortable with the various types of G-tubes and jejunal tubes, the supporting types of apparatus, and the complications inherent in the use of these lifesaving enteral feeding devices.

Pathophysiology

Gastrostomies tubes are inserted via open gastrostomy, or percutaneous endoscopic gastrostomy (PEG). In the open gastrostomy technique, a left upper quadrant or midline incision is used to place the gastrostomy tube through the abdominal wall. The G-tube then passes through a purse string silk suture placed on the anterior wall of the stomach and into the lumen of the stomach at the level of the fundus. The purse string suture is then tightened around the tube to prevent gastric leakage, and the wall of the stomach around the suture is sewn to the abdominal wall where the tube makes its exit. PEG is the most popular of the non-surgical procedures for placing a gastrostomy tube. This technique involves placing a tube into the stomach through a percutaneous hole in the anterior abdominal wall. An endoscope is used to provide light at the exact site on the anterior abdominal wall as a guide for needle puncture. A long guidewire with a feeding tube and pointed dilator is then passed through the mouth and distally until the pointed dilator is seen pushing its way through the skin of the anterior abdominal wall. A cuff system is used to bring the stomach and anterior abdominal wall closely together.

Jejunostomy can be performed via an open technique or percutaneously. Jejunal feeding can also be accomplished by placing a jejunal tube via the gastrostomy. This method allows jejunal feeding and enables venting of gastric air.

Equipment

Gastrostomy Tubes

Several types of gastrostomy tubes are available. Most are made of polyurethane, silicone, or rubber. These devices may vary in length, the number of ports, the type of catheter tip, the number of lumens, and the manner of securing to the patient's skin (Fig. 127.9). The *mushroom* types (Button by Bard Interventional Products Division, Billerica, Massachusetts) have soft flexible tips that require an obturator or stylet to stretch the tip. These devices have a single lumen. The *collapsible wings* tubes (Malecot, St. Louis, Missouri) are not as available but function in a similar manner. The *balloon tip devices* (MIC-KEY, Medical Innovations Corporation, Draper, Utah) have become popular and have begun to replace the mushroom tip and collapsible wings devices. The inflatable balloon is located at the tip, similar to a urinary Foley catheter. They are easy to secure and do not dislodge as easily. These tubes may have multiple ports and lumina. The most recent advance in gastrostomy tubes is the introduction of the low profile G-tube, commonly referred to as buttons (Fig. 127.10). The advantage of this type of apparatus is that no long piece of tubing arises from the stoma. They may have either mushroom or balloon tips. Replacement devices need to be matched for both the size of the stoma (the external diameter of the tube) and the length of the stoma tract. These buttons have uni-directional anti-reflux valves

that are fragile.

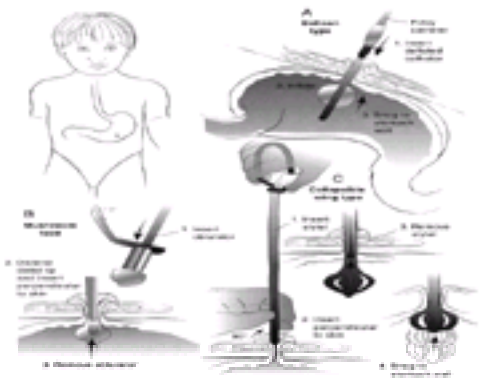

FIGURE 127.9. Gastrostomy tube replacement for balloon-type **(A),** mushroom-type **(B),** and collapsible **(C)** wing-type catheters.

FIGURE 127.10. The button: replacement gastrostomy device.

Jejunal Tubes

Jejunal tubes that pass through the gastrostomy are usually small diameter tubes (8 Fr), an example of which is the Frederick Miller feeding tube set manufactured by Cook (Bloomington, Indiana). These tubes have a small mercury weight at the distal tip and are placed under fluoroscopy. Several types of surgical jejunostomy feeding tubes are available, including Malecot and MIC-KEY jejunal tubes.

Clinical Findings/Management

Patients with gastrostomy tubes (G-tubes) or jejunostomy tubes (J-tubes) who present with symptoms that seem to be related to the tube still require a full evaluation. If the problem is directly related to the G-tube or J-tube, the emergency physician can offer efficient evaluation and therapy if he or she is familiar with potential complications. Complications related to gastrostomy and jejunostomy can be divided into mechanical tube-related problems and stomal problems.

Tube-Related Problems

Dislodgement. Dislodgement is one of the most common complications of gastrostomy tubes. This situation can occur as a result of a traumatic event, such as accidental tension on the external tubing, occult balloon deflation, or rupture of the balloon leading to extrusion of the entire tube. When G-tube dislodgement precipitates an ED visit, many parents either remember the size of the tube or bring one along to the ED. If neither of these events occurs, the patient's medical record usually provides the most recent tube size.

The patient with tube dislodgement may present with a benign stoma or with active bleeding secondary to trauma. If the tube size is unknown or if various tube sizes are not available, the most common temporizing method of replacement is insertion of a Foley catheter. A crucial consideration is the interval of time since the dislodgement. If hours have elapsed, the stoma may be constricted and require insertion of a smaller replacement tube.

The interval since *initial* placement of the gastrostomy is important. Peri-operative displacement (within 1 month of initial placement) is treated differently than dislodgement of a mature stomal tube. If a G-tube dislodges, temporary replacement with a smaller Foley catheter may prevent pushing the recently fixed stomach away from the anterior abdominal wall. A series of progressively larger Foley catheters, beginning with one to two sizes smaller than the original tube, may be used to dilate the stoma if it is partially closed. A gastroenterologist or surgeon should then be consulted for definitive care. An older tube that has dislodged should be replaced urgently with the same size and type of tube to avoid narrowing of the stoma. The physician must use caution when reinserting a G-tube because extreme force can lead to tube insertion into the peritoneal cavity through a false tract.

A jejunal tube that has dislodged needs to be replaced by the subspecialist who placed it initially. For example, a J-tube that was inserted via the gastrostomy should be replaced by the interventional radiologist under fluoroscopy. A surgical

jejunostomy tube should be replaced by a surgeon.

Clogging. Clogging or obstruction of the lumen of the G-tube or J-tube can occur as a result of dried, solidified formula or twisting or kinking of the tube. Tube obstruction is discovered when the caretakers cannot infuse fluids. If formula is suspected as the cause, aspiration of the clot should be attempted as well as gentle flushing of the lumen. Warm water is recommended as the most effective fluid. Despite reports of the success of various carbonated drinks in this situation, their effectiveness is controversial. When the G-tube becomes clogged, insertion of a stylet is not recommended because this technique may result in perforation of the tubing beneath the skin level. Repositioning of the tube should be attempted next; if this procedure is not effective, removal and replacement are necessary. If the gastrostomy is fresh (within 1 month), the surgeon or gastroenterologist should be consulted before removal of the clogged tube. Caretakers should be reminded of the need for proper flushing with each use.

Leaking. Leaking can occur directly from the lumen of the tube or from the peristomal area. Leaking from the stoma often indicates that the stoma has widened and now exceeds the size of the tube. Determining whether the leaking substance is formula, pus, or gastric fluid is crucial. If purulent drainage is coming from the stoma, the physician needs to look further for signs of stomal cellulitis or peristomal abscess (see following section). If formula is leaking from the lumen of the tube, the physician must assess the tube position and check the balloon. In the case of a leaking button, problems with valve patency could occur. If fluid is leaking from the stoma, the stoma may have become larger than the tube. One approach to this problem is removing the tube for a short period, allowing constriction of the stoma to occur. Alternatively, the existing tube can be replaced with a larger one. The stoma may also have become disrupted and, therefore, requires surgical evaluation.

Reflux. Gastroesophageal reflux may be a complication of gastrostomy placement. An increase in prior reflux disease can occur when a Nissen fundoplication is not performed simultaneously. The patient may present with an increase in vomiting and symptoms of esophageal irritation after gastrostomy placement. Patients in this category may benefit from continuous enteral feedings. If continuous feedings are not effective in reducing symptomatic reflux, fundoplication may be indicated.

Gastric Ulceration. Gastric irritation leading to ulceration may occur as a complication of gastrostomy in several scenarios. If the tip of the gastrostomy tube is too long, it may abrade the opposite surface of the stomach mucosa, resulting in traumatic ulceration. Similarly, the balloon may accidentally become overinflated and cause friction, especially when the stomach is empty. Balloon overdilation can occur if medications or flushes are erroneously administered via the balloon port.

A patient with gastric ulcer caused by mechanical trauma presents with symptoms similar to other ulcer patients. Common findings are abdominal pain, irritability, hematemesis, hematochezia, and coffee ground gastric drainage from the G-tube lumen. Saline lavage should be performed. If the fluid obtained is non-bloody, medications such as H_2-blockers, antacids, and Carafate may be administered, and upper endoscopy should be scheduled. The gastrostomy tube should be changed and the patient's symptoms should be monitored carefully.

Gastric Outlet Obstruction. Gastric outlet obstruction is a rare but serious complication of gastrostomy tubes. It is usually the result of the migration of the tube tip into the pyloric channel. Occasionally, the gastrostomy tube can migrate superiorly and block the esophagus. In rare cases, the entire apparatus can migrate distally, resulting in gastric outlet obstruction. The child complains of retching or sudden onset of emesis, and appears uncomfortable. The G-tube needs to be pulled back to its proper location until it is snug against the abdominal wall. If this procedure is not successful, the tube must be removed completely.

Stomal Complications

Irritant Dermatitis/Allergic Hypersensitivity. Skin irritation around the stoma may result from chronic leakage of gastric or jejunal fluid around the tube. If the stoma widens, the leakage may become excessive, resulting in more significant dermatitis. Various brands of adhesives and cleansing solutions may result in an allergic rash around the stoma.

The peristomal skin should be thoroughly cleansed and dried before assessment. Small vesicular lesions with surrounding erythema suggest irritant dermatitis. Local treatment includes keeping the area as dry as possible and using barrier creams to protect the skin from further breakdown. Stomahesive Power (Convatec, Princeton, New Jersey) is useful to mold to the skin surface and keep the area dry and free of debris. In addition, identifying and treating the cause of the leakage is important. If the leakage is caused by an enlarged stoma, surgical intervention may be required in the near future.

Granulation. Children with gastrostomy tubes may develop granulomatous tissue in the peristomal area. These lesions are harmless but occasionally become infected or bleed. Granulomatous tissue typically appears in the borders of the stoma and may begin to cause occlusion of the stoma. The most effective treatment is silver nitrate swabs.

Cellulitis. When peristomal skin surrounding the G-tube or J-tube is irritated by recurrent or intermittent exposure to drainage, cellulitis may occur. The infection may begin as a superficial skin irritation or contact dermatitis and then evolve into a deeper infection. The surrounding peristomal area may become reddened, warm, tender, and edematous. These symptoms and signs may be accompanied by systemic symptoms and fever. The patient with a G-tube may become resistant to gastrostomy feedings because of the discomfort associated with manipulation of the apparatus. Once cellulitis is present, the patient requires systemic antibiotics for resolution of this infection. The common organisms, staphylococci and streptococci usually respond to a first-generation cephalosporin. Occasionally, the cellulitis can be complicated by a peristomal abscess, heralded by a localized area of fluctuance. This abscess requires incision and drainage before antibiotic administration.

Fungal Infection. Recurrent moisture caused by gastric or jejunal leakage in the stomal area can predispose the patient

to fungal infection. The most common causal organism is *Candida albicans,* appearing as fiery red plaques at the stoma site. Topical clotrimazole is curative in most situations. Keeping the area as dry as possible is imperative.

Gastrointestinal and Genitourinary Diversion

Background

Pediatric patients may have a GI or genitourinary diversion for one of many reasons. Congenital causes include Hirschsprung's disease, imperforate anus, cloacal exstrophy, bladder exstrophy, meningomyelocele with a neurogenic bladder, and posterior urethral valves. Acquired lesions may include ulcerative colitis, Crohn's disease, and necrotizing enterocolitis. Traumatic injuries leading to GI or genitourinary diversion include penetrating wounds and falls.

GI diversions consist primarily of colostomy and ileostomy. Colostomy brings the colon to the skin. Patients with colostomies usually have semi-formed stools because the absorptive and storage function of the bowel is preserved. An ileostomy brings the ileum to the skin. Ileostomy patients do not possess large bowel function and consequently have a watery, frequent stooling pattern.

The major forms of urinary diversions consist of ureterostomy, vesicostomy, nephrostomy, and ileal conduits. Ureterostomy brings the dilated ureter to the level of the skin, whereas an ileal conduit implies that the ureters are attached to the ileum, which is then externalized. A vesicostomy opens the bladder to the skin. Nephrostomy indicates that a tube is placed into the renal pelvis percutaneously, usually by an interventional radiologist. Urinary undiversion indicates that the patient has undergone a surgical procedure that internalizes the urine passage via a “neobladder.”

In general, a stomatherapist is crucial to the physicians and families of all patients with stomal sites and appliances. However, patients continue to present to the ED with ostomy-related problems and the emergency physician should become facile with the various types of GI and genitourinary diversions and their specific complications.

Pathophysiology

The nature of the disease and the location of the lesion(s) guide the surgeon when choosing the type of diversion to use. Ileostomy is usually performed in newborns for conditions such as meconium ileus, necrotizing enterocolitis, and intestinal atresia. It may be required in older children and adolescents because of ulcerative colitis or polyposis. The surgical method used depends on the predicted length of time required for the ostomy, as well as the location of the disease.

Colostomy in infants is required for complications of colonic atresia, high forms of imperforate anus, and Hirshsprung's disease. The level of the colostomy is related to the disease type and to anticipated future procedures. Some of the ostomy complications that occur in patients with genitourinary and GI diversions are similar. In both types of diversions, many complications relate to the actual stoma. These conditions are discussed in the previous enteral feeding section. Other complications are metabolic or mechanical in nature.

Vesicostomy is usually performed for patients with myelomeningocele, posterior urethral valves, prune belly syndrome, and spinal cord injury resulting in a neurogenic bladder. This procedure is protective of the upper urinary tract.

Ureterostomy is accomplished by bringing the ureter to the surface of the skin either in the groin (low) or in the flank (high). Most high ureterostomies are of the loop variety, in which a loop of ureter is incised on one side, and passed upward to allow the edges to be anastomosed to the skin. This path allows ureteral continuity from the kidney to the bladder, with a vent to the skin. Low ureterostomies are more common, performed for obstructed ureters such as ectopic ureters or megaureters. To decompress an obstructed system and prevent urinary tract infection, the ureter is divided, the distal end is ligated, and the proximal edges are anastomosed to the skin.

Ileal loop conduits are created using a resected 10 to 20 cm bowel segment of ileum and anastomosing both ureters to one end. The other end of the bowel loop is brought out to the skin. Ileal loop conduits are preferable in older children who can wear an appliance to collect the urine.

Equipment

Standard ostomies are commonly managed by placing an ostomy pouch over the stoma to collect the effluent. In young infants, sigmoid colostomies may be managed without an external pouch if the effluent is not caustic to the skin, and fluid is therefore collected in the diaper. Urinary flow can also be collected in a diaper, and may be preferable because some appliances do not adhere well to the skin for long periods.

Ostomy pouches for children are manufactured in a variety of sizes. One- and two-piece configurations are available, and pouches may be soft or rigid. Supplemental adhesives are crucial to enhance adhesion. In general, the more liquid the effluent, the greater the need for adhesive substances.

Clinical Findings/Management

Gastrointestinal Diversions

Patients with colostomies and ileostomies may present with complications that are common to both types of ostomies. Ileostomies also have metabolic complications that are specific to this type of ostomy.

Cutaneous Complications. Peristomal cutaneous complications are common in patients with ostomies, and stem from

the effect of chronic stool and other drainage on the peristomal skin. This chronic drainage compromises the skin integrity surrounding the stoma. The most effective management is the maintenance of a good seal between the ostomy pouch and the stoma. Contact dermatitis may occur from leakage around the stoma or from allergy to stomal materials such as tape or pouches. This condition is often successfully treated by removing the offending material. Infection with *C. albicans* is fairly common because of the persistent moisture and the frequent use of prophylactic antibiotics. Treatment with antifungal agents such as clotrimazole, especially powders, is effective. The powder can be mixed with a small amount of water and painted onto the skin to enhance adherence of the pouch. Ointments and creams should be avoided in fungal infections. Skin bleeding resulting from mechanical trauma from the stomal tube is usually minor. The cellulitis that can occur if the skin excoriation worsens is treated with systemic antibiotics.

Stomal Complications. Stomal stenosis is not always detectable to the parent or practitioner and may present with reduced or absent output, diarrhea, or cramping abdominal pain. When severe stenosis occurs, it usually presents as obstruction. To assess the degree of stenosis, the physician should gently examine the stoma digitally unless the stoma is too small. In this case, a catheter should be carefully passed. If abdominal obstruction is suspected, radiographs of the abdomen and urgent surgical consultation are indicated.

Prolapse of stoma occurs in greater than 20% of patients with stomas and is usually not an emergency. However, skin excoriation, bleeding, and incarceration of the bowel may occur. The situation becomes more urgent if the prolapse is associated with pain, decreased output, or a dusky stoma color that represents circulatory compromise. Management includes easing the prolapsed contents back into the stoma using both hands. This procedure may need to be done repetitively.

Retraction of the stoma because of excessive tension may cause the stoma to recede beneath the skin. This condition occurs more often than prolapse in patients with ileostomies. Stomal retraction makes it difficult for a pouch to adhere to the skin. Retraction can also result in cellulitis or even peritonitis depending on the location of the detachment and the flow of the effluent. Management usually includes antibiotics. Surgical correction may be required if the stomal detachment reaches the fascial layer.

A hernia of the peristomal contents occurs when there is a protrusion of the colon or ileum into the subcutaneous layers of skin surrounding the stoma. This complication may impede adherence of the ostomy pouch but does not represent an emergency.

Complications Specific to Ileostomy. Patients with ileostomies occasionally develop metabolic derangements. In the face of large volume losses, children tend to become salt and water depleted. If large fluid losses persist, the biochemical profiles of these patients are significantly altered. Determining the cause of the exceptionally high fluid losses from the ileostomy is crucial. Some possibilities are obstruction, gastroenteritis, and dietary indiscretion. Treatment is aimed at restoring normal fluid and electrolyte balance and may require hospital admission.

Patients with ileostomies are prone to acquiring urinary stones. The chemical composition of stones in this scenario is different than that in normal patients; uric acid stones constitute 60% and calcium oxalate makes up the remainder. Treatment is directed at decreasing ileostomy output and increasing urine output.

Urinary Diversions

Vesicostomy. In patients with vesicostomy, eversion of a large portion of the bladder can occur and appear like an exstrophy. When the posterior aspect of the bladder prolapses through the stoma, the patient presents with a red mass, which may progress to purple if not treated promptly. This condition may be managed by applying an index fingertip to the bladder and gently pushing inward. Non-latex gloves are used because children with urologic abnormalities are often allergic to latex. Sedatives may be required to facilitate reduction of the prolapse. A prolapsed vesicostomy should be surgically revised if the manual reduction is unsuccessful.

Patients with stomal stenosis of the vesicostomy usually present with a palpable bladder, a history of unwanted urethral voiding, or with symptoms of urinary tract infection. As the bladder fails to empty at low pressures, the mean storage pressure rises and the chance for seeding bacteria into the upper urinary tract increases. These patients often have a pinpoint opening to the bladder, and the parents usually comment on how much smaller the stoma has become over time. If possible, these patients should have a catheter placed via the vesicostomy using a small (6 or 8 Fr) catheter. The urethra should be catheterized for urine to establish a diagnosis of urinary tract infection. If the vesicostomy is successfully catheterized, the catheter should be left in place until surgical revision may be carried out.

All vesicostomies are colonized with bacteria via stomal contamination. Therefore, a catheterized specimen through the stoma is unreliable. Patients with constitutional symptoms such as fever are treated. Otherwise, a positive culture may represent asymptomatic bacteriuria and is not always of concern.

Skin irritation in the area of the vesicostomy is unusual. The most important preventive measure is frequent diaper changes, even if highly absorbent diapers are used. If urine seeps onto the patient's clothes repetitively, skin breakdown may ensue. In severe cases, temporary urinary diversion with a Foley catheter while applying a barrier ointment allows time for healing.

Ureterostomy. Stenosis is the most common complication in the patient with a ureterostomy. These patients often present with fever and symptoms suggestive of pyelonephritis. The stoma should be catheterized with an 8-Fr catheter, and urine should be sent for culture. Ureterostomy prolapse is rare.

Ileal Loop Conduits. Inflammation of the peristomal skin arises when the appliance fits poorly around this bud of ileum, allowing urine to seep under the protective wafer. Prolonged contact with skin causes irritation and ulceration. The use of paste to create a better seal around the bud is often all that is needed to avoid such a complication. In some cases,

surgical revision is necessary, especially when the bud has retracted.

Prolapse of the ileum occurs occasionally and can be striking, especially if too long of a segment was used in creating the loop initially. Prolapsed segments 20 to 30 cm long have been seen and require surgical revision. If the prolapse is minor, the clinician should perform the same gentle manual reduction technique described in the previous section on stomal complications of GI diversions.

Peristomal hernia can occur when fascial defects adjacent to the ileal loop allow loops of bowel to herniate outside the abdominal wall. This condition requires urgent surgical consultation.

Stenosis of the ileal stoma may occur in these patients. Symptoms may include pain, but the usual presenting complaint for these patients is fever. This finding necessitates a workup for pyelonephritis. Stomal stenosis can also lead to formation of urinary calculi.

Urinary Undiversions. As the child with a vesicostomy or ileal loop grows older, the social stigma of a diaper motivates many of these patients to seek urinary continence. For patients with spina bifida or exstrophy, this goal may be achieved by the use of an intestinal segment to augment bladder capacity as well as a procedure to tighten the bladder neck and create a resistance to leakage. For all spina bifida patients, and most exstrophy patients, continence comes at the expense of daily clean intermittent catheterization for the rest of their lives. Careful patient and family selection is necessary for this procedure; compliance with clean intermittent catheterization is crucial. Nevertheless, the enhanced self-esteem and improved quality of life these patients report is gratifying.

Perforation is the worst complication of intestinal augmentations to create neobladders. Most bladder perforations result from overdistension of the augmented bladder, which then diminishes perfusion to the bowel segment. In addition, the urine in these neobladders is chronically colonized secondary to the use of intermittent catheterization. Patients may present anywhere from 1 month to many years after surgery with a history of acute abdominal pain. Fever may be present within a few hours of perforation. Because many spina bifida patients have decreased or absent abdominal sensation, peritonitis may be fairly advanced before pain is experienced. The presence of abdominal pain in a patient with a urinary diversion should prompt an immediate call to the patient's urologist. The urologic evaluation generally consists of a fluoroscopic gravity cystogram with views during filling and emptying. Small perforations may be obscured with the full bladder and become apparent only during bladder emptying. Prophylactic antibiotics should be administered before the cystogram. Once this diagnosis is established, the patient is prepared for emergency laparotomy.

Patients may present to the ED with a sudden inability to pass a catheter into their neobladder. This situation may be because the appendiceal conduit through which they pass their catheter contains a false passage. A fluoroscopic study is warranted to delineate the passage and allow catheterization under radiographic control. The same situation is often true for patients catheterizing per urethra. In some cases, the urologists may opt to take a patient to the operating room for emergency endoscopy to define the obstruction point. When all else fails and the patient's bladder continues to distend, it is safest to pass a suprapubic drainage catheter into the neobladder.

Because the creation of a neobladder is an intraperitoneal operation, these patients are all at risk for developing small bowel obstructions. A patient with abdominal pain and a neobladder merits radiographic evaluation.

Up to 30% of patients with a neobladder develop stones within their pouch and require either endoscopic or open surgical removal. These stones rarely cause pain by obstruction, but rather they produce a foul urine that can be so irritating to the neobladder that the patient presents with a vague lower abdominal pain. These stones are calcified and show up on an abdominal radiograph. Treatment with antibiotics is palliative until surgical removal is undertaken.

The insertion of bowel segments into the urinary tract carries with it certain fluid and electrolyte complications that may not be a problem under normal circumstances. However, with a GI virus and superimposed diarrhea and dehydration, the patient may not be able to compensate. For example, a patient with a gastric augmentation who presents with diarrhea and lethargy may prove to have a severe hypochloremic hyponatremic metabolic alkalosis. Thus, any patient with a bladder augmented with bowel who is obtunded requires careful consideration of an electrolyte disturbance as the underlying cause.

Suggested Readings

ACUTE CARE OF CHRONICALLY ILL CHILDREN

Rappo P. The care of children with chronic illness in primary care practice: implications for the pediatric generalist. Pediatr Ann 1997;26:687–695.

Reynolds S, Desguin B, Uyeda A, Davis AT. Children with chronic conditions in a pediatric emergency department. Pediatr Emerg Care 1996; 12:166–168.

Schonfeld N, McMorrow M. ED evaluation of the multiply handicapped child. Pediatr Trauma and Acute Care 1990;3:25–26.

TRACHEOSTOMY CARE

Brook I. Bacterial colonization, tracheobronchitis, and pneumonia following tracheostomy and long-term intubation in pediatric patients. Chest 1979;76:420–424.

Downes JJ, Schreiner MS. Tracheostomy tubes and attachments in infants and children. Int Anesthesiol Clin 1985;23:37–60.

Fitton CM. Nursing management of the child with a tracheotomy. Pediatr Clin North Am 1994;41:513–523.

Kallis JM. Replacement of a tracheostomy cannula. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:871–876.

Katz RL. Tracheotomy care. In: Roberts JR, Hedges JR, eds. Clinical Procedures in Emergency Medicine. Philadelphia: WB Saunders, 1991:60–64.

Niederman MS, Ferranti RD, Zeigler A, et al. Respiratory infection complicating long-term tracheostomy. Chest 1984;85:39–44.

Panitch HB, Downes JJ, Kennedy JS, et al. Guidelines for home care of children with chronic respiratory insufficiency. Pediatr Pulmonol 1996;21:52–56.

Pilmer SL. Prolonged mechanical ventilation in children. Pediatr Clin North Am 1994;41:473–512.

Quint RD, Chesterman E, Crain LS, et al. Home care for ventilator-dependent children. Am J Dis Child 1990;144:1238–1241.

Schreiner MS, Downes JJ, Kettrick RG, et al. Chronic respiratory failure in infants with prolonged ventilator dependency. JAMA 1987;258: 3398–3404.

Schreiner MS, Kettrick RG, Balderson P, et al. Swivel-connector system for pediatric tracheostomy: an improvement. Respir Care 1986;31:109–112.

CEREBROSPINAL FLUID SHUNTS

Ammirati M, Raimondi AJ. Cerebrospinal fluid shunt infections in children. A study on the relationship between the etiology of hydrocephalus, age at the time of shunt placement, and infection rate. Child Nerv Sys 1987; 3:106–109.

Bondurant CP, Jiminez DF. Epidemiology of cerebrospinal fluid shunting. Pediatr Neurosurg 1995;23:254–259.

Borgbjerg BM, Gjerris F, Albeck MJ, Borgesen SE. Risk of infection after cerebrospinal fluid shunt: an analysis of 884 first-time shunts. Acta Neurochirurgica 1995;136:1–7.

Borgbjerg BM, Gjerris F, Albeck MJ, et al. Frequency and causes of shunt revisions in different cerebrospinal fluid shunt types. Acta Neurochirurgica 1995;136:189–194.

Duhaime AC, Wiley JF II. Ventricular shunt and burr hole puncture. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:559–565.

Ersahin Y, Mutluer S, Guzelbag E. Cerebrospinal fluid shunt infections. J Neurosurg Sci 1994;38:161–165.

Guertin SR. Cerebrospinal fluid shunts: evaluation, complications, and crisis management. Pediatr Clin North Am 1987;34:203–217.

Iskandar BJ, McLaughlin C, Mapstone TB, et al. Pitfalls in the diagnosis of ventricular shunt dysfunction: radiology reports and ventricular size. Pediatrics 1998;101:1031–1036.

Johnson DL, Conry J, O'Donnell R. Epileptic seizure as a sign of cerebrospinal fluid shunt malfunction. Pediatr Neurosurg 1996;24:223–227.

Key CB, Rothrock SG, Falk JL. Cerebrospinal fluid shunt complications: an emergency medicine perspective. Pediatr Emerg Care 1995;11:265–273.

Madikians A, Conway EE Jr. Cerebrospinal fluid shunt problems in pediatric patients. Pediatr Ann 1997;26:613–620.

McLaurin RL. Ventricular shunts: complications and results. Pediatr Neurosurg 1989;2:219–229.

Meirovitch J, Kitai-Cohen Y, Keren G, et al. Cerebrospinal fluid shunt infections in children. Pediatr Infect Dis J 1987;6:921–924.

Morissette I, Gourdeau M, Francoeur J. CSF shunt infections: a fifteen-year experience with emphasis on management and outcome. Can J Neurol Sci 1993;20:118–122.

Nelson JD. Cerebrospinal fluid shunt infections. Pediatr Infect Dis 1984; 3:30–32.

Odio C, McCracken GH Jr, Nelson JD. CSF shunt infections in pediatrics. A seven-year experience. Am J Dis Child 1984;138:1103–1108.

Piatt JH. Physical examination of patients with cerebrospinal fluid shunts: is there useful information in pumping the shunt? Pediatrics 1992; 89:470–473.

Ronan A, Hogg GG, Dlug GL. Cerebrospinal fluid shunt infections in children. Pediatr Infect Dis J 1995;14:782–786.

Schwartz G, Seamens C. Medical hardware inside software: CSF shunts and indwelling devices in children. Pediatr Emerg Med Reports 1997;2:39–50.

Walker M, Fried A, Petronio J. Diagnosis and treatment of the slit ventricle syndrome. Neurosurg Clin North Am 1993;4:701–714.

Walters BC. Cerebrospinal fluid shunt infection. Neurosurg Clin North Am 1992;3:387–401.

Woodward GA, Carey CM. Evaluation of ventricular shunt function. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997.

Yogev R. Cerebrospinal fluid shunt infections: a personal view. Pediatr Infect Dis 1985;4:113–118.

INDWELLING CATHETERS

Dyer BJ, Weiman MG, Ludwig S. Central venous catheters in the emergency department: access, utilization, and problem solving. Pediatr Emerg Care 1995;11:112–117.

Fuchs SM. Accessing indwelling central lines. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Procedures. Baltimore: Williams & Wilkins, 1997:811–820.

Greene FL, Moor W, Strickland G, et al. Comparison of a totally implantable access device for chemotherapy (Port-A-Cath) and long-term percutaneous catheterization (Broviac). South Med J 1988;81:580–583.

Hartman GE, Schochat SJ. Management of septic complications associated with Silastic catheters in childhood malignancy. J Pediatr Infect Dis 1987;6:1042–1047.

Howell JM. Accessing indwelling lines. In: Roberts JR, Hedges JR, eds. Clinical Procedures in Emergency Medicine. 2nd ed. Philadelphia: WB Saunders, 1991:357–363.

Mirro J, Rao B, Kumar M, et al. A comparison of placement techniques and complications of externalized catheters and implantable port use in children with cancer. J Pediatr Surg 1990;25:120–124.

Nahata MC, King DR, Powell DA, et al. Management of catheter-related infections in pediatric patients. J Parent Ent Nutr 1988;12:58–59.

Reynolds S, Desguin B, Uyeda A, Davis TA. Children with chronic conditions in a pediatric emergency department. Pediatr Emerg Care 1996;12:166–168.

Stokes DC, Rao BN, Mirro J, et al. Early detection and simplified management of obstructed Hickman and Broviac catheters. J Pediatr Surg 1989;24:257–262.

Wagman LD, Konrad P, Schmit P. Internal fracture of a pediatric Broviac catheter. J Parent and Ent Nutr 1989;13:560–561.

ENTERAL FEEDING

American Gastroenterological Association. Technical review on tube feeding for enteral nutrition. Gastroenterology 1995;108:1282–1290.

Browne B, Kauffman B, Brown C. Internal displacement of a gastrostomy button: an unusual case of gastric outlet obstruction. J Pediatr Surg 1993;28:1575–1576.

Clevenger FW, Rodriguez D. Decision making for enteral feeding administration: the why behind where and how. Nutr Clin Practice 1995;10:104–113.

Gauderer MWL, Olsen MM, Stellato TA, Dokler ML. Feeding gastrostomy button: experience and recommendations. J Pediatr Surg 1988; 23:24–28.

Graneto JW. Gastrostomy tube replacement. In: Henretig FM, King C, eds. Textbook of Pediatric Emergency Medicine Procedures. Baltimore: Williams & Wilkins, 1997:915–920.

Kazi S, Gunasekaran TS, Berman JH, et al. Gastric mucosal injuries in children from inflatable low-profile gastrostomy tubes. J Pediatr Gastroenterol and Nutri 1997;24:75–79.

Minard G. Enteral access. Nutri in Clin Practice 1994;9:172–182.

Mollitt DL, Golladay ES, Seibert JJ. Symptomatic gastroesophageal reflux following gastrostomy in neurologically impaired patients. Pediatrics 1985;75:1124–1126.

Paarlberg J, Balink JP. Gastrostomy tubes: practical guidelines for home care. Pediatr Nurs 1985;11:99–102.

Steele NF. The button: replacement gastrostomy device. J Pediatr Nurs 1991;6:421–424.

Tsarouhas N. Tube placement. In: Dieckmann RA, Fiser DH, Selbst SM, eds. Illustrated Textbook of Pediatric Emergency and Critical Care Procedures. St Louis: Mosby, 1997:366–369.

GASTROINTESTINAL DIVERSION

Garvin G. Caring for children with ostomies. Pediatr Surg Nurs 1994; 29:645–654.

Hellman J, Lago C. Dermatologic complications in colostomy and ileostomy patients. Int J Dermatol 1990;29:129–133.

Hill GL. Part 3: metabolic complications of ileostomy. Clin Gastroenterol 1982;11:260–268.

Todd IP. Part 4: mechanical complications of ileostomy. Clin Gastroenterol 1982;11:268–272.

Walton SA, Hopkins J, Garvin G, Grisham J. Ostomy complications in pediatrics. In: Dieckmann RA, Fiser DH, Selbst SM, eds. Illustrated Textbook of Pediatric and Critical Care Procedures. St Louis: Mosby, 1997:699–700.

GENITOURINARY DIVERSION

Benson MC, Olsson CA. Urinary diversion. Urol Clin North Am 1992; 19:779–795.

Cass AS, Luxenberg M, Johnson CF, Gleich P. Management of the neurogenic bladder in 413 children. J Urol 1984;132:521–525.

Duckett JW, Ziylan O. Uses and abuses of vesicostomy. Updates 1995; 14:130–135.

Mitchell M, Rink RC. Pediatric urinary diversion and undiversion. Pediatr Clin North Am 1992;34:1319–1332.

Noe HN. Complications of cutaneous vesicostomy and ureterostomy. In: Marshall FF, ed. Urologic Complications: Medical and Surgical; Adult and Pediatric. Chicago: Year Book Medical, 1986:341–350.

Noe HN, Jenkins GR. Cutaneous vesicostomy experience in infants and children. J Urol 1985;134:301–303.

Ring KS, Hensle TW. Urinary diversion. In: Kelalis P, King L, Belman EB, eds. Clinical Pediatric Urology. Philadelphia: WB Saunders, 1992: 865–903.

CHAPTER 128

Child Abuse

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Child abuse is the single diagnostic term used to describe a range of behaviors from somewhat harsh discipline to intentional repetitive torture. This phenomenon is complex and results from a combination of individual, familial, and societal factors. The common pathway for all these factors is parental behavior destructive to the process of normal growth, development, and well-being of the child. Abuse may be subdivided into four broad categories: 1) physical abuse, 2) sexual abuse, 3) neglect, and 4) emotional abuse. Each form of abuse has individual characteristics of family dynamics, clinical manifestations, and management.

The task of the emergency physician is difficult. The physician must first maintain an open mind to the possibility that abuse not only occurs, but occurs commonly. Thus, abuse should be included in the differential diagnosis of any injury or any unusual physical or psychological complaint. Second, the physician must identify signs and symptoms of suspected abuse. Next, the family crisis must be managed to protect the child yet maintain the abusive parents' motivation for help. Finally, the legal requirements for reporting abuse to the proper social service or police authority should be thoroughly understood.

The demands of managing a case of child abuse may be lessened by sharing the responsibility with other health care professionals. The skills of nursing and social work staff are invaluable. The child abuse field has been a model for multidisciplinary collaboration, which is most productive if begun in the initial phases of case management in the emergency department (ED). Establishing an institutional or departmental protocol for the management of abuse cases is also helpful. This protocol relieves the emergency physician from having to reconstruct a complete management plan for each new case. Having a standard protocol to follow allows the physician more time to concentrate on the individual needs of the patient and parents.

To the unfamiliar observer, the easy solution to all abuse cases is to “take away the child and put the parents in jail.” This commonly held treatment philosophy would be practiced more if it were truly a panacea. However, the alternative forms of child care (i.e., institutional care, foster care, extended family care) are each fraught with their own hazard. With the use of well-organized community services, abusive behavior can be controlled while the child and family receive therapy.

There may be support for the notion that parents who bring their abused child to the ED are motivated to seek help for their child and for themselves. Most parents remorse about their abusive behavior. The severity of injuries inflicted is often overestimated as a result of parental guilt. The emergency physician must neither overlook nor mismanage the opportunity to identify abuse and control the parent's behavior. Sharp focus must be maintained on the dual goals of case management—protect the child and use the crisis to strengthen and preserve family life.

PHYSICAL ABUSE

Background

Physical abuse is the most often reported form of child abuse. Definitions of physical abuse vary from state to state. Operationally, the definitions vary from institution to institution, and indeed from person to person. Even the definition of physical abuse is a definition in transition. Over the past century, many advances in the “rights of the children” have been made. For example, the enactment of child labor and compulsory education laws has been an important step forward. As the history of abuse is traced through the centuries, the forms and the definitions of abuse have changed. Definitions currently used are likely to continue to change with time. The present widespread medical interest in abuse was stimulated by C. Henry Kempe with the introduction of the term *“battered child syndrome”* in 1962. It was only as recently as 1968 that the last of the 50 states enacted child abuse legislation. Many states are now using their second or third generation of child abuse laws.

The Child Abuse Prevention and Treatment Act (CAPTA), as amended and reauthorized in October 1996 (Public Law 104-235, Section 111;42 U.S.C. 5106g), defines child abuse and neglect as, at a minimum, any recent act or failure to act:

- Resulting in imminent risk of serious harm, death, serious physical or emotional harm, sexual abuse, or exploitation
- Of a child (a person under the age of 18, unless the child protection law of the State in which the child resides specifies a younger age for cases not involving sexual abuse)
- By a parent or caretaker (including any employee of a residential facility or any staff person providing out-of-home

care) who is responsible for the child's welfare.

There are four major types of child maltreatment: 1) physical abuse, 2) child neglect, 3) sexual abuse, and 4) emotional abuse. Physical abuse is the infliction of physical injury as a result of punching, beating, kicking, biting, burning, shaking, or otherwise harming a child. The parent or caretaker may not have intended to hurt the child, but rather the injury may have resulted from over-discipline or physical punishment.

The true incidence of abuse is unknown. Many cases go unrecognized and unreported. Estimates for all forms of abuse vary from 500,000 to 4 million, cases per year in the United States. Three national incidence studies have been conducted. The incidence across the three studies is shown in Figure 128.1. The incidence rates for abuse and neglect is 15 per 1,000 American children. A Gallup poll estimated as many as 49 per 1,000 children were physically abused and 19 per 1,000 were sexually abused. Physical abuse accounts for 25% of reported cases. Several studies have estimated that 10% of children less than 5 years old brought to the ED with traumatic injury are victims of child abuse. There may be as many as 2000 abuse-related deaths per year in the United States.

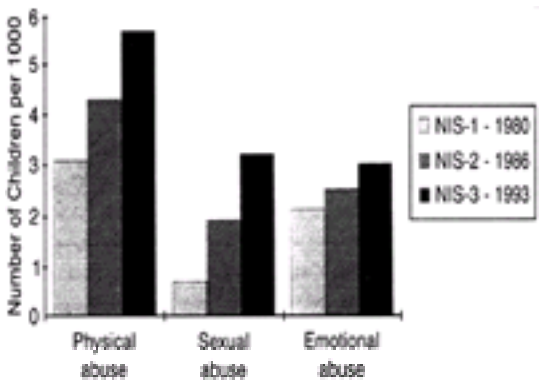


FIGURE 128.1. Incidence of child abuse per 1000 children by abuse type determined on three National Incidence Studies. (Reprinted with permission from the U.S.

Homicide is now the fifth leading cause of death in children aged 1 to 4 years and the fourth leading cause of death in children aged 5 to 14 years. There are 2,000 to 5,000 deaths annually or an incidence of 5.4 per 100,000 children aged 4 and younger. The incidence of child homicide has steadily increased. The rate of homicide in the 1- to 4-year-old age group has increased sixfold since 1925, according to a review by the Centers for Disease Control and Prevention. The perpetrators in these cases of child homicide are most often adults who are known by their child victims.

Although the true incidence of abuse is in question, the number of reports has certainly escalated. Figure 128.2 shows the number of substantiated cases from 1976 to 1997 in Pennsylvania. Pennsylvania established a statewide reporting registry in 1975. Most authorities believe the increase in reports is attributable to an increase in awareness and ease of reporting, as well as to a rise in the true incidence of abuse.

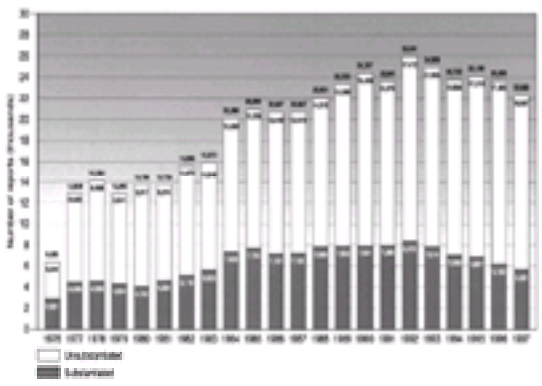


FIGURE 128.2. Child abuse from 1976 to 1997 in Pennsylvania. (Reprinted with permission from the Pennsylvania Department of Public Welfare Office of Children Youth and Families. 1997 Child Abuse Report.)

Dynamics

Many factors contribute to the reasons a parent abuses a child. Helfer's formulation of the necessary elements is shown in Figure 128.3. The factors include a parent who is capable of abuse, a child who actively or passively becomes the target, and a crisis that triggers the angry response. Frederick Green has added to this triad the concept that the process must exist in a society that unknowingly condones or even encourages violence, and in particular violence against children. Some of the factors that contribute to the parents' abusive potential are listed in Figure 128.4.

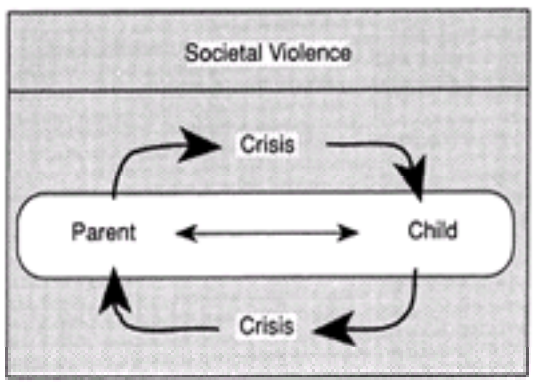


FIGURE 128.3. Essential elements of child abuse. (Modified with permission from Helfer RE. Why most physicians don't get involved in child abuse cases and what to do about it. Child Today 1975;4:28.)

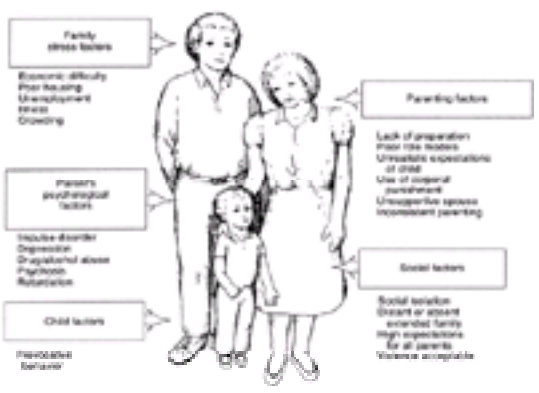


FIGURE 128.4. Risk factors that contribute to abuse and neglect.

Stress and lack of specific child-rearing information and experience play dominant roles. The combination of these factors causes many parents to misread normal childhood behavior as defiant or provocative and to react with a violent, destructive response. The typical example is the parent who is angered by the 1-year-old child's refusal to become toilet-trained. The child's contribution to abuse may be real, as in the case of negative behavior or disparate temperament, or imagined by the parent (e.g., “he's just like his father”). Children with prolonged neonatal hospitalization, handicapping conditions, or developmental delays are at increased risk. Living situations in which nonbiologic parents are present are also high risk. For example, the adopted child or the child living with a parent's paramour may be the target of abusive behavior. The crisis that initiates abuse varies tremendously. It may be unrelated to the child, such as the stress of a family member's death or economic disappointment. However, crisis often occurs because the child's behavior does not meet parental expectations. The crisis is identifiable as the spark that ignites the existing potential for abuse.

Manifestations

The manifestations of physical abuse may affect any body system. Thus, the emergency physician must be prepared to recognize a variety of signs and symptoms. Abuse may also be seen by any specialist physician. National data show neglect to be most common (Fig. 128.5). Injuries seen at The Children's Hospital of Philadelphia between 1976 and 1979 are shown in Figure 128.6. These proportions have remained constant.

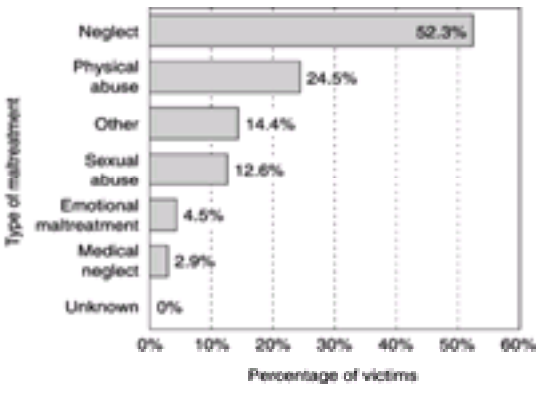


FIGURE 128.5. Types of maltreatment as a percentage of victims (1995) based on surveys in 49 states. *N* = 1,000,502 victims in 49 states. (Note: Percentages total more than 100% because some states reported more than one type of maltreatment per victim.)

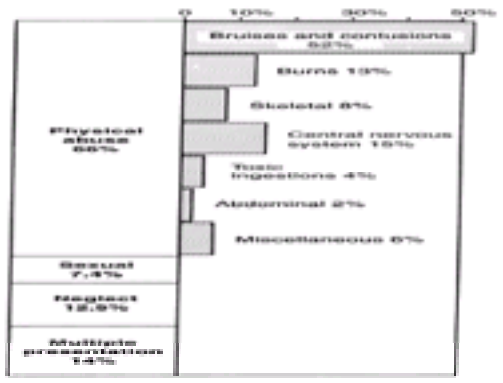


FIGURE 128.6. Types of child abuse injuries seen at Children's Hospital of Philadelphia from 1976 to 1979. (Courtesy of Dr. M. Zeigler.)

Integument

The skin is the most commonly injured body organ. Cutaneous injuries may be divided into nonspecific and specific traumatic lesions, burns, and hair loss. Of the nonspecific traumatic injuries, the bruise or contusion is the most commonly seen. Although bruises are also common in children who are not abused, accidental bruises usually have a different distribution and appearance. Accidental injuries occur most commonly on the extremities and forehead. As bruising moves centrally and becomes extensive, the likelihood of abuse rises. Contusions undergo recognizable stages of healing. In the first 24 hours, the size of the bruise increases slightly if careful measurements are made. The process of resolution is variable. The bruise should be dated and compared with the history provided. A prothrombin time, partial thromboplastin time, bleeding time, and platelet count should be obtained if the issue of “easy bruisability” has been offered as a possible explanation.

Other nonspecific cutaneous injuries include lacerations, punctures, and abrasions. The following criteria are important for the evaluation of any nonspecific injury: 1) the history of injury, 2) the child's age and developmental level, 3) the presence of other old or new injuries, 4) the interaction between the parents and child, and 5) the interaction between the parents and the ED staff.

Specific skin injuries are those that clearly reflect the method or object used to inflict the trauma. Loop-shaped marks are readily seen after a beating with an electric cord or wire. Linear marks may be seen from a belt or paddle injury. Rope burns result in circumferential marks on the wrists, ankles, or around the neck when a child has been bound. Another common specific integument lesion is a hand print on the side of the face or symmetrically on the upper arms. The lesion produced by a slap leaves ecchymotic areas in the location of the interphalangeal spaces. Human bites appear as circular lesions 1 to 2 inches in diameter. Forensic dentistry is able to match the skin lesion with the dentition of the alleged perpetrator. Some specific skin lesions are shown in Figure 128.7.

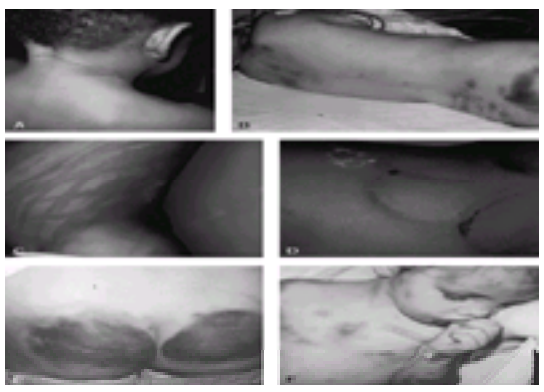


FIGURE 128.7. Cutaneous manifestations of child abuse. **A.** Strangulation mark. **B.** Bruises at various stages of healing. **C.** Linear loop-shaped marks. **D.** Multiple loop-shaped marks. **E.** Buttocks bruises as a cause of myoglobinuria. **F.** Multiple bruises in a central pattern.

Burns of the skin may be caused by abuse or neglect. Burns account for 5% of cases of physical abuse. In particular, tap water scald burns that occur in an immersion pattern (Fig. 128.8A) are often the result of intentional trauma. Immersion burns are likely to be inflicted by an abusive parent when they occur on a child who is being toilet-trained. Other indications of abuse are 1) a delay in seeking treatment, 2) a history of the child being unsupervised, and 3) the child being brought to the hospital by the parent who was not present at the time the burn occurred.

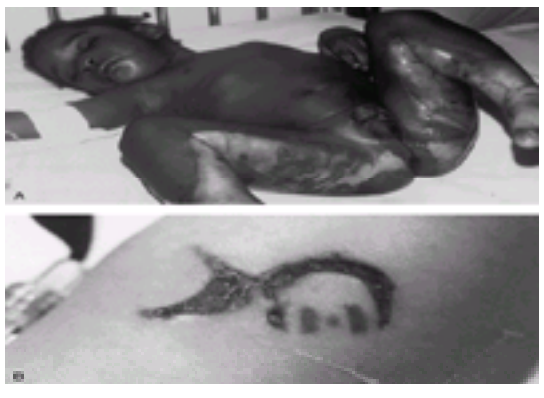


FIGURE 128.8. A. Hot water burn in an immersion pattern. **B.** Pattern burn from cigarette lighter.

In attempting to match the physical findings of the burn with the available history, several factors must be appreciated. The extent of the burn depends on the temperature of the water, duration of exposure, thickness of the skin involved, and presence or absence of clothing. Water temperature 54°C (130°F) or greater causes a full-thickness burn with less than a 30-second exposure. Because palms and soles are thick, they are often spared. Clothing tends to keep the hot water in contact with the skin and causes more severe burns. Burns presumably caused by falling or thrown fluids should produce a droplet or splash pattern. When the child has several small bullous lesions, the main differential diagnosis is a second-degree burn versus bullous impetigo. This differentiation is easily made by Gram stain and culture of a bulla.

Other burns may occur through contact with a hot solid rather than a hot fluid. Cigarette burns are the most common of this type. If the history given is of a child brushing against a cigarette or of hot ashes falling on the child, the resulting injury should be a nonspecific first- or second-degree burn. When a cigarette is extinguished on the child's skin, the injury is a burn that is 8 to 10 mm in diameter and indurated at its margin. A healed cigarette burn is indistinguishable from any other circular skin lesion such as impetigo, abscess, or vesicles. Burns from radiators, hot plates, cigarette lighters (Fig. 128.8B), curling irons, or standard irons imprint the shape of the hot object. Recently, there have been reports of children burned by microwave ovens.

The final category of integumental injury is injury to the hair. Traction alopecia is seen when a parent pulls the child by the hair. The scalp is usually clear, differentiating this lesion from tinea capitis, seborrhea, and scalp eczema. Alopecia areata produces a lesion in which the hair is uniformly absent. In the cases of traction or traumatic alopecia, patches of broken hair remain.

Skeletal System

The skeletal system is also commonly traumatized when children are physically abused. As previously mentioned, matching the history of injury with the physical findings is important. Considering the mobility and strength of the child is also important to identify suspicious injuries. The radiologist needs to review the patient's past radiographs to identify the child with multiple visits to the hospital for fractures. When suspicion of abuse is high, a radiographic skeletal survey should be obtained to ascertain the condition of the entire skeletal system.

Support for the use of radioisotope scans as a more sensitive and immediate way of demonstrating bone injury is increasing. However, radionuclide scans are still second-line studies. Some of the indications for a radiographic skeletal survey or bone scans are listed in the “Management” section of this chapter. Skeletal surveys are often performed on a young child with an obvious fracture that then reveal multiple old fractures. The skeletal survey is the preferred radiographic study because it provides information on the type, location, and age of fractures as well as presence or absence of bone diseases.

Bone injuries may be of several types, including simple transverse fractures, impacted fractures, spiral fractures, metaphyseal fractures, or subperiosteal hematomas. Radiographs of some of these injuries are shown in Figure 128.9. To explain a transverse fracture, the history should be that of direct force applied to the bone. Differentiating the true cause of this type of fracture is often difficult. The impacted fracture should have an accompanying history of force along the long axis of the bone, such as the child's falling on his or her outstretched hand. In the case of a spiral fracture, a history of twisting or torque during the traumatic event should be present. Metaphyseal chip fractures occur when the extremity is pulled or yanked; the periosteum is most tightly adherent at the metaphysis, causing small bone fragments to avulse. Metaphyseal chip fractures are almost exclusively caused by abuse. Subperiosteal hematomas produce a characteristic radiograph. The elevation of the periosteum is seen as a linear opacification running parallel to the bone surface. Subperiosteal hematomas are produced by direct trauma to the bone. However, in up to 10% of small and premature infants, symmetric periosteal elevation that is not caused by abuse may occur along the tibia or humerus. The reason for this finding is unknown, but it should not be confused with abuse.

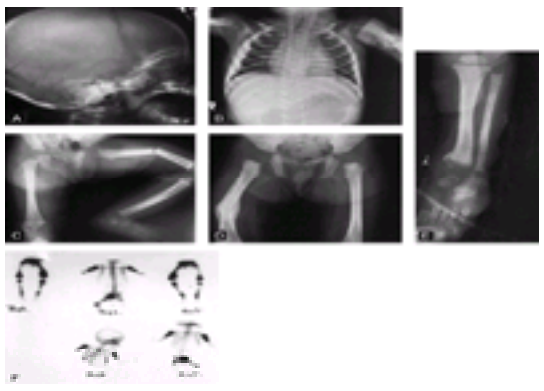


FIGURE 128.9. Radiographic findings of child abuse. **A.** Multiple skull fractures in an infant. **B.** Left humerus fracture and multiple old healing rib fractures. **C.** Left femur fracture and metaphyseal chip avulsion fractures of the right distal femur. **D.** Healing fracture of the right femur with callus formation, and new periosteal bone formation. **E.** “Bucket-handle” deformity of healing distal tibial epiphyseal fracture. **F.** Bone scan showing multiple areas of increased uptake caused by trauma. Some of these areas appeared normal on the original radiographs.

The location of the fracture is important in the identification of abuse. The fracture of a clavicle or the dislocation of a radial head are common noninflicted injuries. However, when the femur of a young child is fractured, or when ribs are fractured, the suspicion of abuse increases. Anderson reported on a series of children with femur fractures. Of 24 children who were less than 2 years old, abuse was proved in 19 cases. In two-thirds of these children, the fracture was the only sign of abuse. Feldman et al. reported on a series of children with rib fractures and noted an obvious history of trauma; for example, motor vehicle accident, an obvious bone disease such as osteogenesis imperfecta, or that the child was abused.

Feldman also reports examining several children who received external cardiac compression and finding that none of them had sustained rib fractures as a result of their cardiopulmonary resuscitation. In a confirmatory report, Schweich and Fleisher found that when the parents could not provide a history for rib fractures, the cause was abuse. The mean age of the group of children who had inflicted trauma was 3 months, whereas the group having accidental rib fractures had a mean age of 8.5 years. Ribs that are fractured along the axillary line are broken by an anterior-posterior force. Ribs that are fractured anteriorly or posteriorly are injured by a side-to-side compression of the thorax. The history of injury must be matched to the physical finding.

Other uncommon and, therefore, suspicious fractures are located in the vertebrae, sternum, pelvis, or scapulae. Uncommon fractures need to be carefully evaluated unless a clear history of significant trauma, such as an automobile injury, is reported.

The age of a fracture may be estimated from the amount of callus formation and bone remodeling seen on the radiograph. Table 128.1 lists fracture landmarks by date. Dating of fractures is not an exact science because, many confounding variables, such as the child's age, location of the fracture, and nutritional status, must be considered. Nonetheless, the child who presents with an acute fracture, and has a second fracture with a callus, stands out as having sustained more than one episode of trauma. The usual long-bone fracture may take 8 to 10 days to form callus and several months to heal completely. In the acute stages of injury, soft-tissue swelling should be seen for 2 to 5 days. Soft-tissue swelling may be clearly seen on standard radiographs. Skull fractures or fractures of other flat bones cannot be dated in the same way.

0–10 Days
Soft-tissue edema
Joint fluid
Visible fracture fragments
Visible fracture lines
10 Days–8 Weeks
Periosteal new bone (layered)
Callus (first subtle and then heavy)
Bone resorption along fracture line makes fracture line more visible
Metaphyseal fragments often more visible
8 Weeks and Over
Periosteal new bone matures, becomes thicker
Callus formation becomes more dense and smoother
Metaphyseal fragments are incorporated into metaphyseal callus and become smoother
Fracture line less visible and then invisible
Deformities and cortical bumps persist

Table 128.1. Dating Fractures

When a young child sustains multiple fractures, the differential diagnosis must be widened beyond accidental trauma and abuse to include osteogenesis imperfecta, infantile cortical hyperostosis, scurvy, syphilis, osteoid osteoma, neoplasms, rickets, hypophosphatasia, and osteomyelitis. Table 128.2 details the distinction between child abuse and osteogenesis imperfecta. All of the other conditions are much more rare than abuse and can be ruled out by the appearance of the bone on the radiograph and by the levels of calcium, phosphorus, and alkaline phosphatase in the serum.

Finding	Osteogenesis Imperfecta	Child Abuse
Incidence	Rare	Common
Positive family history	Common	Common
Blue sclerae	Common	Rare
Abnormal teeth	Common	Rare
Hearing impairment	Common	Uncommon
Osteoporosis	Common	Rare
Abnormal fracture healing	Common	Rare
Wormian bones	Common	Rare
Joint laxity	Common	Rare
Short stature	Common	Occasional
Fracture recurrence in protected environment	Common	Rare
In utero fracture	Occasional	Rare
Biochemical studies	Abnormal	Normal

Table 128.2. Osteogenesis Imperfecta versus Child Abuse

Central Nervous System

Injuries to the central nervous system (CNS) are the main cause of child abuse deaths. These injuries may be subdivided into two categories: direct trauma and shaking injuries. Direct trauma is inflicted by striking the child with an object or by dropping or throwing the child against a wall or onto the floor. The extent of the resulting trauma depends on the amount of force used, the surface contacted, and the child's age. The child may be brought to the ED with a small subgaleal hematoma or in coma. Injuries may vary from scalp contusions to intracerebral hematomas.

A history of a young infant's falling off a bed or dressing table is often presented. The precise extent of injury from this type of fall is unknown, but several reports suggest that even uncomplicated skull fractures are as uncommon as 1 to 2% of cases. If the injury is more severe and the only history is of a fall from less than 8 to 10 feet, abuse should be suspected. Another scenario is that of a child who sustained trauma 1 week before the ED visit. The visit is prompted by the parent's noticing a soft spot on the child's cranium. This sequence may occur when the initial scalp hematoma so rapidly expanded that it had a bony consistency. Only with degradation and softening of the mass does the parent now perceive the hematoma. Although a delay in seeking treatment is a well-recognized red flag for child abuse injuries, this case provides a plausible exception. In all children younger than 1 year of age who have a history of head trauma, skull radiographs are recommended. Infants tend to sustain skull fracture more easily and are more vulnerable to serious sequelae. If a fracture does exist and abuse is suspected, a skeletal survey should also be obtained. For the diagnostic methods to be used for more serious head injuries, refer to Chapter 105.

Shaking injuries characteristically cause serious central nervous system damage without evidence of external trauma. The infant's relatively large head size and weak neck muscles are predisposing factors for whiplash injury. Whether the injury is caused by shaking alone or shaking followed by an impact is controversial. In most fatal cases, minor bruising to the scalp is apparent, although such scalp injuries may not be apparent until the scalp is reflected during the autopsy.

The shearing and contusive forces that result from shaking the infant produce this type of injury. Specific lesions that occur include hematomas, subarachnoid hemorrhages, or brain contusions, particularly in the frontal and occipital lobe. The child may present with lethargy and a “septic” appearance, with seizures, or in a coma. The physical examination is otherwise unremarkable except for retinal hemorrhages (Fig. 128.10A). Occasionally, bruises on the upper arms or shoulders indicate the sites where the child has been grasped. Lumbar puncture produces grossly bloody or xanthochromic spinal fluid. If computed axial tomography is available, it shows the characteristic findings of occipital contusion and intrahemispheric blood (Fig. 128.10B). This form of abusive behavior by the parent is usually triggered by the infant's persistent crying. Occasionally, excessively rough forms of play or misguided resuscitative efforts may result in shaking injuries.

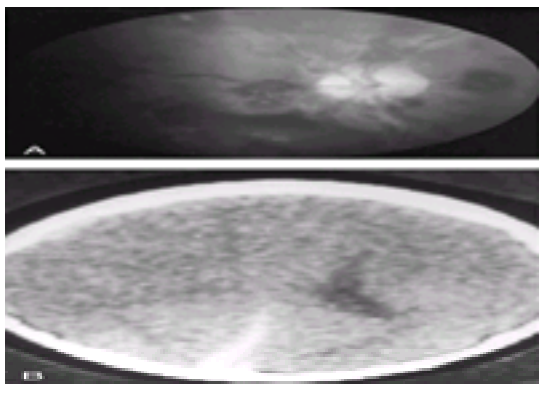


FIGURE 128.10. Manifestations of the whiplash shaking injury. **A.** Retinal hemorrhages as seen on fundoscopic examination. **B.** Computed tomogram showing intrahemispheric subdural bleeding and right cortical brain swelling.

Gastrointestinal System

Gastrointestinal (GI) injuries are relatively uncommon abuse manifestations but, similar to CNS injuries, account for a significant percentage of fatal injuries. Of all GI injuries, mouth trauma is perhaps the most common. Small infants may sustain a tear of the frenulum resulting from “bottle jamming.” In the older child, dental trauma may be a sign of abuse.

Other GI system manifestations are more medically serious and generally result from blunt trauma to the abdominal contents. Rupture of the spleen or laceration of the liver causes the child to present with elevated liver enzymes, with an acute abdomen, or in shock, with no external source of bleeding and with absent or only minor bruising of the abdominal wall. The identification and management of these emergencies is covered in Section IV. A less acute presentation is the afebrile child with persistent bilious vomiting from a duodenal hematoma with small-bowel obstruction. Documenting an elevated serum amylase or lipase or increased liver enzymes is important in providing tangible evidence of abdominal trauma in cases that lack any radiographic finding or abdominal wall bruising. Elevation of the serum amylase may also identify those cases that should be followed for possible development of a pancreatic pseudocyst.

Cardiopulmonary System

Abuse may be manifested in cardiac or pulmonary trauma with no injuries that are characteristically induced by abuse. Pulmonary contusion, pneumothorax, hemothorax, cardiac tamponade, and myocardial contusion may all occur occasionally. Specifics of identification and management of these problems are covered in Chapter 107.

Genitourinary Systems

Common genitourinary complaints, such as hematuria, dysuria, urgency, frequency, and enuresis, may be the initial sign of abuse. These problems may result from direct trauma, sexually transmitted infections, or emotional abuse. Some aspects of genitourinary manifestation are covered subsequently under the “Sexual Abuse” section. As for direct trauma, any part of the genitourinary system may be involved, from the renal parenchyma to the urethral meatus. Penile trauma that does not have an adequate explanation may be an alerting sign of abuse. Traumatic hematuria is managed as described in Chapter 109.

A life-threatening renal manifestation may be the occurrence of rhabdomyolysis and myoglobinuria. With extensive deep soft-tissue and muscle trauma, myoglobin may be liberated in quantities sufficient to cause acute renal failure. Such children have dark or tea-colored urine that tests positive for blood with urine dipstick, but has no visible red blood cells on microscopic examination. Serum myoglobin levels confirm the diagnosis, and the serum creatine phosphokinase reaches extremely high values. Before using hypertonic intravenous contrast materials in the child with heme-positive urine, myoglobinuria should be considered and ruled out. The patient with possible myoglobinuria and acute renal failure must not be given potassium-containing intravenous solutions.

Sensory

All of the sensory organs are vulnerable to physical abuse, including ocular, nasal, and otic injuries. The eye may sustain several different forms of injury, including periorbital ecchymosis, corneal abrasion, subconjunctival hemorrhage, hyphema, dislocated lens, retinal hemorrhages, or detached retina. Each of these lesions is discussed in Chapter 110, Chapter 111, Chapter 112 and Chapter 113. A careful history of injury is important when treating any of these conditions. Injury to the nose may result in simple hemorrhage or fracture and disfigurement of the nasal structures. The external ear may show evidence of contusion. In particular, ecchymosis on the internal surface of the pinna may result from “boxing” the ear and crushing it against the skull.

A direct blow to the ear may also cause hemotympanum and perforation of the tympanic membrane. In such cases, hemotympanum on the basis of basilar skull fracture should also be considered. The presence of discoloration behind the ear (Battle's sign) may be a further indication of a basilar skull fracture. Refer to Chapter 105, which deals specifically with these aspects of emergency care.

Unusual Manifestations

Rarely, the emergency physician is confronted by one of the unusual abuse manifestations. Cases of toxic and nontoxic ingestions, electrolyte disorders such as hyponatremia and hypernatremia, foreign bodies, bathtub drowning, and multiple serious infections may be the result of abuse. In all these situations, the parent actively abuses the child by feeding, instilling, or injecting harmful substances or objects into the child's body. Some children with a toxic ingestion reveal that their parents forced them to ingest the substance. The most common toxic ingestants of this type are alcoholic beverages that are given to or forced on the child to either quiet the child or to demonstrate “manly” qualities. Other drugs may be used to poison the child. Most recent reports are of cocaine ingestions.

Several cases of parents who have placed their children on high-salt, water-only, or pepper diets as a form of punishment have been reported. Such children may present with signs of hypernatremia or hyponatremia, possibly with seizures. Foreign bodies have been found in every orifice, as well as under the skin and in fingernail beds. Several cases of Munchausen syndrome by proxy have occurred in which a parent has inflicted illness on the child rather than feigning or inducing illness (Table 128.3). Cases of fictitious fever, hematuria, and even sepsis have resulted from this form of abuse. Although rare, the unusual manifestations of abuse should be considered when more common causes of these problems cannot be identified.

Difficult to understand medical situation with recurrent episodes
Failure of other centers to arrive at diagnosis—doctor shopping
Unsupportive or "absent" marital relationship
Compliant, cooperative, overinvolved mother
Medical knowledge in parent's background
Findings abort with surveillance of child
Findings correlate to presence of parent
Extensive medical care in parent's background

Table 128.3. Characteristics of Munchausen Syndrome by Proxy

Management

The management of a child abuse case is difficult unless the emergency physician has a previously prepared, well-structured protocol. If reports of abuse are not a daily occurrence, an institutional policy serves as an important guide to the mechanics of management. Consultants from different disciplines, such as nursing and social work, provide invaluable assistance. A multidisciplinary approach simplifies the initial decision-making and subsequent case management. The steps in the protocol are shown in Figure 128.11.

FIGURE 128.11. Procedure for emergency department management of suspected physical abuse.

Suspect Abuse

The first step is to decide whether a reasonable likelihood of abuse exists. Many shades of suspicion make the term *abuse* imprecise. Although every traumatic injury should be suspected as abuse, the physician has the onerous task of deciding how much suspicion is necessary to take some action (i.e., report). To establish the level of suspicion, data are gathered by obtaining a complete history, performing a thorough physical examination, comparing the history and physical examination, observing interactions, and obtaining laboratory studies and/or radiographs. Then, the physician can formulate a differential diagnosis and assign a rank to abuse. Indications of abuse in the history and physical examination and observational data must be used like building blocks that are added until they achieve a certain threshold of suspicion. As demonstrated in Figure 128.12, when the threshold is reached, a report of suspected abuse must follow. In the example of case 1, all of the building blocks must be used to build a level of suspicion; in case 2, the physical injury is sufficient to make the diagnosis.

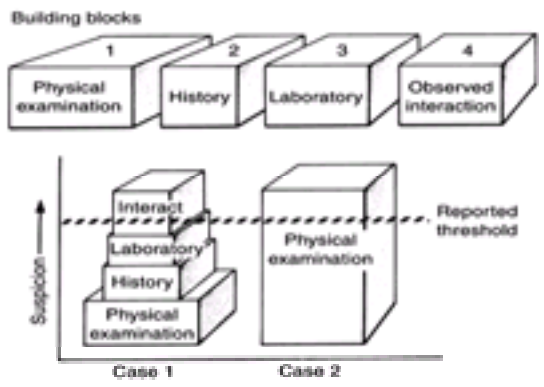


FIGURE 128.12. Building a level of suspicion.

A detailed history is always important. As in many other medical situations, this process is initiated by asking some general open-ended questions about “what happened?” If the child has sufficient verbal skills, the first questions are directed at him or her. General inquiries must then be followed with specific requests for information; however, a harsh

interrogation only alienates the family. Some specific historical indications are listed in Table 128.4.

1. Is the history one of inflicted injury?
2. Is there an absence of history, a "magical" injury?
3. Could the injury have been avoided by better care and supervision?
4. Are there inconsistencies or changes in the history?
5. Is there a history of repeated injury or hospitalization?
6. Was there a delay in seeking medical care?
7. Does the history overestimate or underestimate the injury?
8. Is there a medical history of prematurity, failure to thrive, failure to receive adequate medical care such as immunization?

Table 128.4. Historical Indicators of Abuse

As with the history, the physical examination must be thorough. The signs of physical abuse have been detailed in the section on “Clinical Manifestations.” A thorough examination serves as a means to uncover these findings. Although the clinician may be tempted to merely glance at a small contusion on a child's face, such a cursory examination fails to reveal possible linear and loop-shaped marks on the upper thighs and buttocks. Table 128.5 lists some of the important physical examination features that are indicators of abuse.

1. Does the injury match the history of injury?
2. Are there pathognomonic injuries such as looped wire marks, cigarette burns, and so on?
3. Are there multiple injuries?
4. Are the injuries at various stages of healing?
5. Are there different injury forms, for example, burns and fractures?
6. Is there evidence of overall poor care?
7. Has poisoning been documented in a young child?
8. Is there evidence of failure to thrive without a history of symptoms or physical findings?
9. Are there any visual or unexplained physical findings?

Table 128.5. Physical Indicators of Abuse

After completion of the history and physical examination, the next step is to compare them. Does the stated history match the physical findings? Does the history make sense? Does the history correlate with the developmental level of the child? Answers to these important questions may add further elements of suspicion.

This step of comparing history and physical examination is completed subconsciously by most practitioners. Physicians often attempt to match the patient's degree of symptoms with the presence or absence of physical findings, particularly in patients with psychosomatic complaints. In child abuse cases, this step should be a conscious and well-defined step because it is vital to establishing suspicion. In some cases, a lack of consistency is obvious, such as a parent's claim that burns on the child's buttocks occurred when the child inserted his or her finger into an electric socket. Other situations may be less clear, such as the injury being attributed to hot plastic seat covers on an automobile. Although the latter explanation has in fact been reported as a case of accidental injury, it rarely explains burns on the buttocks.

Laboratory data and radiographs are another source of indicators of abuse. The laboratory studies used are few and, for the most part, document the obvious or rule out other disease states. Biochemical, hematologic, and urinary studies that are used appear in Table 128.6 along with their indications. Radiographs document a specific bony or soft-tissue injury. They may provide a comprehensive and longitudinal record of osseous injury at any site in the skeletal system. Although no precise indications for ordering a skeletal survey exist, some relative indications are: 1) any child less than 1 year old presenting with a fracture, 2) any child with severe or extensive fractures, 3) any child who has a history of more than one fracture, and 4) a history in the child or the family of “soft” or easily broken bones.

Table 128.6. Laboratory/Diagnostic Evaluation of the Physically Abused Child

During the time occupied by the history, physical examination, and performance of laboratory studies, the physician should be cognizant of the interactions among family members and between the parents, the child, and the ED staff. Such an awareness often uncovers subtle indicators of abuse. The observation of parents arguing vehemently on the way to the radiology department may be a clue. The parent who appears to be distant from both the child and the physician is also suspect. Although the parent who is intoxicated or incoherent never fails to gain staff attention, such individuals are in the minority of abusive parents. Observation of the child is important as well. All abused children are not withdrawn, passive, and depressed. On the contrary, some are competent, outgoing, or “pseudomature.”

The observed state of the child depends on several factors: 1) the length, frequency, and severity of abuse; 2) the child's developmental level and age; and 3) the amount of positive interaction the child's parents and extended family have had between abusive episodes. Physicians are often surprised that the child does not immediately state the nature and extent of the abuse and ask for asylum. Such statements by children are actually rare and occur mainly in adolescent patients. Children are loyal to their parents. Abusive parents may be only episodically abusive and at other times nurturing and loving. Young children may have no framework for comparison and may accept the abuse as the norm. Somewhat older children may understand and dislike the abuse, but may fear the consequence of reporting it even more. In the child's mind, it may be better to live with the pain of abuse than to face the unknown of institutional or foster placement.

The final step in establishing a threshold level of suspicion is to review the differential diagnosis. At this point in the management scheme, the physician must add up the indicators and arrive at a judgment. If the process does not lead to a clear determination, most state laws imply that reporting suspected abuse is more prudent than not. Physicians are asked to report suspected, not proven, abuse. The major differentiation is between accidental and nonaccidental trauma. The other elements of the differential diagnosis are all uncommon diseases, including: 1) bone diseases such as osteogenesis imperfecta, osteoid osteoma, hypophosphatasia; 2) hematologic disorders such as idiopathic thrombocytopenic purpura and hemophilia; 3) neoplasms; 4) metabolic disorders such as rickets or scurvy; 5) infections such as syphilis or osteomyelitis; and 6) syndromes in which pain sensation is absent, such as spina bifida or congenital indifference to pain. All of these diseases occur with much less frequency than abuse, but deserve consideration; simple laboratory and radiographic studies confirm or deny these diagnoses.

A special note should be made concerning the child less than 6 months old who is brought to the hospital dead. In this situation, the central differential diagnosis exists between sudden infant death syndrome (SIDS) and child abuse. Other rare causes of sudden death include hypoglycemia, medium-chain fatty acid defects (MCAD), mitochonrial defects, intoxication, and smothering. Victims of SIDS may appear to have bruising as a postmortem change. Clearly, their parents have no adequate explanation for the death. In this situation, the presumption should always be SIDS. Most localities require an autopsy to be performed. If not required, the physician should insist on a postmortem examination and wait for the autopsy to ultimately make the differentiation. Interrogating parents in cases of SIDS about the possibility of abuse can produce unnecessary psychological harm. With the death of a child, supportive ED treatment becomes paramount and suspicions of abuse can be pursued by the medical examiner and law enforcement personnel if warranted. If two or three SIDS deaths have occurred in the immediate family, the level of suspicion for abuse should be elevated.

Multidisciplinary Consultation

If consultation with a nurse, social worker, or physician with more extensive experience in the management of child abuse is available, it should be obtained. The advantages of consultations are many. They allow for: 1) information sharing, 2) joint decision-making, 3) planning, and 4) mutual support. Planning an approach to the family and subsequent case management is useful. This brief consultation enables the physician to be more secure in making decisions about matters that are generally unfamiliar and often value-laden. Joint interviewing is not only time-efficient, but gives the family a uniform approach from the professional staff.

Reporting

Once the suspicion of abuse has been established and consultations obtained, the next step is reporting. Although laws vary from state to state, most have common elements. The emergency physician should become familiar with his or her current state law. The definition of abuse is central to each of the reporting laws. A stated age defines a child. The laws also specify who must report (mandated reporters) and who may report (nonmandated reporters). For most mandated reports, the law requires a specific penalty (as well as malpractice liability) for failure to report, and provides protection from liability if the report of suspected abuse turns out to be unfounded once investigated. Finally, the law dictates to whom and how the report should be made. Generally, reports are made to child protective services (CPS) agencies, to police departments, or to some combination of law enforcement and social work personnel. Many states now have statewide central registries for receiving reports.

Notifying the Parents

An important, but often avoided, step in case management is notification of the parents. This step is often forgotten because it is a difficult interpersonal task; nonetheless, it must be done. Nothing makes parents more resistant to change than completing a “routine” ED visit, only to later receive notification that the physician has filed a suspected child abuse report. Some specific guidelines are helpful in avoiding this breech of trust. The overall approach to the parents must be based on concern for the child. Concern for the child, not accusation should be stressed. The physician should not confront the parents or attempt to seek an admission of guilt. Many times, the parent in the ED may not be the abusive

parent and may know as little about the episode as the hospital staff. The physician should explain the requirement for a mandated reporter to report all suspected cases. However, stating the requirement should not be used as an excuse. The desire to report should also be stated.

In many states, the reporter is required to report all injuries that are not fully explained. This requirement may also be stated to the parent. Using the words *"child abuse report"* is important. This situation is not a time to "soft pedal." However, child abuse does represent a range of behaviors, from the parent who overvigorously disciplines to the parent who sadistically tortures. Parents often have not seen themselves as abusers, and an explanation of the range of abuse is helpful in demonstrating how a child abuse report applies to them. Parents are fearful of what a child abuse report means and of what will happen. Therefore, the consequences of the report should be explained (e.g., "a social worker will call and come visit you in 1 or 2 days"). The physician's natural fear is that the parent will have a dramatic and hostile reaction.

The emergency physician can expect a wide variety of reactions, from hostility to appreciation. To minimize the angry reactions, the physician should stress the focus as being concern for the child. This perspective puts the physician on common ground with the parent. An angry reaction is more likely if guilt or fear is increased. This reaction may be seen as a feedback relationship. Stoking the fire by increasing the parents' guilt or fear results in a flare of angry emotion directed at the staff, the child, or the other spouse. Figure 128.13 illustrates this relationship.

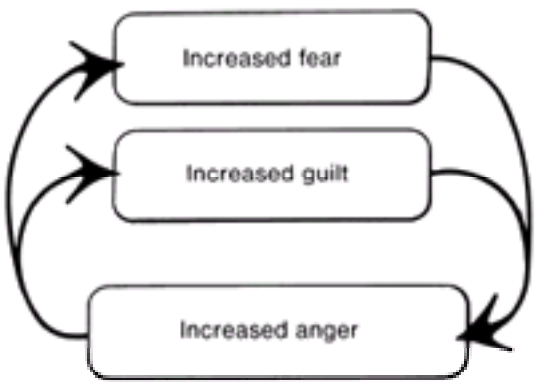


FIGURE 128.13. The cycle of fear, guilt, and anger.

Although the child does not need to be formally notified, the child may often be aware of what is happening and may even ask some pointed questions. The physician may wish to discuss the report with the child. The physician should stress the outcomes of protection and help for the child and family. The ED staff should never be accusatory or belittling of the parent.

Crisis Hospitalization

In some cases of abuse, the family crisis is at such an acute level that hospitalization is necessary. The physician must ask, "Is the home safe?" If the child's environment poses a potential danger, the child should be admitted, no matter what the extent of injury. Some state laws have included protective custody sections that allow physicians virtual police powers in detaining a child for protection. In other states, the physician may need to obtain parental consent for admission.

This task is also difficult. As with reporting, the approach to the parents must be honest and nonaccusatory. The reasons for admission are to observe the child and allow time to evaluate the possibility of such an injury happening again. The focus must be the physician's concern for the child's health. In hospitalizing the child, the physician also makes a statement about the seriousness of the situation and the depth of his or her concern. Most hospitals do not have the resources to hospitalize every abused child, nor is it necessary or advisable to do so in every case. Factors that favor sending the child home are: 1) a concerned relative or neighbor available to support the family, 2) a solution to the inciting crisis, 3) parental acceptance of responsibility, 4) prompt (less than 24 hours) (CPS) or police response, 5) a first episode or minor degree of abuse, and 6) an alternative environment (e.g., grandmother's house). If the physician is unable to hospitalize the child and has lingering doubts, a specific agreed-upon return appointment for the next day may be a solution. If the family is unable to keep the appointment, there should be grave concern, and emergency CPS or police involvement should be requested.

Documentation

Throughout each step of case management, documentation is important. The record of this visit is both a medical and a legal document. A precise description of the injuries enhances the document's value, and small sketches are also helpful. Photographs are invaluable in documenting extensive injuries. Some states require parental permission for photographs, whereas other jurisdictions allow photographs to be taken without parental consent. Photographs may be used in court to illustrate, for a judge or jury, injuries that would be difficult for most witnesses to describe. Even if the photographs are not admissible in court, they refresh the memory of the physician for testimony. Often, court proceedings may not take place for several months after the ED visit. If photographs are taken, their quality must be good. Poor-quality photographs can damage a case by failing to show all of the pertinent findings.

Written descriptions of an injury are adequate for less extensive trauma, but as much detail as possible should be included. The record of the history should be as factual as possible; conclusions or impressions should not be listed. For example, rather than recording "the child looked fearful," describing the physical components or behaviors that prompted that judgment, under standard courtroom procedure, is more appropriate. Direct quotes from the child or parents should

be included because quotes from another person are generally considered hearsay and are disallowed. When such quotes are included in the text of a business (medical) record, they may become more acceptable. General legibility and, in particular, a clearly written signature are always important record requirements. At the time of the ED visit, detailed record-keeping seems to be a burden and waste of time. When a case record requires review before court testimony, good documentation is a necessity. Upon completion of the record, the physician should read it again as if he or she were the defendant's attorney.

Staff Reactions

Child abuse triggers many emotions in the physician and in the ED staff. Sadness and pity for the child and, often, anger for the parent are exhibited. The staff may have a general feeling of disbelief, “How can this happen?” The case management plan previously described requires that each ED staff member be professional and in control of these emotions. The offhand comment of a clerk or the whispering between nurses can undermine the work being done with the family. If a staff member is too upset by abuse, he or she should be relieved of the responsibility of working in such situations. Physicians or nurses who openly attack the family or who are blatantly accusatory should not be involved in these types of cases. Only after having experience with large numbers of cases can an ED staff member develop an appreciation of abuse as a negative, but understandable human response. With such a perspective, the physician can become less angry and more zealous in attempting to provide constructive case management.

If problems relating to the staff reaction exist, as they often will, several corrective actions should be taken. At times, a brief meeting of the involved staff members can be called after the family has been discharged from the ED. This meeting allows the staff an immediate opportunity to ventilate their feelings. In-service education serves the same purpose, providing a forum for the staff to discuss their personal feelings about abuse, as well as their past experiences. ED staff are always involved in the negative aspects of identifying and reporting abuse and never see the long-term treatment and rehabilitation phase. If a therapist, or better yet, a family member, can present this side of abuse to the staff, it engenders more positive attitudes. Without learning about the successes, the staff often displace this anger, charging “incompetence” of the CPS agency or “leniency” of a family court judge. Scapegoating should be put into the proper perspective. Again, in-service education and case reviews are useful. The integrity of the entire treatment team, both in the hospital and in the community, must be maintained to accomplish the difficult goals of protecting the child and preserving family life.

SEXUAL ABUSE

The sexually abused child is another difficult psychosocial emergency for the emergency physician. The state-of-the-art method in identifying and managing cases of sexual abuse has developed rapidly in the last decade. Most child abuse centers are reporting dramatically increasing numbers of sexually abused children. The Children's Hospital of Philadelphia currently sees more than 10 times the number of sexually abused children than were reported 10 years ago. The number of sexual abuse cases reported now equals the number of physical abuse cases. Of all societal taboos, those that prohibit incest are the strongest. This belief leads to denial and makes even basic recognition of the problem more difficult.

As with the physically abused, the sexually abused child engenders a great deal of emotion from the health care professionals in the ED. Treatment issues for both the child and the perpetrator are more complex. Working in a multidisciplinary fashion with nursing and social work staff is important. The effects of this form of abuse may clearly be profound, but may not be expressed as symptoms for many years. Prompt diagnosis, humane emergency management, and referral to long-term treatment resources are the goals of the emergency physician. Many centers have adopted strategies to use the ED for recognition and screening of acute (less than 72 hours) episodes of sexual abuse as well as developing programs for comprehensive evaluation outside the ED.

Background

The term *“sexual abuse”* conjures up images of a violent, forced sexual attack and rape. Although most people may think of the psychopathic criminal luring children on their way home from school by offering them candy, such stereotypes are the exception. This situation has prompted Sgroi et al. to suggest that the term *“sexual abuse”* be changed to *“sexual misuse.”* This new term presents a more realistic image of what occurs in most sexual abuse cases. Commonly, a relationship exists between the perpetrator and the victim. The misuse of that relationship is central to the sexual abuse of the child. The relationship may be a familial one, such as father–daughter, or a household relationship, such as mother's live-in paramour and child, or a more casual relationship, such as that with a neighbor, teacher, or friend of the family. Most often, no overt violence is perpetrated, although harsh threats of violence as a consequence of the child's revealing the act to another person may be issued. The term *sexual misuse* is also more encompassing because it includes the misuse of children for prostitution and pornography. Although the suggested terminology change is based on several sound points, most state laws use the term *“sexual abuse”;* thus, this chapter conforms to that tradition.

CAPTA defines sexual abuse as: (1) employment, use, persuasion, inducement, enticement, or coercion of any child to engage in, or assist any other person to engage in, any sexually explicit conduct or any simulation of such conduct for the purpose of producing any visual depiction or such conduct; or (2) rape, and in cases of caretaker or inter-familial relationships, statutory rape, molestation, prostitution, or other form of sexual exploitation of children, or incest with children. Sexual abuse includes fondling a child's genitals, intercourse, incest, rape, sodomy, exhibitionism, and commercial exploitation through prostitution or the production of pornographic materials. Many experts believe that sexual abuse is the most underreported form of child maltreatment because of the secrecy or “conspiracy of silence” that so often characterizes these cases. Sexual abuse has traditionally been more the domain of police and other law enforcement personnel. Several specific legal definitions should be understood and used precisely. Each state defines these terms independently as well as legislates the age of consent. Emergency physicians must be aware of their local laws. In Pennsylvania, for example, the age of consent is 18 years. Sexual abuse terms are defined as follows:

Rape

A person commits a felony when he or she engages in sexual intercourse with another person not his or her spouse:

1. By forcible compulsion
2. By threat of forcible compulsion that would prevent resistance by a person of reasonable resolution
3. Who is unconscious
4. Who is so mentally deranged or deficient that such person is incapable of consent

Statutory Rape

A person who is 18 years of age or older commits a felony when he or she engages in sexual intercourse with another person not his or her spouse who is less than 14 years of age.

Involuntary Deviate Sexual Intercourse

A person commits a felony when he or she engages in deviate sexual intercourse per anus, or per os, or any form of sexual intercourse with another person:

1. By forcible compulsion
2. By threat of forcible compulsion
3. Who is unconscious
4. Who is mentally deranged or deficient
5. Who is less than 16 years of age

Indecent Assault

A person who has indecent contact (any touching of the sexual or other intimate parts of the person for the purpose of arousing or gratifying sexual desire in either person) with another not his or her spouse, or causes such other to have indecent contact with him or her is guilty of indecent assault.

Incest

A person is guilty of incest if he or she knowingly marries or cohabits or has sexual intercourse with an ancestor or descendant, a brother or sister of the whole or half blood, or an uncle, aunt, nephew, or niece of the whole blood. These relationships include blood relationships without regard to legitimacy and relationship of parent and child by adoption.

Promoting Prostitution

A person who knowingly induces or encourages a child to engage in prostitution is guilty of promoting prostitution.

The true incidence of sexual abuse is unknown. There is a recent documented upward trend in number of reports. The National Center on Child Abuse/Neglect estimates that the current annual incidence of sexual abuse is between 75,000 and 250,000 cases per year. Most estimates do not include children who are victims of pornographic exploitation and child prostitution.

Dynamics

Sexual abuse encompasses several different sexual acts committed by different perpetrators for different reasons. Thus, no single theory can explain the dynamics. In an effort to simplify the interactions and make them understandable in light of behaviors seen in the ED, the physician should consider intrafamilial versus extrafamilial sexual abuse. The intrafamilial category includes incest in all of its forms as well as sexual abuse by significant, although perhaps not legal, members of the family. Sexual abuse between a girl and her mother's paramour would be included in this category. Extrafamilial abuse occurs between adults and children of adolescents and children who have no familial relationship.

The dynamics of intrafamilial abuse are controversial. Professionals are divided into at least two theoretical camps. One group theorizes that sexual abuse is the sole responsibility of the perpetrator, usually a male adult. Advocates of this position contend that the abuse results from an inability of the individual to control his sexual impulses or to establish age-appropriate adult relationships. This perpetrator is the pedophile. A second group sees the problem more as a family responsibility. In this model, the disturbed relationship between the adults expresses itself in the male parent's (or parent equivalent's) crossing generational lines for sexual gratification. Theorists in this camp often point to the mother's passive sanction of the abuse, even to the point of being informed about the incest and still allowing it to continue over long periods.

Obviously, the two theories point to different treatment strategies. According to the first, the solution is to simply remove the male perpetrator to jail or a mental hospital and to concentrate rehabilitation efforts on the perpetrator. The second group would prescribe therapy for the entire family to reorder the relationships so that the adults are able to meet their own needs and the children are protected. These professionals would contend that, when the male adult is removed, the mother soon finds a replacement.

The dynamics of extrafamilial abuse are less well understood. In some cases, the rape of a child by a stranger, similar to the rape of an adult, is a crime of violence. Such attacks are triggered by extreme anger, and a child may be selected as more easy prey. In other cases of extrafamilial abuse, some of the dynamics are still based on the misuse of a

relationship, although in this case the relationships are more casual. Perhaps the most common extrafamilial sexual abuse occurs between neighbors or between a babysitter and the child being watched. These episodes often involve adolescent perpetrators. The abuse dynamic may be an abuse of a power relationship, an uncontrolled sexual curiosity, or a combination of these factors. As the age difference between the child and the perpetrator widens, the more pathologic the dynamic becomes. As the perpetrator increases in age, socialization is expected to instill more self-control. Certainly, the sexual exploration that occurs between children of the same age is not sexual abuse, although on occasions, an uninformed parent may consider it as such.

Clinical Manifestations

The manifestations of sexual abuse may occur at a time shortly after the abuse has occurred or at a time more distant from the event. The manifestations may be influenced by a single episode or by a pattern of repeated encounters. Finally, the manifestations may depend on the child's age and maturity. The manifestations may be divided into four categories, as shown in Table 128.7. These categories are specific physical findings, specific behavioral manifestations, nonspecific physical complaints, and nonspecific behavioral complaints.

Physical Complaints	Behavioral Complaints
Specific	*Specific*
Genital injury	Explicit assertions of sexual contact
Bruises	Inappropriate knowledge of adult sexual behavior
Lacerations	Compulsive masturbation
Rectal laceration, fissures	Excessive sexual curiosity, sexual acting out
Sexually transmitted disease	*Nonspecific*
Pregnancy	Excessive fears, phobias
Nonspecific	Refusal to sleep alone, nightmares
Anorexia	Runaways
Abdominal pain	Encopresis
Enuresis	Aggressive behavior
Dysuria	Attempted suicide
Evidence of physical abuse in genital area	Any abrupt change in behavior
Vaginal discharge	
Urethral discharge	
Rectal pain	

Table 128.7. Identification of Sexual Abuse

Specific Physical Complaints

Bruising on the upper thigh, lower abdomen, or genitalia is a rare finding in childhood sexual abuse. The child is not usually injured because he or she is often used for stimulation, masturbation, or genital contact that involves no force. Nonetheless, a physical injury to the genitalia should elicit a suspicion of sexual abuse. For children with even small vaginal lacerations, a detailed history of injury should be obtained. Straddle injuries do produce genital trauma and are the most common form of accidental genital injury to young girls. In males, accidental penile trauma may occur from zipper accidents or from a toilet seat that falls. Beyond these common accidental situations, the emergency physician should scrutinize the history given. The premenstrual child who presents with vaginal hemorrhage may be bleeding from a vaginal laceration that is not visible on external examination. Accidental injuries such as straddle injuries are always able to be seen. Prompt surgical or gynecologic consultation should be obtained to identify and repair unseen sites of trauma.

The presence of sexually transmitted disease in a prepubertal child is a specific finding of sexual abuse until proven otherwise. Studies by Branch and Paxton and others have shown that when instances of prepubertal gonorrhea were carefully investigated for cause, the source of the infection was through sexual contact, most often in the child's home or in a relative's home. Gonorrhea may occur in the genitourinary tract, rectum, or oropharynx. When gonorrhea is culture proven, it should be pursued as sexual abuse, according to the Centers for Disease Control and Prevention. The American Academy of Pediatrics has issued guidelines for the diagnosis of gonorrhea. Parents may bring their child to the ED for the complaint of vaginal discharge. Gonorrhea may also appear in cases of less well-defined symptoms, such as vaginal pain, itching, urinary frequency, or enuresis. Recent studies indicate that only children with vaginal discharge need to be cultured.

The Centers for Disease Control and Prevention indicate in their treatment guidelines that “Any sexually transmitted infection in a child should be considered as evidence of sexual abuse until proven otherwise.” At present, knowledge about sexually transmitted diseases is limited. Any sexually transmitted disease in a prepubertal child is suspicious of abuse; however, basic understanding about disease transmission places these infections in three categories (Table 128.8). In the first category are those infections that virtually always are transmitted through sexual contact; for example, syphilis and gonococcal infections. In the second category are those infections that to the best of clinical knowledge are usually transmitted sexually. It must be noted, however, that exact scientific information is not present. For example, Kaplan reported on six cases of genital herpes simplex, but in only four cases could sexual abuse be documented. Category three includes diseases that are suspicious of abuse but that may be transmitted by nonsexual contact. The pregnant adolescent may be a victim of incest. The physician should try to obtain a specific history of conception. Often, the focus of case management centers on how the adolescent plans to notify her parents or whether she considers abortion or adoption as options. If the issue of paternity is not pursued, instances of sexual abuse escape detection.

Always	Usually	Possibly
Neisseria gonorrhoeae	Herpes simplex	Condylomata
Syphilis	*Chlamydia trachomatis*	Scabies
	Trichomoniasis	Pediculosis
		Gardnerella vaginalis

Table 128.8. Sexually Transmitted Diseases and Their Probability of Being Caused by Child Sexual Abuse

Specific Behavioral Complaints

The most common clinical manifestation of sexual abuse is a positive history. The child who gives a clear detailed story of sexual encounter with an adult has a specific behavioral manifestation. Reports of suspected sexual abuse may be based on history alone because, children do not make up such allegations. Most children who are not abused are not knowledgeable in the details of sexual encounters. Thus, when a child offers the specifics of an encounter, he or she must be believed. The detail of the history varies with the child's age and language development, but even children of 3 or 4 years are able to make simple yet credible statements about someone touching their genitals.

Some children manifest behaviors in their play or in their conversation that indicate that they have been exposed to sexual experiences, and perhaps abused. These signs are less specific than a clearly stated history, but are significant enough to require further explanation. For example, the young child who discusses urogenital contact may be demonstrating a specific behavioral manifestation. Children who wish to fondle their parents' genitals as an expression of affection are cause for concern. These behaviors are usually learned. All children manifest sexual curiosity and may engage in some form of masturbation, but when either of these behaviors appears in excess, it deserves investigation. Sexual abuse may be the cause.

Recently, several films, videotapes, books, and school programs have been developed as prevention tools in sexual abuse situations. These instructional materials are also helpful in opening discussions between parents and children and thereby promote disclosure of past events experienced by the child.

Nonspecific Physical Complaints

The physician should keep sexual abuse in the differential diagnosis for many complaints. Sexual abuse may be related to cases that present with pain in the abdomen, thighs, or genitals; dysuria; pain on defecation; hematuria; or hematochezia. Abuse may manifest as a change in habits, such as urinary frequency, enuresis, constipation, or encopresis; other complaints may be vaginal discharge or chronic sore throat. The cause of each of these complaints may be any number of things. For example, in studying a group of children with enuresis, sexual abuse is an uncommon cause of the complaint. Nonetheless, sexually abused children are regularly brought to EDs with nonspecific complaints. If sexual abuse is not considered, it goes unnoticed.

Nonspecific Behavioral Complaints

The final group of clinical manifestations includes unexplained changes in the child's behavior. In this group are relatively minor behavioral changes, such as the recent acquisition of nightmares or phobias, or major changes, such as school truancy and adolescent runaways. Children who bear no physical evidence of their abuse and in whom no physical symptoms develop may express themselves behaviorally. Many children demonstrate change in one or more of the important spheres of their life: at home, in school, or with peers. This situation is exemplified by a 5-year-old girl who begins avoiding contact with her father and other male relatives after an abusive episode with a male friend of the family. A sudden change in school performance unexplained by the teacher, social withdrawal, and isolation may also be nonspecific behavioral manifestations. Similar to the nonspecific physical complaints, these behavioral complaints may be caused by several other things as well. Sexual abuse is likely to produce a behavioral change in children old enough to comprehend the wrongness and shamefulness of the situation.

Management

The primary goals in case management of the sexually abused child are to identify and report the abuse and to avoid the secondary abuse phenomenon. *Secondary abuse phenomenon* refers to the physical examination that is so overzealous that it assumes a rape-like quality in the mind of the child. Also to be avoided are parental or staff reactions that make the child feel responsible or blamed for the abuse. Many centers have moved to performing a screening function. If the child has been abused in the past 72 hours or more and no acute symptoms (e.g., bleeding, signs of sexually transmitted disease) are present, the child is referred to a sexual assault center provided such a community service exists. In places where no center is functioning, the entire evaluation must be done in the ED. The following sections offer techniques in management to identify suspected sexual abuse and to gather enough documentation for legal purposes in a manner that is humane for the child and supportive for the family. Chapter 130 details the management of adolescent rape victims.

Interviewing the Parent

Parents may present the problem of sexual abuse either directly or indirectly. For the parent who is direct (i.e., “My child's been abused”), it is important to provide a controlled, quiet environment because he or she will be upset and angry. It may be necessary to limit what is said in front of the child. With such parents, the interviewer's tasks are calming, limiting, and clarifying. In an example of the indirect presentation, the parent brings the child for complaints such as those detailed in the sections on nonspecific physical or behavioral manifestations. With this parent, the task of the interviewer is to bring the possibility of sexual abuse into the open. Once the topic is nominally broached, it becomes apparent that the parent has often already given it consideration. With both types of parents, exploring both their concerns and their information in detail is important.

Recent public media attention to the problem of child sexual abuse has contributed significantly to raising the level of awareness about this problem. The increased number of reported cases may be directly linked to growing public and parental concern. Most of the parents bringing their child to the ED initiate their visit based on a real observation or a strong feeling that something has happened to their child. The emergency physician must help clarify what the initiating cause may have been. In a rare situation, the parent may have responded to pure anxiety that their child may have been abused; however, these parents can provide no substantive cause for their concern. Arrangements should be made for parents in this latter group to have at least one follow-up visit to a sexual abuse center to again explore their motivation before putting their concerns to rest.

Interviewing the Child

Beyond standard history-taking from the parents, the emergency physician must always obtain a history from the child. This task is difficult for several reasons: 1) the child's level of language development, 2) the child's level of psychosexual development, 3) the desire not to contaminate what may be important evidence, 4) the apprehension of the child and the parent, and 5) the awkwardness and apprehension felt by the interviewer in discussing sexual matters with a child. The first steps are for the physician to obtain a quiet, private place and to decide whether he or she wishes the parent to be present. If possible, the physician may wish to defer the comprehensive examination to a more appropriate time and place. Based on previous history-taking from the parent, the physician can gauge the parent's level of emotional composure. This criterion is useful to decide whether parents should be present. If the parents are excluded, another third party (e.g., a nurse or social worker), should be present. An initial discussion of topics, other than the alleged abuse, comforts the child and encourages him or her to talk to the interviewer. Information about school, peers, and family adjustment is important in looking for nonspecific behavioral manifestations, and this preliminary conversation also helps evaluate the child's developmental level. Table 128.9 briefly outlines normal sexual developmental stages and appropriate interviewing techniques for each level.

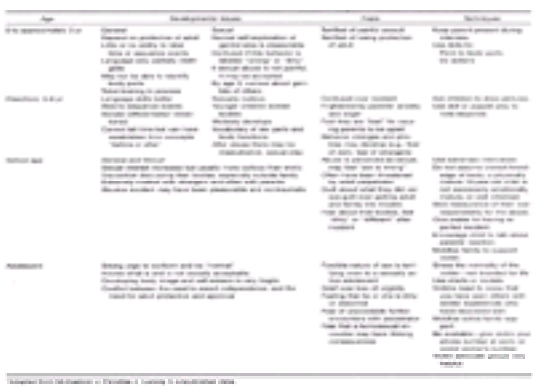


Table 128.9. Developmental Issues in Managing the Sexually Abused Child

In focusing the conversation on the abuse, one technique may be to ask the child why his or her parents brought him or her to the hospital. Another approach that is more appropriate for younger children is to establish common vocabulary by asking the child the term used for his or her genitalia. Children offer a rich variety of terms and may have no understanding of the words *“vagina”* and *“penis.’* One 4-year-old girl stated “He tried to put his pencil in my pocketbook.” In eliciting and using common language, the physician gets to the point of the interview more easily.

If the parental history and surrounding circumstances are credible, inquiries should be phrased to obtain the details of the abuse rather than to ask the child to make the initial allegation. For example, the physician may want to directly ask “How did Uncle Tommy touch your pee pee (vagina)?” rather than “Did Uncle Tommy touch you?” If questions are phrased in a yes-or-no format, a one-word response will be given. Obtaining detail is important to add credibility to the history. The history is also important in guiding the physician to significant aspects of physical examination, evidence collection, and treatment.

With a preverbal child, using anatomically correct dolls may be helpful (Table 128.9). These dolls allow the child to play out the episode. Similarly, some children may choose to draw a picture that tells the story or that may be used by the interviewer to initiate the interview. These techniques are fraught with hazard as far as legally sound methodology, but when simple interviewing fails, making a diagnosis and protecting the child is more important than protecting the legal process.

Emotional Support

Throughout the interview and in all contacts with the child, the rightness of his or her decision to discuss the abuse should be stressed. A child experiences conflict about revealing a secret, especially a long-standing secret. The patient

may also feel conflict in sensing that his or her actions may be provoking a great deal of emotional turmoil. Often, the child has a relationship with the perpetrator and realizes that this admission may alter or end the relationship. At times, the child is aware or is made aware of getting the perpetrator “in trouble.” Reaffirm the importance of what the child has revealed and focus the wrongdoing on the perpetrator. The child may have been threatened not to tell. Thus, bringing the nature of the threats into the open and offering protection to the child are important concerns. Finally, many children have fears about the abuse. Common fears are shown in Table 128.9. The physician may anticipate and address these fears based on the child's development.

Physical Examination

The physical examination may be a point of significant trauma for the child. The examination should be conducted in a standard fashion with all parts of the body examined. The position of the child depends on age and comfort. Many young children want to be examined while sitting in their parent's lap. In examining the genitalia of young girls, two positions are recommended. One is a frog-leg posture while sitting on an adult lap. Alternatively, the child can lie prone with knees tucked under the thorax (i.e., the knee–chest position).

In the prepubertal girl, only the external genitalia need be examined. If even minimal vaginal bleeding appears to be coming from a more internal source, exploration and possible surgical repair is best done under general anesthesia in the operating room. Examination in the ED should be deferred. In the pubertal child, a full genital examination should be performed. This examination may be modified if it is a girl's first speculum examination and it proves too difficult. Chapter 94 details physical examination techniques.

Examination of the rectum and oropharynx needs to be carefully performed, particularly if the history suggests that these were sites of sexual contact. Other physical findings to note carefully are any contusions, abrasions, or lacerations in nongenital areas. Common sites for these signs of trauma are the upper thighs, buttocks, and upper arms. Figure 128.14 shows some of the visible findings of child sexual abuse.

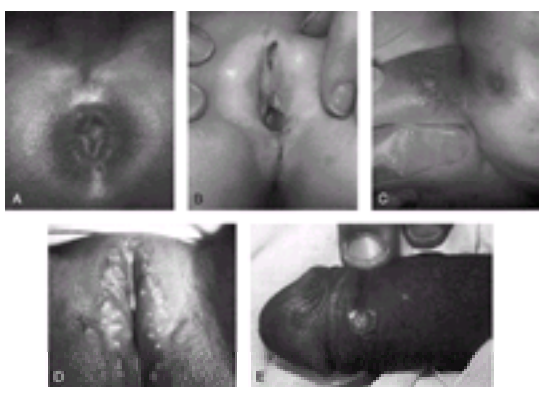


FIGURE 128.14. Physical signs of child sexual abuse. **A.** Rectal dilation and multiple lacerations after sodomy. **B.** Multiple vaginal and paraclitoral lacerations. **C.** Herpes virus infection in perirectal area. **D.** Herpes virus vaginitis. **E.** Syphilis in a sexually abused adolescent.

If the physical examination proves too traumatic for the child, the physician is faced with a significant dilemma. The choices are to further traumatize the child or to perform an incomplete examination and collect inadequate evidence. As with all dilemmas, the best choice is not obvious. Physically or psychologically traumatizing the child should be avoided. Often, no physical evidence is present, and the history, if detailed enough, may be sufficient. The guiding principle should be primum non nocere (first do no harm).

Evidence Collection

The type of evidence to be collected, the collection methods used, and the procedures for processing the results vary by locale. The specimen-collecting procedures at the Children's Hospital of Philadelphia have been reviewed by the Philadelphia Police Department and District Attorney. The protocol is listed in Table 128.10. Whatever the specifics of a particular jurisdiction, some general principles should be followed. Establishing a standard protocol is important so that each new case does not force the emergency physician to reformulate the entire process. The ED should have on hand either standard “rape kits” or some modification thereof. The kits should contain all of the necessary tubes, slides, swabs, and supplies. Evidence collection should be performed with another health care professional present, either a nurse or a social worker. A standard for marking the specimens should be established, including the patient's name and medical record number. Finally, the protocol should include a procedure for a specified person to take the specimens to the laboratory and to officially have them received or logged in by the laboratory. This detail becomes important in the court proceedings against the perpetrator. For example, nothing is more unsatisfying than seeing an alleged perpetrator go unconvicted because the hospital cannot be legally sure that a positive gonorrheal culture belongs to the victim in question.

1. Child's history in detail
2. History of observers
3. Documentation of general physical examination; note signs of force (e.g., bruises)
4. Documentation of genital injury (colposcopy)
5. Documentation of sexual contact
   - Presence of sperm or semen (e.g., on patient's clothing, linens)
   - Sexually transmitted disease
   - Pregnancy
   - Foreign material
6. Documentation of perpetrator
   - Sperm: motile/nonmotile
   - Seminal fluid
     - Genetic marker (blood group antigens)
     - Acid phosphatases
   - PSG glycoprotein
   - Blood
   - Hair analysis
   - DNA matching

Adapted from Tellbrug (personal communication).

Table 128.10. Evidence for Child Sexual Abuse

Consider a Differential Diagnosis

In any consideration of abuse, the physician must always consider the question, “What else could it be?” There may be plausible explanations such as accidental injuries to the genitals as in straddle injuries. Other important alternatives to consider are: 1) infections such as streptococcal, *Haemophilus influenzae,* and monilial; 2) congenital anomalies such as hydrometrocolpos, hemangioma, and perineural clefts and pits; 3) foreign bodies of the rectum and vagina; and 4) dermatologic conditions such as lichen sclerosus et atrophicus, diaper dermatitis, contact dermatitis, Ehlers-Danlos syndrome, and phytodermatitis. Perhaps the most common mistaken perineal finding is prolapsed urethra, which appears as a hemorrhagic mass covering the upper vaginal area.

Documentation

Careful record-keeping cannot be stressed too strongly. As with the collection and processing of evidence, ED records can make or break a case. The Children's Hospital of Philadelphia has developed a separate form that guides the examining physician to include all of the pertinent information (Fig. 128.15). All of the aspects of record-keeping mentioned in the section on physical abuse apply. In particular, what the child said in his or her own words should be carefully recorded. Such questions may be the mainstay of any legal actions to be taken. Good records not only help the police and lawyers involved, but also help the physician review the case before a hearing that may not take place for 6 months. In some jurisdictions, legal provisions allow videotaping patient interviews. This tool is particularly helpful to the victim in that he or she may not have to repeat the history so many times. It may also be helpful to the physician by serving as another form of documentation. When such tapes are shown during court proceedings, they are most helpful.

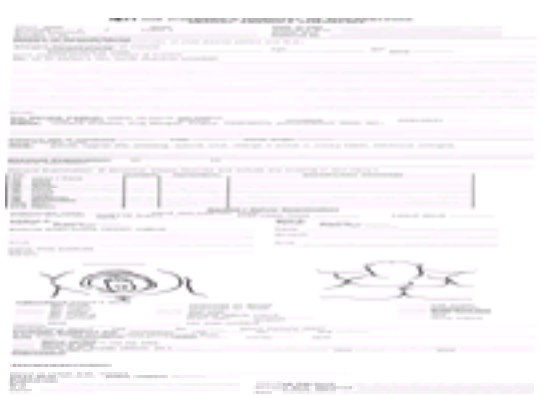


FIGURE 128.15. Sexual assault checklist.

Diagnosis

The diagnosis of sexual abuse should be based on a composite of the history, physical examination, and laboratory findings. Many centers have begun to use a four-category classification of assessment developed by Adams and Harper (Table 128.11) using a classification of physician findings noted in Table 128.12.

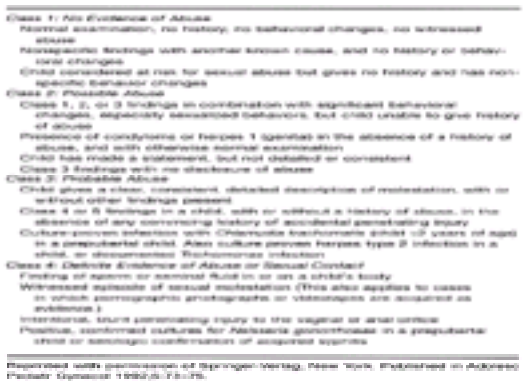


Table 128.11. Overall Assessment of the Likelihood of Sexual Abuse

Table 128.12. Proposed Classification of Anogenital Findings in Children

Reporting

In most jurisdictions, sexual abuse is a criminal offense. Thus, all cases are reported to the police department. In some jurisdictions, when the abuse has occurred in the home or during a time when parental supervision was lax, a civil report to the CPS agency may also be required. This detail needs to be specified according to local guidelines and included in an ED procedure. In the event of a criminal (police) report, a civil (child abuse) report, or both, the parent needs to be informed that such reports are being made. The physician or social worker must spell out the practical consequences of the reports for the parent.

Preparing the Parent

Beyond notifying the parent about reporting the sexual abuse, additional preparation must be given. Many workers believe that, for the young child, the parental reaction to sexual abuse may have as important a role as the abuse itself in producing subsequent manifestations. Long-term follow-up studies of sexually abused children performed using a case control methodology show that the sexually abused child is at long-term risk for a variety of psychological and behavioral consequences. Parents need to be aware of this correlation. All parents will be upset. All parents will be angry. Some parents may express disbelief or the feeling that “this could not be happening to me.” Parental reactions vary depending on whether the abuse is intrafamilial or extrafamilial. Social worker consultation and collaboration for this aspect of case management are essential.

The first step is to focus the parent's attention on the child. Especially in situations of father–daughter incest or surrogate father–daughter incest, the maternal reaction may initially be more self-centered. In directing the parent's attention to the child, the physician returns the parent to a more comfortable traditional role and raises many important issues to which the parent must attend. The physician should review with the parent that manifestations of sexual abuse may be physical and/or psychological. The physical manifestation may seem minor to the physician, but parents must be specifically told that whatever the injury, it can be repaired with no impairment of the victim's sexual functions as an adult. The physician should stress that soft tissues can be repaired, and gonorrhea, if present, can be treated.

Parents must be told that the psychological outcome, in part, relates to their reactions to the situation. Their role must be to provide comfort, support, and reassurance to the child. Methods of supporting the child have been raised previously in this section. These techniques must be used by the parents. Unfortunately, parental anger and blame may sometimes be displaced onto the child, who has set off an emotional time bomb by revealing the abuse. For example, parents may openly say, “I told you not to play with those children down the street,” in a case of extrafamilial abuse. In cases of intrafamilial abuse, the parent may have an even more damaging perception that the child lured the adult into the abuse. Statements such as “I knew she would end up no good” or “I told her never to walk around the house in her underwear” are not uncommon. All attempts to place responsibility or blame on the victim need to be eliminated. The ultimate responsibility was that of the perpetrator, no matter what the behavior of the child.

Some parents may focus their anger clearly on the perpetrator but wish to “take the law into their own hands.” Parents should be cautioned about leaving the ED to find and confront the perpetrator. This action is a police responsibility. The parental role is to provide safety for the child. Parental ire can be cooled by pointing out that a parent's arrest for assault of the perpetrator will not benefit the child.

Yet another common parental reaction is to want to institute several lifestyle changes. Parents may want to change their place of residence, change the child's school, or quit their jobs to be able to guard the child 24 hours a day. The ED staff should stress that returning to as normal a lifestyle as possible is best for the child. Change is always difficult for children, and the stress of entering a new school or meeting a new set of friends is a burden the sexual abuse victim does not need. Parents should be cautioned about limiting the amount of open conversation with friends and relatives that the child may overhear. The victim's desire to discuss the abuse should regulate how much abuse-related conversation should take place between family members. Some children may want repeated reassurances about their parents' approval and about their future safety. Other children may wish to let the episode be forgotten and return to school and play. If the sexual abuse triggers disruption and chaos in the parents' life and disapproval of the child, it will surely have psychological ramifications. The best prognosis results if the parents can show their concern in a way that assures the

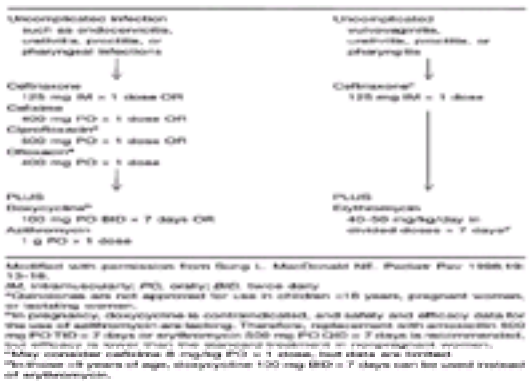
child of approval, protection, and resumption of a normal lifestyle.

Hospitalization

Two indications for hospitalizing the sexually abused child are: 1) severe injury requiring treatment and 2) an unsafe home. However, outpatient management of sexual abuse victims is always preferable. The rationale is to avoid victimizing the child twice. If an adult male has been the intrafamilial perpetrator, he should be removed from the home so that the child may return. Children who are hospitalized because the home is unsafe believe that they are being sent away for their wrongdoing. Another message hospitalization may transmit is that the parent is incapable of providing protection. Both of these messages are harmful to the child's psychological adjustment.

Treatment

Whether the child is hospitalized or discharged from the ED, three additional issues should be considered: 1) gonococcal prophylaxis, 2) human immunodeficiency virus (HIV) testing, and 3) pregnancy prevention. If the abuse has occurred less than 48 hours before the hospital visit, gonococcal prophylaxis is recommended. Within this time period, cultures for *Neisseria gonorrhoeae* may prove negative, even if a true infection is incubating. In abuse that has occurred more than 48 hours before the visit, the choices are either to treat all children prophylactically or to culture the genitalia, anus, and throat and await culture results. This choice depends in part on the reliability of the microbiology laboratory in recovering *N. gonorrhoeae* (which is a fastidious organism) and the ability to provide follow-up treatment for positive cultures. Recommended treatment regimens are shown in Table 128.13 (see also Chapter 94).



Modified with permission from Sung L, MacDonald NE. Pediatr Rev 1998;19:13–18.
IM, intramuscularly; PO, orally; BID, twice daily.
*Ciprofloxacin are not approved for use in children <18 years, pregnant women, or lactating women.
†In pregnancy, doxycycline is contraindicated, and safety and efficacy data for the use of azithromycin are lacking. Therefore, replacement with amoxicillin 500 mg PO TID × 7 days or erythromycin 500 mg PO QID × 7 days is recommended, but efficacy is lower than the standard treatment in nonpregnant women.
‡May consider cefixime 8 mg/kg PO × 1 dose, but data are limited.
§In those <8 years of age, doxycycline 100 mg BID × 7 days can be used instead of erythromycin.

Table 128.13. Treatment Guidelines—Gonorrhea

HIV testing is already an issue on the minds of most parents of sexual abuse victims. Transmission of HIV during child sexual abuse contacts has been reported. Routine testing for HIV may depend on the nature and extent of sexual contact, regional rates of HIV infection, ability to test the alleged perpetrator, and parental desires for testing. All HIV testing should be preceded by written consent and should be done in a manner that results in proper notification, counseling, and follow-up. See Chapter 85 for more on HIV infections.

Most children can be eliminated from consideration for pregnancy prophylaxis if they have not yet reached menarche. Another group may be eliminated based on the nature of the abuse. If the abuse took the form of fondling, and no ejaculate was produced by the perpetrator, prophylaxis does not need to be used. In the pubertal child not using contraception, in whom active penetration occurred at a midmenstrual cycle time, pregnancy prophylaxis should be offered. Documenting that the patient does not have an existing pregnancy is always important. The Children's Hospital of Philadelphia's current therapy is a singleday treatment with oral contraceptive pills (see Chapter 130).

Referral and Follow-Up

All sexually abused children need some form of referral and need careful follow-up care. Referral may initially be to the hospital social worker for monitoring of the child's symptoms and the family's ability to cope with this stress. In some locales, volunteer self-help groups organized for women who have been raped may provide support to the child victim and parent. Referral for more in-depth mental health counseling depends on the 1) symptoms manifested by the child, 2) state of family organization, 3) length of time the abuse has occurred, and 4) the child's age. In general, the older the child and the longer the abuse has occurred, the more likely he or she may have or may develop a serious mental health problem. All children should be referred to mental health services. Follow-up health care visits should be arranged with an informed and sympathetic practitioner who can continue the humane and supportive care initiated in the ED.

False Allegation and Unfounded Reports

At times, the emergency physician is called on to establish the diagnosis of sexual abuse without adequate data. Two situations may be at play: a false allegation or an unfounded report. False allegations refer to situations in which a child sexual abuse complaint is being used by one adult against an estranged wife or husband. These situations are extremely difficult to manage from the ED. Beyond taking a detailed history and performing a physical examination to rule out acute injury, these cases are best referred to a child abuse center. False allegations are often identified by weekend presentation, lack of both history from the child and physician findings, upcoming custody hearing, and bitter divorce situation. Unfounded reports often occur because of undue parental concern stimulated by a media presentation or public discussion of sexual abuse or an unwarranted fear stemming from a parent's own childhood sexual abuse experience. Historical indications by the child or physical findings may be absent. Unfounded reports should also be referred to the

local child sexual abuse center for further evaluation if parental concerns cannot be satisfied.

NEGLECT

Child neglect is by far the most prevalent form of child abuse. When neglect is blatant, it is easily recognized and reported. More often, neglect is not obvious and goes undetected for long periods. Although the manifestations of neglect are less dramatic than those of physical abuse, the long-term effects may be more destructive to the child. The indolent nature of child neglect makes it a serious public health problem. For the ED staff, neglect cases are difficult because they require that certain value judgments be made. The balance between supporting the independent rights of the child and maintaining the privacy and sanctity of family rights is delicate. With neglected children, the questions are: 1) How much should the family be doing? 2) How much are they capable of doing? 3) How much support from the community or society do they require? and 4) How much support or help are they willing to accept on behalf of their child? As with the other forms of child abuse, the management of child neglect cases is made easier by working with a multidisciplinary team. In the ED, the team would generally consist of the physician, nurse, and social worker. Particularly in situations in which the line between adequate child care and neglect needs to be drawn, the diversity of personal and professional opinions adds credibility to decision making and lessens the burden on the single practitioner.

Background

The definition of child neglect is difficult because no societal standards for child care are explicitly stated. This vagueness creates a situation in which parents and professionals are left to define their own standards that produce many problems. Defining *"abandonment"* (the ultimate neglect) is an excellent example of the difficulties encountered in setting standards. No societal norm is established for either the age of the child left alone or the duration of time. Thus, if a parent leaves a 3-year-old child unattended in a home for several hours, most neighbors and professionals would consider that neglect. But what if the child were 10 years old, and the parent was gone for 20 minutes? Then the situation becomes less clear.

Many states have tried to partially define the condition by making it depend on a physical manifestation in the child. Thus, the child left alone who stays out of trouble is not neglected; the one who burns himself or herself and cries to the point that the neighbors become aware is neglected. The National Center for Child Abuse and Neglect uses the following definition: child neglect is characterized by failure to provide for the child's basic needs. Neglect can be physical, educational, or emotional. Physical neglect includes refusal of or delay in seeking health care, abandonment, expulsion from the home or refusal to allow a runaway to return home, and inadequate supervision. Educational neglect includes the allowance of chronic truancy, failure to enroll a child of mandatory school age in school, and failure to attend to a special educational need. Emotional neglect includes actions such as significant inattention to the child's needs for affection, refusal of or failure to provide needed psychological care, spouse abuse in the child's presence, and permission for drug or alcohol use by the child. The assessment of child neglect requires consideration of cultural values and standards of care as well as recognition that the failure to provide the necessities of life may be related to poverty.

Other apparent societal conflicts are inherent in defining neglect. One conflict centers on the relationship between neglect and poverty. This situation relates to the questions asked previously and the issue of what the family should be doing versus what they are financially capable of doing. Certainly, at times, poverty may be coincident with neglect, but the issues are distinctly separate. Most poor families find a way to provide the material essentials and, more important, the emotional essentials for their children. The contrasting example is that of families who are in the middle or upper socioeconomic scale and who could provide for their children but who are neglectful. Another conflict is based on the failure to recognize the evils of excess. The child who is underfed and wasted is promptly labeled *"failure to thrive."* The child who is overfed to the point of obesity may face as many serious consequences. The child who is developmentally delayed from understimulation may be labeled *neglected.* The child who has psychosomatic illness from being stressed to overachieve also deserves identification and help. The term *neglect* generally refers to underprovision on the part of the parent. Overprovision may be as deleterious, which presents a serious conflict to the definition of neglect. The true incidence of neglect is not ascertainable.

Dynamics

The dynamics of child neglect have been explained by several different theorists. Some theories are based on purely individual dynamics and point to the immature, overwhelmed, overstressed parent who responds by withdrawal. Polansky has well documented and labeled this condition as the "apathy futility syndrome." Theories that have a more social orientation point to the societal pressures and to the existence of poverty. David Gil considers all children existing on a welfare stipend as neglected and recommends a change in the distribution of societal resources as a solution. Most theories would support the notion that the neglectful parent does not see himself or herself as such. Most parents are caught up in a neglectful lifestyle that is self-perpetuating. Most neglect is not purposeful, it just happens. Studies have clearly shown that women who fail to seek prenatal care are also likely to not obtain health care for their children. In the parents' view, it is a pattern of living that seems to be the norm. This facet of the dynamics of neglect is important to the ED management.

Manifestations

The manifestations of neglect are countless. The manifestation may be tangible, such as the weight loss of a child whose diet has been inadequate, or intangible, such as the psychological effects of unsatisfactory relationships. A categorization of neglect can be made by looking at the standard functions of the family and then considering the failure to fulfill these functions. A family must 1) provide and distribute material goods—food, clothing, and shelter; 2) ensure health; 3) promote safety; 4) socialize and educate; and 5) provide emotional support, security, and love. The manifestations of neglect may occur in one or more of these functional areas. The most commonly reported manifestations are: 1) nonorganic failure to thrive—a lack of food and feeding skills; 2) medical neglect—a failure to provide needed health care; 3) abandonment—total neglect, generally viewed as a lack of supervision and as a safety

hazard; and 4) truancy and school avoidance.

Failure to Thrive

The term *failure to thrive* has been used as a diagnostic term to group several diseases and disorders that result in growth failure. Growth failure is generally measured in weight, length, and head circumference as compared with standard growth curves for these parameters. Growth failure may be defined as measurements that fall below 2 standard deviations for age or patterns that cross percentile lines (i.e., move 2 standard deviations) and do not follow the normal lines of growth. Patients diagnosed as failure to thrive may be subcategorized into three groups: 1) organic, 2) nonorganic, and 3) mixed group. *Organic* refers to children whose failure to thrive is based on a physical cause such as congenital heart disease, renal disease, or a genetic abnormality. *Nonorganic* refers to the group whose growth failure is environmentally related. When these children are hospitalized and fed standard diets, they grow rapidly and thrive. The group of patients with nonorganic failure-to-thrive includes a substantial number of neglected children who may be brought to the ED for care. The mixed group refers to patients who have a combination of physical and environmental factors. An example might be a physical condition that so overstresses a family that they cannot function, and thus neglect the child in some aspect of the feeding process.

In recognizing the patients with nonorganic failureto-thrive, the following factors are suggestive: History: 1) an idealized feeding history; 2) a chief complaint and history that do not identify the child's growth pattern as a problem; 3) no description of losses, such as vomiting or diarrhea; and 4) failure to give a history of a schedule or scheduled pattern of feeding (e.g., baby eats about every 4 hours). Physical examination: 1) measurements in which weight is more depressed than length, which is more depressed than head circumference; 2) other signs of neglect such as poor hygiene, diaper rash, and flat and balding occiput; 3) dull, apathetic facies; 4) body posture of an understimulated child; 5) excessive oral self-stimulation; and 6) developmental delay, particularly in the social adaptive and language areas. Parental observation: the parent who 1) has an uninterested attitude; 2) does not respond to child's needs (e.g., react to crying); 3) lacks concern about health issues; and 4) appears to be a drug or alcohol abuser. These factors are shown in Figure 128.16.

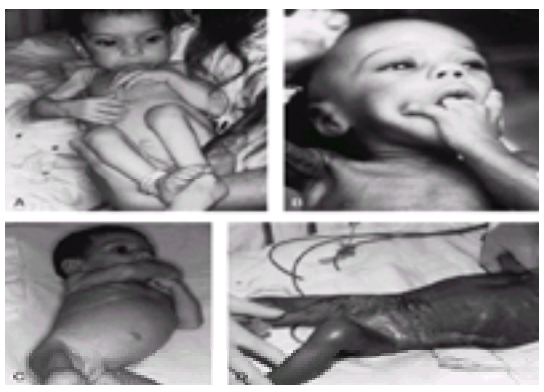


FIGURE 128.16. Physical signs of failure to thrive. **A.** Dull, apathetic eyes that avoid eye contact. **B.** Oral self-stimulatory behavior. **C.** Wasted extremities and protuberant abdomen. **D.** Severe diaper rash as a sign of overall neglect.

Medical Neglect

The differentiation between medical neglect and noncompliance is often difficult. The key to differentiating them is to ask, “Has identifiable harm come to the child?” If a parent fails to complete a course of therapy prescribed by a physician, noncompliance exists. However, if the failure to give medication results in further illness in the child, medical neglect exists. The manifestations of medical neglect can be documented and reported as such. Noncompliance merely results in a worsening doctor-patient relationship. Proving that the failure to give medication, attend follow-up appointments, or obtain a procedure directly resulted in damage to the child's health may be difficult. Intervening variables, such as the complexity of the disease (e.g., the exacerbations of an asthma attack), or the proven efficacy of the treatment often exist. The ED is often the central place for identifying the manifestations of medical neglect. Good documentation of prescribed treatments and good communication with the source of the child's ongoing health care are important.

Abandonment

Local jurisdictions may dictate the length of time a child must be without supervision before he or she is declared legally abandoned. These cases often come to the ED as the result of a neighbor's call or the initiative of a relative who is aware of the neglect. At times, the situation may become apparent as the ED attempts to obtain permission to treat a child and has difficulty locating a parent or responsible adult. Manifestations of abandonment include: 1) physical findings such as excessively dirty diapers, poor hygiene, or hoarse cry; 2) excessive hunger documented by unusual intake; and 3) dehydration as documented by urine specific gravity or blood urea nitrogen. Other manifestations may relate to lack of supervision and protection and may include burns, ingestions, or repeated accidents. Children with all of these manifestations may be brought to the ED for treatment. Good case management results in their identification.

Truancy

Truancy as a manifestation of neglect may be less commonly recognized in the ED. The section on school avoidance (see Chapter 129) details many of the aspects of this complex psychosocial emergency. The emergency physician may recognize truancy as a neglect problem when the truant child presents with multiple somatic complaints. As the complaints are explored and no organic basis is found, the parent may be instructed to return the child to school. A

failure to comply with this aspect of treatment constitutes medical neglect. For the child who makes frequent visits to the ED, neglect needs to be considered.

Management

The management of cases of child neglect follows the principles detailed in the “Physical Abuse” section. The steps are to: 1) suspect and recognize neglect manifestations; 2) obtain multidisciplinary consultation; 3) report the neglect; 4) inform the parents; 5) determine the need for hospitalization; and 6) arrange follow-up. These steps are reviewed in the following sections to underscore those aspects unique to neglect.

Suspect Neglect

As with other forms of abuse, the open mind of the physician allows neglect to be recognized. Because the manifestations are more subtle than with physical abuse, recognition is more difficult. The physician can overcome this difficulty only by obtaining a more detailed history and observing the parent-child interaction. In-depth social work evaluation can often uncover previous reports of neglect or involvement with child welfare agencies. Piling the building blocks of suspicion to the height of a threshold point may be more difficult because the size of each block may be smaller and less dramatic than in physical abuse.

Multidisciplinary Consultation

Because much of defining neglect is value laden, using a multidisciplinary consultation is vital. Such consultation can broaden perspectives on “normal” lifestyles and child-rearing practices. The difficulty any one professional feels in making a value judgment can be shared by a group. The multidisciplinary consultation may be made with someone outside the ED. Speaking with a school teacher, nurse, or counselor may be informative. A public health nurse or visiting nurse may have worked in the family's home and have excellent insights. Any multidisciplinary consultations are of value.

Reporting

Most states provide that reports of child neglect go to the CPS agency. Some states have joint police and social work (CPS) reporting. Criminal charges under the rubric of “endangering the welfare of another” may be brought against some neglectful parents. However, the greatest number of neglect cases come under the supervision of CPS. Police involvement becomes almost essential in cases of abandonment, and police have special skills in locating a missing parent.

Informing Parents

Informing the parents is more difficult in cases of neglect. Responses may be either active or passive. The term *neglect* often triggers an active and angry response. This reaction occurs because neglectful parents believe they are trying to parent as best as they can. Their perceptions of the neglect are different from those of the ED staff. This difference invariably creates conflict and evokes guilt and anger. When informing the parents, the focus should be on the child. The physician may need to verbally recognize the positive efforts of the parent. Nonetheless, if the result in the child is inadequate, action needs to be taken to help the child. Taking this approach often directs the parents' energy toward the child and sufficiently quiets emotional reactions.

The passive response may be equally disquieting to the physician. It may be seen in parents who are overwhelmed, have inadequate personalities, or are depressed, intoxicated, or retarded. Separating these parental problems from each other may be difficult. The physician may incorrectly assume that the parent does not care what happens to the child because of the lack of any response. With extremely passive parents, even engaging them in conversations may be difficult. Approaching the withdrawn parent with simple questions that do not directly relate to the neglect may be helpful. Often, by initiating a neutral conversation, the physician can learn about the parental problem. Asking the parent to perform a task that requires reading or writing may also be instructive. When informing the parent of the neglect report does not trigger active resistance, the physician may be left with an uneasy feeling that the communication was not clearly understood. However, repeat explanations may not stimulate more parental response.

Hospitalization

The need for hospitalization depends on several factors. Certainly, if the child's physical condition warrants treatment, hospitalization is indicated. For example, a child failing to thrive in the first 6 months of life should be hospitalized. Another indication relates to the degree of parental dysfunction. If the parents are assessed as being so overwhelmed, withdrawn, depressed, or inadequate that they are unable to assume parental responsibility, the child should be protected regardless of his or her current physical status. A third indication stems from the chronicity of neglect. Because of the indolent nature of neglect, the physician may need to make a point of hospitalizing a child to dramatize a chronic or recurrent situation. The final factor in determining the need for hospitalization is the time required for community agency response. If the report of neglect triggers an immediate investigation and institution of therapeutic services, the need for hospitalization is diminished. The response to neglect reports varies by community.

Follow-Up

In cases of neglect, arranging follow-up care by a physician or clinic is important. Often, the step of informing the receiving physician is overlooked. The staff providing the long-term care is expected to have the capacity to closely monitor patients and to become aggressive about correcting failure to keep appointments. The treatment of neglected children is often a long and frustrating process. Thus, special referral resources should be sought. Attempts to provide treatment in standard health care facilities may be doomed because the passive and indolent character of neglect may

escape health care providers and thereby injure the child.

EMOTIONAL ABUSE

Emotional abuse is the form of child abuse that most seriously and most often affects children. With every episode of physical or sexual abuse, a negative psychological message is being inflicted. The child is told, “You are bad!” and often comes to believe this statement. When the bruises, burns, and broken bones are healed, psychological injury may remain untreated. The neglected child also feels devalued and unloved. Emotional abuse always accompanies other forms of abuse and at times is inflicted independently.

Yet, this form of abuse is the least well understood. Furthermore, a report of suspected abuse is rarely based solely on emotional abuse. Gathering enough objective data to prove that emotional abuse has occurred is difficult. Courts and legal authorities remain unconvinced that a given parental behavior or set of behaviors can be shown to be responsible for effects in the child. Emotional abuse rarely results in a psychosocial emergency. More often, similar to neglect, it is a chronic impediment to normal growth and development.

Background

Emotional abuse has been defined in many different ways. The National Center for Child Abuse and Neglect defines it simply: “Child abuse which results in impaired psychological growth and development. Frequently occurs as verbal abuse or excessive demands on a child's performance and results in a negative self-image on the part of the child and disturbed child behavior.” The Pennsylvania Child Abuse Law provides for reporting serious mental injury, which is defined as “…a psychological condition determined by a psychologist, psychiatrist, or pediatrician, and caused by the acts or omissions of a parent or person responsible for the child's welfare, which: (1) renders the child chronically and severely anxious, agitated, depressed, socially withdrawn, or in reasonable fear of his/her life; (2) makes it extremely likely that the child will be chronically and severely anxious, agitated, depressed, socially withdrawn, or be in reasonable fear that his/her life is threatened; or (3) seriously interferes with the child's ability to accomplish age-appropriate developmental milestones or school or community tasks or peer relations.”

The Pennsylvania law identifies some of the potential manifestations of emotional abuse, but does not define the difficult issue of a child's behavior as “caused by” an act or omission on the part of the parent. The issue of causality hampers the reporting of emotional abuse. The incidence of emotional abuse is unknown, and reasonable estimates of its frequency are not available. Although neglect is reported most often, emotional abuse is the most common form of abuse because it occurs along with all other forms of abuse and neglect as well as independently.

Dynamics

Emotional abuse occurs when a parent attacks, belittles, humiliates, or devalues a child. Such behavior is not the infrequent reaction of a normal angry parent, it is an often repeated message to the child. An emotionally abusive parent appears to be determined to verbally destroy his or her child. The reasons for this behavior are many. All of the dynamics responsible for physical abuse may be operative for emotional abuse. Often, poor parental self-image is a strong component. The child either threatens the parent or reflects unwanted parental characteristics. In either case, the parent is unable to withstand the stress and attacks the child. When there are many children in a family, a parent may scapegoat the one child with whom he or she most identifies. At times, the child may be identified with another family member, such as an estranged father or a grandparent.

Helfer has capsulized the intergenerational nature of emotional and physical abuse in his “World of Abnormal Rearing” formulation (Fig. 128.17). The child who is told repeatedly that he or she is “bad” and “worthless” eventually accepts it. Once the belief that “I'm no damn good” exists, the child's behavior matches the expectation. Ultimately, the poor self-image is reflected in the inability to choose friends or a mate and to accept help. The cycle is completed with poor parenting and emotional abuse of the next generation of children. The intergenerational potential for emotional abuse makes it a serious public health problem.

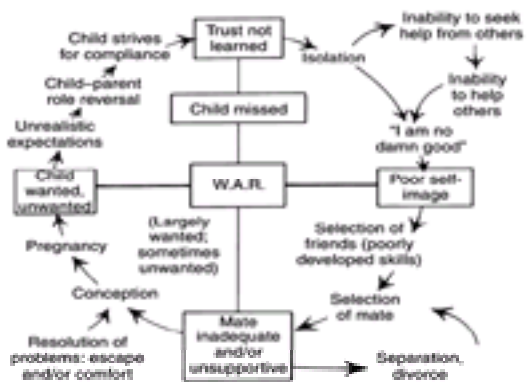


FIGURE 128.17. World of Abnormal Rearing (W.A.R.). (Modified with permission from R.E. Helfer.)

Manifestations

Emotional abuse manifests in many forms, including children who are excessively withdrawn and passive and those who are aggressive and act out. The manifestations are so varied that it is difficult to identify any constant characteristics of

the emotionally abused child. To identify emotional abuse, the physician must witness family interactions on repeated occasions; however, the emergency physician does not often have the opportunity for such comprehensive evaluation.

Rarely, more direct presentations of emotional abuse may be seen in the ED. Some children may seek hospital asylum because of excessive fear of their parents. Adolescent “runaways” may include a subset of children who are emotionally abused (see Chapter 129, Chapter 130, and Chapter 131). Developmental delay may be recognized in the ED, but the cause of the delay can rarely be identified. Finally, children with drug or alcohol abuse may be in a high-risk group for prior emotional abuse. The effects of emotional abuse closely parallel the findings in children with substance abuse (e.g., poor self-image, difficulty in establishing relationships).

Another group of emotionally abused children frequently seen in the ED is youngsters caught in the conflict between estranged parents. These victims have been called “yo-yo” children because they are pulled between arguing parents. Sometimes the children are virtually kidnapped by one parent. These presentations to the ED are based on one parent's attempt to document the poor parenting or neglect of the other. The misuse of the child as a pawn in the marital dispute constitutes emotional abuse. Despite the credibility and motivation of the parent in the ED, emotional abuse should be reported to remove the child from the middle.

Management

The management principles for emotional abuse are the same as those for other forms of abuse. To substantiate emotional abuse, documentation of behavior must be precise. The physician, nurse, and social worker must record observed interactions in behavioral terms using a minimum of subjective assessments and personal conclusions. Recording significant statements from the parents and child is good documentation. Citing a pattern of abuse or repeated episodes of abuse may be necessary to strengthen a report.

It is often difficult and painful for parents to see themselves as emotionally abusive. Thus, informing the parent in a constructive and sensitive way is also difficult. The informant must keep the discussion child-focused and nonaccusatory. Child welfare agencies and the family court system also have difficulty in identifying and treating emotional abuse. As the rights of children become more well-established and standards for child care more widely accepted, management of cases of emotional abuse will become less difficult.

Suggested Readings

GENERAL

A Nation's Shame: Fatal Child Abuse and Neglect in the United States. Washington, DC: US Advisory Board on Child Abuse and Neglect, US Department of Health and Human Services, 1995.

Centers for Disease Control and Prevention. Child homicide—United States. Morbid Mortal Weekly Rep 1982;31:292.

Child Maltreatment 1995: Reports from the States to the National Child Abuse and Neglect Data System. Washington, DC: US Department of Health and Human Services, National Center on Child Abuse and Neglect, 1997.

Christoffel KK, Zieserel EJ, Chiarmonte J. Should child abuse and neglect be considered when a child dies unexpectedly? Am J Dis Child 1985;139:876–880.

Disciplining Children in America: A Gallup Poll Report, 1995.

Gil DG. Violence Against Children: Physical Abuse in the U.S. Cambridge: Harvard University Press, 1970.

Green FC. Child abuse and neglect: a priority problem for the private physician. Pediatr Clin North Am 1975;22:329.

Helfer RE. Why most physicians don't get involved in child abuse cases and what to do about it. Child Today 1975;4:28.

Hodge D, Ludwig S. Child homicide: emergency department recognition. Pediatr Emerg Care 1985;1:3.

Kempe CH, Helfer RE. The Battered Child. 4th ed. Chicago: University of Chicago Press, 1987.

Kempe CH, Silverman FH, Steele BF, et al. The battered child syndrome. JAMA 1962;181:17.

Kleinman PK. Diagnostic Imaging of Child Abuse. 2nd ed. Baltimore: Williams & Wilkins, 1998.

Ludwig S. A multidisciplinary approach to child abuse. Nurs Clin North Am 1981;16:161.

Ludwig S, Kornberg AE. Child Abuse: A Medical Reference. New York: Churchill-Livingstone, 1992.

McClain P, Sacks J, Frohlke R. Estimates of fatal child abuse and neglect, United States, 1979 through 1988. Pediatrics 1993;91:338–343.

Smith JR, Brooks-Gunn J. Correlates and consequences of harsh discipline for young children. Arch Pediatr Adolesc Med 1997;151:777–786.

Straus MA, Sugarman DB, Giles-Sims J. Spanking by parents and subsequent antisocial behavior of children. Arch Pediatr Adolesc Med 1997;151:761–767.

Third National Incidence Study of Child Abuse and Neglect: Final Report (NIS-3). Washington, DC: US Department of Health and Human Services, National Center on Child Abuse and Neglect, 1996.

PHYSICAL ABUSE

Arkovitz MS, Johnson N, Garcia VF. Pancreatic trauma in children: mechanisms of injury. J Trauma: Inj, Inf, and Crit Care 1997;42(1):49–53.

Banco L, Lapidus G, Zavoski R, et al. Burn injuries among children in an urban emergency department. Pediatr Emerg Care 1994;10:98–101.

Berkowitz CD. Pediatric abuse: new patterns of injury. Emerg Med Clin North Am 1995;13:321–341.

Billmire ME, Myers PA. Serious head injury in infants: accident or abuse. Pediatrics 1985;75:340–348.

Brewster A, Nelson J, Hymel K, et al. Victim, perpetrator, family and incident characteristics of 32 infant maltreatment deaths in the United States Air Force. Child Abuse Negl 1998;22:91–101.

Bruce DA, Zimmerman RA. Shaken impact syndrome. Pediatr Ann 1987; 18:482–487.

Bryk M, Siegel PT. My mother caused my illness: the story of a survivor of Munchausen by proxy syndrome. Pediatrics 1997;100:1–7.

Cameron CM, Lazoritz S, Calhoun AD. Blunt abdominal injury: simultaneously occurring liver and pancreatic injury in child abuse. Pediatr Emerg Care 1997;13:334–336.

Chadwick DL, Chin S, Salerno C, et al. Deaths from falls in children: how far is fatal? J Trauma 1991;31:1353–1355.

Clark KD, Tepper D, Jenny C. Effect of a screening profile on the diagnosis of nonaccidental burns in children. Pediatr Emerg Care 1997;13(4):259–261.

Coant PN, Kornberg AE, Brody AS, et al. Markers for occult liver injury in cases of physical abuse in children. Pediatrics 1992;89(2):274–278.

Cooper A, Floyd T, Barlow B, et al. Major blunt abdominal trauma due to child abuse. J Trauma 1988;28:1483–1489.

da Fonseca MA, Feigal RJ, ten Bensel RW. Dental aspects of 1248 cases of child maltreatment on file at a major county hospital. Pediatr Dent 1992;14:152–157.

Dailey JC, Bowers CM. Aging of bite marks: a literature review. J Forensic Sci 1997;42:792–795.

Duhaime AC, Christian CW, Rorke LB, et al. Nonaccidental head injury in infants—the “shaken-baby syndrome.” N Engl J Med 1998;338(25):1822–1829.

Feldman KW, Brewer DK. Child abuse, cardiopulmonary resuscitation, and rib fractures. Pediatrics 1984;73:339.

Feldman KW, Schaller RT, Feldman JA, et al. Tap water scald burns in children. Pediatrics 1978;62:1.

Finkel MA, Ricci LR. Documentation and preservation of visual evidence in child abuse. Child Maltreatment 1997;2:322–330.

Gilliland MGF, Folberg R. Shaken babies—some have no impact injuries. J Forens Sci 1996;41:114–116.

Greist KF, Zumwalt RE. Child abuse by drowning. Pediatrics 1989;83:41–47.

Harris VJ, Lorand MA, Fitzpatrick JJ, et al. Radiographic Atlas of Child Abuse: A Case Studies Approach. New York: Igaku-Shoin, 1996.

Hymel KP, Rumack CM, Hay TC, et al. Comparison of intracranial computer tomographic (CT) findings in pediatric abusive and accidental head trauma. Pediatr Radiol 1997;27(9):743–747.

Joffe M, Ludwig S. Stairway injuries in children. Pediatrics 1988;82:457–472.

Kapoor S, Schiffman J, Tang R, et al. The significance of white-centered retinal hemorrhages in the shaken baby syndrome. Pediatr Emerg Care 1997;13(3):183–185.

Kleinman PK, Schlesinger AE. Mechanical factors associated with posterior rib fractures: laboratory and case studies. Pediatr Radiol 1997;27:87–91.

Leavitt EB, Pincus RL, Bukachevsky R. Otolaryngologic manifestations of child abuse. Arch Otolaryngol Head Neck Surg 1992;118:629–631.

Musemeche CA, Varthel M, et al. Pediatric falls from heights. J Trauma 1991;31:1347–1349.

Nashelsky MB, Dix JD. The time interval between lethal infant shaking and onset of symptoms: a review of the shaken baby syndrome literature. Am J Forens Med Pathol 1995;16(2):154–157.

Ng CS, Hall CM, Shaw DG. The range of visceral manifestations of non-accidental injury. Arch Dis Child 1997;77:167–174.

Nichols JS, Elger C, Hemminger L, et al. Magnetic resonance imaging: utilization in the management of central nervous system trauma. J Trauma 1997;42:520–524.

Odom A, Christ E, Kerr N, et al. Prevalence of retinal hemorrhages in pediatric patients after in-hospital cardiopulmonary resuscitation: a prospective study. Pediatrics 1997;99(6):e3.

Reece RM. Unusual manifestation of child abuse. Pediatr Clin North Am 1990;34(4):905–921.

Schwengel D, Ludwig S. Rhabdomyolysis and myoglobinuria as manifestations of child abuse. Pediatr Emerg Care 1985;1:194.

Sirotnak AP, Krugman RD. Physical abuse of children: an update. Pediatr Rev 1994;15:394–399.

Sivit CJ, Taylor GA, Eichelberger MJ. Visceral injury in battered children. Radiology 1989;173:759–761.

Slosberg E, Ludwig S, Duckett J, et al. Penile trauma as a sign of child abuse. Am J Dis Child 1978;132:719.

Spevak MR, Kleinman PK, Belanger PL, et al. Cardiopulmonary resuscitation and rib fractures in infants. JAMA 1994;272(8):617–618.

Starling S, Holden JR, Jenny C. Abusive head trauma: the relationship of perpetrators to their victims. Pediatrics 1995;95(2):259–262.

Southall DR, Plunkett MCB, Banks MW, et al. Covert video recordings of life-threatening child abuse: lessons for child protection. Pediatrics 1997;100:735–760.

Swischuk LE. Radiographic signs of skeletal trauma. In: Ludwig S, Kornberg A, eds. Child Abuse Neglect: A Medical Reference. 2nd ed. New York: Churchill-Livingstone, 1992.

Thomas SA, Rosenfend NS, Leventhal JM, et al. Long-bone fractures in young child distinguishing accidental injuries from child abuse. Pediatrics

1991;88(3):471–476.

Williams RA. Injuries in infants and small children resulting from witnessed and corroborated free falls. J Trauma 1992;31:1350–1352.

Wyatt JP, McLeod L, Beard D, et al. Timing of paediatric deaths after trauma. Br Med J 1997;314(7084):868.

Zimmerman RA, Bilaniuk LT. Pediatric head trauma. Neuroimaging Clin North Am 1994;4:349–366.

SEXUAL ABUSE

Adams JA, Harper K, Knudson S, Revilla J. Examination findings in legally confirmed child sexual abuse: it's normal to be normal. Pediatrics 1994;94:310–317.

Allard JE. The collection of data from findings in cases of sexual assault and the significance of spermatozoa on vaginal, anal and oral swabs. Sci Justice 1997;37(2):99–108.

Bays J, Jenny C. Genital and anal conditions confused with child sexual abuse trauma. Am J Dis Child 1990;144:1319–1322.

Berkowitz CD. Child sexual abuse. Pediatr Rev 1992;13:443–452.

Branch G, Paxton R. A study of gonococcal infections among infants and children. Public Health Rep 1965;80:4.

Cheasty M, Clare AW, Collins C. Relation between sexual abuse in childhood and adult depression: case-control study. Br Med J 1998;316:198–201.

Christian CW, Singer ML, Crawford JE, et al. Perianal herpes zoster presenting as suspected child abuse. Pediatrics 1997;99:608–610.

Committee on Child Abuse and Neglect, American Academy of Pediatrics. Gonorrhea in prepubertal children. Pediatrics 1998;101:134–135.

Committee on Infectious Diseases. Gonococcal infections. In: Report of the Committee on Infectious Diseases. 24th ed. Elk Grove Village, IL: American Academy of Pediatrics, 1997.

Cox RA. *Haemophilus influenzae:* an underrated cause of vulvovaginitis in young girls. J Clin Pathol 1997;50:765–768.

DeJong AR, Rose M. Frequency and significance of physical evidence in legally proven cases of child sexual abuse. Pediatrics 1989;84:1022–1026.

DeJong AR, Rose M. Legal proof of child sexual abuse in the absence of physical evidence. Pediatrics 1991;88(3):506–511.

Finkel MA. Anogenital trauma in sexually abused children. Pediatrics 1989;84:317–322.

Fost N. Ethical considerations in testing victims of sexual abuse for HIV infection. Child Abuse Negl 1990;14:5–7.

Friedrich WN, Fisher J, Broughton D, et al. Normative sexual behavior in children: a contemporary sample. Pediatrics 1998;101:e9.

Giardino AP, Finkel MA, Giardino ER, et al. A practical guide to the Evaluation of Sexual Abuse in the Prepubertal Child. Newbury Park: Sage, 1992.

Goldberg CL, Yates A. The use of anatomically correct dolls in the evaluation of sexually abused children. Am J Dis Child 1990;144:1334–1336.

Gutman LT, St. Claire KK, Weedy C, et al. Human immunodeficiency virus transmission by child sexual abuse. Am J Dis Child 1991;145:137–141.

Kamarashev JA, Vassileva SG. Dermatologic diseases of the vulva. Clin Dermatol 1997;15:53–65.

Kaplan KM, Fleisher GR, Paradise JE, et al. Social relevance of genital herpes simplex in children. Am J Dis Child 1984;138:872.

Krugman RD. Recognition of sexual abuse in children. Pediatr Rev 1986; 8:25–30.

McCann J, Voris J, Simon M. Labial adhesions and posterior fourchette injuries in childhood sexual abuse. Am J Dis Child 1990;144:242–244.

McCann J, Voris J, Simon M, et al. Comparison of genital examination techniques in prepubertal girls. Pediatrics 1990;86:182–187.

McCauley J, Kern DE, Kolodner K, et al. Clinical characteristics of women with a history of childhood abuse: unhealed wounds. JAMA 1997;227:1362–1368.

Obalek S, Jablonska S, Orth G. Anogenital warts in children. Clin Dermatol 1997;15:369–376.

Paradise J. The medical evaluation of the sexually abused child. Pediatr Clin North Am 1990;39(4):839–862.

Paradise JE, Finkel MA, Beiser AS, et al. Assessment of girls' genital findings and the likelihood of sexual abuse: Agreement among physicians self-rated as skilled. Arch Pediatr Adolesc Med 1997;151:883–891.

Paradise JE, Rostain AL, Nathanson M. Substantiation of sexual abuse charges when parents dispute custody or visitation. Pediatrics 1988;81:835–839.

Peters JJ. Children who are victims of sexual assault and the psychology of offenders. Am J Psychother 1978;30:398.

Pettigrew J, Burcham J. Effects of childhood sexual abuse in adult female psychiatric patients. Aust NZ J Psych 1997;31:208–213.

Sgroi SM. Sexual molestation of children. Child Today May-June 18, 1975.

Sinal S, Lawless M, Rainey D, et al. Clinician agreement on physical findings in child sexual abuse cases. Arch Pediatr Adolesc Med 1997;151:497–501.

Starling SP. Syphilis in infants and young children. Pediatr Ann 1994;23(7):334–340.

Sung L, MacDonald NE. Gonorrhea: a pediatric perspective. Pediatr Rev 1998;19(1):13–16.

Sung L, MacDonald NE. Syphilis: a pediatric perspective. Pediatr Rev 1998;19(1):17–22.

Swanston HY, Tebbutt JS, O'Toole BI, et al. Sexually abused children 5 years after presentation: A case-control study. Pediatrics 1997;100:600–608.

Widom CS, Ames MA. Criminal Consequences of Child Sexual Victimization. Child Abuse Negl 1994;18:303–318.

NEGLECT

Frank DA, Silva M, Needlman R. Failure to thrive: mystery, myth, and method. Contemp Pediatr 1993;Feb:114–133.

Harrington D, Black MM, Starr Jr RH, et al. Child neglect: relation to child temperament and family context. Am J Orthopsych 1998;68:108–116.

Helfer RE. The neglect of our children. Pediatr Clin North Am 1990;37(4):923–942.

Homer C, Ludwig S. Categorization of etiology of failure to thrive. Am J Dis Child 1981;135:848.

Hufton IW, Oates RK. Non-organic failure to thrive; a long-term follow up. Pediatrics 1977;59:73.

Ludwig S. Failure to thrive/starvation. In: Ludwig S, Kornberg A, eds. Child Abuse Neglect: A Medical Reference. 2nd ed. New York: Churchill-Livingstone, 1992.

Margolin L. Fatal child neglect. Child Welfare 1990;69:309–319.

Polansky NA, Borgman RD, DeSaix C. Roots of Futility. San Francisco, Jossey-Bass, 1972.

Polansky NA, Chalmers MA, Buttenwieser E, Williams DP. Damaged Parents: An Anatomy of Child Neglect. Chicago: University of Chicago Press, 1981.

PSYCHOLOGICAL/EMOTIONAL

Cavaiola A, Schiff M. Behavioral sequelae of physical and/or sexual abuse in adolescents. Child Abuse Neglect 1988;12:181–187.

Garbarino J. The psychologically battered child. Pediatr Ann 1989;18: 502–510.

Kilpatrick KL, Litt M, Williams LM. Post-traumatic stress disorder in child witnesses to domestic violence. Am J Orthopsych 1997;67:639–644.

Ludwig S, Rostain A. Family dysfunction. In: Levine MD, Carey WB, Crocker AC, eds. Developmental Behavioral Pediatrics. 2nd ed. Philadelphia: WB Saunders, 1992.

Skuse DH. Emotional abuse and neglect. Br Med J 1989;298:1692–1693.

van der Kolk BA. The psychobiology of posttraumatic stress disorder. J Clin Psychiatry 1997;58:16–24.

Widom C. The cycle of violence. Science 1989;244:160–164.

CHAPTER 129

Psychiatric Emergencies

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INTRODUCTION

In contemporary American society, the emergency department (ED) of the hospital has become a major community resource. It is open 24 hours each day, professionally staffed, and used by the community for assistance in dealing with many physical, social, and emotional problems. The ED is the setting for the initial evaluation of a variety of difficulties of children and their families, including acute and chronic illnesses with their emotional sequelae, psychophysiologic conditions, family crises, and the entire spectrum of emotional and behavioral disorders. ED visits of children with social and emotional difficulties continue to increase, especially for families without a designated primary care physician. Although the medical staff and the community may disagree concerning appropriate uses of the ED, the ED staff must, nevertheless, respond to the problems that are brought to it. Therefore, physicians working in EDs must master the skills of psychiatric diagnosis, crisis intervention, and disposition planning.

Crisis and Crisis Intervention

Psychiatric emergencies are best understood as crisis situations. Crisis involves the acute development of circumstances or events that render the usual coping and adaptive patterns of an individual or social unit inadequate. Most crisis situations during childhood are handled effectively by the child's family. However, in some instances, the child's behavior or the overall situation becomes unmanageable. At this point, professional help is sought, and the child may be brought to the ED. Typically, the family members appear overwhelmed and helpless. Severe anxiety, disagreements among various caretakers, and disruption of previously stable family relationships may have occurred and be ongoing. In other cases, relationships may have been unstable for some time, with the acute superimposition of a crisis.

Psychiatric emergencies in childhood may be defined as crises in which the adults around the child are no longer able to help the child control his or her anxiety and can no longer provide adequate support and control of the child's emotional reactions and behavior. Any psychiatric emergency in childhood implies a limitation of effective interaction of the child and his or her caretakers. For example, the suicidal child is seen by his or her family as being uncontrollably self-destructive and needing professional help. However, the child's suicidal state also reflects the family's inability to assist him or her in developing alternatives other than suicide. Comprehensive assessment and treatment of psychiatric emergencies in children should involve the participation of the child's family. In this way, the family, with professional support, can resume its role in guiding the child's emotional and behavioral development.

Psychiatric crises are not just times of intolerable stress for the family, but also unique opportunities to bring about

change. When a family brings a child to the ED with an emotional problem, the crisis created by the problem has overcome the usual tendency to deny difficulties and maintain the status quo. Because family coping patterns are failing and denial is lessened, intervention at this time can be used to modify nonadaptive patterns and to create alternative solutions. Thus, family crises are nodal points at which the possibility for family change is maximized and during which interventions can extend beyond the amelioration of the concern at hand to have lasting impact.

It follows that the goal of the emergency physician is not to merely alleviate the crisis per se, but to use the crisis to create meaningful change. The physician needs to maintain within the family a sense of urgency that enables it to respond positively. Therefore, the emergency physician needs to be able to assess the immediate crisis and to appreciate through observation and history-taking how the family has become ineffective. Crisis intervention techniques in the ED address not only the complaint, but also the underlying problems that have given rise to it.

Requirements of the Emergency Department

The ability to respond effectively to psychiatric emergencies of children and families requires special capacities of the ED staff. Clinicians in the ED should have a preexisting relationship with a mental health team that is committed to providing child psychiatric consultation at all times. The emergency staff must be capable of collaborating effectively with this mental health team. Requests for psychiatric consultation should provide a clear statement of the problem and the goal of the consultation. To enable adequate response, the degree of urgency of each request should be clearly stated. When the consultant arrives, the ED staff should describe in detail its findings and any interventions made thus far. The emergency physician and the consultant should discuss how to proceed throughout the remainder of the ED visit. Such clarification not only is useful in the current situation, but also fosters mutual respect in the future.

Adequate physical space is another requirement of the ED. A place where interruptions are uncommon should be designated for psychiatric emergencies to be assessed. This room should have seats for each family member and the emergency physician. The availability of a separate room for psychiatric emergencies enables a mood of concern and deliberation to be achieved. The ED should also have a holding or observation room where certain children, such as those recovering from overdoses or those being stabilized with psychotropic medication, can be observed and evaluated regularly. Such a room should be adequately staffed by nurses as well as security personnel, when indicated, and the capacity for using restraints should also be present.

The ED should also have relationships with various psychiatric inpatient units so that hospitalization, when needed, can be arranged efficiently. Relationships with brief, crisis-oriented inpatient units that involve the child and the family in problem-focused treatment are of particular value. The staff should be thoroughly familiar with the procedures for psychiatric hospitalization, including the specific legal requirements for involuntary commitment. In certain situations, such as children recovering from medically serious suicide attempts, medical hospitalization may be necessary. The hospital should have specific guidelines for the management of psychiatric patients on medical floors.

Finally, the ED should have relationships with other social agencies and an awareness of relevant laws. The police should be aware of which children to bring to the ED for psychiatric assessment and should be prepared to remain in the ED until adequate security has been arranged. Relationships should be developed with mental health base service units, temporary shelters, and other crisis intervention centers, ensuring effective referrals when necessary. Staff should be aware of child protection laws and the procedures for emergency intervention in situations of abuse and neglect.

Physician Responsibilities and Skills

The responsibilities of the emergency physician with psychiatric emergencies are shown in Table 129.1. To effectively fulfill these responsibilities, the physician must possess a variety of clinical skills and the ability to block out other concerns when responding to psychiatric emergencies. The physician must be able to display empathy for the child's and family's distress. Once the family senses the physician's concern, it will be more responsive. The physician needs to handle the family's anxiety and uncertainty by approaching the family crisis calmly and systematically. In doing so, the physician establishes the leadership and authority that enables the family to discuss its problems freely and to consider and act on recommendations. Throughout the ED visit, the physician needs to foster the belief among family members that improvement in their situation will be achieved through appropriate changes in family members' behavior and relationships.

Obtain necessary information for database	Develop specific crisis interventions
Rapidly identify crisis situation	Acute medical management
Assess nature and degree of child and family stress	Acute psychiatric management
	Psychiatric consultation
	Independent disposition planning

Table 129.1. Childhood/Adolescent Psychiatric Emergencies: Emergency Physician Responsibilities

Another important skill of the emergency physician involves the ability to obtain and assess relevant information about the child, family, and their community supports. This topic is covered subsequently in the section “Evaluation of

Psychiatric Emergencies.”

Family Responsibilities

A childhood psychiatric emergency implies a limitation of effective interaction between the child and his or her caretakers. In our society, the child's caretakers have the major responsibility for promoting growth and development. In many instances, the child's caretakers are his or her parents, but, in some situations, grandparents, extended kin, foster families, and state agencies have legal custody of the child. The emergency physician must establish who the child's actual caretakers are and try to involve as many of them as possible in the ED. When evaluating the child, the caretakers, and their relationships with each other, the emergency physician should assess the degree to which the parents (or other caretakers) are meeting the following responsibilities:

1. Ensure the physical and emotional safety of the child. Parents need to protect the child, as much as possible, from external danger (e.g., getting lost, walking into traffic, going off with strange adults) and from internal family danger (e.g., neglect, physical and emotional abuse, sexual abuse). If safety is not provided, the existence of the child may be jeopardized.
2. Provide support and nurturance, especially to younger children, such that an emotional bond is established between child and parent. When such an attachment is present, the child desires parental approval and usually responds to parental authority.
3. Provide enough socialization to set limits on the child's behavior. The physician should assess whether the limits chosen are appropriate, too rigid, or too weak.
4. Promote the child's efforts in age-appropriate tasks, including consistent school attendance and performance, learning to relate to peers, and assuming greater autonomy within the family as the child grows older.
5. Instill a sense of competence and mastery in the child at each developmental level. This accomplishment allows the child to successfully integrate new knowledge and new experiences.
6. Assist the child in coping with unexpected failures and losses, including academic disappointment, family disruption, and disability resulting from physical illness.

By keeping these family responsibilities in mind, the emergency physician can assess families in crisis, determining which functions are being met and which need to be supported.

Working with Strengths

The emergency physician working with a family in crisis must look for problem areas, as well as areas of competence in both the child and family. These areas of strength form the basis for a successful treatment plan that enables the family to master the crisis. Typically, families in a crisis do not use their existing abilities enough as they pursue a narrow range of responses to the problem at hand. Through history-taking, observation of family interaction, and assessment of the way that the family relates to the physician, the physician can identify the family's assets. Once recognized, these skills enable the parents to be more confident and competent in dealing with their child. A family that has successfully reared its children until now should also be able to respond effectively to new challenges that the children present. A family that has successfully dealt with aggressive behavior in a child in the past can use some of that previous experience in responding to the current crisis. A family with a disruptive or destructive child that successfully has brought the child to the ED, where he or she sits quietly, has more parenting skills than members may realize. The emergency physician should help the family recognize its capabilities at a time when confidence is at its lowest level.

EVALUATION OF PSYCHIATRIC EMERGENCIES

The evaluation of acute psychosocial emergencies can be divided into five sections (Table 129.2). Orienting data and relevant history indicates the general living situation and previous psychosocial adaptation of the child or adolescent patient. It also provides a complete description of the current crisis, including apparent precipitants. Medical history and physical evaluation determine the child's current physical status. The mental status examination of the child provides information about the patient's current psychological well-being. A family evaluation, using both history and observation of the family's behavior during the ED visit, enables the physician to determine the family's ability to respond to the child's distress. By integrating these sources of information, the emergency physician is well-equipped to understand the crisis and to pursue appropriate treatment alternatives.

1. Orienting data
2. Relevant history
3. Medical history and physical examination
4. Mental status of the child
5. Family evaluation

Table 129.2. Childhood/Adolescent Psychiatric Emergencies: Categories of Necessary Information

Orienting Data and Relevant History

Psychosocial orienting data, as shown in Table 129.3, provide information that enables the physician to appreciate the basic living situation of the child and family. This information can be quickly obtained and includes the child's age, sex, and race; the child's grade in school and type of classroom setting; and the address and type of neighborhood where the family lives. Family composition includes who lives at home, what their relationships are to each other and to the identified patient, and who are the primary and secondary caretakers.

Age of child, sex, race
Grade in school, name of school, classroom setting
Family address, type of neighborhood, parental occupations
Family composition
One- or two-parent family; approximate ages of parents
Siblings of patient and their ages
Other family members, if any, living in the home
Other significant relatives and caretakers

Table 129.3. Childhood/Adolescent Psychiatric Emergencies: Orienting Data

Relevant history, as shown in Table 129.4, builds on identifying information to provide a more complete description of the problem at hand. Historical information should include a thorough understanding of the current crisis and its apparent precipitants, as well as similar problems in the past and previous psychiatric involvement for either child or family. The recent school performance of the child and the adequacy of his or her relationships with peers and with family members should also be determined.

History of presenting crisis and apparent precipitants
Past episodes or other major psychosocial problems
Psychiatric treatment, past or current, for child or family member
School performance of child
Child's relationships with siblings and peers

Table 129.4. Childhood/Adolescent Psychiatric Emergencies: Relevant History

The history of the current crisis should be obtained directly by asking family members in turn to give their account. Usually, beginning with the parents and other adults in the room is easiest. The physician must also obtain the child's version of the current difficulties. If the family does not provide a coherent history, the physician should guide the interview by interrupting respectfully and asking relevant questions. The physician can ensure a more complete understanding of the problem by periodically summarizing what family members have said and then checking for accuracy. When accounts and opinions differ among family members, this disparity should be made explicit. When important issues such as suicidal thinking and severe depression are not brought up, the physician should inquire about them directly. This inquiry reassures the family that its distress is understood and enables the emergency physician to have all of the relevant information needed for assessment. As the physician gains control of the interview and obtains the necessary information, his or her own sense of confidence in dealing with the crisis will increase.

Medical History and Physical Examination

The child's physical status must be determined during the emergency assessment. Many diseases, as well as acute intoxications, can mimic psychiatric disorders. In addition, psychiatrically impaired children may also have concomitant medical problems. Therefore, the emergency physician must obtain a thorough medical history of the child, including current medication and possible medicines available to the child, followed by a complete physical examination, including assessment of neurologic functioning. This information is used in conjunction with the psychological aspects of the assessment to provide a complete picture of the child.

Mental Status of the Child

Evaluation of the child's mental status takes place throughout the entire ED visit. The mental status examination provides a psychological profile of the child at the same time that it assists in determination of a psychiatric diagnosis. Generally, the physician does not need to perform a formal mental status examination of the child because most of the relevant data emerge from history, the physical examination, and the interactions that the child has with family members and with the physician during the emergency assessment. However, the emergency physician should have a systematic and thorough understanding of the mental status examination, and should follow up areas of concern with more specific questions.

Table 129.5 lists the major categories of the mental status examination. These categories are described as they apply to emergency psychiatric assessment.

Orientation	Speech
Appearance	Affect
Memory	Thought content and process
Cognition	Insight and judgment
Behavior	Strengths
Relating ability	Synthesis of evaluation

Table 129.5. Childhood/Adolescent Psychiatric Emergencies: Child Mental Status Examination

Orientation

The level of consciousness and orientation of the child is the first area of assessment. The child not under the influence of drugs or with severe organic illness should be oriented in all spheres: person, place, time, and situation.

Appearance

The physical appearance of the child reveals important information about both the way the child feels about and cares for himself or herself and the supervising care by the family. The examiner should carefully observe factors such as physical size, personal hygiene, choice of clothes, neatness, grooming, posture, and gait.

Memory

The child's memory can be evaluated while listening to the history and through direct questioning. Impairment of memory in a child is a strong indication that his or her emotional and behavioral disturbance may have an organic cause.

Cognition

Intelligence, fund of knowledge, and the ability to think and reason are evaluated while talking with the child. Intelligence and fund of knowledge need be categorized only as adequate or inadequate for the child's age.

Behavior

The child's behavior can be observed throughout the visit. Activity level may be at the appropriate age level and goal-directed, too rapid and random (hyperactive), or too slow and diffuse (psychomotor retarded). The child may appear well-focused, or may be distractible. Behavioral tendencies are revealed in the child's talking with the examiner and in interactions with various family members. Psychotic youngsters may respond to people as objects and use objects in non-directed, bizarre ways. Nonpsychotic children may behave in angry, aggressive ways that can usually be distinguished from the behavior of psychotic children by its negative or resistant nature. The child's ability to control his or her behavior in response to the examiner's or family's request should be carefully noted.

Relating Ability

The child's capacity to relate to the examiner is a key element in the mental status evaluation. In a sense, the examiner is a window to the outside world, and the degree to which a positive relationship can develop during the assessment suggests the child's current capacity for forming relationships in general. The examiner should be concerned with what occurs at any moment during the evaluation and, even more important, how the interaction evolves during the course of the visit. The following questions should be considered: 1) To what degree does the child offer eye contact and speak spontaneously? 2) How trusting does the child appear to be, and to what degree does the child appear to desire the examiner's approval? and 3) On the other hand, is the child too friendly and open, suggesting extreme neediness? The child's cooperativeness and tendency to alter his or her mood in response to the examiner's encouragement are important components of his or her capacity to relate.

Speech

Speech includes elements such as spontaneity, coherence, articulation, and vocabulary. As such, the category of speech overlaps with the capacity to relate, the quality of thought processes, and the level of intelligence. Poor vocabulary and articulation may suggest mental retardation, psychosocial deprivation, specific language disabilities, or combinations of these.

Affect

The child's affect, as the external manifestation of predominant feeling states, is assessed informally during the course of the interview. Fluctuations of affect according to changes in content and interactions should be carefully observed, with

more serious concern raised by children whose affect does not change as different subjects are discussed. Depressed children may show both sad and angry affect, which suggests the way in which the child sees both self and the external world. Some angry children express their anger directly, even in the form of rage. Other children become so well-defended that their affect appears flat and constricted. Frankly psychotic children, in addition to blunted affect, show an inappropriate response to internal and external events, such as smiling while serious topics are discussed.

Thoughts

Thoughts include both thought processes and thought content. The evaluation of the preceding categories necessarily yields much information on thinking. Thought process involves the coherence and goal directedness of verbal communication. Evasiveness and guardedness must be distinguished from the looseness of associations of the psychotic child or adolescent. Loose associations have no logical coherence or connection with previous statements. Flight of ideas, as found in bipolar disorder, involves rapid shifting from one topic to another, often triggered by the patient's ongoing monologue. Thought content involves the major themes that emerge as the child talks spontaneously and responsively to the examiner. If themes of violence and insecurity are present, are other more hopeful and positive themes also present? Such information can often be obtained by eliciting fantasy material, such as three wishes, personal goals, and views of the future. Self-concept, when low, may become apparent as persistent themes and fantasies are pursued. Thorough screening also involves determining the possible presence of psychotic phenomena (hallucinations, delusions, grandiosity, and ideas of reference) and present or past tendencies toward suicide or homicide.

Insight and Judgment

Insight involves the degree of recognition and acknowledgment of current problems by the child. A child with a high degree of insight can also identify possible precipitating factors. Judgment involves the child's ability to think before acting. Over the course of the interview in the ED, the examiner can assess these elements informally.

Strengths

The purpose of any child's mental status examination is not only to screen for possible deficits, but also to search for strengths and areas of competence in the child. Thus, the examiner must determine areas of interest, competence, and motivation of the child. These strengths may go undiscovered unless specifically looked for. Thus, the role of the evaluation extends beyond assessment; it also involves, through the identification of strengths, the beginnings of positive interventions.

Synthesis

After the component parts of the mental status examination have been determined, the physician should integrate them into a comprehensive picture of the child. For example, a 14-year-old boy presents to the ED fully alert and oriented, but disheveled and malnourished. His cognitive abilities appear to be intact, but his actions are slow and labored. The child's thinking shows no evidence of incoherence, but themes of disappointment emerge from the conversation. The boy relates to the physician in a withdrawn manner, and appears to be preoccupied. The data from this mental status examination suggest that the adolescent described is depressed. This impression should then be integrated with historical, medical, and family information as the examiner plans appropriate treatment.

Family Evaluation

Physicians, while performing emergency assessments, explaining results of evaluations, and developing treatment plans, typically deal with both the ill child and his or her family. Recommendations depend in part on the physician's impression of the family and of how effectively the family will carry out the required treatment. To assess families, the physician needs to have an organized framework to guide the evaluation process (Table 129.6). The goal of a family evaluation for childhood psychiatric emergencies is to determine the methods that the family uses to help its members when distressed, the adequacy of these efforts, and the possibilities for new alternatives that will help the family cope successfully with the current crisis. In obtaining the history from the family and proceeding with the assessment, the emergency physician should keep in mind these specific aspects of family functioning so that he or she can evaluate the family systematically during the ED visit. When conducting the interview with child and family, the emergency physician is encouraged to remember that, despite the disruption caused by the crisis, families know their child the best and can often collaborate effectively with the medical staff. When the physician approaches parents as partners, the likelihood of positive outcomes is maximized.

Signs of Competence and Strength	*Danger Signs with Parents/Caretakers*
Level of concern	Psychosis
Verbal communication	Intoxication/drug abuse
Problem-solving ability	Depression
Relationships	Violence
Parents and child	History of abuse (physical, emotional, sexual) and/or neglect
Parents or caretakers	
Parents and physician	

Table 129.6. Child Adolescent Psychiatric Emergencies: Family Assessment

Family Mental Status

Just as it is important to know the child's mental status, the emergency physician must also determine the mental status of the rest of the family. This task can be accomplished as the physician observes the family members and listens to their presentation of the history. The history should be coherent and logical, and should follow a temporal sequence. Families that do not present an organized history may have serious difficulties resolving crises. Family members under the influence of drugs or alcohol may not be fully alert and oriented. Their history may not be clear. Depressed parents appear withdrawn and downcast. They may be so preoccupied with their depression that they do not focus effectively on the child's problem, or they may describe the problem in extremely hopeless terms.

Although anxiety, distress, and even anger may be appropriate responses to a psychiatric emergency, parents should be able to use the physician's support to control these responses so that the crisis can be approached systematically. When this cooperation does not occur, the emergency physician should consider psychiatric consultation. Other indications for psychiatric consultation include the presence of psychosis or other severe psychiatric disturbance in a parent or caretaker. When the family presents a disorganized history, the physician can indicate that he or she is confused and ask for clarification. The physician can also suggest that only one person talk at a time, and can repeat the history given and ask the family to confirm it. When these attempts to provide structure to the family fail, psychiatric consultation is needed.

Hierarchy and Leadership in the Family

The family is a social system that requires acknowledged leadership that is consistent and whose direction is followed. In American nuclear families, the parents are generally the acknowledged leaders, with the responsibility to set rules that are respected and followed by the children. In many families, grandparents live nearby and help with the children, but they are expected to defer to the parents' plans and approaches. In other families, however, especially single-parent families, a grandparent may function as caretaker while the parent is away at work and the parenting responsibility is shared. For effective collaboration to occur, the specific roles for each caretaker must be explicit and agreed on.

In two-parent families, specific parental roles and expectations must also be made explicit. In this way, undermining of one parent by the other is avoided and family rules are enforced consistently. Children are allowed to voice complaints, with the understanding that the parents are the final arbiters. In providing leadership, the parents use both closeness and distance in relation to the child at different times. The relationship between any parent and child involves closeness—the ability to be loving, nurturing, and supportive—and also involves distance—the ability to set and enforce limits and the willingness to allow the child some independence.

The emergency physician should be concerned when either excessive closeness or excessive distance characterizes the relationship between parent and child. An overly close relationship between parent and child may interfere with the parent's ability to establish and enforce rules. The parent may hold back either because he or she is unable to get angry at the child or because he or she fears upsetting the child by taking a firm stand. Overly close relationships are common in single-parent families, but may occur in two-parent families as well. Such relationships may be revealed in the ED when child and overinvolved parent are sitting close to each other (and apart from other family members, if present), when the parent answers for the child or describes how the child feels rather than encouraging the child to speak for himself or herself, and when the parent uses the pronoun “we” while describing difficulties pertaining to the child individually.

The emergency physician can assess an overly close parent–child relationship in several ways. He or she can suggest that child and parent not sit so close to each other or can have the child switch seats with another family member. The physician can ask parental permission to speak with the child in the presence of the family, with only the child speaking for himself or herself. The physician can point out the nature of the overly close relationship to the other parent and ask whether this situation is of concern to the less involved parent. When these interventions fail, psychiatric consultation is in order.

Excessively distant relationships occur in some disorganized families when parents are so involved with their own problems that the needs of the child are overlooked. In such families, the child is given more autonomy or responsibility than is appropriate for his or her age, and rules are either nonexistent or enforced inconsistently. Such families may be unable to focus on the child's problem. The parents may be primarily concerned about the effect of the child's problem on their lives, and less concerned about the child's distress. The child may be scapegoated by the parents as the source of all the problems. Statements by such parents include, “Why are you doing this to me?” “I have better things to do than to be in the emergency room with you,” and “If it weren't for you, things would be going smoothly.” Disengaged or underinvolved parents may also appear apathetic and unresponsive to the child's disturbed behavior.

In responding to an underinvolved family, the emergency physician should create an intensely serious mood and emphasize the gravity and danger of the situation. The physician can appeal to positive parental instincts, conveying a belief that the parents want to do what is best for their child. In some cases, this approach creates a tone that enables family members to actively respond to the child's distress. In other situations, however, the caretakers remain unresponsive. When this happens, psychiatric consultation is required and hospitalization may be necessary.

Protectiveness versus Autonomy

All relationships can be placed on a continuum of involvement, with intimacy at one pole and disengagement at the other. Effective parenting requires avoidance of the extremes and the capacity to shift from one position to another at different times and in different situations. As previously described, some families have an overly close relationship between the child and one parent. This situation may occur in any family in which significant conflict occurs between the parents or between parent and other caretakers (e.g., grandmother). In addition, overinvolvement with a child may occur in families

with a special child, such as one with a physical handicap or chronic physical illness. Parents in such families may be overprotective of the child, assuming functions and speaking for the child. Overprotectiveness toward the child may involve primarily one parent or may involve the entire family. In such families, conflict is often avoided so the child does not get upset. The child also may avoid disagreement out of concern for his or her parents. A consequence of parental overprotectiveness is diminished autonomy on the part of the child, who tends to be more involved with adults than with peers. In such situations, the child's development of independence is often significantly delayed.

Underinvolved families, on the other hand, provide insufficient protection and support for the child. The child learns not to expect parental support and may present as under-socialized, with a conduct disorder, or as depressed and possibly suicidal. The child may also present as an overwhelmed “little adult.”

With overprotective families, the emergency physician can assess the family's flexibility by suggesting that the child speak for himself or herself and that the parents increase their expectations. When the overprotectiveness is rigid and severe, the physician may request psychiatric consultation as a way of initiating ongoing therapy for the family. With disengaged families that fail to respond to the physician's statement of urgency, psychiatric intervention should be pursued.

Conflict Resolution versus Conflict Avoidance

All families have disagreements among their members. In some families, disagreements are acknowledged and confronted directly, whereas in others, potential conflict is consistently avoided. Other families disagree openly, but are unable to reach a constructive resolution. The capacity of the family for conflict resolution is an important area for the emergency physician to assess because unresolved disagreements typically lead to chronic hostility, undermining of relationships, and ineffective parenting. Families that tolerate and resolve disagreements are usually more open in their discussion of family problems, whereas conflict-avoiding families use a great deal of denial. In some families, the presence of conflict becomes apparent as the history is given. Family members provide differing accounts of the history and may interrupt and contradict each other. The tension in the room increases, and the physician may start to feel anxious. In such circumstances, the emergency physician can indicate the need to hear from each family member one at a time and suggest that everyone take a turn. The physician can also summarize the family's account as he or she goes along, confirming information at the same time that tension in the room is controlled.

Families that are unable to resolve conflict often have significant marital problems. The parents, unable to deal effectively with each other, instead become overinvolved with one of the children. The child may have a chronic illness, may be either the oldest or the youngest, or may be chosen for another reason. The child gets caught in the marital struggle of the parents, in part through their efforts and in part through his or her own desire to remain close to the parents and keep them together. This child often develops physical and psychiatric symptoms and may present to the ED with the family.

The emergency physician may observe several possible patterns of conflict avoidance. The parents may agree that the only problem in the family is the child, and that, were it not for him or her, everything would be fine. However, the physician notes that the parents do not look at each other or talk to each other. Their one common ground of agreement is the scapegoated child and his or her symptoms. In a related pattern, the parents suppress all conflict by focusing excessive concern on the symptomatic child, who is seen as vulnerable and weak. This reaction often occurs in families with a child with psychosomatic symptoms, where the child is overprotected and his or her symptoms are typically exacerbated by family conflict. The third pattern involves parental focusing on the child and his or her symptoms as the battleground for overt parental and spouse disagreements. The parents deny the existence of any disagreements except those related to the identified patient, about whom they disagree openly and angrily.

The emergency physician should request psychiatric consultation when inability to resolve conflict has seriously impaired the functioning of the family and the child. Psychiatric involvement should also be sought when patterns of conflict avoidance occur in conjunction with other aspects of family adaptational problems described in this section.

Capacity for Problem Solving

Families in crisis usually try to solve their problems by attempting the same solutions over and over again. They perceive other remedies as currently inaccessible or impossible. As a result, the family's repertoire of behaviors becomes extremely limited and responses to problems become stereotyped and rigid. As the physician pursues historical information and observes the family, he or she looks for signs of flexibility and strength. The physician asks about the family's previous attempts to solve the presenting problem and seeks the family's ideas on how to approach the difficulty now. When offering observations, the physician notes whether the ideas are accepted or rejected. When asking family members to behave differently now, the physician observes how hard they try to change and evaluates the probability that these changes will enable the family to manage the crisis.

Disagreeing parents must find a way to put aside their differences in the interest of the child and the family. These parents must develop a mutually acceptable plan for responding to their child's problematic behavior. Thus, the parents must soften rigid and polarized positions, when these are present. For example, a father who harshly states that the child needs strong punishment and a mother who is adamant about not upsetting the child must both change their positions. The emergency physician tries to help the parents find an acceptable middle ground so that they can resume control of their family. When these attempts fail, psychiatric consultation should be sought.

Relating to the Physician

The emergency physician should rely substantially on his or her overall impression of the family and how members relate to him or her. Do family members maintain eye contact with the physician, or do they avoid looking at her or him and display guardedness or a sense of hostility? Families that accept the physician's expression of interest and concern are more likely to benefit from the ED visit. Families that are suspicious of the physician's motives and unresponsive to his or

her input probably gain less from emergency interventions. Emergency physicians need to recognize that those families that antagonize them the most may also be the families most in need of the physician's (and the psychiatrist's) interventions. When feeling anger toward the family, the physician must decide if these feelings should be expressed. For example, the physician may feel offended by a family that repeatedly criticizes a child whom he or she likes. He or she may want to rebuke the family for being unfair and want to defend the victimized child. However, such feelings, if acted on, would only compound the problem because the family might disqualify the physician and single out the scapegoated child even more when the physician is no longer present. On the other hand, in other circumstances, expressing feelings may be extremely helpful. For example, the physician can convey to parents a concern that his or her ideas are being disregarded and suggest that they be considered more fully. Such a statement may lead the family to reconsider the validity of the physician's statements.

Using Social Support

Some families come to the ED feeling isolated, overwhelmed, and exhausted. Often, such families have not used all of the family and community resources available to them. Effective crisis intervention for psychiatric emergencies involves not only emergency treatment but also effective disposition planning for the family. The ED staff should determine what community resources are available, or potentially available, to the family. The parents should be asked about relatives or neighbors who might be able to help them. Families that have previously mobilized community supports should be commended by the emergency physician for their competence and resourcefulness. This support by the physician often stimulates the family to recognize its capacity to address the crisis.

Integration

In following the aforementioned guidelines for family evaluation, the emergency physician obtains the necessary data for understanding the family in crisis. By integrating the history, the physical examination, the child's mental status, and the family assessment, the emergency physician is better able to understand the nature of the current crisis and the appropriate responses. However, effective intervention involves more than understanding. The emergency physician needs to offer information and recommendations in a respectful manner that conveys his or her recognition of the family's competence and its desire to help the child.

DEPRESSION

Background

The appreciation of depression in children and adolescents as a highly significant problem has increased greatly in recent years. *Depression* can refer to the symptom of feeling sad but most appropriately describes a symptom complex or syndrome that includes cognitive and physiologic components in addition to affective ones. Depression implies more than momentary sadness, and involves a pervasive inflexibility of sad mood, accompanied frequently by self-deprecation and suicidal ideation. Depression also implies a change in functioning from an earlier state of relatively good adjustment, rather than a temperamental or personality type.

Because no generally agreed-on definition of depression exists, incidence figures vary according to the definition used, as well as the nature of the population studied. In one study of high school students aged 11 to 15 years in suburban Boston, 33% of these early adolescents were believed to have moderate to severe symptoms of depression. Other estimates put the incidence of depression in children and adolescents in the 20% range. The incidence of depression is higher in children with school problems (including learning disabilities and attention deficit disorder), and in children with significant medical problems. Because most children with depression come to the ED with another chief complaint (e.g., somatic symptoms, school problems, behavior problems), the physician must keep in mind the possibility of depression in all children seen with recurrent or vague somatic complaints.

Considerable evidence suggests that a genetic predisposition exists for depression, particularly severe depression. Depressive episodes may be triggered by environmental events of significance to the child.

The depressed child typically experiences a profound sense of helplessness, feeling unable to improve an unsatisfactory situation. The child may be experiencing frustration and failure at home, at school, and with peers. Negative outcomes reinforce the child's negative self-image, which contributes in circular fashion to more negative outcomes.

Clinical Manifestations

Depression appears differently at different stages of development. In infancy, depression is usually the result of loss of mother and/or lack of nurturance, and is seen as a global interference of normal growth and physiologic functioning. Thus, some of the manifestations of depression in infancy include apathy and listlessness, staring, hypoactivity, poor feeding and weight loss, and increased susceptibility to infection.

During latency, depression can appear as part of a syndrome, or may be masked by other symptoms. Petti describes the two key features in childhood depression as dysphoric mood and self-deprecatory ideation. Dysphoric mood is manifested by looking or feeling sad and forlorn, being moody and irritable, and crying easily. Self-deprecatory thoughts are reflected by low self-esteem, feelings of worthlessness, and suicidal ideation. Depression in this age can also appear as other common symptoms, including multiple somatic complaints, school avoidance or underachievement (including learning-disabled children or children with attention deficit disorder), angry outbursts, runaway behavior, phobias, and fire-setting.

Depression during adolescence is more similar to adult-onset depression. The major symptom is a sad, unhappy mood, and/or a pervasive loss of interest and pleasure. Other symptoms may include a change in appetite, change in a sleep behavior, and psychomotor retardation or agitation. Also present in many depressed teenagers are loss of energy,

feelings of worthlessness or excessive guilt, decreased ability to concentrate, indecisiveness, and recurrent thoughts of death or suicide. Depressed teenagers can also present with somatic complaints, academic problems, promiscuity, drug or alcohol use, aggressive behavior, and stealing. Many teenagers with behaviors such as these are unaware of their depression because it is not on the surface. Others simply deny the painful depressive affect. In talking with these patients about their lives at home, at school, and with peers, the underlying depression usually becomes apparent.

Management

Once any initial medical concerns have been dealt with, the three major goals in the management of depression involve 1) determining suicidal potential, 2) uncovering acute precipitants, and 3) making an appropriate disposition.

The emergency treatment of depression can usefully be thought of as the prevention of suicide attempts. The task of the physician is to carefully determine whether any suicide attempts have been made and whether suicidal ideation is present. The physician should not be hesitant about asking the child about suicidal deeds, thoughts, or wishes. Such questions represent a positive confrontation of the problem of depression and are unlikely to catalyze a subsequent suicide attempt. In fact, questions about suicide may actually provide a sense of relief for the depressed child.

The physician should attempt to determine possible acute precipitants of the current depression to guide subsequent recommendations. The duration of the depression should be determined, as well as the family response. Assessing overall adjustment at home, in school, and with peers is important, as well as looking for the strengths of child and family for use in the treatment plan.

When suicidal ideation is present, the emergency physician should request psychiatric consultation. A decision can then be jointly made regarding outpatient or inpatient treatment. Whether or not suicide is an imminent danger, the task of the physician is to create a sense of hope that things will improve. To achieve this goal, the physician must form a solid doctor–patient relationship with child and family. Outpatient management can be used when adequate social support is present. The parents must first acknowledge the existence of depression in the child and then come to understand that the solution involves a strong commitment on their part, including at times their participation in family therapy. The physician can begin the process by helping the family create a list of areas involving child and family that need to be addressed. Another area commonly needing attention with a depressed child is school. School consultation as therapy begins may provide valuable information about the child and can help the child obtain academic remediation when needed. The emergency physician can inform child and family that school difficulties will be addressed in outpatient therapy.

Psychotropic medication, although not approved by the Federal Drug Administration (FDA) for use in children and adolescents, has been increasingly used in the treatment of childhood and adolescent depression. As an acute intervention, however, the emergency physician should not prescribe antidepressant medication because its desired mood-elevating effects generally require up to 1 month to take effect, and the act of prescribing medication in the ED may decrease the likelihood of successful referral for follow-up mental health treatment.

The emergency physician should be familiar with commonly used antidepressants, which are sometimes used adjunctively in the treatment of depression. In recent years, use of the selective serotonin reuptake inhibitors (the SSRIs) has displaced the tricyclic antidepressants as first-line medications. SSRIs include Prozac (fluoxetine), Paxil (paroxetine), Zoloft (sertraline), and Luvox (fluvoxamine). Advantages of these agents over tricyclic antidepressants include a decreased likelihood of cardiotoxicity, the absence of anticholinergic side effects, and the relative safety of these medications when used in overdose. These medications all require approximately 1 month to take effect. Another commonly prescribed antidepressant is Wellbutrin (bupropion), which is chemically distinct from other agents. A side effect of potential concern with Wellbutrin involves seizures.

Because the older tricyclic antidepressants are still prescribed at times for children and adolescents, and may more commonly be prescribed to a parent by a family physician, emergency physicians should be familiar with these medications, which include Tofranil (imipramine), Elavil (amitriptyline), Pamelor (nortriptyline), Norpramin (desipramine), and Anafranil (clomipramine). If the emergency physician learns that any of these medications have been used in overdose, especially in doses exceeding 10 mg/kg of body weight of the child, the possibility of cardiac arrhythmias should be considered. Other common side effects of tricyclic antidepressants are drowsiness and those resulting from the anticholinergic effects of these drugs, which include dry mouth, constipation, and blurred vision. Cardiac arrhythmias are reportedly less common with Sinequan (doxepin), Ascending (amoxapine), and Ludiomil (maprotiline), none of which are tricyclic compounds. However, drowsiness and variable amounts of anticholinergic effects may occur. Chapter 88 covers the management of specific toxic ingestions.

SUICIDE ATTEMPTS

Background

Suicidal behavior involves thoughts or actions that may lead to self-inflicted death or serious injury. A distinction is made between suicidal ideation and suicidal attempts in which a deliberate attempt to take one's own life occurred.

The increasing trend toward suicidal behavior by children and adolescents is alarming. Table 129.7 provides information on the nature and scope of this problem.

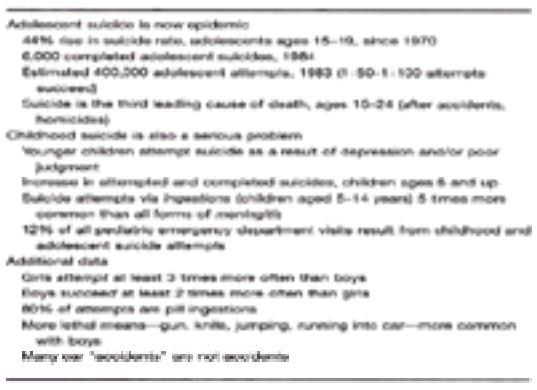

Table 129.7. Childhood and Adolescent Suicide: Nature of the Problem

Suicide can be seen as the final common pathway for a variety of situations in which the child experiences a pervasive sense of helplessness, with a perceived absence of alternative solutions. To the distressed child, suicide appears to be the only solution to his or her problems and also to the family's problems. Most suicide attempts occur in depressed children. Others occur with children experiencing major losses, such as serious illness or death in the family. Still others occur in children with depression in association with problems of impulsivity. A small but significant percentage of suicide attempts occur in psychotic children and adolescents. These attempts may be deliberate (stemming from hopelessness) or accidental (impaired judgment and reality testing). Table 129.8 outlines the potential sources of adolescent suicide attempts.

Table 129.8. Potential Sources of Adolescent Suicide Attempts

A factor that complicates the discussion of suicide in children is their differing conceptions of death at various ages. Up to age 5, death is seen as a reversible process in which the activities of life still occur. From 5 to 9 years, the irreversibility of death is beginning to be understood but death is personified, rather than seen as an independent event. It is not until about age 9 that death is seen as irreversible in the adult sense of being both final and inevitable. Even then, however, the child may imagine his or her own death as being reversible. Under such circumstances, a suicide attempt may have a different meaning from an adult, where suicide corresponds to a definite end of one's life.

Clinical Manifestations

In latency-aged children, certain risk factors have been identified that distinguish children with suicidal behavior from other children with emotional problems (Table 129.9). Suicidal children are likely to be depressed and hopeless. Self-esteem is low, and they see themselves as worthless. The wish to die is present, as are preoccupations with death. The family history may include past episodes of parental depression and suicidal behavior. Suicidal children tend to view death as temporary and pleasant rather than irreversible.

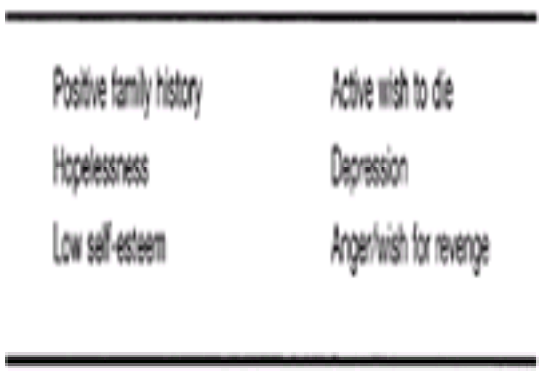

Table 129.9. Characteristics Associated with Childhood and Adolescent Suicide Attempts

Teicher has offered a longitudinal perspective of adolescent suicide attempts, which are usually not simply impulsive

acts. Before the suicide attempt, problems have been present in the family for at least 5 years. These problems include a parent or close relative attempting suicide, many residential and environmental changes, and unexpected separations from meaningful relationships (divorce, separation, or death). With the onset of adolescence, an escalation phase occurs in which frustration results from the teenager's desire for autonomy and the belief that his or her parents do not understand. The teenager withdraws or rebels, becoming alienated from his or her parents at a time when they are really still needed. The scene is then set for the final stage, in which some precipitating event leads to the suicide attempt. The precipitating event may be a peer rejection, the breakup of a romance, an unwanted pregnancy, or problems in school. (It was found that 36% of children attempting suicide were not enrolled in school at the time of the suicide attempt.)

Table 129.10 indicates the high-risk situations for suicidal behavior in which direct questioning about suicide should occur. The first two situations immediately alert the physician to the danger of suicidal behavior. The other situations involve a different chief complaint, masking possible suicidal ideation or behavior. All accidental ingestions should be screened for the possibility of a suicide attempt. Overtly depressed children are at risk for suicide, as are depressed children who present with somatic complaints. Children who have acted violently are also at risk because violence can be turned inward. Psychotic children present a special problem and may present with inadvertent suicide attempts as the result of impaired judgment, hallucinations, and delusions of persecution. The isolated, withdrawn child may harbor suicidal thoughts that are uncovered only by direct questioning.

Suicide attempt just made	Aggressive violent behavior
Suicidal threat made	Psychotic child
"Accidental" ingestion	Significant withdrawal by child
Child complains of depression	
Medical concerns, but child appears depressed	

Table 129.10. Childhood and Adolescent Suicide: High-Risk Situations for Suicide Attempts

Management

Assessment

The emergency physician should specifically ask about suicidal thinking in all high-risk children. When an actual attempt has taken place, the physician should attend to any medical needs such as suturing, lavage, and managing physical repercussions of overdoses. Then he or she should conduct an assessment as well as requesting psychiatric consultation.

The dichotomy sometimes drawn between suicide "attempts" and suicide "gestures" is ill conceived. All suicidal behavior should be regarded as suicide attempts, which are best evaluated by appreciating the medical lethality of the act, the suicidal intent of the child, the impulsivity of the act, and the strengths and supports within the family (Table 129.11). The lethality of a suicide attempt by itself may be misleading because suicidal children may miscalculate, causing at times greater harm than was intended and at other times less harm than was intended (Table 129.12). As an example, the child who takes 10 tablets of his mother's tricyclic antidepressant medication, Tofranil 100 mg, could make a fatal miscalculation, as 1000 mg of Tofranil can cause fatal arrhythmias in children. On the other hand, a child who takes 10 aspirins may be far more suicidal than the lethality of his or her ingestion would suggest. In general, more violent methods of attempted suicide (e.g., hanging, shooting, jumping) usually reflect greater suicidal intent. However, the physician cannot conclude that attempts with low lethality are not serious attempts until he or she has specifically asked about and assessed the child's suicidal intent, that is, just how seriously the child wanted to end his or her life (Table 129.13).

Medical lethality	Impulsivity
Suicidal intent	Strengths/supports

Table 129.11. Assessing Childhood/Adolescent Suicide Attempts: Four Major Dimensions

Vital signs
Level of consciousness
Evidence of drug/alcohol intoxication (e.g., pupils, smell on breath)
Need for emesis, lavage, or catharsis
Acute medical complications (cardiac, respiratory, renal, neurologic)
Indications for medical hospitalization, including intensive care
Residual abnormalities

Table 129.12. Childhood and Adolescent Suicide: Assessing Medical Lethality

Circumstances of Suicide Attempt
Nature of suicide attempt (pills versus violent means)
Use of multiple methods
Method used to extreme (all versus some pills ingested)
Suicide note written
Secrecy of attempt (attempt concealed versus revealed)
Premeditation (long-planned versus impulsive attempt)
History of prior attempts
Child Self-Report
Premeditation of attempt
Anticipation of death
Desire for death
Attempt to conceal attempt
Nature of precipitating stressors
Child's Mental Status
Orientation/cognitive intactness
Presence/absence of psychosis
Manner of relating to physician
Current suicidality
Response to being saved/being unsuccessful in attempt
Active plan for another attempt
Readiness to discuss stressors
Readiness to accept external and family support
Nature of orientation toward future

Table 129.13. Childhood and Adolescent Suicide: Assessing Suicide Intent

In addition to asking directly about suicidal intent (“When you took those pills, what were you hoping would happen?”), the physician should gather as much information as possible about the attempt itself to help infer the degree of suicidal intent on the part of the child. Did the child take all of the pills that were available; did he or she expect to wake up; did he or she tell anyone after taking the pills; did he or she leave a suicide note? Now that he or she is awake, is the child pleased or displeased to be alive? Does he or she intend to try again?

Children who threaten suicide without making an actual attempt should also be questioned carefully about suicidal intent. How long has the child considered suicide; what methods are planned; when will this take place? Has the child ever made previous attempts? How about other family members? Psychotic and depressed children, especially when the parents appear unable to supervise the child, should elicit particular concern.

Assessment of the child's level of impulsivity is also important (Table 129.14). Does the attempt appear to have been impulsive rather than planned? Is there a history of prior impulsive behaviors? Is there evidence of impulsivity during the ED interview?

Evidence of impulsive suicide attempt	Evidence of impulsivity during
History of prior impulsive behaviors	interview

Table 129.14. Childhood and Adolescent Suicide: Assessing Impulsivity

The physician should ask the child and family about possible precipitating events to determine what changes in the environment may be needed. The strengths of the family should be assessed to determine whether sufficient social support exists to allow for outpatient management (Table 129.15).

Strengths and Assets of Child	Nature of External Supports
Ability to relate to physician	Outpatient psychiatric/family physician
Ability to rely on parents in crisis	Extended family
Ability to acknowledge problem	Neighbors/other significant adults
Positive orientation toward future	Religious community
Strengths and Assets of Family	Self-help groups
Commitment to child	
Ability to unite during crisis	
Problem-solving abilities	
Capacity to supervise child (support and limits)	
Ability to use external supports	

Table 129.15. Childhood and Adolescent Suicide: Assessing Strengths and Supports

Evaluation for Hospitalization

No universally agreed-on criteria have been established for when to hospitalize a child with suicidal behavior and when to manage him or her on an outpatient basis. Garfinkel and Golombek have identified seven areas to assess to determine whether hospitalization is indicated (Table 129.16).

Social set	Stress
Intent	Mental status
Method	Support
History	

Table 129.16. Areas to Assess Following a Suicide Attempt

Social set involves the degree of privacy that the child arranged at the time of the attempt. Did he or she tell anyone before or after the attempt, or were pains taken to set up a situation in which detection was unlikely? Intent may be reflected in a suicide note left by the child, by the degree of detail of the suicide plans, and by direct questioning of the child regarding his or her suicidal intent at the time of the attempt and at the time of examination. The choice of method also helps in the assessment of the suicide attempter. Was a method with high lethality used or desired? Did the child understand the likely outcome of the method used? With an ingestion, were all available pills consumed? The history reveals both the presence of past suicide attempts by the child and past attempts by other family members. The evaluation of the stressful precipitating events is important in planning disposition, as is the mental status of the child in the present and in comparison with the past. Finally, the degree of support expected from within the family and outside the immediate family (extended family, neighbors, peers, and teachers) must be assessed. To what degree can the family unconditionally commit itself to support the child's safety and well-being? Are the resources present for the family and larger network to implement this commitment? The decision to hospitalize the child is made when the child's safety is still in doubt after these questions have been answered.

In general, any suicide attempt deserves a thorough assessment by the emergency physician and a complete psychiatric consultation. Hospitalization should be used in the circumstances listed in Table 129.17: 1) the physician has had difficulty in gaining the cooperation of the child and the family, 2) the child has made a serious suicide attempt, 3) the child is continuing to be actively suicidal, 4) the child is unwilling/unable to provide a no-suicide commitment to the parents, 5) the child is psychotic, 6) the family appears unable to provide necessary supervision and support to the child, and 7) the child and family rapidly deny the significance of a serious suicide attempt.

1. Failure of rapport among physician, child, and family
2. Serious suicide attempt (lethality and intent)
3. Continuing active suicidality
4. Inability to provide no suicide commitment to parents
5. Psychosis of child
6. Divisive/disturbed family, incapable of support and supervision
7. Rapid denial of significance of suicide attempt

Table 129.17. Indications for Psychiatric Hospitalization Following Childhood/ Adolescent Suicide Attempt

Initiating Treatment

The critical goal in dealing with suicidal behavior in a child is to create a context for living—an immediate response to the crisis that increases the likelihood that the child remains alive. The emergency physician creates a context for living through his or her thorough assessment of child and family, the eventual disposition, and the encouragement of family and child to increase communication and develop alternative solutions to problems that have arisen.

The parents should be encouraged to tell the child that they want him or her to live and that suicidal behavior is forbidden. Parents can be tender in expressing their love for the child, but they need to be firm in establishing the rule that self-destructive behavior is an unacceptable response to problems. The child should also be told that he or she has a responsibility to himself or herself and the family to keep himself or herself alive. The emergency physician may need to remind tentative parents that, whether or not hospitalization is used, they still have primary responsibility for their child.

A critical moment occurs when the parents, guided by the emergency physician, ask the child to make a no-suicide commitment, also known as a safety contract. This commitment signifies the child's promise to the parents that he or she will not try to harm himself or herself again, no matter how upset he or she is. Instead, the child will seek out the parents or another responsible adult for assistance.

The emergency physician should recognize the common tendency toward denial by the child in the ED after the actual suicide attempt. As a result of this denial, or in an effort simply to appease the parents, the child may make an insincere no-suicide commitment. Therefore, when this commitment is being made, parents and child should discuss it so that parents can determine the real intentions of the child and convey the urgency of the no-suicide commitment.

Although a no-suicide commitment is not always sufficient to avoid psychiatric hospitalization, sending any child home when an earnest no-suicide commitment has not been given and accepted is potentially hazardous.

If inpatient treatment is required, the child and family should be informed about how the hospital operates and what to expect. The goals of the hospitalization should be discussed, and the active role of the family in the treatment emphasized. In many states, voluntary consent forms need to be signed. In instances in which the child or parents do not agree to hospitalization, involuntary commitment may need to be used, although every effort should be made to enlist the concurrence of the parents first. When possible, the child and family should be accompanied to the psychiatric hospital by the consultant psychiatrist or an involved social worker so that the transition to the psychiatric facility is made smoother.

Outpatient management of suicidal behavior becomes feasible when 1) the child and family are cooperative and engageable; 2) the attempt is determined not to have been too serious, in terms of intent and medical lethality; 3) the child is not actively suicidal or psychotic at the time of the evaluation; 4) the child provides an earnest no-suicide commitment; and 5) the family can take responsibility for the child until formal psychiatric treatment is begun the next work day, and appears capable of managing the child within the home setting as mental health treatment is provided. Before sending a family home, the psychiatrist or emergency physician should have the family formulate a concrete plan concerning how it will manage the child. The expectations and responsibilities of each family member, including the suicidal child, should be spelled out.

Outpatient psychotherapy can begin immediately with emergencies that occur during the work day. When outpatient treatment cannot begin until the next day, the physician should give the family a therapist's name or the name of the “intake person” at the mental health agency. This information personalizes the agency and increases the chances that the family will follow through. The family should be instructed to use the physician's name as the source of referral and should be reassured that the physician will contact the agency before the family's call. At least one parent and the child, if an adolescent, should be asked to sign a release of confidentiality to authorize communication between the physician and the mental health agency. This release also enables the agency or the psychiatrist to contact the family if the family fails to follow through in making an appointment to be seen. Any discussion of suicide must contain careful consideration of prevention. Parents should be given guidelines for the prevention of suicide (Table 129.18) as well as instruction in the early warning signs (Table 129.19).

Understand nature of parent-child dilemma during adolescence
Maintain physical contact—be around, combat tendency toward isolation
Maintain emotional contact—stay involved, show positive regard
Listen to child before responding—promote safety in talking
Respond to child, once child has finished—take child seriously, do not dismiss or attack
Encourage choices by adolescent
Acknowledge child and provide respect

Table 129.18. Prevention of Childhood and Adolescent Suicide: Guidelines for Parents

Withdrawal (peers, parents, siblings)
Somatic complaints
Irritability
Crying
Diminished school performance
Sad or anxious appearance
Significant loss (rejection by peer group, break-up of romance, poor grades, failure to achieve important goal)
Major event or change within family
Casual mention of suicide or being "better off dead"
Explicit suicide threat
Minor, seemingly unimportant suicide "gestures"
Apparent "accidents"
Other unusual behavior pattern—housebound behavior, breaking curfew, running away, drug or alcohol abuse, bizarre or antisocial actions

Table 129.19. Prevention of Childhood and Adolescent Suicide: Warning Signs for Parents

PSYCHOSIS

Background

Psychosis is the term used to describe severe disturbances in a patient's mental functioning. It is manifested by significant aberrations in cognition, perception, mood, impulses, and reality testing. Thoughts and feelings are not well-integrated, and acted upon, perceptions may become distorted so that the world is seen as threatening, and mood may become ecstatic or despondent. Behavior may also become extremely agitated and potentially violent, or excessively withdrawn to the point where the patient does not recognize and attend to his or her physical needs. Psychotic patients are actively attempting to regain control over their mental capacities and are trying to understand and deal with highly unusual thoughts, perceptions, and impulses. Their subjective experience often is one of helplessness and extreme anxiety.

Psychosis in children and adolescents can be divided into two groups based on cause: organic psychosis and psychiatrically based psychosis. Psychiatrically based psychosis in children and adolescents has four major causes: 1) autism with onset before 30 months of age, 2) other pervasive developmental disorders with onset between 30 months and 12 years of age, 3) adult-type schizophrenia with onset in adolescence, 4) acute reactive psychosis, and 5) bipolar or manic–depressive illness with onset in late childhood or adolescence. Emergency management of organic psychosis and the four major types of psychiatrically based psychosis is described in the following sections.

Organic Psychosis

Differentiation of organic psychosis as a separate class does not imply that other (psychiatrically based) psychosis is completely independent of brain processes. On the contrary, all psychosis is assumed to be associated with aberrant brain function. The term *organic psychosis* merely implies that the cause of the aberrations in mental functioning is known and resolution of the psychosis depends on improvement in the underlying organic problems. Psychiatrically based psychoses, on the other hand, are those in which specific organic causes have not yet been determined (Table 129.20). The causes of organic psychoses can be acute or chronic illnesses, trauma, or intoxications with an exogenous substance (Table 129.21, Table 129.22 and Table 129.23).

Assessment Feature	Organic Psychosis	Psychiatrically Based Psychosis
History		
Nature of onset	Acute	Insidious
Past illness/drug use	Prior illness/drug use	Prior psychiatric history (self or family)
Medical evaluation		
Vital signs	May be impaired	Usually normal
Level of consciousness	May be impaired	Normal
Pathologic autonomic signs	May be present	Normal
Laboratory studies	May be abnormal	Normal
Mental status evaluation		
Orientation	May be impaired	Intact
Recent memory	May be impaired	Intact
Cognitive/intellectual functioning	May be impaired	Intact
Nature of hallucinations	Usually not auditory (e.g., visual, tactile)	Auditory
Response to support and reinstitution	Often dramatic	Often limited

Table 129.20. Organic versus Psychiatrically Based Psychosis: Major Differentiating Features

Medical conditions (acute and chronic)
Trauma (acute and chronic)
Prescribed medications (toxicity/side effects/withdrawal)
Drug intoxications
Accidental, including misuse of proprietary medication
Drug abuse/experimentation
Alcohol abuse (alone or with drugs)
Deliberate suicide attempt

Table 129.21. Causes of Organic Psychosis

Central Nervous System Lesions	Adrenal disease (hyper and hypo)
Tumors	Uremia
Brain abscess	Hepatic failure
Cerebral hemorrhage	Diabetes mellitus
Meningitis or encephalitis	Porphyria
Temporal lobe epilepsy	*Rheumatic Diseases*
Cerebral Hypoxia	Systemic lupus erythematosus
Pulmonary insufficiency	Polyarteritis nodosa
Severe anemia	*Infections*
Cardiac failure	Malaria
Carbon monoxide poisoning	Typhoid fever
Metabolic and Endocrine Disorders	Subacute bacterial endocarditis
Electrolyte imbalance	*Miscellaneous Conditions*
Hypoglycemia	Wilson's disease
Hypocalcemia	Reye syndrome
Thyroid disease (hyper and hypo)	

Table 129.22. Medical Conditions That May Lead to Psychosis

Alcohol	Quaalude
Barbiturates	Anticholinergic compounds
Antipsychotics (e.g., phenothiazines)	Heavy metals
Amphetamines	Cocaine and crack
Hallucinogens—LSD, peyote, mescaline	Corticosteroids
Marijuana	Reserpine
Phencyclidine (PCP)	Opiates (e.g., heroin, methadone)

Table 129.23. Exogenous Substances That Cause Psychosis Following Ingestion of Significant Quantity

Clinical Manifestations

The child or adolescent with an organic psychosis presents to the ED in an agitated and confused state. The child's orientation to time and place is often disturbed, and he or she may be highly distractible, with significant disturbance of recent memory. Evidence of bizarre and distorted thoughts is apparent, and disconnected ideas may be juxtaposed. The child may also have significant difficulty controlling behavior and may persist in activities without regard for personal safety. The child may get up to leave the room without saying where he or she is going or why he or she needs to leave. Intellectual functioning may also be impaired, and the child may be unable to concentrate on simple reading or arithmetic tasks.

The child with an organic psychosis may experience visual hallucinations, which may be frightening in nature. Tactile hallucinations may be present. Auditory hallucinations, more common in schizophrenia and manic–depressive illness, are rare in organic psychoses but may occur. As a result of impaired reality testing, organically psychotic children and adolescents are often extremely difficult to control and may strike out at family or staff when attempts are made to control their behavior.

An accurate and thorough history is essential in the evaluation of any child or adolescent for psychosis and is also helpful in appreciating its underlying cause. A complete medical history helps determine whether the organic psychosis is a concomitant feature of an already existing chronic illness (e.g., lupus cerebritis), a result of medication prescribed to treat an ongoing disease (e.g., steroids for lupus erythematosus), or a result of a drug ingestion (e.g., amphetamine psychosis). Typically, an acute intoxication or drug ingestion causes the acute onset of psychosis and represents an abrupt change from the child's previous psychological functioning. The possibility of alcohol use must also be considered in the cause of organic psychosis, and the history should explore the possibility of trauma. In general, no specific features of the mental status examination differentiate the various causes of organic psychosis.

The physical examination is often extremely helpful in both differentiating organic from psychiatrically based psychosis and in determining the underlying cause of an acute organic psychosis. Fever is likely to be present in infections, and tachycardia is often associated with chronic illness or intoxication. The general physical examination gives indications of pulmonary, cardiac, liver, or autoimmune disease, and the neurologic examination assists in the diagnosis of central nervous system disease. Abnormalities of reflexes or of motor, sensory, or coordination systems always require complete neurologic evaluation. Signs of increased intracranial pressure may be indicative of a cerebral vascular accident, central nervous system tumor, or cerebral edema. Signs of autonomic dysfunction, such as pupillary abnormalities, are often indicative of acute intoxication.

In instances of suspected organic psychosis, laboratory evaluation should include a complete blood count, urinalysis, serum electrolytes, calcium, blood urea nitrogen, blood glucose, and complete drug and alcohol screens. Serum, urine, and gastric aspirate should be obtained for toxicology screening. Other laboratory and radiologic studies depend on

abnormalities noted in the history and physical examination. If central nervous disease is suspected, skull radiographs, computed tomography, and a lumbar puncture may be necessary. Liver function studies, thyroid studies, and other specialized and specific laboratory tests may be obtained as required.

Management

Management of the child or adolescent with organic psychosis involves several steps (Table 129.24). First and foremost is diagnosing the underlying cause. Medical treatment is then pursued as indicated for the specific organic condition. Any child with a suspected organic psychosis should be admitted to a medical inpatient unit for diagnostic evaluation and treatment. This treatment is especially important because organic psychosis may be a transitory condition in a child or adolescent whose illness or intoxication is progressive and life-threatening.

Diagnose underlying cause.
Request *immediate psychiatric consultation*, with all psychiatrically based psychosis.
Use *medical hospitalization*, if clinically indicated, with organic psychosis.
Request *psychiatric consultation* with psychotic drug intoxications, either *immediately* or when mental status stabilizes.
Use *quiet room*, *family* and friends, and *constant medical supervision*.
Avoid administration of antipsychotic medication for psychiatrically based psychosis, in emergency department, when possible.
Use *restraints*, if necessary.
Recognize clinical variations of *extrapyramidal reactions* to antipsychotic medications (Table 122.26)

Table 129.24. Guidelines for Management of Acute Adolescent Psychosis

Other important components of the management of a psychotic child involve controlling the child's behavior, preventing injury to himself or herself or others, and alleviating the child's fear and anxiety. This goal should be attempted first through supportive statements indicating the physician's appreciation of the child's condition and his or her distress. Specific instructions to the child (e.g., “Try to relax and look at your mother”) may also be effective. Often, such interventions calm the child, but since the child is distractible and anxious, instructions may need to be repeated frequently.

Antipsychotic and sedative medications affect the child's neurologic status and should therefore be used only when the medical diagnosis is known with certainty and when it is clear that the medication will not worsen the underlying disease process or potentiate the intoxication. (Specific medications and dosages for psychosis are discussed under “Management,” see Table 129.25.) In most instances, when direct behavior control is essential, the child should be placed in arm and leg restraints. While in restraints, the patient should be attended by staff or family members and provided with frequent orienting statements and explanations of the need for restraint.

Generic Name	Brand Name	Estimated Equivalent Dosage (mg)	Total Daily Dosage
Phenothiazones			
Chlorpromazine	Thorazine	100	50–1000
Thioridazine	Mellaril	100	50–800
Trifluoperazine	Stelazine	5	5–30
Fluphenazine	Prolixin	2	1–20
Butyrophenone			
Haloperidol	Haldol	2	2–40
New Atypical Neuroleptics			
Clozapine	Clozaril	75	300–450
Risperidone	Risperdal	—	1–6
Olanzapine	Zyprexa	—	2.5–17.5
Quetiapine	Seroquel	—	150–300

Table 129.25. Antipsychotic Medications

Autism and Other Pervasive Developmental Disorders of Childhood

These disorders are extremely rare, and are approximately three times more common in boys than in girls. Because of the infrequency of these disorders and the chronicity of their course, it is unusual for children with either infantile autism or other pervasive developmental disorders to present in an ED undiagnosed. However, children with these disorders may present in the ED for the treatment of intercurrent illnesses or an acute exacerbation of the child's behavior.

Autism

According to the current psychiatric diagnostic nomenclature (*DSM* IV), autism is one specific type of pervasive developmental disorder (PDD) of childhood. The major differentiating feature between autism and other forms of PDD is the age of onset. Autism always has an onset before 30 months of age. Children with autism have a generalized lack of responsiveness to other people and a failure to develop normal attachment behavior. They do not develop relationships and instead play alone, often showing stereotyped behavior and using objects in bizarre, inappropriate ways. The autistic child becomes extremely upset if objects in his or her environment are disturbed or changed. Language development is

impaired or absent. Only 30% of autistic children have an IQ (intelligence quotient) greater than 70. Some autistic children have underlying illnesses, such as maternal rubella syndrome or previous encephalitis or meningitis, but in many cases the cause is unknown. Many autistic children have coexisting seizure disorders. The course of infantile autism is generally chronic, with two-thirds of all autistic children remaining severely handicapped throughout life.

A comprehensive educational and socialization program with psychiatric monitoring is essential for autistic children. If an autistic child seen in the ED is not participating in such a program, outpatient psychiatric referral is indicated. Medication management of autism at times may involve careful use of psychotropic medication, including neuroleptics, antidepressants, and a-adrenergic agents. Such psychotropic strategies should be used only in conjunction with ongoing psychiatric treatment. Clear justification must be present for psychotropic medication use, not just the diagnosis of autism. In general, acute psychiatric hospitalization is rarely necessary with autism. In instances of extremely disturbing behavior or acute agitation, sedation with either diphenhydramine (1 mg/kg) or chloral hydrate (30 mg/kg) may be helpful. If the parents are distressed by their child's immediate behavior and the child is receiving psychiatric treatment, phone contact with the psychiatrist may be helpful to both the emergency physician and the family. In the absence of ongoing care, a psychiatric consultation should be requested.

Other Pervasive Developmental Disorders

Pervasive developmental disorder of childhood is a generic term that includes other developmental impairments in which an incapacity to form reciprocal relationships with others results in severe, sustained impairment of attachment and social relationships. Other features may include extreme anxiety and severe emotional reactions to minor difficulties, with inappropriate affect and extreme mood lability. Abnormalities of speech, hypersensitivity to sensory stimuli, peculiar posturing, and self-mutilation may also occur. PDD other than autism has onset after 30 months and before 12 years of age.

The term *PDD* incompletely incorporates entities such as childhood schizophrenia, symbiotic psychosis, and atypical psychosis, as well as other recently added conditions. One type of PDD with which the emergency physician should be familiar is Asperger's disorder. Children with this disorder typically have normal or above average intelligence, with a well-developed capacity for speech and language. The impairment is in the capacity to form reciprocal relationships, and emotional rigidity, idiosyncratic thinking, and intense pursuit of a narrow range of interests may be present. Children with Asperger's disorder may be confusing to the emergency physician, because they present as higher functioning than other children with PDD and are not psychotic, yet they may appear to be significantly strange.

All children with PDD, including children with autism, require comprehensive psychiatric and educational treatment. Parents of children with autism and PDD should be given appropriate referrals, because children who receive services early in their development are believed to have an improved prognosis. When necessary, the same acute pharmacologic approaches for children with autism are also relevant for other PDDs. Low-dose thioridazine, a sedating neuroleptic, may also be used. With the acute exacerbation of a child with PDD, psychiatric hospitalization may be necessary, both to provide assistance to the parents and to develop or modify a comprehensive treatment program. Families with acute concerns about their child's behavior should receive psychiatric consultation.

Schizophrenia

Schizophrenia often has its onset in adolescence and occurs in approximately 0.5% of the population. This disorder is equally common in males and females, and is more prevalent among family members of known individuals with the disease. An excess of dopamine activity in the brain is believed to be one feature of schizophrenia.

Clinical Manifestations

Symptoms of schizophrenia involve impairment of basic psychological processes, including perception, thinking, affect, capacity to relate, and behavior (Table 129.26). Impaired thought content includes delusions (strongly held beliefs involving the self with no basis in reality), such as delusions of persecution and external control. For example, an adolescent with schizophrenia may think that others can read and insert thoughts into his or her mind. Significantly illogical thinking occurs. Speech is often characterized by loose associations, in which ideas shift from one subject to another entirely unrelated subject without the speaker recognizing that the topics are not connected. Auditory hallucinations are common and may include direct commands for suicide or for violence to others. Typically, but not always, the voices talk to the patient in the third person, with a highly critical and demeaning message. Affect may be blunted and flat, or inappropriate and bizarre. Sudden and unpredictable changes in mood may occur. These teenagers may appear extremely agitated or may be withdrawn, speaking only in monosyllables and describing only concrete objects. Schizophrenic patients typically have significant distortions of their identity and their abilities, and demonstrate behavior that is not goal-directed.

Flat affect
(Patient uninvolved and without emotion)
Auditory hallucinations
(Physician: "Have you been hearing voices even when no one is there?")
Thoughts spoken aloud
(Physician: "Can other people read your mind? Can you read their minds?")
Delusions of external control
(Physician: "Is anyone trying to kill you? . . . trying to control your mind or your body?")

Table 129.26. Acute Schizophrenia in Adolescence: Most Common Features

Although classified as a psychiatrically based psychosis, schizophrenia often has a strong family history and is considered to be an organic disorder. Families with schizophrenic adolescents may experience difficulties in communication, and relationships between parents and the affected teenager may be superficial and distant. Parents may have noted progressive emotional difficulties in the adolescent before seeking assistance and may be frightened by the teenager's bizarre behavior. At other times, the family may have been concerned about the child for many years, with no definitive understanding emerging before the acute onset of psychosis. Family members require clear explanations of the child's condition and proposed treatment, the longitudinal course of the disorder, and the essential need for their active participation.

The history often reveals a prodromal phase that includes social withdrawal, peculiar behavior, failure to look after one's appearance, and significant reduction in performance in school or work. This phase is followed by an acute phase in which the previously described symptoms develop, sometimes as a result of an acutely stressful event. The overall course of schizophrenia is often chronic and associated with remissions and exacerbations. Exacerbations often occur when treatment, including medication, is suspended. However, other individuals experience a schizophrenic-like acute psychosis and recover completely with appropriate treatment, experiencing no further deterioration.

Management

The management of an acute schizophrenic episode should always take place in collaboration with psychiatric consultation. Patients with suicidal or homicidal ideation should receive psychiatric hospitalization. Psychotic patients from disorganized home environments should also be hospitalized for initial treatment. In general, the approach to the psychotic patient in the ED depends on the condition of the patient and the anticipated site of the ongoing treatment. For agitation and dangerousness, approaches include reassurance and a quiet setting, psychotropic medication, and/or physical restraint. Medication involves a choice between a calming, sedating medication such as diphenhydramine or use of an antipsychotic medication. If the child requires an additional psychiatric assessment at a site different than the ED, such as at a designated psychiatric emergency facility as a precondition for psychiatric hospitalization, antipsychotic medication should be used sparingly, if at all, at the pediatric ED. Use of antipsychotic medication can alter the child's mental status such that, by the time of assessment at the emergency psychiatric facility, the child no longer appears in need of psychiatric hospitalization and instead may be inappropriately sent home.

If the ED is associated with a psychiatric inpatient unit where the child can be admitted, antipsychotic medication can be used in the ED, resulting either in psychiatric hospitalization or the child's return home with the parents with outpatient treatment, depending on the child's clinical response. Haldol (haloperidol) should be used in these circumstances. A conservative approach involves a single dose of 5 to 10 mg haloperidol orally or intramuscularly for adult-size adolescents (approximately 70 kg) or 2 to 5 mg haloperidol (orally or intramuscularly) for smaller children. A more aggressive approach involves the use of these same doses, administered every 3 to 60 minutes up to a maximum dose of 40 mg, until the patient's state of agitation lessens or until he or she becomes sedated. The patient's vital signs, general condition, and possible side effects should be monitored frequently. If the patient does not respond to this latter medication regimen, inpatient psychiatric hospitalization is necessary. If significant improvement occurs, suicidality and homicidality are absent, and side effects do not occur, the patient can be considered for discharge to outpatient psychiatric treatment with careful follow-up, so long as the parents or caretakers are well-organized, appreciate the child's condition, and feel capable of managing the child at home.

Commonly used antipsychotic medications, their trade names, relative potency, and usual dosage ranges are listed in Table 129.25. In addition to long-standing antipsychotic medications (now called typical antipsychotics), which include both phenothiazines such as Stelazine (trifluoperazine) and non-phenothiazines such as Haldol (haloperidol), a new class of antipsychotic medications, called atypical neuroleptics, is being used. Included in this growing class are Risperdal (risperidone), Clozaril (clozapine), Zyprexa (olanzapine), and Seroquel (quetiapine). Clinical advantages offered by this new class of medications include clinical effects on the “positive symptoms” of schizophrenia (e.g., an improvement in the ability of the individual to relate to the environment and to others, not just a positive effect on hallucinations and delusions) and a decreased likelihood of extrapyramidal side effects and long-term tardive dyskinesia. The emergency physician should become familiar with the use of antipsychotic medications in emergency situations. With typical antipsychotics, high-potency agents, such as haloperidol or trifluoperazine, rapidly reach therapeutic blood levels and produce less sedation than low-potency drugs, such as chlorpromazine and thioridazine. However, high-potency antipsychotics have a greater incidence of extrapyramidal effects that can be reversed by administration of the appropriate medication (see following section).

The major side effects of typical antipsychotic medications are extrapyramidal symptoms, including acute dystonic reactions (abnormal muscle tone or posturing), akathisia (motor restlessness), and parkinsonian effects (rigidity, tremor, slowed movement, and loss of balance). Acute dystonic reactions are best treated by the intravenous or intramuscular administration of diphenhydramine (25 to 50 mg), followed by a daily maintenance dose of benztropine (1 to 2 mg/day orally). Adolescents begun on high-potency antipsychotic medication on an outpatient basis should be treated prophylactically with benztropine (1 to 2 mg/day orally) to prevent side effects. Patients placed on antipsychotic medication should be clearly informed of possible side effects and instructed to return to the ED or to the responsible mental health professional should such symptoms develop. Attempts can be made later in treatment to lower the dosage of antipsychotic medication and discontinue use of the antiparkinsonian agent.

Acute Reactive Psychosis

Acute reactive psychosis, a relatively uncommon psychiatrically based psychosis, involves a time-limited loss of reality caused by the accumulated effects of externally imposed traumatic events. Although vulnerability may vary from child to child, children and teenagers can develop acute psychotic symptoms in response to trauma. The diagnosis of reactive psychosis can be made partly by history, but only after a complete medical and psychiatric evaluation has eliminated organic and other psychiatrically based psychoses. The acuteness of the clinical presentation and its precipitating events differentiates acute reactive psychosis from posttraumatic stress disorder.

The clinical picture of acute reactive psychosis varies, in some instances resembling schizophrenia and in others a less defined disorganized state characterized by loss of contact with reality, panic, and specific hallucinations (usually auditory or visual).

Different traumatic experiences, including physical or sexual abuse, rape, homelessness, and running away, may elicit a reactive psychosis. All such situations impose stress on the child and may also disrupt usual patterns of living. Confronted with a new environment and a new reality, the child's familiar cues are absent and confusion or frank psychosis may occur.

The emergency physician should appreciate that most children who present with acute reactive psychosis do not have a permanent psychiatric disorder. The emergency management is similar to that of other psychotic states. Physical and emotional protection of the child is the first priority. The child should be given support and time to reconstitute. Efforts to avoid antipsychotic medication should be made in the beginning, but, when necessary, low-dose antipsychotic medication can be used. When the parents or other caretakers are not implicated in the traumatic events, emergency staff should encourage their active involvement with the child. When the parents are implicated in the trauma or when the facts are unclear, immediate investigation should take place and contact made with appropriate child protection authorities, if indicated.

After emergency treatment, the prognosis of the child depends in large measure on the restoration—or creation—of a safe and dependable family support system. Referral for outpatient family therapy should be made unless the child requires psychiatric hospitalization for further evaluation or treatment. In the absence of adequate family support, some of these children may eventually require foster placement, residential treatment, or other placements.

Manic–Depressive or Bipolar Disorder

Manic–depressive or bipolar disorder occurs in approximately 0.5% of the population. Onset is usually before 30 years of age and occurs during late childhood and adolescence. Because depression is discussed in detail elsewhere in this chapter, this section deals only with manic psychosis, most commonly observed in adolescence, and with the childhood form of bipolar disorder.

Clinical Manifestations of Manic Psychosis

The patient with mania has a distinct period of predominantly elevated, expansive, and irritable mood (Table 129.27). The child has a significant decrease in need for sleep, high distractibility, hyperactivity and pressured speech, and emotional lability. These patients also exhibit what is called flight of ideas—a nearly continuous flow of accelerated speech with abrupt changes from topic to topic, usually based on understandable associations, distractions, or plays on words. Unlike the loose associations of the schizophrenic, the flight of ideas of a manic patient retains logical connection from one idea to the next, but moves quickly from one topic to another. The manic patient may at times have a remarkably inflated self-esteem, with uncritical self-confidence and significant grandiosity. This grandiosity may also include delusional ideas. The individual may be aggressive and combative. He or she may go on buying sprees or pursue other reckless behaviors. Sleep patterns may be significantly impaired, the individual reporting limited or no need to sleep. Manic patients usually have a history of previous depressive episodes, but an acute manic episode in adolescence may be the initial presentation of the disorder. A family history of psychiatric disturbance usually exists in patients with manic–depressive disorder. Typically, manic patients report feeling extremely well, and they are brought to the ED against their will. However, underneath this superficial presentation, the patient has extremely low self-esteem and self-confidence.

Pressured speech	Euphoria
Grandiosity	Anxiety/irritability
Apparent "high" (euphoria)	Combativeness/panic
Rapid shifts of emotion	

Table 129.27. Acute Mania in Adolescence: Most Common Features

The differentiation of mania and schizophrenia in an initial episode of psychosis in adolescence may at times be difficult. Hyperactivity, distractibility, and expansive and euphoric mood are often helpful in identifying manic individuals. Both groups may have auditory hallucinations and delusions, but someone listening to the speech of the manic adolescent

should recognize the flight of ideas and their connection with each other.

Clinical Manifestations of Childhood Bipolar Disorder

Bipolar disorder that occurs in childhood and early adolescence usually looks different than the later adolescent and adult form just described. The child typically presents in the beginning with depression rather than mania and with remarkable shifts in mood, involving sudden changes from depressed to irritable or happy, then back to irritable or depressed. These changes can be disorienting to parents, who cannot understand why the child changes so much and so dramatically, possibly even several times the same day. The sudden shifts in mood and functioning are the reason that childhood bipolar disorder is often referred to as “rapid cycling.” Unlike the older adolescent, the child often does not have a clear recovery from identified episodes but rather may continue to present in at least a mildly unstable way with irritability and anger for much of the time. Approximately 90% of children with bipolar disorder have concurrent symptoms of attention deficit hyperactivity disorder, which may in fact present before the onset of the mood instability.

Management

When an adolescent is suspected of having manic–depressive illness or an acute manic episode, psychiatric consultation should be obtained and psychiatric hospitalization initiated. Involuntary commitment may be necessary. Because the treatment of mania often includes the long-term use of lithium carbonate, which takes time to take effect and which requires careful blood monitoring to assure therapeutic levels, psychiatric hospitalization is necessary. Initial emergency treatment of the agitated manic patient may require the use of restraints and the acute administration of antipsychotic agents, such as haloperidol, in doses equivalent to those used for schizophrenic adolescents. The physician should also be aware that some patients receiving lithium may present with signs of lithium overdose, including nausea, vomiting, muscle weakness, ataxia, tremor, slurred speech, blurred vision, and confusion or somnolence.

Management of the childhood form of bipolar disorder, once suspected and diagnosed, is less likely to involve the need for psychiatric hospitalization, although this is not invariably the case. In the ED, psychiatric consultation should be obtained because these children do best when they receive ongoing individual and family psychotherapy with psychotropic medication. Younger children often respond more favorably to anti-seizure mood stabilizing medications, such as Depakote (valproic acid) and Tegretol (carbamazepine), than to lithium carbonate. However, some children respond more favorably to lithium, so the treatment must be individualized to the child, with careful longitudinal monitoring of blood levels and clinical responsiveness.

POSTTRAUMATIC STRESS DISORDERS

In part to acknowledge the accumulated effect of stress on individuals and also to avoid more invasive diagnoses, psychiatry has invoked the concept of posttraumatic stress disorder (PTSD) with increasing frequency in recent years. Used in the 1970s and 1980s to explain some of the maladjustments of some Vietnam War veterans, PTSD can also occur in childhood and adolescence, typically based on the experience of severe trauma during earlier years. Either the reemergence of the old trauma (or the emergence of a new similar one) or the recollection of the original trauma can activate a PTSD.

A summary of the official description of PTSD in the psychiatric nomenclature (*DSM*-IV) is helpful in understanding this concept:

> The person has been exposed to a traumatic event in which the person experienced, witnessed, or was confronted with an event that involved actual or threatened death or serious injury, or a threat to the physical integrity of self or others. The person's response involved intense fear, helplessness or horror, or, in children, disorganized or agitated behavior. In addition, the traumatic event is persistently re-experienced in one or more ways, there is persistent avoidance of stimuli associated with the trauma and numbing of general responsiveness, and there are persistent symptoms of increased arousal.

Highly stressful situations that may precipitate severe emotional reactions by the child and PTSD include: physical beatings and other violence, repeated threats and belittling by adults, long-standing hunger and poverty, sexual abuse, and rape. In some cases, a separate dissociative reaction may occur instead (see following section). With the child, PTSD probably emerges through a combination of traumatic events along with a silent or nonaccepting environment that fails to provide the child with adequate protection and support.

Symptomatically, the child may persistently reexperience the traumatic event in many ways, including recurrent and distressing recollections of the event, which may be observed through repetitive play by young children in which themes or aspects of the trauma are expressed. Recurrent and distressing dreams of the event may also occur. Hallucinations and flashbacks may follow the child's sudden reliving of the experience. In addition, events that symbolize or resemble some aspect of the traumatic event may produce intense anxiety and distress; the connection between precipitating event and distress is not always evident to parents or child.

Other PTSD symptoms experienced by children include generalized numbing of responsiveness to events and people. Stimuli associated with the trauma may be consistently avoided. The emergency physician should also be alert for signs of increased arousal—anxiety and agitation, difficulty falling asleep, irritability or anger, suspiciousness, difficulty concentrating—and various physiologic complaints in response to events that resemble or symbolize the traumatic event.

The key task for the emergency physician is to recognize PTSD in the differential diagnosis of an agitated, confused, or even psychotic child or adolescent. A careful history usually provides clues to this diagnosis. Supportive management in the ED, including using family and friends, often is sufficient. Low-dose antipsychotic medication should be reserved for children who are frankly psychotic and who do not respond to reality-based support. Often, an antihistamine or anxiolytic

medication may suffice.

When parents dismiss or doubt the child's symptoms or worries, the emergency physician can encourage the parents to respond supportively to their child. When the physician suspects parental abuse, this concern must be addressed directly with the family with appropriate authorities brought in, if justified.

Many children with PTSD benefit significantly from individual and family therapy. If child and family are not already in treatment, a referral is appropriate.

DISSOCIATIVE DISORDERS

Some children develop a dissociative disorder in response to extreme trauma. In a dissociative disorder, the child separates the usually integrated functions of identity, memory, and consciousness. As a result, the child's affect appears split off from the rest of the person. Specific symptoms vary, but in most cases, the child appears distant, even weird, but is not psychotic. Dissociative disorders occur most commonly in females, with sexual abuse a common original trauma.

The function of dissociative reactions is believed to decrease the child's awareness of emotional pain caused by the trauma. The process of splitting off the affect from the body may help a severely traumatized child deal with and survive the assault. This response probably begins at the time of the trauma, especially if it occurs repeatedly, then is continued afterward as a form of coping. However, a consequence is that the child may continue to split off full emotional responsiveness to daily experiences, creating a profound isolation. This process may continue into adulthood.

The emergency physician may encounter a child with depersonalization, a feeling of detachment from one's self or a feeling of being an automaton or in a dream. Another dissociative response is psychogenic amnesia, the sudden inability to recall important personal information (or even know one's own identity). Some runaway adolescents may present with a psychogenic fugue, another dissociative disorder. In a fugue state, the individual leaves home unexpectedly with no apparent justification and may at times assume a partial or complete new identity.

The most extreme—and increasingly common—form of dissociative disorder is multiple personality disorder (MPD). In this condition, the child has two or more personalities and appears puzzling to parents, teachers, and physicians. At least two of these personalities recurrently take full control of the child's behavior, with the child unaware of the process. Children with MPD are aptly described as erratic, inconsistent, even mercurial.

The emergency physician should consider the possibility of MPD or other dissociative disorders in all children and adolescents who present in a confused and confusing way. None of these children are psychotic; in fact, an entirely different emotional process is operating. A thorough history is most rewarding and may reveal a female with repeated sexual abuse who often appears far off into her own world. Psychiatric referral is appropriate for patients with MPD. The emergency physician should also determine any possible ongoing abuse before releasing the child to the family.

SCHOOL REFUSAL

Background

School refusal, also called school avoidance and school phobia, entails a child's not attending school and expressing somatic complaints that keep him or her at home. Usually, some somatic complaint is used as the justification for school absence, with the child just “too sick” to be in school. School refusal involves the knowledge and complicity of the family. A parent, usually the mother, is aware of the child's school absence and has endorsed his or her being home, in part because the parent may consider the child to be physically ill.

School refusal is an important condition with which the emergency physician should be familiar. Usually, it is not the initial complaint. Typically, one or more physical complaints bring the child to the ED, and information about school attendance is not offered. The physician must maintain an “index of suspicion” in a child with recurring complaints for which no organic cause is apparent.

On rare occasions, school refusal may be the initial concern of a parent. The mother may bring her child, stating that for a specified time she has been unable to get her child to go to school. Attempts to force the child have been ineffective, either because the child refused to go, got sick in school and was sent home, or caused such family upheaval that eventually the parents gave in. In such a circumstance, school refusal represents a true emergency and should be responded to as such.

There are no specific prevalence figures for school refusal, but the condition is not uncommon. It probably occurs more often than it is diagnosed and may be easily missed. The problem is not confined to a specific population or socioeconomic group and can occur across ethnic, religious, and class lines.

Chronically ill children have been considered high risk for school refusal, although many physically ill children are determined to attend school regularly despite their disability. The child with vague, undiagnosable somatic complaints is more likely to fit the school refusal pattern. So-called vulnerable children and special children may also be in this category. Vulnerable children were ill at an earlier point in their lives, but parental overconcern remains. Special children, such as those born to an older couple who had problems conceiving, were wanted by their parents. The love and attention given to the child can easily yield to overprotectiveness, setting the stage for the development of school refusal. School refusal reportedly tends to occur in families with an orientation toward illness and disability, so it is not uncommon for one or more parents to have past or present illnesses, physical complaints, or hospitalizations. The presence of marital problems may also contribute to the development of school refusal.

Clinical Manifestations

Certain school attendance patterns are suggestive of but not necessary for the diagnosis of school refusal. More absences occur in the fall, when school begins, than in the spring. The child often exhibits a reluctance to return to school after weekends and holidays. There may be a lessening of somatic complaints on weekends and over the summer. Similar sporadic attendance patterns may often be elicited at some other time in the child's past. In other instances, however, school refusal may develop in a child who has previously given no cause for concern. Only specific questions asked routinely of every child and parent about school attendance yield the necessary information. Such questions should be specific, addressing not just recent school attendance but the pattern over the entire year. In this way, the diagnosis is made on the child who misses several days of school per week but still maintains good grades, as well as the more easily diagnosed child who has missed several consecutive weeks or months of school.

Schmitt has formulated a diagnostic triad of the clinical manifestations of school refusal: 1) vague physical symptoms, 2) normal physical and laboratory findings, and 3) poor school attendance. The child may have one or more complaints. Schmitt has also pointed out that many of the symptoms are reflective of depression and anxiety. This finding is consistent with the fact that many children with school refusal are also depressed. The symptoms that may be present include fatigue, insomnia, transient pallor, a smothering feeling (hyperventilation), palpitations, tension headaches, dizziness, syncope, and a variety of gastrointestinal complaints. Most common is nonspecific abdominal pain. Anorexia in the absence of weight loss may occur, as may diarrhea. Nausea may occur with or without associated vomiting. A clue to the psychogenic nature of the vomiting is that it sometimes follows stressful events. The serious complaint of vomiting should not mislead the physician into an overly extensive workup that iatrogenically reinforces disability. Similar considerations apply to the skeletal complaints that may occur—bone pain, joint pain, and back pain. Such tentative diagnoses as “possible rheumatoid arthritis” should be avoided. Other possible symptoms that may mask the school refusal syndrome include chest pain, dysmenorrhea, muscle weakness, coughing, tics, and recurrent sore throats.

Certain characteristics of the family with school refusal have been identified. Berger described four important elements: 1) An overprotective infantilizing attitude toward the patient exists. The child is excused from family responsibilities, and his or her wishes are quickly granted. 2) There is a belief in the physical or emotional vulnerability of the mother. This belief is fostered by the mother herself, who may voice fear of an emotional breakdown and complain of various physical symptoms. 3) The fathers in such families tend to be isolated and devalued. They are seen as disinterested and unreliable, perhaps even as violent. There may be underlying marital tension. 4) It has also been found that, in many families with school refusal, a major change in family composition, often the departure from home of an older sibling, has occurred. This situation focuses new attention on the identified patient.

Other characteristics of families with school refusal have been noted. An illness orientation is revealed by physical complaints in other family members as well as frequent somatic references in verbal communication. The closeness between the mother and child may be manifested by the mother's frequent use of “we” when talking about the child. Active undermining by the parents may at times be observed.

Management

The major responsibility of the emergency physician is the detection of school refusal. The diagnosis is made by having a higher index of suspicion for children with vague somatic complaints and by routinely inquiring about school attendance patterns.

Although the emergency physician cannot guide the entire treatment of school refusal, he or she can get the process going. The physical examination should be done in the presence of the parents in a thorough manner, with the physician emphasizing the absence of physical findings. Appropriate, but not excessive, laboratory work should be performed, and medication should not be prescribed. After acknowledging the genuineness of the child's symptom so that there is no misunderstanding that the child is “faking it,” the physician should provide a firm and unequivocal statement to the family that the child has no serious illness. He or she should then make sure that the family understands what has been said and accepts it. In this way, misunderstandings or disagreements can be confronted directly, thereby decreasing the likelihood of subsequent “doctor-shopping” by the family. The emphasis is then placed on the child's learning to function in spite of his or her symptoms.

Once school refusal is recognized and the possibility of organic disease ruled out, the principal goals in the treatment of school refusal are: 1) getting the child back to school as soon as possible, 2) ensuring continuity of medical care, and 3) addressing underlying individual and family issues that contributed to the development of the problem.

It may be helpful for the parents rather than the physician tell the child that he or she needs to return to school. In this way, the family takes responsibility for the resolution of the problem from the beginning of the intervention. The parents should be encouraged to work closely together to achieve their desired goal.

The emergency physician varies his or her recommendations concerning the child's return to school according to the severity of the problem. Uncomplicated cases of childhood or preadolescent school refusal can be addressed by the emergency physician and psychiatric consultant, and referred to the primary care physician or to a pediatric colleague. With severely depressed or psychotic teenagers, or in situations in which the problem has been long-standing, psychiatric consultation should be followed by a specific mental health referral. The emergency physician should ask the family to identify the responsible physician who will monitor the child's physical well-being over time and collaborate with the therapist. Ideally, minimally disturbed, younger children should return to school within several days' time. A follow-up appointment should be scheduled with either the primary care physician or the mental health professional for the day that the child returns to school. For longer term, more complicated cases of school refusal, careful planning of the child's return to school may be necessary. In all cases, the emergency physician should resist any request by child or family to

sanction or underwrite the child's continued absence from school.

CONDUCT DISORDERS

Background

A child with a disorder of conduct engages in repetitive, socially unacceptable behavior, without evidence of medical or other psychiatric disorder. The diagnosis of conduct disorder implies a continuing pattern of disruptive or deviant behavior, rather than isolated antisocial acts. The behavior may involve violence and aggression (e.g., vandalism, mugging, assault, and rape), or may involve behavior that is socially unacceptable but nonaggressive (e.g., truancy, running away, lying, stealing, substance abuse). Therefore, a disorder of conduct involves more serious behavior than ordinary mischief and pranks of children and adolescents. Because violent and other unacceptable behaviors may be performed by children with medical illnesses and intoxications, these causes must be ruled out before the diagnosis of conduct disorder can be made. Similarly, because children with psychosis and depression can also behave in socially unacceptable ways, these serious psychiatric disorders must also be considered and eliminated before diagnosing a conduct disorder (see Chapter 20). However, even with primary medical and psychiatric causes of socially unacceptable behavior ruled out, some youth with conduct disorder may have ill-defined physiologic predispositions that contribute to its emergence.

Viewed longitudinally, the incidence of youth violence has increased substantially since the 1970s, particularly among African-American males and females, and Hispanic- and Native-American males. The number of youth in detention facilities has increased proportionately. Approximately one in every nine children appear in juvenile court before their 18th birthdays. Delinquent behavior is five times more common in males than in females, and males are more likely to demonstrate violent behavior directed against others.

Society disagrees about whether to regard children and adolescents with conduct disorders as psychiatrically impaired and needing treatment, or as delinquent and needing detention or incarceration. Probably only a small percentage of violent and aggressive children are brought to the ED for psychiatric evaluation. Many are taken by police to detention centers and others engage in their unacceptable behavior without receiving legal or medical attention. No consistent agreement exists about the appropriate criteria for taking such children to an ED as opposed to a juvenile center. In actual practice, certain factors probably influence the choice of disposition, such as age (younger children are more likely to receive medical evaluation), socioeconomic level (middle- and upper-level income children are more likely to be taken to an ED), race (Caucasian children are more likely to be taken to an ED than African-American or other minority children), and nature of the infraction (children with aggressive acts directed outside the family are more likely to be taken to a detention center). Aggressive children should always undergo an emergency medical and psychiatric evaluation any time an intoxication, underlying medical condition, or other psychiatric disorder is suspected.

Clinical Manifestations

Children with conduct disorders typically have poor adjustment at home and in the community. Peer relationships are superficial, based more on what the child can get from the other person than on a sense of empathy. The child thinks primarily about himself or herself, trying to manipulate situations to personal advantage without significant concern for the feelings and needs of others. The child with a conduct disorder is unlikely to extend himself or herself for others when no immediate advantage can be gained. When the child is apprehended, little sense of remorse or guilt is exhibited, but rather a sense of anger at being detected and detained. Such children rarely accept responsibility for their own actions, and instead tend to blame others for their mistakes.

School attendance of children with a conduct disorder is often sporadic, and academic performance is often poor. This may be caused by a variety of factors, including lack of interest and discipline, but may also be caused by specific learning disabilities and a concurrent attention deficit disorder, diagnoses that are remediable but often missed.

The child or adolescent with a conduct disorder shows low frustration tolerance, irritability, and temper outbursts. He or she may be reckless in behavior and project an image of “toughness.” Smoking, drinking, drug use, and precocious sexual activity may all occur. In addition to possible legal difficulties, the child may have other problems, including school suspensions, drug dependence, sexually transmitted disease, pregnancy, and physical injury from accidents and fights.

The presence of a conduct disorder implies a failure of the child's environment to instill familial and societal values and to implement their rules effectively. As a result, the child comes to believe that he or she can act as he or she chooses and does not develop control of impulses. The specific pattern of inadequate limit-setting varies, but families share an inconsistency in enforcing rules and do not hold the child accountable for his or her behavior. In some families, discipline may fluctuate from being perfunctory at times to being harsh and even physically abusive at other times. Parental role models may show poor impulse control themselves and disregard societal norms. In some families, one parent is the enforcer and the other tends to protect and excuse the child; in other families, the parents may shift roles in protecting the child. In these situations, mutual parental undermining enables the child to persist in avoiding responsibility for his or her behavior. As a result, the child or adolescent comes to expect that, somehow, he or she will survive or be excused from any repercussions of his or her behavior.

In addition to inconsistent limit-setting, families of children with conduct disorders tend to be poorly organized, with the roles and expectations of various family members often unclear. Parental separations and divorce, mental illness, and alcohol or drug abuse may also be factors. Parental criminality and incarceration occur in some families. Families with aggressive and impulsive children often do not know how to effectively use social service resources and may consider themselves helpless in controlling their child and in dealing with the world at large.

When taken to the ED, predictably by some outside agent (police, parent, or other caretaker) against his or her will, children with a conduct disorder have variable presentations. For example, the child or adolescent may be angry, hostile,

uncooperative, and even violent, refusing to answer questions directed to him or her, but quick to interrupt to defend himself or herself when others speak. Alternatively, the child may present with a superficially smooth and pleasant facade, hoping to persuade the physician and authorities of his or her innocence. Often, once the child realizes that he or she will not be permitted to act out or manipulate in the ED, he or she may settle down and cooperate more fully. At other times, the child maintains an essentially impenetrable persona.

Management

The goals for managing aggressive and disruptive children in the ED are 1) to ensure the safety of the child, family, and staff; 2) to rule out possible medical conditions and severe psychiatric disorders before making the diagnosis of conduct disorder; and 3) to gather sufficient information to make an appropriate disposition.

The safety of the child and staff and control of the child's unacceptable behavior must be achieved in the ED. In many instances, the disruptive behavior occurred and ended before the child's coming to the ED and gaining the child's cooperation is not a problem. In other instances, however, the child may remain combative and aggressive in the ED. Dealing with such a problem requires the presence of adequate security staff and a quiet space where attempts to control the patient do not disrupt the remainder of the ED. The patient should be told firmly that he or she is in the hospital for medical and psychiatric evaluation and will not be permitted to harm himself or herself or others. The child should be informed of the need to cooperate with the staff and to control his or her behavior. The child's parents, if present, should be asked to assist in controlling the child. The child can be reassured that he or she will get a chance to tell his or her side of the story completely. These interventions usually are sufficient to gain the child's cooperation.

When the child remains aggressive and threatening in the ED, an adequate number of security staff should be summoned. If the child still cannot be calmed, physical restraints should be used. This task is accomplished by several security personnel laying the child down on a cart and applying leather arm and leg restraints with soft cuffs. Often, the external control established by the use of restraints helps the child or adolescent to feel safe and regain personal control. Nonpsychotic aggressive patients do not usually require psychotropic medication, and none should be given until possible medical conditions and intoxications have been ruled out. When needed for children 12 years and older, chlorpromazine (25 mg intramuscularly) can be used, and psychiatric consultation should be sought.

The history and physical examination assist in ruling out medical conditions and intoxications. The presence of ongoing medical conditions should be specifically asked about, as should any recent alcohol use or drug ingestion because substance abuse is common. As indicated, specimens for toxicologic screening should be obtained. Epilepsy can be ruled out as the cause of the abnormal behavior in the presence of a normal neurologic examination and the absence of an aura, abnormal neurologic signs, or postictal phenomena.

The history and mental status examination enables the diagnosis of psychosis or depression, when present, to be made. A history of withdrawal and social deterioration, including disregard of rules and possible cruelty or firesetting, suggests emotional antecedents of the current disturbed behavior. Hallucinations, delusions, and loose associations suggest psychosis, whereas sad affect, crying, and possible suicidal ideation indicate the presence of severe depression.

With this information available, the emergency physician can now consider appropriate treatment and disposition of the patient. Children with medical conditions and acute intoxications are best managed through medical hospitalization. The presence of psychosis or depression requires psychiatric consultation and possible psychiatric hospitalization. Psychiatric consultation should be obtained for children with presumptive conduct disorder and no underlying medical condition. Less severe and complicated cases can be managed through referral for outpatient therapy. More severe cases may require more intensive community-based services, such as partial hospitalization, or even psychiatric hospitalization, which should be considered in cases of severe, chronic conduct disorders, especially when the child's behaviors are escalating and treatment to date has been ineffective. For intervention to be effective with such children, the family must be willing to participate actively, with the goal of altering persistent patterns of disturbed behavior.

The child should agree to follow the rules of the institution and attempt to control violent impulses. The hospitalization can be explained as an opportunity for the child to learn to behave in more socially acceptable ways while the family learns to set limits for the child more effectively.

Involuntary hospitalization may be necessary when the child's condition continues to pose a threat to himself or herself or others, or when overt homicidal or suicidal ideation is present. When the child is not suicidal or homicidal and refuses to make a commitment to work in psychotherapy, and when the family does not support the proposed psychiatric hospitalization, problematic behaviors are more likely to continue and the child may eventually enter the juvenile justice system.

ATTENTION DEFICIT HYPERACTIVITY DISORDER

Background

Attention deficit hyperactivity disorder (ADHD) refers to a syndrome found in school-age children, characterized by a pervasive difficulty in maintaining attention and goal-directed behavior. The condition, previously called hyperkinetic syndrome and minimal brain dysfunction, has been relabeled, “ADHD” because the primary source of difficulty is believed to be inattention. ADHD has an incidence of between 5 and 10% of school-age children, occurring 10 times more often in boys than girls. It is presumed to have an underlying neurologic cause. Some cases of ADHD may be inherited, whereas others may be a consequence of prenatal or perinatal difficulties or of unknown cause. ADHD occurs in up to 50% of latency-age children receiving psychiatric treatment, making it the most common cause of chronic behavioral problems for this age group. The peak age range for referral of ADHD children is between the ages of 8 and 10.

The emergency physician should be familiar with ADHD, because many children so affected become depressed and may

make suicide attempts. ADHD is also common in children with bipolar disorder, and may in fact initially mask the bipolar disorder. During adolescence, ADHD may itself be masked by antisocial behavior, so the physician should always consider the possible concurrence of ADHD in the presence of conduct disorders. Because identification and treatment can produce significant improvements in the clinical picture, the diagnosis of ADHD should not be missed.

Clinical Manifestations

Although some children with attentional problems present without hyperactivity, most children have hyperactivity in association with inattention and impulsivity and therefore fit within the full ADHD umbrella. Wender has described the various possible components of the ADHD picture. Attentional difficulties occur both at home and in school, and are often more severe in school. The child has difficulty fixating attention. This problem manifests itself as an inability to persist for long periods in any activity and, in extreme form, may involve a frenetic movement from activity to activity. Rather than persisting in schoolwork and other tasks, the child often appears not to be listening to the teacher, and discipline may be a problem.

Impulsivity is another essential characteristic of ADHD. The child has difficulty with self-control, exhibiting behaviors that get him or her in trouble with parents, siblings, teachers, and peers. At home, the child typically has outbursts and temper tantrums, and enforcement of discipline may be difficult. Lack of self-control may also manifest through stealing, lying, playing with matches, and other forms of acting out.

Hyperactivity is usually part of the clinical picture, although some children with a subtype of attentional problems are inattentive but not hyperactive or impulsive. As with attentional difficulties and impulsivity, hyperactivity tends to be worse in a group situation than at home or in one-to-one interactions. This characteristic at times creates difficulty in the diagnosis of ADHD because the telltale signs are least likely to occur during an individual assessment by the physician. In one study, 75% of children referred for hyperactivity did not exhibit this behavior in the doctor's office. Therefore, the most important diagnostic clue is the history as related by parents, school teacher, and child. However, the aimless, non–goal-directed quality of the activity rather than the absolute amount of activity distinguishes the ADHD child from a normally active child.

The child with ADHD may have associated neurologic deficits or “soft signs,” but these signs are not necessary for the diagnosis. Up to two-thirds of children with ADHD may have motor incoordination as manifested by deficits in gross and fine motor skills. Impairment of fine motor skills may manifest in inability to tie shoelaces and poor handwriting. Poor eye–hand coordination may be reflected in difficulties with sports, and difficulty riding a bicycle suggests an impaired sense of balance. Various perceptual problems, including right–left discrimination problems, problems with spatial orientation, problems of sensory integration, and problems in processing various types of information, all may be present and may contribute to learning problems. Specialized psychological and educational evaluations are used to detect these deficits.

The child with ADHD may be labile, with fluctuations in mood and a tendency toward overreaction and temper tantrums, but such responses are not always extreme. Appreciating the low self-esteem and possible depression that may be present in these children, as a result of academic failure, conflicts at home, and peer and sibling rejection is important. In acute situations, the depression may find expression as suicide attempts or violent behavior. ADHD children are a high-risk group for self-destructive behavior. The physician often perceives the child's sense of sadness beyond a cocky bravado that masks feelings of frustration and inadequacy.

Family problems are typically found in the families of ADHD children, if only as a consequence of the child's impulsivity and challenging behaviors. The child may provoke the parents, interrupt family members, and fail to learn consistently from experience. The child is often difficult to discipline effectively. Conflicts with siblings may occur. If the parents become overly frustrated by the child, marital strain and blaming may also occur.

Management

The principal responsibility of the emergency physician is to recognize the possibility of ADHD in children who present with other problems—including depression, mood instability, and conduct disorder—and to consider the diagnosis. The physician is then in a position to clarify the meaning of this disorder with the family and to restore hope for the child's improved behavior and adaptation by making a psychiatric referral when indicated. The history is the most reliable diagnostic indicator. Once a presumptive diagnosis is made, appropriate referral and treatment can follow.

The treatment approach to ADHD should be comprehensive and multimodal and is best managed by a child psychiatrist who has a long-term relationship with the child and family. Psychostimulant medication is often helpful in alleviating the symptoms of ADHD. In addition, academic achievement and emotional well-being require attention to the child's individual style of learning and the maintenance of a supportive interpersonal environment. The child should be monitored carefully over time, since some children who present initially with behavior consistent with ADHD develop the childhood or adolescent form of bipolar disorder.

Although psychostimulant medication should not be prescribed in the ED, the emergency physician should have a familiarity with the commonly used drugs, including methylphenidate (Ritalin), D-amphetamine (Dexedrine), pemoline (Cylert), and Adderall (a mixed salt of a single-entity amphetamine product). Tricyclic antidepressants and Wellbutrin (bupropion) have also been successfully used in the treatment of ADHD, but constitute a second line of medication and are not approved by the FDA for this purpose. In general, response rates to the stimulant medications are higher than 75%. Methylphenidate may be given in median effective dosages of 40 mg/day, but dosages as low as 0.3 mg/kg per/day (10 to 15 mg/day) have been effective. A typical dose of D-amphetamine is 20 mg/day. The principal short-term side effects of the stimulants are appetite suppression and insomnia. The principal concerns of long-term use of stimulants are suppression of weight gain and linear growth. However, rebound growth appears to occur when the medication is discontinued. Because the best indicator of dosage adequacy is classroom performance and not behavior at home, close

communication must be maintained with the child's teacher.

Beyond the use of medication, the effective treatment of ADHD involves a multimodal approach with specific attention to identified areas of concern to child and family. If the child is far behind academically, tutoring or other remediation should be pursued. If a diagnostic evaluation discloses a specific learning disability, individualized educational approaches in the regular classroom or in a resource room may be indicated. Individual and family therapy can help the child learn to make accommodations to the ADHD and can help the parents more effectively normalize family functioning while also improving the child's self-esteem and social skills.

DISORDERS OF INFANCY

Excessive Crying

Excessive crying in an infant is a common presenting complaint in the ED. Parents usually learn to discriminate among the various cries of their baby and to respond with appropriate relief and comfort. By the age of 3 months, crying is generally of short duration and a baby is readily comforted. However, in a few infants, crying without underlying physiologic difficulty continues to be severe and intractable. Parents dealing with this symptom are often exhausted and frustrated, and may also describe difficulties in feeding their infants and significant disruptions in their child's sleep patterns.

An appropriate emergency evaluation includes complete medical history and physical examination to rule out underlying physical causes of distress for the infant, as described in Chapter 17. While examining the baby, the physician may note that the child is easily excitable and difficult to comfort.

In young infants, excessive crying has been called colic. Typically, colic can be expected to decrease with time, ceasing by 3 to 4 months of age. In some infants, however, the crying may persist, often with serious disruptions in parent–infant relationships. Parents in these families may be insecure, and their inability to effectively feed or calm their infants may lead to further insecurity, discouragement, and tension. Parents may be further troubled by impulses to physically harm their babies in response to their frustration. Prematurity and prolonged separation between mother and infant may have been underlying factors in the development of the symptom. Families presenting in the ED may feel isolated and have limited support. Other underlying parental difficulties may include disturbed marital relationships, parental depression, and unsupported single parenthood. The disorder itself is recognized by the child's excitability and the parents' anxiety and tension during the ED evaluation.

Management

Appropriate treatment involves first and foremost an appreciation of the situation and the level of difficulty felt by the parents. Supportive acknowledgment by the emergency physician of the stress confronting the parents and of their genuine desire to be effective with their child is usually greatly appreciated. Statements about the specific temperamental qualities of the child and about how common excessive crying is in infancy are also reassuring.

When the parents are not too exhausted and overwhelmed, or when alternative sources of support and caretaking are available to them, the problem can often be managed on an outpatient basis. Parents should be told not to overstimulate their baby and can be shown effective ways of handling and feeding the child. Follow-up well child care should be arranged so that the family receives intensive support from the primary care physician and allied professionals such as nurse practitioners, visiting nurses, and social workers. With this support, the family can become more effective in interrupting the vicious cycle of infant crying, parents becoming tense, and infant crying again.

In other cases, when a high degree of parental tension and exhaustion from the crying is present, a brief hospitalization of the infant may be necessary. Hospitalization should be proposed in a positive way rather than as a sign of parental defeat. The mother should be asked if she wishes to stay with her infant. If she feels an immediate need for rest at home, that is acceptable as long as the mother agrees to participate actively during the remainder of the child's hospitalization. In general, formula changes and the use of medication are not recommended in the emergency treatment of excessive crying.

Sleep Disorders

Infants and young children have varying patterns of sleeping. Whereas most children during infancy and early childhood sleep through the night, some are noted to wake easily and others to need only small amounts of sleep. It has been estimated that up to 20% of 1-year-old children wake regularly during the night. The exact causes of sleep disruption in young children remain undetermined, but low sensory threshold, high activity level, increased irritability, and adverse emotional factors in the home may lead to the development of disturbed sleep patterns. Concern about an infant's sleep pattern may be an underlying reason for ED visits with minor physical complaints. The emergency physician should be alert to the possibility of sleep disorders when evaluating young children in the ED. Because this problem causes significant anxiety and exhaustion in the parents and can lead to disruptions of maternal–infant attachment, sleep disorders should be appreciated and appropriate treatment should be initiated.

Infants with sleep disorders often have difficulties going to bed or getting to sleep and may often be found to sleep in the parents' bed. These children are also noted to be highly active, extremely intense, irregular in their behavior patterns, and difficult to comfort. The physical examination and medical history is noncontributory with an isolated sleep disturbance; however, when evaluating children with sleep problems, the physician must rule out any associated physical difficulties or underlying medical illness. In some cases, a history of perinatal difficulties or prolonged hospitalization following birth may be obtained. Parental depression and insecurity may also be noted during the ED evaluation.

Management

As in the case of persistent crying, the most important part of the emergency evaluation is identifying the problem and conveying to the parents that, with time and appropriate treatment, the child's sleeping behavior will become more regular and less disruptive to family living. Usually, the identification of the problem and the commitment to develop an effective treatment plan are extremely helpful to parents in the ED. Children with isolated sleep disorders rarely require hospitalization, and the initial approach to therapy should again be referral to ongoing primary care with effective social support. Underlying family difficulties or parental depression may require mental health treatment.

Most children with sleep disorders awaken during the night and return to sleep after comforting. Parents should be advised to avoid overstimulation of the child during the night, and sedative medication is rarely necessary. If extreme parental exhaustion is a problem, the possibility of using alternative caretakers should be investigated, and methods of the parents' sharing the care might be suggested. If sedation is viewed as an absolute necessity by the physician, chloral hydrate (30 mg/kg) or diphenhydramine (1 mg/kg) may be administered to the child. Sustained use of sedative medication should be used only as part of an ongoing treatment program for the infant and family. Chapter 131 includes a discussion of other sleep disorders.

Attachment Disorders

Occasionally, infants will be seen in the ED who are noted to be withdrawn and apathetic. These infants demonstrate severe disturbances of attachment with their primary caretakers, often have feeding disturbances, and may fail to thrive. The most significant disability of these children is a dramatic failure of social development. They do not track with their eyes, and they smile rarely. They do not interact with caretakers in age-appropriate fashion, and facial responsiveness may be entirely absent. The child may be noted to be weak, have poor muscle tone, and a feeble cry. The child demonstrates little spontaneous activity, sleeps excessively, and has a generalized lack of interest in the environment.

The cause of attachment disorders in infants is a continuing lack of adequate caretaking. Features that interfere with maternal-infant bonding are often noted in the history. These may include significant maternal depression and isolation, other maternal incapacitation including substance abuse, maternal indifference toward the infant, history of prolonged separation between mother and infant following birth because of perinatal difficulties, and actual physical abuse. Infants who are temperamentally placid, who make their needs known quietly, often have more difficulties in the presence of maternal depression or maternal preoccupation than more active and responsive infants.

Underlying chronic illness may also lead to the development of social withdrawal and apathy. Also, children with physical problems in infancy may be more difficult to care for and parental reactions to the child's illness may interfere with attachment. Children with mental retardation, although they develop slowly, do not generally demonstrate the profound apathy of the child with an attachment disorder. Furthermore, children with mental retardation receive generally adequate caretaking and do not fail to thrive.

Management

Children with attachment disorders require complete medical evaluation together with careful assessment of their environment. Such children are often seen in the ED for minor physical complaints and may not be receiving regular pediatric care. Thus, the emergency physician must recognize attachment disorders and make effective referrals for ongoing health care for child and family. When parental apathy accompanies severe failure to thrive, hospitalization may be necessary to initiate needed changes and to plan continuing treatment. The physician should recognize that attachment disorders and the associated failure to thrive are often reversible once adequate caretaking is instituted and maintained. If the physician suspects that the child's problems are a result of actual abuse or neglect, this belief should be reported to the appropriate agencies. (Child neglect is discussed in detail in Chapter 128.)

FIRE SETTING

Virtually all of the children in our society develop a fascination with fire and may experiment with it at a relatively early age. For most children, this experimentation is transient and consists mainly of playing with matches or lighting small fires. However, some children may persist with fire-setting behavior and may actually plan to set larger fires that are destructive to both people and property. At this point, the child is demonstrating evidence of a significant psychiatric disorder and requires intensive treatment. The exact incidence of fire setting is unknown, but serious repetitive fire setting is believed to be uncommon in children and adolescents. In general, fire setting is a symptom of serious underlying emotional difficulty and is often associated with other disturbances of behavior and impulse control. Fire setting is also associated with significant anger and aggressiveness on the part of the child. The background of fire-setting children is likely to be highly unstable. These children may have had multiple contacts with social agencies in the past, and some may have been placed outside the home in foster homes or institutions. Although the exact percentage of fire setters with underlying ADHD is unknown, many of these children are described as having been hyperactive, with significant learning problems and long-standing truancy from school.

Management

The clinical manifestations of fire setters in the ED are similar to those of other children with conduct disorders as previously described. Effective ED evaluation should always consider psychiatric hospitalization. As psychiatric hospitalization is arranged, the facility must be informed about the child's previous fire-setting behavior so that appropriate behavior monitoring and safety measures can be employed during the admission.

Suggested Readings

GENERAL

American Psychiatric Association. Quick Reference to the Diagnostic Criteria from DSM-IV. Washington DC: American Psychiatric Association Press, 1994.

Anderson WH. Psychiatric emergencies. In: Wilkins EW, Dineen JJ, Monlure AC, Gross PL, eds. Massachusetts General Hospital Textbook of Emergency Medicine. Baltimore: Williams & Wilkins, 1978.

Bristol JH, Giller E, Dougherty JP. Trends in emergency psychiatry in the last two decades. Am J Psychiatry 1981;138:623.

Dubin W, Weiss K. Handbook of Psychiatric Emergencies. Torrance, CA: Homestead School, 1997.

Gerson S, Bassuk E. Psychiatric emergencies: an overview. Am J Psychiatry 1980;137:1.

Glick RA, Myerson AT, Robbins E, Talbot JA, eds. Psychiatric Emergencies. New York: Grune & Stratton, 1976.

Golan N. Short-term crisis intervention: an approach to serving children and their families. Child Welfare 1972;50:101.

Goodman JD, Sours JA. The Child Mental Status Examination. New York: Basic Books, 1967.

Green WH. Child and Adolescent Clinical Psychopharmacology. 2nd ed. Baltimore: Williams & Wilkins, 1995.

Greenspan SI. The Clinical Interview of the Child. New York: McGraw-Hill, 1981.

Khan AU. Psychiatric Emergencies in Pediatrics. Chicago: Year Book, 1979.

Langsley DG. Crisis intervention. Am J Psychiatry 1972;129:725.

Langsley DG, Kaplan DM. Treatment of Families in Crisis. New York: Grune & Stratton, 1968.

McDaniel KD. Pharmacologic treatment of psychiatric and neurodevelopmental disorders in children and adolescents (3 parts). Clin Pediatr 1986;25:65–71, 143–146, 198–204.

Minuchin S. Families and Family Therapy. Cambridge, MA: Harvard University Press, 1974.

Morrison GC, ed. Emergencies in Child Psychiatry. Springfield, IL: Charles C Thomas, 1975.

Pittman F. Turning Points: Treating Families in Transition and Crisis. New York: Norton, 1987.

Rappaport J. Community Psychology: Values, Research, and Action. New York: Holt, Rinehart & Winston, 1977.

Riddle MA. Child and Adolescent Psychiatric Clinics of North America: Pediatric Psychopharmacology I. Philadelphia: WB Saunders, 1995;4(1).

Riddle MA. Child and Adolescent Psychiatric Clinics of North America: Pediatric Psychopharmacology II. Philadelphia: WB Saunders, 1995;4(2).

Rosenbaum CP, Beeke JE. Psychiatric Treatment: Crisis, Clinic, Consultation. New York: McGraw-Hill, 1975.

Rotenberg MB. Psychiatric emergencies in pediatrics. Pediatr Digest 1979;21:33.

Shader R, ed. Manual of Psychiatric Therapeutics. Boston: Little, Brown, 1975.

Simons J. Psychiatric Examination of Children. 2nd ed. Philadelphia: Lea & Febiger, 1974.

DEPRESSION

Birmaher B, Ryan N, Williamson D, et al. Childhood and adolescent depression: a review of the past 10 years, Part I. J Am Acad Child Adol Psychiatry 1996;35:11, 1427–1439.

Carlson G, Cantwell D. Unmasking masked depression in children and adolescents. Am J Psychiatry 1980;137:445–449.

Cytryn L, McNew D. Proposed classification of childhood depression. Am J Psychiatry 1980;137:22–25.

Cytryn L, McKnew DH Jr, Bunney WE Jr. Diagnosis of depression in children: a reassessment. Am J Psychiatry 1981;138:143–153.

Garrison CZ, Jackson KL, Marsteller F, McKeown R, Addy C. A longitudinal study of depressive symptomatology in young adolescents. J Am Acad Child Adol Psychiatry 1990;29:581–585.

Kashani JH, Husain A, Shekim WO, Hodges KK, Cytryn L. Current perspectives of childhood depression: an overview. Am J Psychiatry 1972;129:149–155.

Kutcher SP, Marton P, Korenblum M. Adolescent bipolar illness and personality disorder. J Am Acad Child Adol Psychiatry 1990;29:355–359.

Petti T. Depression in children, a significant disorder. Psychosomatics 1981;22:444–447.

Renshaw D. Suicide and depression in children. J School Health 1974; 44:487–489.

SUICIDE

Brown LK, Overholser J, Spirito A, Fritz GK. The correlates of planning in adolescent suicide attempts. J Am Acad Child Adol Psychiatry 1991;30:95–99.

Duncan J. The immediate management of suicide attempts in children and adolescents: psychological aspects. J Fam Pract 1977;4:77–80.

Gardner S, with Rosenberg G. Teenage Suicide. New York: Julian Messner (Teen Survival Library), 1985.

Garfinkel BD, Golombek H. Suicide and depression in childhood and adolescence. Can Med Assoc 1974;110:1278–1281.

Henden H. Psychodynamics of suicide, with particular reference to the young. Am J Psychiatry 1991;148:1150–1158.

Hofman A. Adolescents in distress: suicide and out of control behavior. Med Clin North Am 1975;59:1429–1437.

Hyde M, Forsyth E. Suicide: The Hidden Epidemic. 3rd ed. Minneapolis: CompCare, 1986.

Leder J. Dead Serious: A Book for Teenagers about Teenage Suicide. New York: Atheneum, 1987.

Mattson A, et al. Suicidal behavior as child psychiatric emergency. Arch Gen Psychiatry 1969;20:100–109.

McAnarney E. Suicidal behavior of children and youth. Pediatr Clin North Am 1975;22:595–604.

McIntire MS, Angle CR, Schlicht ML. Suicide and self-poisoning in pediatrics. Adv Pediatr 1977;24:291–309.

Pfeffer C. Suicidal behavior of children: a review with implications for research and practice. Am J Psychiatry 1981;138:154–159.

Pfeffer CR, Conte HR, Plutchik R, Jerrett I. Suicidal behavior in latency-age children. J Child Psychol Psychiatry 1981;18:679–692.

Teicher J. A solution to the chronic problem of living: adolescent attempted suicide. In: Schoolar J, ed. Current Issues in Adolescent Psychiatry. New York: Brunner Mazel, 1973;129–147.

Wells C, Stuart I, eds. Self Destructive Behavior in Children. New York: Von Nostrand Reinhold, 1981.

PSYCHOSIS

Geller B, Luby J. Pediatric bipolarity: a review of the past 10 years. J Am Acad Child Adol Psychiatry 1997;36:1168–1176.

Kessler K, Waletsky JP. Clinical use of the antipsychotics. Am J Psychiatry 1981;138:202.

Volkmar FR. Childhood and adolescent psychosis: a review of the past 10 years. J Am Acad Child Adol Psychiatry 1996;35:843–851.

Werry JS, ed. Pediatric Psychopharmacology: The Use of Behavior Modifying Drugs in Children. New York: Brunner Mazel, 1978.

Wiener JM. Psychopharmacology in Childhood and Adolescence. New York: Basic Books, 1977.

POSTTRAUMATIC STRESS DISORDER

Nader K, Pynoos R, Fairbanks L, Frederick C. Children's PTSD reactions one year after a sniper attack at their school. Am J Psychiatry 1990;147:1526–1530.

Pfefferbaum B. Posttraumatic stress disorder in children: a review of the past 10 years. J Am Acad Child Adol Psychiatry 1997;36:1503–1511.

Terr L. Childhood traumas: an outline and overview. Am J Psychiatry 1991;148:10–20.

DISSOCIATIVE DISORDERS

Chu J, Dill D. Dissociative symptoms in relation to childhood physical and sexual abuse. Am J Psychiatry 1990;147:887–892.

Deel P, Eisenhower J. Adolescent multiple personality disorders: a preliminary study of 11 cases. J Am Acad Child Adol Psychiatry 1990; 29:359–366.

SCHOOL REFUSAL

Berger H. Somatic pain and school avoidance. Clin Pediatr 1974;13:819–826.

Bernstein G, Svingen PH, Garfield BD. School phobia: patterns of family functioning. J Am Acad Child Adol Psychiatry 1990;29:24–30.

Last C, Strauss C. School refusal in anxiety-disordered children and adolescents. J Am Acad Child Adol Psychiatry 1990;29:31–35.

Nader PR, Bullock D, Caldwell B. School phobia. Pediatr Clin North Am 1975;22:605–616.

Schmitt B. School phobia—the great imitator: a pediatrician's viewpoint. Pediatrics 1971;48:433–442.

CONDUCT DISORDERS

Boyle M, Offord D. Primary prevention of conduct disorders: issues and prospects. J Am Acad Child Adol Psychiatry 1990;29:227–233.

Coccaro E, Kavoussi R. Fluoxetine and impulsive aggressive behavior in personality-disordered subjects. Arch Gen Psychiatry 1997;54: 1081–1089.

Cox-Jones C, Lubetsky MJ, Fultz SA, Kolko DJ. Inpatient psychiatric treatment of a young recidivist firesetter. J Am Acad Child Adol Psychiatry 1990;29:936–941.

Gruber AR, Heck EG, Mintzer E. Children who set fires: some background and behavioral characteristics. Am J Orthopsychiatry 1981;51:484.

Kashani JH, Husain A, Robins AJ, Reid JC, Wooderson PC. Patterns of delinquency in girls and boys. J Am Acad Child Adol Psychiatry 1980; 19:300.

Klein RG, Abikoff H, Klass E, et al. Clinical efficacy of methylphenidate in conduct disorder with and without attention deficit hyperactivity disorder. Arch Gen Psychiatry 1997;54:1073–1080.

Lewis DO, Shanok SS, Pincus JH, Glaser GH. Violent juvenile delinquents: psychiatric, neurological, psychosocial and abuse factors. J Am Acad Child Adol Psychiatry 1980;17:307.

Loeber R. Antisocial behavior: more enduring than changeable? J Am Acad Child Adol Psychiatry 1991;30:393–397.

Prothrow-Stith D. Deadly Consequences: How Violence is Destroying Our Teenage Population and a Plan to Begin Solving the Problem. New

York: HarperPerennial, 1993.

Rada RT. The violent patient: rapid assessment and management. Psychosomatics 1981;22:101.

Rey J, Plapp J. Quality of perceived parenting in oppositional and conduct disordered adolescents. J Am Acad Child Adol Psychiatry 1990; 29:382–385.

ATTENTION DEFICIT HYPERACTIVITY DISORDERS

Barkley RA. Attention Deficit Hyperactivity Disorder: A Handbook for Diagnosis and Treatment. New York: Guilford Press, 1990.

Barkley RA. Taking Charge of ADHD: The Complete Authoritative Guide for Parents. New York: Guilford Press, 1995.

Cantwell DP. Attention deficit disorder: a review of the past 10 years. Am Acad Child Adolesc Psychiatry 1996;35:978–987.

Saccar C. Drug therapy in the treatment of minimal brain dysfunction. Am J Hosp Pharm 1978;35:544–552.

Safer D, Allen R. Hyperactive Children: Diagnosis and Management. Baltimore: University Park Press, 1976.

Silver LB. Attention-Deficit Hyperactivity Disorder: A Clinical Guide to Diagnosis and Treatment. Washington, DC: American Psychiatric Association Press, 1992.

Wender P. Minimal brain dysfunction in children: diagnosis and management. Pediatr Clin North Am 1973;20:187–202.

INFANCY

Campbell M, Schopler E, Cueva J, Hallin A. Treatment of autistic disorder. J Am Acad Child Adol Psychiatry 1996;35:134–143.

Goldberg C. Outcome in autism and autistic-like conditions. J Am Acad Child Adol Psychiatry 1991;30:375–382.

Rogers S, DiLalla D. Age of symptom onset in young children with pervasive developmental disorders. J Am Acad Child Adol Psychiatry 1990; 29:863–872.

OTHER

Pruett KS, Leonard WF. The screaming baby: treatment of a psychophysiological disorder of infancy. J Am Acad Child Adol Psychiatry 1978;17:289.

Richman N. A community survey of characteristics of one- to two-year-olds with sleep disruptions. J Am Acad Child Adol Psychiatry 1981;20:281.

CHAPTER 130

Adolescent Emergencies

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OVERVIEW

The transition from childhood to adulthood is often a tumultuous process. Adolescents struggle to become independent from parents, to adopt a peer group identity, to think abstractly, and to adopt moral and philosophical values. In addition, they strive to accept a sexual identity, to establish meaningful interpersonal relationships, and to work toward long-term life goals. At the same time, dramatic physical growth occurs because of the complex changes in the hypothalamic–pituitary axis and subsequent increased production of the sex steroids. It is not surprising that when so many psychologic and somatic changes occur simultaneously in one individual presentation for emergency medical attention is likely. Although 80% of adolescents cope well with their development, having only occasional need of emergency medical services, 20% have serious behavioral or medical problems, including conditions that can cause serious injury and threaten their lives.

Adolescents account for approximately 10 to 15% of all emergency department (ED) visits; of these, 13% require acute hospitalization. In one report, 78% of these visits were triaged to the acute and urgent categories. A recent survey of hospital discharges revealed that more than half of all discharges in patients 15 to 24 years of age were a result of pregnancy and its complications, poisoning (substance abuse and suicide gestures/attempts), and trauma (including physical and sexual abuse). Other reasons for ED visits include minor trauma, sexually transmitted diseases, mental health disorders not associated with suicide, and routine visits for acute and chronic illness.

The changes in the health care needs of adolescents during the recent decades is reflected in the causes of their mortality. The death rate for adolescents is 150 per 100,000 with a male:female ratio of 2:1. A decrease in deaths caused by nonsexually transmitted communicable diseases has been accompanied by an increase in deaths associated with “social morbidities” (substance abuse, sexually transmitted diseases, accidents, homicides, suicides, mental health disorders, and eating disorders). As a result of this shift in cause, particularly the rise of violent deaths, no decrease in adolescent mortality has been observed.

Of all deaths of individuals between 15 and 24 years of age, 75% are the result of accidents, homicide, and suicide, with an additional 15 million or more nonfatal injuries. The last three decades have seen a doubling in adolescent homicide and a tripling in the number of suicides. The leading agents of these injuries include motor vehicles, drowning, poisoning, firearms, fires, and falls. Surveys of teens have revealed some insight about the extent they are at risk for some of these health problems. For example, 25% of 12- to 13-year-olds surveyed engaged in at least one health risk such as fighting or tobacco use. One-third of high school students smoke and one-third drink; 25% use marijuana and 3% use cocaine. Alcohol abuse plays a role in 20% of fatalities resulting from motor vehicle collisions, 25% of motor vehicle–pedestrian collisions, and 40% of drownings. In addition, teenagers are estimated to be the victims of 16 to 30% of all perpetrated physical abuse. Recent trends and changes in American family life, including increased incidence of unwed teenage pregnancy, high divorce rate, the deterioration of public education in some segments of our society, and unemployment, undoubtedly contribute to the health care problems of adolescents. These findings indicate that adolescent death and injury are not random events but have an epidemic cause that may be altered by timely appropriate interventions.

Another important influence on the acute medical problems of adolescents is their lack of access to the health care system. The National Health Interview Survey revealed that one in seven adolescents have no health insurance. Adolescents from families that are poor, near poor, or nonwhite families are more likely to be uninsured, as are those from families with little formal education. Economic reasons were most commonly cited for the lack of coverage.

Ten percent of adolescents have no regular source of health care, and 18% identify EDs, outpatient clinics, and city clinics as their only source of health care. When adolescents do have a regular physician, it is usually a family practitioner; the remaining are cared for by internists or pediatricians. Unfortunately, studies indicate that most of these primary care physicians perceive themselves to be deficient in experience, knowledge, and training in the care of

adolescents.

The adolescent may personally contribute to the poor access to health care. Inexperience, denial, and fear may delay the recognition of disease symptoms and the timely seeking of medical care. The anonymous setting of the ED is often preferred by adolescents because they can be treated in certain instances without parental consent and are rarely, if ever, refused. Although the adolescent's acute health care needs may be well served this way, proper follow-up and recognition of chronic problems may be less than adequate.

Thus, for multiple reasons ranging from the individual adolescent's development to physician availability and physician education and training to the adolescent's economic status, many teenagers have difficulty gaining access to the health care system. Adolescents are a medically underserved segment of our population.

Just as children are not just small adults, adolescents are not just large children. Adolescents are prone to a distinct group of diseases and have special medical needs that are much different from those confronted by younger children or adults. Tolmas has eloquently listed some important considerations for medical professionals preparing to care for this group of patients: “a knowledge of the growth and developmental tasks that young people address, a personal interest, respect for confidentiality, honesty and pragmatism in all aspects of behavior and conversation, and a genuine attempt to keep from projecting one's own moral code.” This chapter addresses these considerations by first briefly reviewing some special considerations in the history and physical examination of adolescents, then, in following sections, focusing on some of the specific adolescent problems, including eating disorders, pregnancy and its complications, rape and sexual abuse, suicide, substance abuse, psychosomatic complaints, and violence.

GENERAL CONSIDERATIONS FOR THE HISTORY AND PHYSICAL EXAMINATION

In taking the history, it is important to assess the developmental age of the adolescent and not to simply treat the adolescent on the basis of his or her “physical age.” As for all patients, anticipating the adolescent's fears is also important. Taking a moment to establish rapport, eye contact, patience, and a caring demeanor aids in conducting a successful interview. Medical staff often grossly underestimate patients' concerns regarding health, but more than 50% of adolescents worry about their health. Teens often seek care for health problems they define as physiologic. They avoid seeking care for social or psychiatric concerns, yet stress, depression, and nervousness associated with this period of physiologic change are often translated into somatic complaints. The reason for the visit is thus often intentionally or unintentionally disguised and presented in conventional or acceptable, nonspecific medical complaints because of the adolescent's embarrassment or inability to think abstractly. The clinician must be certain to find the “hidden agenda.”

Because nonspecific complaints may be the harbinger of pregnancy, sexual abuse or incest, physical abuse, significant depression, fear of sexually transmitted disease or homosexuality, or fear of serious illness, at some point during the patient evaluation, the patient should be interviewed without the presence of the parent or other caretaker. The physician must always assure the patient that confidentiality will be maintained but that openness and honesty between patient and physician is a prerequisite for good medical care. This is best accomplished by defining confidentiality at the start of the interview in front of the patient and parent/guardian. The clinician should be familiar with state laws regarding the treatment of minors. Confidentiality is not unqualified. If a teen is in imminent danger, he or she should be informed of the need to disclose this important information to a responsible adult either alone or with the clinician's help. Patients less than 14 years of age should be allowed to choose which responsible adult will be involved in their care plans. The primary physician and/or social work counselor should always be involved when possible to ensure necessary follow-up.

General questions about the adolescent's home and school life are helpful in assessing overall “wellness” of the teen. Table 130.I lists the components of the psychosocial interview, “HEADSS,” providing a framework to identify health risk behaviors. Reviewing components of this list may well be pertinent to the chief complaint that brings the teen to the ED.

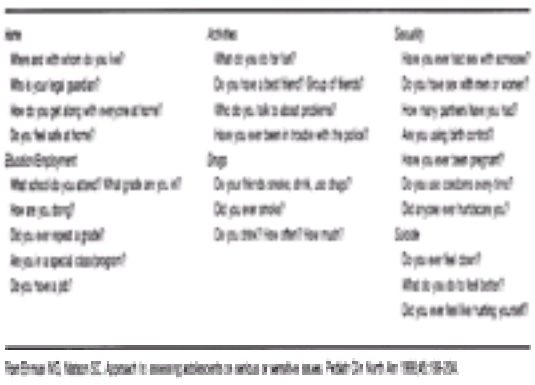


Table 130.1. The Psychosocial Interview

A brief sexual history must be taken for almost every complaint (see Chapter 85 and Chapter 94). Interviewers obtain more reliable information if questions are asked in a matter-of-fact fashion. For example, “What are you using for birth control?” is more likely to be effective in comparison to “Are you sexually active?” or worse, “Are you a virgin?” Adolescents may try to give the “proper” or perceived “desired” answer to such questions.

The physical examination pertinent to the chief complaint should be done with a low threshold for performing a pelvic examination (see Chapter 94). The sexual maturity rating of adolescents set forth by Marshall and Tanner is reviewed in Table 130.2. Figure 130.1, Figure 130.2, Figure 130.3 and Figure 130.4 show the normal progression of healthy adolescent growth. Abnormal progression is a sign of underlying organic disease and requires thorough evaluation.

Therefore, maturity rating becomes important in the evaluation of some adolescent complaints.

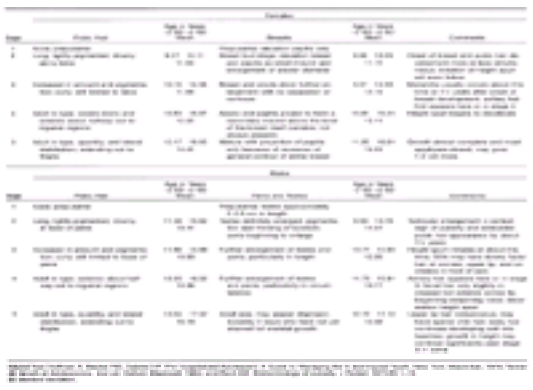


Table 130.2. Pubertal Development Scale

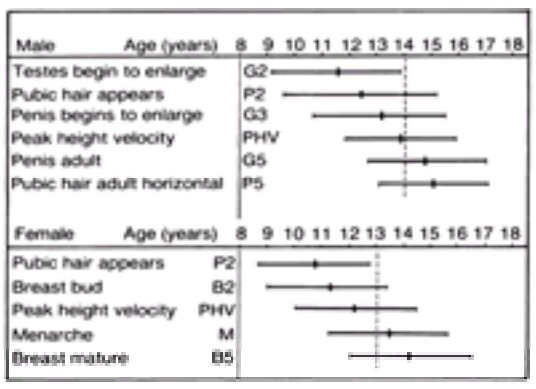


FIGURE 130.1. Normal timing of puberty in boy *(top)* and in girls *(bottom)* in the Zurich Longitudinal Growth study. *Horizontal bars* represent ±2 standard deviations; *vertical marks* represent mean age of stage. *PHV,* peak height velocity. (Reprinted with permission from Rapp C. The adolescent patient. Ann Intern Med 1983;99:52–60.)

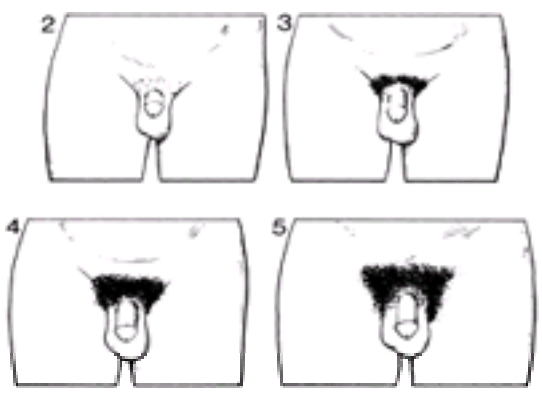


FIGURE 130.2. Stages of pubic hair growth and development of the external genitalia. Numbers in left-hand corner refer to staging according to Tanner. Description of stages of pubic hair: *1,* no pubic hair (not shown); *2,* long, downy pigmented hair at and lateral to the base of the penis; *3,* dark, coarse, curled hair at and lateral to the base of the penis; *4,* abundant adult-type sexual hair limited to the pubic region with no extension to the thighs; *5,* sexual hair is adult-type in quantity and distribution with spread to the medial aspects of the thighs. Description of genitalia stages: *1,* prepubertal; *2,* enlargement of the testes and scrotum, with pigmentation and thinning of the scrotum; *3,* lengthening of the penis, further enlargement of the testes and scrotum; *4,* increase in width and length of the penis, further enlargement of the testes and scrotum, increased pigmentation of the scrotum; *5,* adult size and shape of genitals. (Modified with permission from Marshall WA, Tanner JM. Variations in patterns of pubertal changes in girls. Arch Dis Child 1969;44:291–303; and Root AW. Endocrinology of puberty: 1. Normal sexual maturation. J Pediatr 1973;83:1–19.)

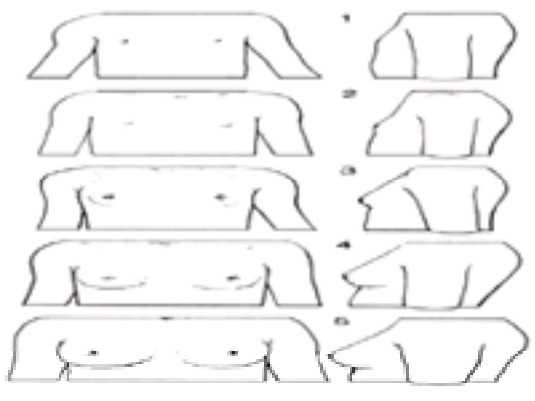


FIGURE 130.3. Stages of breast development in girls. Numbers refer to staging according to Marshall and Tanner. Description of stages: *1,* no breast development; *2,* breast budding widening or areola and elevation on mound of

subareolar tissue, erect papilla; *3,* continued enlargement of breast and widening of areola without separation of their contours; *4,* areola and papilla project above the plane of enlarging breast; *5,* mature breast, areola and breast in same plane, erect papilla. (Modified with permission from Marshall WA, Tanner JM. Variations in patterns of pubertal changes in girls. Arch Dis Child 1969;44:291–303; and Root AW. Endocrinology of puberty: 1. Normal sexual maturation. J Pediatr 1973;83:1–19.)

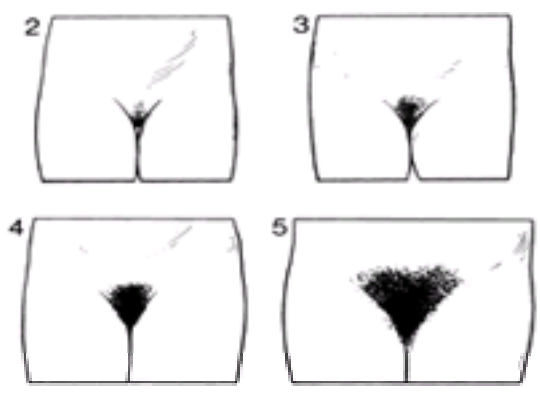


FIGURE 130.4. Stages of pubic hair growth in girls. Numbers in left-hand corner refer to staging according to Marshall and Tanner. Description of stages: *1,* no pubic hair (not shown); *2,* long, pigmented hair over mons veneris or labia majora; *3,* dark, coarse, curled hair spread sparsely over the mons veneris; *4,* abundant, adult-type sexual hair limited to the mons veneris; *5,* sexual hair is adult-type in quantity and distribution with spread to the medial aspect of the thighs. (Modified with permission from Marshall WA, Tanner JM. Variations in patterns of pubertal changes in girls. Arch Dis Child 1969;44:291–303; and Root AW. Endocrinology of puberty: 1. Normal sexual maturation. J Pediatr 1973:83:1–19.)

SPECIFIC DISORDERS

Eating Disorders (Anorexia Nervosa, Bulimia Nervosa)

Background

The prevalence of anorexia nervosa and bulimia among young women appears to be 0.5% and 2%, respectively. These disorders occur in males at one-tenth this rate. All socioeconomic, racial, and ethnic groups are affected.

Although the precise cause of these disorders remains unknown, they are likely a result of a combination of psychologic, social, and biologic factors. Predisposing factors include society's emphasis on thinness, family dysfunction and impaired conflict resolution, a history of sexual or physical abuse, obsessive–compulsive or schizoid personality traits, weight preoccupation, perceptual disturbances, and family history of obesity and/or eating disorders. Precipitating factors include family death, dieting, teasing about weight, demands of puberty, and diminished self-esteem or self-control. The vulnerable individual responds by dieting, which enhances low self-esteem. As starvation continues, performance in school, other activities, and relationships suffer, perpetuating dieting behavior and further reducing sense of control. Binge eating occurs as a response to dieting, reinforcing low self-esteem, loss of control, and more restrictive dieting. Attempts are then made to reduce weight gain from binge eating by self-induced vomiting, laxative and diuretic use, and exercise.

Anorexia nervosa and bulimia carry some of the greatest morbidity and mortality rates of all psychiatric illnesses. Pediatricians and emergency physicians must recognize these diagnoses and be familiar with the management of their medical complications, which are caused by starvation, malnutrition, hypovolemia, electrolyte abnormalities, and purging.

Clinical Manifestations

Anorexia nervosa is characterized by severe weight loss achieved through rigid caloric restriction, a disturbed body perception, and hyperactivity. Bulimia is characterized by secretive eating binges followed by purging (self-induced vomiting and abuse of ipecac, diuretics, and laxatives). In both disorders, the adolescent exhibits an intense preoccupation with food and body weight, fear of fatness, aggressive pursuit of thinness, and loss of normal appetite and impulse control. Diagnoses are confirmed through fulfillment of specific criteria, appearing in the *DSM* IV (1994), rather than through exclusion of other disorders (Table 130.3 and Table 130.4).


A. Refusal to maintain body weight at or above a minimally normal weight for age and height (e.g., weight loss leading to maintenance of body weight less than 85% of that expected; or failure to make expected weight gain during period of growth, leading to body weight less than 85% of that expected).
B. Intense fear of gaining weight or becoming fat, even though underweight.
C. Disturbance in the way in which one's body weight or shape is experienced, undue influence of body weight or shape on self-evaluation, or denial of the seriousness of the current low body weight.
D. In postmenarchal females, amenorrhea (i.e., the absence of at least three consecutive menstrual cycles; a woman is considered to have amenorrhea if her periods occur only following hormone, for example, estrogen, administration.)

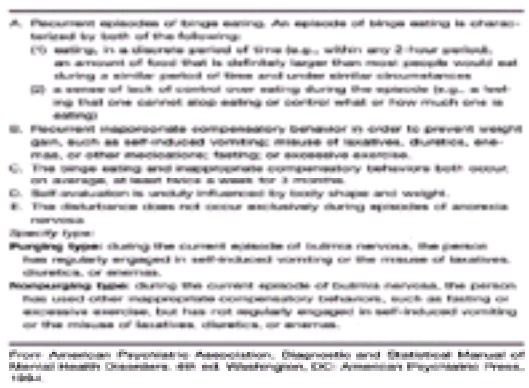
Specify type:

Restricting type: during the current episode of anorexia nervosa, the person has not regularly engaged in binge-eating or purging behavior (i.e., self-induced vomiting or the misuse of laxatives, diuretics, or enemas)

Binge-eating/purging type: during the current episode of anorexia nervosa, the person has regularly engaged in binge eating or purging behavior (i.e., self-induced vomiting or the misuse of laxatives, diuretics, or enemas)

From American Psychiatric Association. Diagnostic and Statistical Manual of Mental Health Disorders, 4th ed. Washington, DC: American Psychiatric Press, 1994.

Table 130.3. DSM-IV Diagnostic Criteria for Anorexia Nervosa



A. Recurrent episodes of binge eating. An episode of binge eating is characterized by both of the following:
   (1) eating, in a discrete period of time (e.g., within any 2-hour period), an amount of food that is definitely larger than most people would eat during a similar period of time and under similar circumstances
   (2) a sense of lack of control over eating during the episode (e.g., a feeling that one cannot stop eating or control what or how much one is eating)
B. Recurrent inappropriate compensatory behavior in order to prevent weight gain, such as self-induced vomiting; misuse of laxatives, diuretics, enemas, or other medications; fasting; or excessive exercise.
C. The binge eating and inappropriate compensatory behaviors both occur, on average, at least twice a week for 3 months.
D. Self-evaluation is unduly influenced by body shape and weight.
E. The disturbance does not occur exclusively during episodes of anorexia nervosa

Specify type:

Purging type: during the current episode of bulimia nervosa, the person has regularly engaged in self-induced vomiting or the misuse of laxatives, diuretics, or enemas.

Nonpurging type: during the current episode of bulimia nervosa, the person has used other inappropriate compensatory behaviors, such as fasting or excessive exercise, but has not regularly engaged in self-induced vomiting or the misuse of laxatives, diuretics, or enemas.

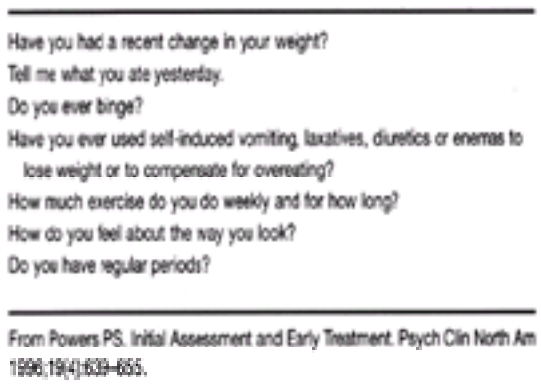
From American Psychiatric Association. Diagnostic and Statistical Manual of Mental Health Disorders. 4th ed. Washington, DC: American Psychiatric Press, 1994.

Table 130.4. DSM-IV Diagnostic Criteria for Bulimia Nervosa

Typically, anorexia nervosa begins in early adolescence; the mean age of onset is 13.75 years. Discrete onset occurs when the individual makes a conscious decision to lose weight, which is then followed by preoccupation with food and the development of profound changes in eating patterns. Classically, the anorexic adolescent is a white female from an upper middle-class family who could be described as intelligent and highly motivated but self-critical and living in a stressful family or social situation. These patients are asexual and detached and often deny their problem by hiding under layers of bulky clothing. The practice of bulimia appears later in adolescence and has a more insidious onset. Multiple attempts at weight reduction lead the patient to become aware of the role of purging in weight control. The bulimic patient is usually heterosexual and outgoing and is distressed by the disorder. Bulimics are more likely to exhibit other behavioral disturbances, such as promiscuity and substance abuse, but they are also more likely to seek and accept help.

Evaluation and Management

Anorexic patients present for medical evaluation with chief complaints related to parental concerns about their behavior and appearance, symptoms associated with the complications of the illness (e.g., syncope or weakness), or complaints that may seem unrelated, such as amenorrhea or pathologic fractures. Alternatively, bulimic adolescents most often seek medical care because of concern for their health, although these concerns are often hidden in a nonspecific complaint such as a mild gastrointestinal disturbance. If an eating disorder is suspected, the diagnosis can be made through careful history taking, focusing on evaluation of eating behavior and weight control. The diagnosis of eating disorders hinges on the presence of an intense fear of fatness and a relentless pursuit of thinness. Case reports of patients diagnosed incorrectly as having eating disorders have lacked these characteristics (Table 130.5).



Have you had a recent change in your weight?
Tell me what you ate yesterday.
Do you ever binge?
Have you ever used self-induced vomiting, laxatives, diuretics or enemas to lose weight or to compensate for overeating?
How much exercise do you do weekly and for how long?
How do you feel about the way you look?
Do you have regular periods?

From Powers PS. Initial Assessment and Early Treatment. Psych Clin North Am 1996;19(4):639–655.

Table 130.5. Important Historical Questions

Other important historical information includes family history of eating and affective disorders and screening for depression. The clinician should query about symptoms associated with purging, including abdominal pain, periodontal disease, sore throat, constipation, diarrhea, polyuria, palpitations, and abdominal discomfort. Other symptoms found in patients with eating disorders include hair loss, bruising, cold intolerance, and light-headedness. The differential diagnosis includes both medical and psychiatric causes. Medical causes of weight loss to be considered include occult malignancy, chronic infection, inflammatory bowel disease, malabsorption, hyperthyroidism, Addison's disease, and diabetes mellitus. Psychiatric causes include affective, obsessive– compulsive, and somatization disorders and schizophrenia.

Patients with eating disorders may manifest a wide variety of physiologic and metabolic derangements (Table 130.6). A complete physical examination is essential and should include height, weight, and orthostatic vital signs. Some physicians prefer the use of body mass index (weight in kg/m^2) for diagnosis in the older adolescent. Body mass index less than 18 is used as criteria for the diagnosis. Bradycardia (heart rate less than 60 beats/minute) and hypotension (blood pressure less than 90/60 mm Hg) is present in 90% of patients with anorexia. Hypothermia and cold intolerance

are not uncommon. The respiratory rate is usually normal.

Neuroendocrine	Delayed puberty Amenorrhea Anovulation Low estrogen state Euthyroid sick state Hypercortisolemia Hypothermia
Cardiovascular	Bradycardia Orthostatic hypotension/shock Arrhythmia Cardiomyopathy Dependent edema
Renal	Hypokalemia Hematuria, proteinuria Acidosis/alkalosis Decreased glomerular filtration rate, elevated blood urea nitrogen Hypercholesterolemia
Gastrointestinal	Constipation Delayed gastric emptying Enamel erosion, palatal trauma Enlarged parotids Esophagitis, Mallory-Weiss
Bone Marrow	Pancytopenia
Musculoskeletal	Weakness Osteopenia/osteoporosis

Adapted from Herzog DB. Eating disorders. N Engl J Med 1988;318:293.

Table 130.6. Physiologic Disturbances in Patients with Eating Disorders

Patients who induce vomiting may have evidence of palatal trauma as well as erosion of the enamel on the lingual surface of the teeth. Non-tender parotid swelling is common in bulimic patients. Skin changes include dry hair, hair loss, dry skin, brittle nails, carotenemia, lanugo, and pre-tibial edema. Patients who induce vomiting may have “Russell's sign,” calluses on the knuckles resulting from chronic trauma (Table 130.7).

	Anorexia	Bulimia
General Characteristics		
Incidence	0.5–1%	2–19%
Age	Early adolescence	Late adolescence
Sex	90–99% female	~80% female
Onset	Distinct	Insidious
Weight	>25% weight loss	Thin to above average
Personality	Model child	Behavioral problems
Social characteristics	Asexual, isolative	Heterosexual, gregarious
Menses	Absent	Absent, irregular
Death	Starvation	Hypokalemia, suicide
Physical Findings		
Skin	Dry, xanthemia	Edema
Hair	Lanugo, scalp and pubic hair loss	Hair loss
Mouth		Enamel erosion Palatal bruising, cuts Enlarged parotid glands Diminished gag reflex Pyorrhea
Endocrinologic	Low estrogen state	
Neurologic		Neuropathy Muscle cramping

Adapted from Herzog DB. Eating disorders. N Engl J Med 1988;318:293.

Table 130.7. Differences In Anorectics and Bulimics

Suggested screening laboratory evaluation includes a complete blood count with indices, electrolytes, blood urea nitrogen and creatinine, calcium, magnesium, phosphates, thyroid function tests, and a baseline electrocardiogram (ECG). More extensive evaluation is dictated by the individual patient's history and physical examination findings. Hematologic abnormalities are common and include leukopenia, normochromic, normocytic anemia, and thrombocytopenia. Electrolyte disorders, including hypokalemia, hypomagnesemia, and hypophosphatemia, are also common. Metabolic alkalosis is present in patients who vomit. Those patients who abuse laxatives may have a metabolic acidosis. Elevated amylase of salivary origin is seen in patients who vomit. ECG abnormalities include sinus bradycardia, non-specific ST-T wave changes, atrial tachycardia, interventricular conduction delay, premature ventricular contractions (PVCs), and prolonged QTc.

Patients who abuse ipecac may develop myopathy as well as cardiomyopathy. Myopathy manifests as weakness, stiffness, and tenderness in peripheral muscles and is reversible. Cardiomyopathy presents as chest pain, tachycardia, hypotension, ECG changes, including T-wave and ST-segment changes, and alterations in the QRS, P-R, and QTc intervals.

Acute medical management includes restoring the intravascular volume with saline solution, correcting symptomatic electrolyte imbalance, and closely monitoring vital signs. Once the diagnosis is confirmed, psychiatric evaluation is necessary with referral to a definitive, multidisciplinary treatment program. Guidelines for hospitalization appear in Table 130.8. This treatment may occur in a pediatric or psychiatric ward, depending on the patient's needs and ward capabilities. Treatment is a multifactorial endeavor, including medical, behavioral, personal, and family therapy. The first and most difficult task is to convince the adolescent that he or she needs help and that treatment will relieve the fears and obsessions, making it easier to return to normal activities.

A. Presence of severe or persistent medical complication that threatens life or health. Presence of at least one major and three minor, or at least six major complications.	
Major Complication	Weight <75% ideal body weight (IBW) Hypoglycemic syncope Severe fluid/electrolyte imbalance Cardiac arrhythmia Severe dehydration
Minor Complication	Weight <85% IBW Recalcitrant vomiting Bradycardia Hypothermia Hypotension Lanugo Amenorrhea for three consecutive cycles Acute starvation Vasomotor instability Minor electrolyte imbalance or hypoglycemia Hypophosphatemia Nutritional anemia Exercise-induced injury Impaired anal function Intestinal atony Failure of 2 months weekly outpatient management

From: Herzog DB, Bierach EV. Anorexia nervosa. In: Mash Lewis, ed. Child & Adolescent Psychiatry: A Comprehensive Textbook. 2nd ed. Baltimore: Williams & Wilkins. 1996.

Table 130.8. Guidelines for Hospitalization of Anorexic Patients

Mortality remains significant. In a 5-year post-hospitalization follow-up of anorexic women, 70 to 75% had moderate symptomatic improvement, 20% were chronically ill, and 5% had died. Depression, suicide attempts, perfectionism, and obsessive thinking and compulsive behaviors were seen. A significant number have affective or anxiety disorders at follow-up.

Pregnancy

Background

Fifty-five percent of 15 to 19 year olds and 8% of girls less than 15 years of age have had sexual intercourse, leaving them at risk for pregnancy and sexually transmitted diseases. In the latter half of the 1980s, there were 95.9 pregnancies per 1000 teenaged girls 15 to 19 years, representing a 9% increase. Abortion rates remained stable at 36.0 per 1000 and birth rates increased by 18% to 59.9 per 1000. Most of these pregnancies are unplanned and unexpected. The United States continues to have one of the highest rates of teenage pregnancy and birth rates of all developed countries.

Pregnancy in the adolescent years has tremendous health and socioeconomic effects on the mother as well as the child. Teen mothers are less likely to complete school and more likely to have low-paying jobs or be unemployed, and they are more likely to live at or below the national poverty level. Children of teen parents are at risk for adverse cognitive and psychologic effects as well as abuse. Health risks to the mother and fetus are most likely a result of inadequate prenatal care secondary to late diagnosis and economic constraints. Adolescents often seek the anonymous setting of the ED for treatment of problems related to pregnancy. The physician caring for these patients should be prepared to diagnose pregnancy, manage its complications, and arrange appropriate follow-up.

Clinical Manifestations

The adolescent may present with a concern of pregnancy or with a variety of chief complaints that often seem unrelated to the underlying pregnancy. Adolescents presenting to the ED are less likely to complain of amenorrhea, breast tenderness or enlargement, urinary frequency, or morning sickness. The information that they offer concerning menstrual cycle, sexual activity, and birth control is often inaccurate. Thus, the evaluating physician should have a low threshold for performing pregnancy tests for postmenarchal females, especially if diagnostic or therapeutic procedures are performed or medications administered. Making an early diagnosis and establishing appropriate referrals for care is in the best interests of the mother and the fetus.

During pregnancy, b-human chorionic gonadotropin hormone (b-hCG) is secreted by the trophoblast 9 to 11 days after ovulation. Concentration of this hormone doubles approximately every 1 to 3 days, reaching about 100 mIU/mL at the time of the first missed menses. An enzyme-linked immunoassay specific for the b subunit of hCG is available and offers virtually 100% sensitivity and specificity. This test is inexpensive, easy to perform, and detects hCG as low as 20 mIU/mL in the urine. The serum b-hCG is slightly more sensitive and should be obtained to confirm equivocal urinary pregnancy test results or when quantitation is necessary.

Evaluation and Management

A physical examination and a pelvic examination should be performed in patients with positive test results. The date of the last menstrual period and uterine size are used to estimate gestational age. Notable findings include enlarged breasts, galactorrhea, palpation of fetal movement and heart tones, palpable fundus on abdominal examination, palpable softening of the corpus (Hegar's sign), softening of the cervix (Goodell's sign), congested, bluish hue to the vaginal mucosa and cervix (Chadwick's sign), and palpable enlargement of the fundus on pelvic examination. The nongravid uterus approximates the size of a lemon. The 8-week gravid uterus approximates the size of an orange, and a 12-week gravid uterus is the size of a grapefruit and is just palpable at the pubic symphysis. At 16 weeks, the uterus is palpable midway between the pubic symphysis and the umbilicus, and at 20 weeks is at the level of the umbilicus. A gestational sac can be detected at 5 to 6 weeks by abdominal ultrasound and by radiography at 16 weeks. Routine pelvic cultures should be sent and hemoglobin and rapid plasma reagin (RPR) should be obtained at the time of the initial diagnosis.

Informing the adolescent of results of pregnancy testing should be done privately, without the parent. During this interview, the clinician should identify a support person for the adolescent, elicit feelings regarding the pregnancy, and attempt to disclose incest or rape. Most often, the adolescent chooses to involve an adult to help her through the crisis. If she refuses, it then becomes an individual decision to break confidentiality. The adolescent should then be introduced to the options regarding the management of her pregnancy, which depend on the estimated gestational dates. These options include keeping the baby, putting the baby up for adoption, or terminating the pregnancy through first (less than 13 weeks) and second (less than 18 weeks) trimester abortion. When possible, the father should be involved in the discussion if the mother so chooses. Above all, the clinician should emphasize that the ultimate decision regarding the pregnancy belongs to the adolescent mother alone.

Appropriate referral and follow-up of this high-risk group of patients is essential in providing the best possible patient care. Ideally, a follow-up within a few days to answer questions and to reach and act on a decision may help keep the adolescent within the health care system. The younger the pregnant adolescent is, the less likely she is to have prenatal care beginning in the first trimester. Therefore, to increase compliance and decrease the decisions that the adolescent has to make, an appointment should be scheduled for the adolescent when possible. This follow-up appointment does not necessarily have to occur in the ED, but ideally it should happen. An experienced, knowledgeable social worker is

invaluable and each ED should establish a reliable follow-up mechanism for these patients. Figure 130.5 provides an example of patient diagnosis and referral.

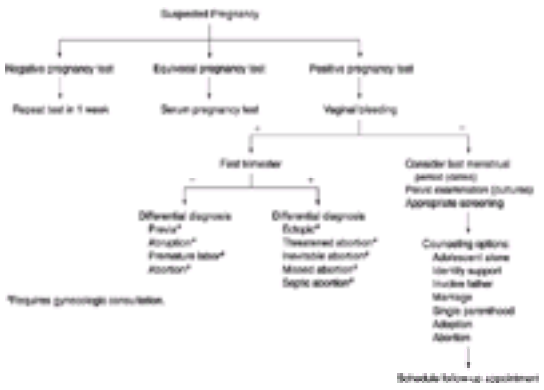


FIGURE 130.5. The pregnant adolescent.

Complications of pregnancy include hyperemesis, premature labor, placenta previa and abruptio, abortion (threatened, inevitable, incomplete, or complete), infection, and ectopic pregnancy. Patients presenting with signs and symptoms of these complications should be thoroughly evaluated by the emergency physician. When necessary, emergency gynecologic evaluation should be performed in the pediatric ED or the patient should be transferred to an appropriate facility in a timely fashion.

Concomitant with the sexual revolution, there has been an epidemic of sexually transmitted diseases, resulting in an increase in ectopic pregnancy. The frequency of this diagnosis has tripled over the past two decades. This increase is presumed to be caused by an increased incidence of tubal infection, a change in microbiology of infection (with the emergence of *Chlamydia*), iatrogenic causes secondary to fertility control (intrauterine device, tubal ligation, minipill), and an increase in recognition. Of patients with ectopic pregnancies, 50% have evidence of chronic salpingitis.

Ectopic pregnancy remains the third leading cause of maternal death, largely because of missed diagnosis. The classic triad of abdominal pain, abnormal vaginal bleeding, and adnexal mass are pathognomonic for this entity but are rarely present, especially during early pregnancy. However, 90% of patients complain of abdominal or pelvic pain and vaginal bleeding.

The earlier these patients present for evaluation, the more subtle their symptoms. The patient presenting with a ruptured ectopic pregnancy usually exhibits signs of intravascular volume depletion or shock, along with signs of intraperitoneal hemorrhage. These patients require acute resuscitation with attention to the ABCs (airway, breathing, and circulation), volume resuscitation, and immediate surgical intervention.

Patients with vaginal bleeding and pregnancy need a thorough evaluation so that the diagnoses of normal versus ectopic pregnancy; threatened, missed, or complete abortion; and placenta previa or abruption can be distinguished from each other (Fig. 130.5). The patient presenting with vaginal bleeding in the third trimester has placental abruption or previa until proven otherwise. No pelvic examination should be done in these patients. Oxygen and intravenous fluids should be administered and immediate gynecologic evaluation should be obtained. Occasionally, a patient in active labor may arrive in the ED and no obstetrician will be available immediately. In most instances, the mother delivers the infant without problems. For abnormal presentations, a textbook of gynecology/obstetrics should be available for reference.

Rape/Sexual Abuse

Background

Rape is defined as genital contact without consent, through use or threat of use of force or fraud, or when the victim is unable to give consent because of physical or mental disability. Statutory rape is intercourse with a female below the age of consent, which is considered to be 16 years in most states. The term *incest* applies to the situation in which the assailant and victim are related and therefore could not legally marry or have a functional situation simulating this relationship. *Sexual abuse* or *molestation,* a broader term, is involvement of the adolescent in activities that he or she does not fully comprehend. These include exhibitionism, fondling, oral–genital contact, and rectal or vaginal penetration. Incest is probably the most common form of adolescent abuse and is the most underreported and difficult to prove.

An estimated one of every four adolescents will be victims of sexual abuse before they reach adulthood. Most reported crimes are committed against females; however, it is estimated that for every two female victims of sexual abuse, there is one male. These figures represent only the tip of the iceberg because less than 50% of these crimes are reported. More than 50% of all rape victims are younger than 19 years, with a peak age from 14 to 17 years. Of all rape/homicides, 25% occur in adolescents younger than 18 years old. In the National Adolescent Student Health Survey (NASHS), 12% of eighth graders and tenth graders reported that someone had raped them or tried to rape them outside of school, and 18% of them said someone tried to force them to have sex. Thus, because of the magnitude of this problem in the pediatric age group, the pediatrician as well as the emergency physician should develop skill in recognition and appropriate workup, and have an organized system for patient follow-up. Most cities have special programs and designated rape and child abuse centers that can assist victims of assault and can offer advice to physicians regarding acute management. Physicians practicing in a particular area should investigate this topic and be knowledgeable of the state laws regarding the care of adolescents.

Clinical Manifestations

Adolescent victims present in one of two ways, either after an acute event with an unknown perpetrator or after an acute stress when an incestual relationship is revealed. All patients who have been assaulted within the previous 72 hours or those with symptoms should be evaluated on an emergency basis. Asymptomatic patients with ongoing chronic abuse (not occurring in the preceding 72 hours) can be scheduled in special outpatient clinics designed to care for this particular group. These patients require a multidisciplinary approach, including physicians, social workers, nurses, and psychologists, to provide optimal care. The aim of the acute intervention is to obtain the details of the incident, to perform a complete medical evaluation, to collect medicolegal evidence, and to provide appropriate medical and psychological follow-up. The acute intervention is not intended to ascertain whether a crime was committed. All suspected cases of abuse/rape must be reported to the police (see Chapter 128).

Evaluation and Management

Consent for interview, examination, and collection of evidence should be obtained from the victim. Accurate and thorough records are of the utmost importance. The victim should be given the control of the situation and has the right to terminate the process at any point. A supportive female or male, or ideally, an experienced rape counselor/nurse, should be present throughout the interview and examination. When possible, this process should take place in a secluded, quiet area of the ED.

Most facilities use a detailed information sheet and protocol for the evaluation of these patients. Figure 130.6 provides a sample from the state of California as an example. The history should include the details of the event, including time and place, the details of the sexual acts (including whether oral, rectal, or vaginal penetration occurred), whether or not ejaculation occurred, the force or threat that was used by the assailant, and the associated use of alcohol or drugs. Hygiene, including bathing, douche, or change of clothes after the event, should be recorded. Medical history, including menstrual history, sexual activity, previous history of sexually transmitted diseases, birth control use, last intercourse, previous obstetric and gynecologic history, and history of health problems, should be documented.

FIGURE 130.6. Example of medical report form for suspected sexual assault. (Reprinted with permission from State of California, Office of Criminal Justice Planning.)

As many as 46% of victims have non-genital injuries, with as many as 15% of them requiring therapy and follow-up. Up to 80% of victims have minor genital injuries, most of which are external and involve the posterior fourchette. These observations emphasize the need for a complete, careful examination. Photographs should be taken with the victim's consent, when possible. The taking of photographs does not preclude documentation of existing injuries on the patient's chart. A description of the patient's general appearance and emotional status, and description of clothing condition begins the physical examination. The victim's clothing should be placed in a paper bag for use as evidence. The physician should explain the parts of the examination before performing them to allow the patient to assume control and to alleviate as much anxiety as possible. A complete physical examination to assess for bruises, scratches, and sexual maturity rating should be performed. Debris under the fingernails should be removed with a wooden curette and placed in a labeled envelope. Dried semen should be removed with saline-moistened swabs and stored in dry test tubes. The use of a Wood's lamp may aid in identifying these areas. Matted pubic hair should be removed and the remaining pubic hair gently combed into a labeled envelope. A sample of the patient's pubic hair should be included in the evidence. A careful external examination is paramount, and the hymen and the posterior forchette should be carefully inspected for areas of laceration.

A speculum examination follows to assess for the presence of vaginal and cervical trauma. Lubricants should be avoided because they affect sperm motility and culture results. If secretions are present in the posterior fornix, they should be aspirated and placed in a sterile container for sperm and acid phosphatase detection. Cotton swabbings taken from the posterior fornix should be used to make slides for detection of motile sperm, acid phosphatase, and blood group antigen. Cultures for gonorrhea and *Chlamydia* and wet mount should also be obtained and the remainder of the pelvic examination be completed in the usual manner (see Chapter 94). The rectum and oral cavity should be carefully examined. If there is history or evidence of trauma, a swab from the gum line and buccal mucosa can also be evaluated for the presence of sperm. Next, the victim should be asked to chew on a piece of filter paper or a cotton ball so a sample of saliva may be obtained. Eighty percent of people secrete blood group antigens in their saliva, sweat, and other body fluids. Finally, serum for syphilis testing should be obtained. An additional serum sample may be obtained and stored and may be of help in the event of future diagnosis of venereal disease.

The risk of venereal disease ranges from 1 to 27%. Studies have shown that 2 to 12% of adolescents at the initial visit

have gonorrhea and 1.5 to 10% have *Chlamydia.* On follow-up visit, 1 to 3% have positive cultures. The risk of trichomonas and bacterial vaginosis ranges from 5 to 25%. Thus, most physicians treat their patients prophylactically. Options include a single-dose ceftriaxone, cefixime, or ciprofloxacin followed by 1 g of azithromax or by a course of doxycycline for 7 to 10 days. Alternatively, a single dose of 2 g of azithromax may be given. The risk of human papilloma-virus (HPV) is unknown; however, the proportion of patients having abnormal Papanicolaou smears ranges from 3 to 27%. The risk of herpes simplex virus (HSV), syphilis, and human immunodeficiency virus (HIV) is low. Follow-up of the patient 7 days after the initial examination is recommended by the Centers for Disease Control and Prevention for repeat vaginal specimens for gonorrhea, *Chlamydia,* and *Trichomonas.* A test for syphilis should be repeated at 6 to 8 weeks. Testing for HIV should occur only after counseling, at 3 to 6 months after the incident, and if negative, again at 1 year. The risk of transmission is low but may increase along with the increasing prevalence of the disease. Azidothymidine prophylaxis may be recommended for high-risk patients, but no standard guidelines currently exist.

All patients should have a sensitive pregnancy test performed. Pregnancy occurs as a result of rape in 1% of victims. If the patient is not at risk for early pregnancy, a protocol for pregnancy prevention may be followed. The most accepted regimen is the use of Ovral, two tablets initially followed by two tablets 12 hours later. This treatment should be given within 72 hours of the event. Finally, all patients should have scheduled medical and psychological follow-up before discharge from the ED.

Teenage Suicide

Background

Every year, 2000 children younger than 19 years of age commit suicide. This statistic represents an underestimation of the true occurrence because of underreporting and lack of recognition. Although suicide rates for adolescents are among the lowest compared with other age groups, the rate of suicide in the age group from 10 to 24 years has tripled in the last three decades and now ranks as the third leading cause of adolescent death. The rate of attempted suicides far exceeds this number; reports range from 50 to 200 attempts per one successful suicide. Males are three times more likely to successfully commit suicide and females are three times more likely to attempt suicide. White males are in the highest risk group. Firearms and explosives are used by 60% of successful male and 40% of female adolescent suicides, poisoning is used in 15 to 20% of male and 25 to 35% of female suicides, and suffocation by hanging, drowning, or strangulation is used in 15% of males and 10% of females. Drug overdose accounts for most suicide attempts presenting to the ED. Although an estimated 1 to 12% of all ED visits are secondary to suicide attempts, the adolescent proportion is closer to the higher estimation. As many as 9% of adolescents surveyed have made a suicide attempt, but less than half of these attempts reach the attention of the psychiatric professional. Thus, many of the existing studies report results in biased population samples; namely, those seeking care.

Many patients seek general medical care shortly before attempting suicide. Hawton reported that 50% of youths saw physicians 1 month prior and 25% within 1 week of attempting suicide. Slap and Vorters recently reported that suicidal adolescents had poorer mental health, impulse control, family relationships, and school performance than their ill counterparts, and they were also most likely to depend on the ED for their care. They identified five risk factors: 1) previous suicide attempts, 2) previous mental health care, 3) poor school performance, 4) marijuana use, and 5) dependence on the ED for primary care in 88% of the suicidal teenagers compared with 55% of other ill adolescents. Perhaps these factors, when prospectively studied, will help identify the adolescents who are at risk.

Much has been written regarding the risk factors having high association with adolescent suicide. The severity of the injury and the likelihood of rescue directly parallel the seriousness of the attempt. Other risk factors include the presence of a chaotic, disorganized family environment, poor support systems, a family history of suicidal behavior, psychiatric disorders or chronic substance abuse, and history of physical or sexual abuse. Patients with an underlying mental disorder have a substantially higher risk of both attempting and committing suicide. Disorders with highest risk include major affective disorders and schizophrenia. Patients who have had a previous hospitalization for their illness or who practice substance abuse also seem to have a higher risk. Male homosexuals as well as lesbians have a higher rate of suicide than their heterosexual counterparts. The attempted suicide rate among male homosexuals has been reported as high as 30%. Previous suicide attempts and continued suicidal ideation are worrisome. Patients who are hopeless and have low self-esteem are at higher risk (see Chapter 129).

Evaluation and Management

Many adolescents arriving in the ED after a suicide attempt require acute stabilization and treatment following purposeful ingestions or self-inflicted violence (see Chapter 88 and Section IV). The primary goals of the emergency physician are assessment, stabilization, recognition, and appropriate disposition. The physician is responsible for preventing the patient from leaving the medical facility before a full evaluation has been completed. In rare instances, patients might require physical restraint. However, most do well with one-on-one observation by a responsible individual. Regardless of the means, patient safety is of utmost importance and requires continued reassessment.

After initial assessment and stabilization, management should focus on 1) establishing whether suicidal ideation persists; 2) the intent of the patient and assessment of lethality of the means, degree of access, and risk:rescue ratio; 3) the patient's current mental status and existing signs of an underlying mental health disorder; and 4) the crisis that precipitated the event and the social situation and support system. Hospitalization may be required for medical treatment before this evaluation. Ideally, all patients should have an evaluation by a psychiatrist and a social worker. High-risk cases may benefit from a brief inpatient hospitalization, whereas others may do well with outpatient treatment. If a psychiatrist is not available to the patient, the emergency physician must rely on the social worker, the primary care physician, and the available mental health services in the area. Before discharge, a safety contract should be discussed with the patient and family. In addition, education directed toward the caretakers to limit access to means of suicide by removing firearms, medications, and so forth from the home environment is crucial. A follow-up system to ensure that the

patient and family have established access to mental health care is also important.

Substance Abuse

Background

Drug abuse is defined as the pathologic use of a substance that results in impairment in social, psychological, physical, or occupational functioning. The adolescent population is a polysubstance abusing group, with the highest users between ages 18 and 25 years. Long-term studies have revealed that the risk for abuse of drugs other than marijuana and alcohol decreases if the use is postponed to the later adolescent years. The National Institute on Drug Abuse (NIDA) has provided cross-sectional surveys from high school seniors since 1975 and has continually documented the magnitude of the problem. Use of illicit drugs increased dramatically in the 1970s and, fortunately, has decreased some recently. However, the use of drugs has been incorporated into our society; new substances continue to appear and overall use remains unacceptably high.

The most common substances abused by adolescents include alcohol, cigarettes, marijuana, stimulants, cocaine, inhalants, hallucinogens, tranquilizers, and sedatives. Adolescents abuse drugs for many reasons, including experimentation, low self-esteem, peer or family use, relief of stress or anxiety, and escape from parental constraints.

Clinical Manifestations

The most common reasons for these patients to come to the ED include acute intoxication, associated trauma, a frightening reaction to the ingested drug, drug-seeking behavior disguised under another complaint, and parental request for drug screening. If the adolescent does not present with symptoms of acute intoxication, only direct questioning regarding abuse may reveal the underlying problem. Again, beginning the discussion by inquiring about the use of drugs at parties, the attitude toward drugs, and the use of drugs by family members and close peers is often helpful. Determining the quantity and frequency of drug use and the degree of life disruption that has occurred is also helpful. Questions regarding physical symptoms of withdrawal, history of blackouts or accidents associated with intoxication, a fall in school performance and in extracurricular activities, arrests or juvenile court records, and conflict with family and peers define the severity of the problem (Table 130.9). Identifying significant depression in this group is also important because this diagnosis changes the management plan for the patient.

Medical	*Legal*
Blackouts	Arrests
Withdrawal symptoms	Friends with criminal background
Adverse reactions	Violence
Injury while intoxicated	*Family/Peers*
School	Social isolation
Fall in performance	Conflict with parents over drug use
Fall in extracurricular activities	Substance abuse in family, peers
Use of drugs in school	
Behavior problems	

From Farrow JA. Adolescent chemical dependency. Med Clin North Am 1990; 74:1265–1274.

Table 130.9. Substance Abuse History

Evaluation and Management

The principles of managing the acutely intoxicated patient are covered in depth in Chapter 88. The patient experiencing a "bad trip" may experience hallucinations or exhibit paranoia. These patients should be cared for in the quietest section of the ED, and the team should approach them in a calm and reassuring fashion. The potentially harmful or "out of control" patient presents another challenge. If the patient is armed, only a trained security guard should assume the task of removing the weapon. If reassuring the patient fails, physical restraint may be necessary. Safety is best achieved in numbers, and the general approach is to assign security guards or other personnel to each limb. The show of numbers is often enough to control the patient's behavior. Four-point leather restraints are used to secure the patient to a stretcher, taking care to avoid cardiorespiratory compromise and undue trauma. Some physicians prefer to restrain a patient on his or her side to avoid airway complications in case of vomiting. If the patient requires restraint, the clinician must remember to perform a thorough physical examination so that other problems that could cause such altered mental status are not overlooked. Occasionally, the patient may require sedation; the medication chosen depends on the drug intoxication (see Chapter 88).

Urine screening at the parent's request should be avoided because this procedure creates an adversarial situation between the medical staff and the adolescent. The possibility of counseling should be offered instead. Unfortunately, many adverse reactions resulting from drug abuse never come to medical attention. For those that do, treatment of the acute intoxication does not constitute definitive treatment. In general, confidentiality is the rule, and information given to the physician during the interview is kept confidential, unless the adolescent allows the physician to disclose the information or the degree of substance abuse poses a significant threat to the child's health.

Counseling begins in the ED. The patient with symptoms of acute intoxication as well as the adolescent presenting with only a minor manifestation of a drug reaction (i.e., a dystonic reaction) undoubtedly need further treatment, with definitive referral to an established program. A skilled social worker, drug counselor, or psychologist can be indispensable in beginning the counseling process in the ED. Before discharge, the patient should be evaluated for symptoms of major

depression or suicidal ideation because these may go hand in hand with substance abuse. The responsibility of the emergency physician and pediatrician includes a knowledge of the trends of adolescent drug abuse, the ability to impart knowledge and identify problem behavior, and a knowledge of the available community resources for drug treatment programs so that a proper referral can be made.

Psychosomatic Complaints

Background

Patients go to see doctors for many reasons. Some seek diagnosis and treatment of physiologic disease and informational needs. Other visits result from psychosocial illness secondary to life stresses, psychiatric disorders, and social isolation. A relationship between stress and recurrent pain has been identified clearly in the pediatric and adolescent population. Adolescents cope with life stresses in many ways. Adapting a sick role provides an acceptable means of receiving attention. Therefore, adolescents often present with chronic complaints that are not attributable to an underlying organic disorder but rather are related to psychosocial factors. Studies conducted in ambulatory care settings have repeatedly shown that up to 68% of these patients have no underlying physical disorder. Because malingering is rare in this age group, the physician's task is often one of diagnosing underlying affective disorders or eliciting current stresses in the patient's life.

Clinical Manifestations

The most common complaints include vague abdominal pain, chest pain, chronic fatigue, and headache.

Evaluation and Treatment

The evaluation of psychosomatic complaints is part of daily medical practice and does not exclude the practice of emergency medicine. The goal of treatment of these patients is screening through thorough medical and psychosocial history and physical examination. The possibility of significant disease is eliminated with a minimum of laboratory tests. A discussion with the adolescent regarding the role of stress in somatic symptoms follows along with gentle reassurance and appropriate referral, allowing the patient to reliably enter the health care system. The initial diagnosis is made with caution, and repeated observation is important to avoid missing other diagnoses.

Screening for known stressors may help in making the diagnosis of psychosomatic illness. These stressors include the onset of symptoms and associated events, the pattern of past visits, including multiple ED visits, a history of many somatic complaints, a strong family history for somatization, and the developmental stage of the adolescent. Unfortunately, these patients often undergo many tests that serve only to eliminate nonpsychosomatic possibilities and reinforce symptoms. The discussion between the physician and adolescent after the initial evaluation should include a summary of the physical examination and laboratory results and the reassurance that nothing serious seems to be wrong. An attempt should be made to link the presence of certain physiologic responses to stresses related to a current situation, along with attempts to recognize and remove the stress or to deal with it in another way. Regular scheduled appointments alleviate the patient's need to develop symptoms to see the doctor. Frequent physical examination and discussion can reduce the amount of costly and/or risky diagnostic tests or procedures.

People cope with life stresses in many ways, and adopting a sick role allows the individual to relinquish responsibility and provides an acceptable means for gaining attention and sympathy. Most patients with diagnosable psychiatric disorders still receive their care from medical physicians in this country. Socially isolated patients visit the doctor for interpersonal exchange to gain a sense of belonging. Patients distressed by physical symptoms often seek information as a method of coping with their symptoms and reducing the associated anxiety.

Chest Pain

Although chest pain is a common complaint in adolescents, it is rarely caused by serious disease. However, a recent prospective study of adolescents with chest pain, evaluated in an ambulatory setting by Pantell and Goodman, revealed that two-thirds of these patients restricted their activities and 40% had absences from school. Causes were found in 57% of the patients and included musculoskeletal pain (31%), hyperventilation (20%), breast-related problems (5%), respiratory disease (2%), gastrointestinal disease (2%), and cardiac disease (1%). No definite cause was found in the remaining 43% of patients. The typical teen complained of intermittent, frequent, sharp, substernal pain that had occurred over several months, and physical examination was normal except for occasional tenderness on chest wall palpation. More than half of the patients experienced significant anxiety concerning their illness, fearing a serious medical disorder. Approximately one-third of the patients had experienced a stressful life event in the preceding months. Multiple other existing reviews of children and adolescents report similar findings.

Most patients with chest pain with an identifiable cause have well-localized, well-defined, sharp pain consistent with anterior chest wall syndrome. This pain is not associated with exertion but rather with movement. This syndrome may be thought of to include costochondritis, Tietze's syndrome, slipping rib syndrome, and muscular strain. Pulmonary-related causes include pneumonia, asthma, pleurodynia, pulmonary hypertension, pulmonary embolus, and pneumothorax. Pain that is exertional or that is associated with palpitations, shortness of breath, dizziness, or syncope suggests a cardiac cause. This event is rare. Conditions to consider include myocarditis/pericarditis, aortic outlet obstruction, mitral valve prolapse, and anomalous coronary artery. Gastrointestinal causes include gastroesophageal reflux, ulcer disease, pancreatitis, and gall bladder or liver disease. Patients without an identifiable cause often have a history of recent stress.

Evaluation of patients presenting to the ED complaining of chest pain includes a history and physical examination and treatment of medical emergencies. The extent of diagnostic workup should be tailored to the individual patient (see Chapter 52 and Chapter 82). The goal of management with the remaining patients is to provide symptomatic relief,

reassurance, and appropriate long-term referral.

Hyperventilation Syndrome

In response to a stress, b-adrenergic stimulation results in hyperventilation and hypocapnic alkalosis. Underlying anxiety and stress are presumed to be the culprit. Some evidence suggests that this disease is biochemical; in susceptible patients, symptoms can be reproduced by infusions of sodium lactate and can be alleviated by b-blockers. The anxiety and fear created by the symptoms of hyperventilation that remain unrecognized by physicians and family are enough of a stress to perpetuate the chronic syndrome.

Clinical manifestations include breathlessness, fatigue, sleep disturbance, poor concentration, paresthesia, syncope, tunnel vision, chest pain, palpitations, tachycardia, bloating, belching, muscle stiffness, cramps, and carpopedal spasms. Observation reveals excessive thoracic muscle use, frequent yawning, coughing, clearing of the throat, and moistening of the lips. The presence of these symptoms should alert the physician to underlying family problems or other stresses leading to undue anxiety in the adolescent. These patients may present with a complaint of syncope or chest pain, and other causes must be considered (see Chapter 52 and Chapter 73).

Symptoms can sometimes be reproduced by using the hyperventilation provocative test or by asking the patient to hold his or her breath. The patient is asked to stand and take deep breaths at a rate of 30 to 40 breaths/minute for 4 to 5 minutes. As the symptoms are reproduced, the patient is asked to describe them. Then the patient is instructed to breathe slowly into a paper bag to demonstrate a method by which he or she can rapidly control the frightening symptoms. The patient is instructed to use this technique to control symptoms in the future. Referral for counseling and/or psychiatric evaluation with focus on the adolescent's underlying anxieties and fears may prevent the patient from becoming an adult with hyperventilation syndrome.

Chronic Fatigue Syndrome

The complaint of fatigue by an adolescent should be taken seriously because this age group typically has an endless supply of energy. The causes of fatigue include demanding, unrealistic activity; poor health habits, stress, anxiety, and depression; or an underlying medical disease. Most cases of fatigue in the adolescent age group are caused by stress and resolve with improved health habits and maintaining realistic scheduling of activities.

A complete history and review of systems, and a thorough physical examination are necessary to define life stresses and eliminate underlying organic disease. The patient who complains of fatigue that is present upon awakening and that is not relieved by rest or sleep most likely has these symptoms because of intercurrent stress and anxiety. Conversely, the patient who experiences increasing fatigue during the day that is relieved by rest often has an underlying medical cause for the fatigue. The presence of other symptoms, such as fever or weight loss, suggests an organic cause. Initial screening laboratory studies may include a complete blood count, sedimentation rate, and urinalysis. Further evaluation should be tailored to the patient's complaints, severity of illness, and physical examination findings. After initial evaluation, if no underlying disease is suspected, the patient should be reassured that his or her symptoms are caused by overactivity and stress and not to any medical disorder. Suggestions regarding health habits, including regular sleep patterns, healthy diet, and regular exercise, may be helpful to some of these patients. If underlying organic disease is suspected, appropriate evaluation and referral should be done. In the instance of underlying emotional problems, social work or psychiatric evaluation with referral is necessary.

Although less common, adolescents may present with a complaint of chronic persistent fatigue. Chronic fatigue syndrome is characterized by acute onset of overwhelming fatigue, malaise, painful lymphadenopathy, sore throat, low-grade or subjective fever, myalgia, arthralgia, headache, and mental exhaustion (Table 130.10). The symptoms are disabling and impact significantly on school attendance and participation in extracurricular activities. Currently, the cause of this syndrome is unknown; most likely, its origin is multifactorial, including viral infection in combination with psychologic predisposition.

1. Persistent fatigue ≥6 months that is new in origin, not the result of ongoing exertion and substantially alleviated by rest, resulting in substantial reduction in previous level of activity

AND

The concurrent occurrence of ≥4 of the following persists/recurring during 6 months:

2. Problems with memory or concentration
   Sore throat
   Tender cervical or axillary nodes
   Muscle pain
   Multi-joint pain with arthritis
   Headaches
   Unrefreshing sleep
   Postexertional malaise

Exclusion:
   Active medical condition that explains fatigue
   Past/current diagnosis of a major depressive disorder, bipolar affective disorder, schizophrenia, delusional disorder, anorexia nervosa, or bulimia nervosa.
   Substance abuse within 2 years of onset

From Fukuda K, Straus SE, Hickie I, et al. The chronic fatigue syndrome: a comprehensive approach to its definition and study. Ann Intern Med 1994; 121:953–959.

Table 130.10. Case Definition for Chronic Fatigue Syndrome

Chronic fatigue syndrome is a diagnosis of exclusion because no reliable physical examination or laboratory markers can indicate its presence. Thus, history and physical examination should focus on eliminating other organic causes, such as collagen vascular disease, malignancy, or chronic infection. Suggested laboratory tests, in addition to those already mentioned, include albumin, globulin, liver function tests, calcium, phosphate electrolytes, blood urea nitrogen, creatinine, glucose, and thyroid function tests. Management includes acknowledging the patient's complaints, explaining that evaluation and tests aid in ruling out other illnesses, and ensuring that long-term outcome for patients with this

disease is excellent. Patients should be referred back to their primary care physician for continued care. Psychological evaluation may prove to be beneficial.

Violence and Abuse

Violent encounters account for many ED visits by adolescents. In a recent report, half of all injuries seen in this age group resulted from violence. Rates of adolescent maltreatment (including physical, sexual, and emotional abuse and neglect) are also high. In 1990, 25% of all cases reported to Child Protective Services were teens between the ages of 12 and 17. Adolescents are less likely to have serious injury or death resulting from these incidents; however, long-term consequences include increased high-risk behaviors, including runaway behavior, violence, and substance abuse. In the ED, violent behavior and maltreatment can be screened for and recognized.

Non-threatening, open-ended interviewing yields maximum information. Barriers in adolescent disclosure include incomplete recall of abusive events as well as their perception that their behavior precipitated the punishment and therefore was deserved. Important information includes the circumstances of the injury, the relationship of the assailant, substance abuse, use of weapons, previous history of violent behavior, and whether the teen plans to seek revenge.

Cases of teen maltreatment should be evaluated by a social worker and reported to Child Protective Services and/or the police. All cases of sexual abuse should be reported to the police. Teens involved in violent behaviors resulting in an ED visit should also be evaluated by a social worker and decisions should be made regarding needs for mental health evaluation and treatment.

Currently, existing child protective and mental health services are sparse. However, this lack should not effect the decision to report.

CONCLUSION

The care of adolescents is both challenging and rewarding. They present to the ED with a wide spectrum of diseases and often require gynecologic evaluation and referral, crisis intervention, and medical or psychological follow-up. Thus, the ED should design an organized approach to the care of the adolescent patient.

Suggested Readings

ADOLESCENT PREGNANCY

Anonymous. Counseling the adolescent about pregnancy options. Pediatrics 1989;83:135–137.

Brenner PF, Roy S, Mishell DR. A study of 300 consecutive surgically treated cases. JAMA 1980;243:7:673–676.

Cates W, Schulz KF, Grimes DA. The risks associated with teenage abortion. N Engl J Med 1983;309:11:621–624.

Emans SJH, Goldstein DP. Teenage pregnancy. In: Pediatric and Adolescent Gynecology. Boston: Little, Brown, 1990.

Grimes DA, Mishell DR, Shoupe D, Lacarra M. Early abortion with a single dose of the antiprogestin RU-486. Am J Obstet Gynecol 1988;158:1307–1312.

Maciak BJ, Spitz AM, Strauss LT, et al. Pregnancy and birth rates among sexually experienced US teenagers—1974, 1980, and 1983. JAMA 1987;258:15:2069–2071.

McAnarney ER. Adolescent pregnancy and childbearing: new data, new challenges. Pediatrics 1985;75:973–975.

McAnarney ER. Young maternal age and adverse neonatal outcome. Am J Dis Child 1987;141:1053–1059.

McAnarney ER, Hendee WR. Adolescent pregnancy and its consequences. JAMA 1989;262:74–77.

McAnarney ER, Hendee WR. The prevention of adolescent pregnancy. JAMA 1989;262:78–82.

Neinstein L, ed. Teenage pregnancy. In: Adolescent Health Care. 2nd ed. Baltimore: Urban & Schwarzenberg, 1991.

Scott VB. Complications of pregnancy. In: Rosen P, ed. Emergency Medicine Concepts and Clinical Practice. 2nd ed. St Louis: Mosby, 1988.

Silber TS, Saadat M. Diagnosis of adolescent pregnancy—1965 vs. 1980. Clin Pediatr 1982;21:556–558.

Spitz AM, Velebil P, Koonin LM, et al. Pregnancy, abortion and birth rate among U.S. adolescents—1980, 1985 and 1990. JAMA 1996;275:989–994.

Spivak H, Weitzman M. Social barriers faced by adolescent parents and their children. JAMA 1987;258:1500–1504.

ADOLESCENT ABUSE/RAPE

Braen GR. Sexual assault. In: Rosen P, ed. Emergency Medicine Concepts and Clinical Practice. 2nd ed. St Louis: Mosby, 1988.

Emans SJH, Goldstein DP, eds. Pediatric and Adolescent Gynecology. Boston: Little, Brown, 1990.

Felice M, Grant J, Reynolds B, et al. Rape and the adolescent. Pediatrics 1988;81:595–597.

Grant LM, Demetriou E. Adolescent sexuality. Pediatr Clin North Am 1988;35:1271–1290.

Holmes K, Sparling PF, Per-Andra M, et al, eds. Sexually Transmitted Diseases. 2nd ed. New York: McGraw-Hill, 1990.

Jenny C. Adolescent risk-taking behavior and the occurrence of sexual assault. Am J Dis Child 1988;142:770–772.

Neinstein L, ed. Rape and sexual abuse. In: Adolescent Health Care. 2nd ed. Baltimore: Urban & Schwarzenberg, 1991.

1997 Red Book. Report of the Committee on Infectious Diseases. 24th ed. Elk Grove, IL: American Academy of Pediatrics, 1997.

Tilelli JA, Turek D, Jaffe AC. Sexual abuse of children: clinical findings and implications for management. N Engl J Med 1980;302:319–323.

Woodling BA, Kossoris PD. Sexual misuse: rape, molestation, and incest. Pediatr Clin North Am 1981;28:481–499.

ADOLESCENT EATING DISORDER

American Psychiatric Association. Diagnostic and Statistical Manual of Mental Health Disorders. 4th ed. Washington, DC: American Psychiatric Press, 1994.

Andersen AE. Anorexia nervosa and bulimia: a spectrum of eating disorders. J Adolesc Health Care 1983;4:15–21.

Arroyo D, Tonkin R. Adolescents with bulimic and nonbulimic eating disorders. J Adolesc Health Care 1985;6:21–24.

Bray GA. Classification and evaluation of the obesities. Med Clin North Am 1989;73:161–184.

Carney CP, Anderson AE. Eating disorders: guide to medical evaluation and complications. Psych Clin North Am 1996;19(4):657–679.

Comerci GD. Medical complications of anorexia nervosa and bulimia nervosa. Med Clin North Am 1990;74:1293–1309.

Eckert ED, Halmis KA, Pierce B, et al. Ten-year follow-up of anorexia nervosa: clinical course and outcome. Psycho Med 1995;25:143–156.

George Hsu LK. Epidemiology of eating disorders. Psych Clin North Am 1996;19(4):681–700.

Halmi KA. Treatment of anorexia nervosa: a discussion. J Adolesc Health Care 1983;4:47–50.

Health and Public Policy Committee—American College of Physicians. Eating disorders: anorexia nervosa and bulimia. Ann Intern Med 1986;105:790–794.

Herzog DB, Beresin EV. Anorexia nervosa. In: Melvin Lewis, ed. Child & Adolescent Psychiatry: A Comprehensive Textbook. 2nd ed. Baltimore: Williams & Wilkins, 1996.

Herzog DB, Copeland PM. Eating disorders. N Engl J Med 1985;313: 295–303.

Herzog DB, Keller MB, Lavar PW. Outcome in anorexia nervosa and bulimia nervosa: a review of the literature. J Nerv Ment Dis 1988;176:131–143.

Isner JM, Roberts WC, Heymsfield SB, et al. Anorexia nervosa and sudden death. Ann Intern Med 1985;102:49–52.

Kulig JW, Siqueira LM. Diagnosis of anorexia nervosa in the adolescent male. J Adolesc Health Care 1983;4:31–34.

Maloney MJ, Klykylo WM. An overview of anorexia nervosa, bulimia, and obesity in children and adolescents. J Am Acad Child Psychiatry 1983;22:99–107.

McAnarney ER, Greydanus DE, Campanella VA, et al. Rib fractures and anorexia nervosa. J Adolesc Health Care 1983;4:40–43.

Mickley DW. Eating disorders. Hosp Pract 1988:58–79.

Milner MR, McCanarney ER, Klish WJ. Metabolic abnormalities in adolescent patients with anorexia nervosa. J Adolesc Health Care 1985; 6:191–195.

Mitchell JE, Seim HC, Colon E, et al. Medical complications and medical management of bulimia. Ann Intern Med 1987;107:71–77.

Palla B, Litt IF. Medical complications of eating disorders in adolescents. Pediatrics 1988;81:613–623.

Palmer EP, Guay AT. Reversible myopathy secondary to abuse of ipecac in patients with major eating disorders. Med Intell 1985;313:1457–1459.

Powers PS. Initial assessment and early treatment options for anorexia nervosa and bulimia nervosa. Psych Clin North Am 1996;19(4):639–655.

Sheridan PH, Collins M. Potentially life-threatening hypophosphatemia in anorexia nervosa. J Adolesc Health Care 1983;4:44–46.

Woodside DB. A review of anorexia nervosa and bulimia nervosa. Curr Prob in Pediatr 1995;25:67–87.

ADOLESCENT SUICIDE

American Academy of Pediatrics—Committee on Adolescence. Suicide and suicide attempts in adolescents and young adults. Pediatrics 1988;81:322–325.

Centers for Disease Control and Prevention. Premature mortality due to suicide and homicide—US 1983. MMWR 1986;35:357.

Centers for Disease Control and Prevention. Youth Suicide—US 1970–1980. MMWR 1987;36:87.

Crumley FE. Adolescent suicide attempts. JAMA 1979;241:2402–2407.

Harkavy J. Suicide attempts in adolescence. Prevalence and implications. N Engl J Med 1985;313:1290.

Hawton K, Cole D, O'Grady J, et al. Motivational aspects of deliberate self-poisoning in adolescents. Br J Psychiatry 1982;141:286–291.

Low BP, Andrews SF. Adolescent suicide. Med Clin North Am 1990;74: 1251–1264.

Marks A. Management of the suicidal adolescent on a nonpsychiatric adolescent unit. J Pediatr 1979;95:305–308.

McIntire MS, Angle CR, Wikoff RL, Schlicht ML. Recurrent adolescent suicidal behavior. Pediatrics 1977;60:605.

McManus BL, Krusesi MJP, Dontes AE, et al. Child and adolescent suicide attempts: an opportunity for ED's to provide injury prevention

education. Am J Emerg Med 1997;15:357–360.

Neinstein L, ed. Suicide. In: Adolescent Health Care. 2nd ed. Baltimore: Urban & Schwarzenberg, 1991:935.

Phillips DP, Carstensen LL. Clustering of teenage suicides after television news stories about suicide. N Engl J Med 1986;315:685–689.

Rund D. Suicide. In: Rosen P, ed. Emergency Medicine. 2nd ed. St Louis: Mosby, 1988:1835.

Slap GB, Vorters DF, Chaudhuri S, et al. Risk factors for attempted suicide during adolescence. Pediatrics 1989;84:762–772.

Smith CW, Bope ET. The primary care physician's role in evaluation and treatment. Suicidal Patient 1986;79:197–202.

ADOLESCENT SUBSTANCE ABUSE

Anglin T. Interviewing guidelines for the clinical evaluation of adolescent substance abuse. Pediatr Clin North Am 1987;34:381.

Anonymous. Drugs that cause psychiatric symptoms. Med Lett 1984;26: 75–78.

Farrow JA. Adolescent chemical dependency. Med Clin North Am 1990;74:1265–1273.

Farrow JA, Deisher R. A practical guide to the office assessment of adolescent substance abuse. Pediatr Ann 1986;15:677–684.

Johnson LD. Illicit drug use by American high school seniors in 1989. News and Information Services of the University of Michigan, February 13, 1990.

Joshi N. Drug use, depression and adolescents. Pediatr Clin North Am 1988;35:349.

Khantzian EJ. Acute toxic and withdrawal reactions associated with drug use and abuse. Ann Intern Med 1979;90:361–372.

Kulberg A. Substance abuse: clinical identification and management. Pediatr Toxicol 1986;33:325–361.

Neinstein L. Drug use and abuse. In: Adolescent Health Care. 2nd ed. Baltimore: Urban & Schwarzenberg, 1990.

Robinson TN, Killen JD, Taylor DB, et al. Perspectives on adolescent substance use: a defined population study. JAMA 1987;257:2072–2076.

Sanders JM. Adolescents and substance abuse. Pediatrics 1985;76:630–632.

PSYCHOSOMATIC ILLNESS

Barsky AJ. Hidden reasons some patients visit doctors. Ann Intern Med 1981;94:492–498.

Brown JT, Mulrow CD, Stoudemire GA. The anxiety disorders. Ann Intern Med 1984;100:558–564.

Carter BD, Marshall GS. New developments: diagnosis and management of chronic fatigue in children and adolescents. Curr Prob Pediatr 1995;25(2):281–293.

Green JW, Walker LS, Hickson G, et al. Stressful life events and somatic complaints in adolescents. Pediatrics 1985;75:1922.

Katon W, Kleinman A, Rosen G. Depression and somatization: a review. Am J Med 1982;72:127–133.

Smith MS. Psychosomatic symptoms in adolescence. Med Clin North Am 1990;74:1121–1133.

Smith GR, Monson RA, Ray DC. Patients with multiple unexplained symptoms: their characteristics, functional health, and health care utilization. Arch Intern Med 1986;146:69–72.

Van Winter JR, Stickler GB. Panic attack syndrome. J Pediatrics 1984;105:661–665.

CHEST PAIN AND HYPERVENTILATION

Bass C. Unexplained chest pain and breathlessness. Med Clin 1991; 75:1157–1173.

Brenner JI, Berman MA. Chest pain in childhood and adolescence. J Adolesc Health Care 1983;3:271–276.

Coleman WL. Recurrent chest pain in children. Pediatr Clin North Am 1984;31:1007–1025.

Feinstein RA, Daniel WA. Chronic chest pain in children and adolescents. Pediatr Ann 1986;15:685–694.

Fyfe DA, Moodie DS. Chest pain in pediatric patients presenting to a cardiac clinic. Clin Pediatr 1984;23:321–324.

Magarian GJ. Hyperventilation syndromes: infrequently recognized common expressions of anxiety and stress. Medicine 1982;61:219–235.

Pantell RH, Goodman BW. Adolescent chest pain: a prospective study. Pediatrics 1983;71:881–887.

Pickering D. Precordial catch syndrome. Am J Dis Child 1981;56:40.

FATIGUE

Holmes GP, Kaplan JE, Gantz NM, et al. Chronic fatigue syndrome: a working case definition. Ann Intern Med 1988;108:387–389.

Katz BZ, Andiman WA. Chronic fatigue syndrome. J Pediatr 1988;113: 944–947.

Neinstein L, ed. Fatigue. In: Adolescent Health Care. 2nd ed. Baltimore: Urban & Schwarzenberg, 1991.

ABDOMINAL PAIN

Lebenthal E. Recurrent abdominal pain in childhood. Am J Dis Child 1980;134:347.

Liebman W. Recurrent abdominal pain in children. Clin Pediatr 1978;17:149.

Neinstein L, ed. Recurrent abdominal pain. In: Adolescent Health Care. 2nd ed. Baltimore: Urban & Schwarzenberg, 1991.

Oster J. Recurrent abdominal pain, headache and limb pain in children and adolescents. Pediatrics 1972;50:429–436.

GENERAL

American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders. 3rd ed. Washington, DC: American Psychiatric Association, 1987.

Blum R. Contemporary threats to adolescent health in the United States. JAMA 1987;257:3390–3395.

Centers for Disease Control. Youth Risk Behavior Surveillance—U.S., 1995. MMWR 1996;45:9–13.

Cheng TL, Savageau JA, Sattler AL, DeWitt TG. Confidentiality in health care: a survey of knowledge, perceptions and attitude among high school students. JAMA 1993;269:1404–1407.

English A. Treating adolescents: legal and ethical consideration. Med Clin North Am 1990;74:1097–1111.

Fox JW. Mothers' influence on their adolescents' tendency to seek medical care. J Adolesc Health Care 1991;12:116–123.

Hofmann A, Becker RD, Garbier HP. The Hospitalized Adolescent: A Guide to Managing the Ill and Injured Youth. New York: Macmillan, 1976.

Krauss BS, Harakal T, Fleisher GR. The spectrum and frequency of illness presenting to a pediatric emergency department. Pediatr Emerg Care 1991;7:67–71.

Marshall WA, Tanner JM. Variations in patterns of pubertal changes in girls. Arch Dis Child 1969;44:291–303.

McManus M, McCarthy E, Kozak LJ, et al. Hospital use by adolescents and young adults. J Adolesc Health Care 1991;12:107–115.

Metzer-Lange M, Lye PS. Adolescent health care in a pediatric emergency department. Ann Emerg Med 1996;27:633–637.

National Center for Health Statistics. Advance Report of Final Mortality Statistics, 1993. Hyattsville, MD: Department of Health and Human Services, Public Health Service, Centers for Disease Control, 1996.

Newacheck PW, McManus MA. Health insurance status of adolescents in the United States. Pediatrics 1989;84:699–708.

Newacheck PW, McManus MA. Health care expenditure patterns for adolescents. J Adolesc Health Care 1990;11:133–140.

Rapp CE. The adolescent patient. Ann Intern Med 1983;99:55–60.

Results from the National Adolescent Student Health Survey. MMWR 1989;38:147–150.

Results from the National Adolescent Student Health Survey. MMWR 1989;261:2025–2031.

Rickert VI, Jay MS, Gottlieb AA. Adolescent wellness. Med Clin North Am 1990;74:1135–1147.

Seidel JS. Emergency medical services and the adolescent patient. J Adolesc Health Care 1991;12:95–100.

Tolmas HC. The role of the private practitioner. In: Strasburger VC, Greydanus DE, eds. Adolescent Medicine: State of the Art Reviews. Philadelphia: Hanley & Belfus, 1990:146–151.

Wood DL, Hayward RA, Corey CR, et al. Access to medical care for children and adolescents in the United States. Pediatrics 1990;86:666–673.

Yunker R, Levine M, Sajid A. Freestanding emergency centers and the health care of adolescents. J Adolesc Health Care 1988;9:321–324.

VIOLENCE

Council on Scientific Affairs—American Medical Association. Adolescents as Victims of Family Violence. JAMA 1993;270:1850–1856.

Ginsburg KR. Teen violence prevention. Physician and Sports Medicine 1997;25(3):69–83.

Kharasch SJ, Yuknek J, Vinci RJ, et al. Violence-related injuries in a pediatric emergency department. Pediatr Emerg Care 1997;13(2):95–97.

Melzer-Lange M, Lye PS. Adolescent health care in a pediatric emergency department. Ann Emerg Med 1996;27:633–637.

CHAPTER 131

Behavioral Emergencies

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This chapter reviews a number of conditions with prominent behavioral and somatic symptoms that may cause an infant or child 131to be brought to the emergency department (ED) (see Table 131.1). Although these conditions rarely require emergency intervention, the associated symptoms, such as stereotyped movements or cyanosis, may mimic other disorders with important physiologic consequences. The conditions reviewed in this chapter are heterogeneous: age of onset, symptoms, and prognosis vary widely. Some are part of normal childhood development, and others likely have an organic or genetic basis. All of these conditions, however, have prominent behavioral features as part of the history that are the key to diagnosis. Familiarity with these disorders aids the emergency practitioner in an appropriate evaluation followed by reassurance of the family and referral, when necessary, to a source for ongoing care that is the basis of treatment.

Cyanosis
Cyanotic breath-holding spells
Syncope
Cyanotic or pallid breath-holding spells
Motor Activity
Tic disorders and Tourette's syndrome
Paroxysmal choreoathetosis
Benign paroxysmal torticollis
Opsoclonus and myoclonus
Spasmus nutans
Hyperventilation Syndrome
Sleep-Related Disorders (see Table 131.2)

Table 131.1. Behavior-Related Problems Presenting with Somatic Symptoms

GENERAL APPROACH

Diagnosis of these disorders depends on obtaining a history of symptoms associated with the child's other activity at the time. Organic conditions such as seizures (see Chapter 70) or syncope (see Chapter 73) are involuntary and usually unrelated to other behaviors. Gathering the necessary information requires a careful history because the child's caretaker may not recognize the behavioral nature of the event, and emotional response may cloud recall. The clinician should review the episode chronologically in detail; key data include any precipitating events (e.g., breath-holding attacks, hyperventilation); the child's level of consciousness before, during, and after the episode (sleep disturbances); and any history of similar symptoms in the child or family members (tics). Having the caretaker act out the episode to demonstrate it and allow for measurement of duration may be useful. Social history and assessment of family dynamics (see Chapter 129) may further strengthen the diagnosis of some of these disorders.

Further evaluation beyond a careful history and physical examination is generally unnecessary for these conditions. The clinical picture may be specific enough to lead to the correct diagnosis. Laboratory values are generally normal or merely document the degree of symptoms (e.g., cyanosis) that has occurred. If the history is unclear or ambiguous, further investigation or referral may be warranted to rule out an important diagnosis as discussed in the appropriate chapter of this text. As in all uncertain clinical scenarios, the degree of evaluation should reflect the seriousness of potential diagnoses, in accordance with the maxim *primum non nocere* (above all, do no harm).

Management of these disorders generally requires referral to a physician with an ongoing relationship with the patient. For some of the conditions that are part of normal development (e.g., night terrors), reassurance may be all that is necessary. Nevertheless, the emergency practitioner may play an important role in this process because he or she may have the best opportunity to obtain important historical information that may be forgotten by the time of a later evaluation. The effect that a correct initial diagnosis may have on preventing unnecessary tests and reducing further anxiety should not be underestimated.

BREATH-HOLDING SPELLS

Background

Breath-holding spells in young children have been described since antiquity; accounts of this condition can be found in the works of Hippocrates, Rousseau, and Dickens, among others. In the modern medical literature, Lombroso and Lerman characterized the clinical syndrome in a group of 225 children with breath-holding spells. These patients were identified in a prospective study of almost 5,000 children, suggesting an incidence of 4.6%. The physicians described episodes of apnea and color change followed by loss of consciousness and postural tone that appeared to be triggered by an inciting event such as pain, fright, or agitation. Breath-holding children were categorized as cyanotic (62%), pallid (19%), or indeterminate (19%) based on the type of color change. These subgroups appeared to have distinctly different clinical features. Cyanotic breath-holding spells were usually preceded by vigorous crying; pallid spells were sudden and more likely to be followed by convulsive activity.

Pathophysiology

Although the cause of breath-holding spells remains unclear, research has refined the understanding of this condition. The diagnostic term itself is a misnomer. *"Breath-holding"* would suggest voluntarily "waiting to exhale," although most episodes appear to be involuntary and to occur at the end of expiration. Studies of breath-holding children have reported associations with autonomic dysfunction and anemia as well as an apparent familial predisposition. The clinical difference observed between pallid and cyanotic spells may have a basis in pathophysiology; pallid breath-holding spells can be reproduced by a vagal stimulus and are associated with significant bradycardia progressing to asystole. This finding suggests a similarity to vasovagal syncope or neurally mediated hypotension as described in adults. Cyanotic breath-holding spells have been attributed to cerebral hypoperfusion caused by hyperventilation followed by an increase in intrathoracic pressure. Further research is needed to validate these hypotheses.

Diagnosis

The diagnosis of breath-holding spells is based on the clinical history. Initial presentation usually occurs within the first 2 years of life and is not associated with other physical, developmental, or behavioral disorders. The typical episode begins with an inciting stimulus such as anger, frustration, fear, or pain. The child begins to cry, either briefly or for a prolonged period, and then suddenly stops in full expiration with mouth wide open. The spell may resolve at this point or proceed to color change followed by loss of consciousness. The child then becomes limp but soon may progress to an opisthotonic posture. Return to consciousness usually occurs within 1 minute. Severe breath-holding episodes may be associated with body jerks or incontinence and a transient recovery period of several minutes.

When a child is evaluated after a breath-holding spell, the clinician should expect to find a normal physical examination. Laboratory tests are also usually normal and add little to the evaluation. Some physicians suggest measuring hemoglobin because anemia has been associated with breath-holding spells. Electroencephalogram (EEG) performed during a spell reflects hypoxemia, but returns to normal afterward.

The differential diagnosis of breath-holding spells includes seizure disorders (see Chapter 70), structural cardiac disease (e.g., Tetralogy of Fallot), arrhythmia (e.g., long QT syndrome, see Chapter 82), syncope (see Chapter 73), and apnea (see Chapter 10) secondary to infection, brain tumor, injury, or congenital causes. A characteristic history in a healthy child with a normal physical examination should be sufficient to make the diagnosis. The key elements of the history are identifying the precipitating event and determining that the color change preceded any motor activity (unlike most seizures). In uncertain cases involving syncope, an electrocardiogram (ECG) is advisable to measure the QT interval corrected for heart rate. Although unlikely to be necessary in the usual clinical setting, serum prolactin measurement has been suggested as an adjunctive test because it is elevated after a seizure but normal in breath-holding spells and syncope.

Management

Once the diagnosis has been made, parents should be reassured that there is no evidence of long-term sequelae from typical childhood breath-holding spells. The relationship between the inciting event and the spell should be explained, as well as the possibility of recurrence. Frequency of recurrence varies from daily to yearly, but most breath-holding children stop having spells by school age. Inciting events such as pain and frustration are to be expected in healthy children, and overzealous attempts at prevention may impair the exploratory behavior and appropriate limit-setting that is part of normal development. If a spell recurs, parents should be instructed to clear the airway and place the child in a lateral, supine position away from other objects. In severe cases, referral to a specialist is indicated because treatment with atropine, theophylline, or other medications has been helpful in selected cases. A recent trial found a reduced frequency of spells in children treated with iron, although the efficacy of this treatment in non-anemic children remains unclear.

TICS AND MOVEMENT DISORDERS

Tics are involuntary, rapid, repetitive movements or vocalizations that may present throughout childhood and be confused with seizures or other disorders. Tic disorders affect 1 to 10 per 10,000 individuals and range from mild, self-limited symptoms to the chronic Giles de la Tourette syndrome, which can be severe and debilitating. Males are affected with tics three times as commonly as females, and familial predisposition has been well documented. Attention deficit hyperactivity disorder and obsessive–compulsive disorder are more common in children with tics.

Diagnosis of tic disorders relies on obtaining a history of stereotypical, involuntary motor activity that most often involves the muscles of the head and neck. Eye movements, head twitches, and shoulder shrugs are common, although complex movements and vocalizations may also occur. Unlike partial seizures, tics are non-rhythmic and partially suppressible,

which may result in their being absent at the time of an evaluation. Tics tend to increase at times of anxiety, stress, and fatigue and to decrease during sleep or relaxation. The differential diagnosis of tic disorders includes chronic diseases affecting the central nervous system, such as Wilson's disease, Sydenham's chorea, and metabolic disorders. A toxic cause should be considered because adverse reactions to many medications, including neuroleptics, metoclopramide, and antihistamines, can present with dystonic symptoms simulating tics.

ED evaluation and management of tics should be limited to establishing the diagnosis by history and ruling out other conditions. Physical examination is usually normal. Laboratory tests, imaging studies, and EEG are also usually normal and generally unnecessary. The emergency practitioner should reassure the family that the tics are not harmful and should encourage further discussion with a continuing care provider. Mild tics in young children often resolve spontaneously; more severe tics may require referral to a neurologist or psychiatrist for consideration of pharmacologic treatment.

Other movement disorders of childhood are listed in Table 131.1. Disorders of paroxysmal choreoathetosis are usually chronic and familial. Opsoclonus–myoclonus is a syndrome of chaotic, irregular eye movements that is associated with neuroblastoma in more than 50% of affected patients. Spasmus nutans is a condition involving head tilt, nodding, and nystagmus that presents in infancy and is associated with optic glioma in some cases. Benign paroxysmal torticollis presents in infancy with recurring episodes lasting minutes to days and must be differentiated from posterior fossa tumors and other conditions.

HYPERVENTILATION SYNDROME

Background

Hyperventilation is defined as ventilation in excess of that required to maintain normal arterial blood partial pressure of oxygen (PaO_2) and partial pressure of carbon dioxide ($Pa\,CO_2$). It may be produced either by an increase in frequency or in depth of respiration. This syndrome is common in adults but has received little attention in the pediatric literature. It can nonetheless be a disabling clinical pediatric problem that is often misdiagnosed and improperly treated.

Pathophysiology and Dynamics

The pathophysiology of the hyperventilation syndrome contains two components: 1) physiologic derangement produced by hyperventilation (Fig. 131.1) and 2) underlying psychiatric disturbance most often including anxiety.

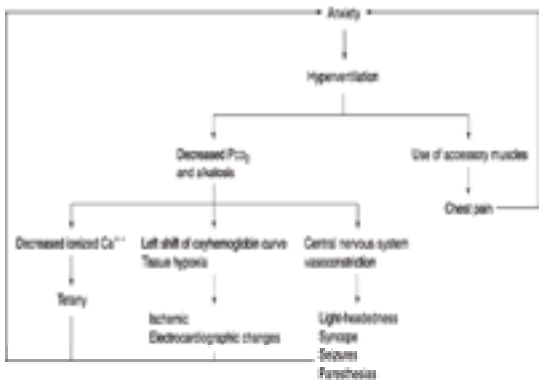

FIGURE 131.1. The pathophysiology of hyperventilation syndrome.

A single deep breath can reduce $PaCO_2$ by 7 to 16 mm Hg, and the $PaCO_2$ can drop to half of normal after only 30 seconds of hyperventilation. Thereafter, only an occasional deep breath superimposed on normal breathing is required to maintain low $PaCO_2$.

When normal subjects voluntarily hyperventilate, they experience relatively few and mild symptoms compared with the exaggerated complaints observed in patients with hyperventilation syndrome. This condition emphasizes the presence of an underlying psychiatric disturbance. Hyperventilation can be a signal of severe anxiety, which may persist into adulthood.

Clinical Manifestations

The age of onset is between 6 and 18 years, with more than half presenting during puberty. The incidence is equal in males and females. Patients almost always report that their symptoms occur in “spells” or “attacks.” In the ED, patients can present with various combinations of symptoms, including: tachypnea, anxiety, breathlessness, light-headedness, paresthesias, coldness of the extremities, tetany, trembling, chest pain, palpitations, blurred vision, feeling of impending doom, syncope, or seizure.

Physical examination may reveal obvious hyperventilation, or, more commonly, the patient may be observed to take periodic deep sighing respirations.

ECG changes, such as ST-segment depression and flattening and inversion of T waves both in the resting and exercise ECG, have been reported in adults who hyperventilate. However, unlike ischemic ECG changes, these changes occur

during early exercise and disappear as exercise continues.

Differential Diagnosis

The manifestations of hyperventilation syndrome are variable and can initially seem worrisome. Organic disorders that require serious consideration in the differential diagnosis include metabolic acidosis, hyperammonemia, hypocalcemia, drug intoxication (including salicylism), hypercapnia, cirrhosis, organic central nervous system disorders, fever, and the response to severe pain. A few paroxysmal disorders, such as hypotensive syncope, Stokes-Adams attacks, epilepsy, migraine, and asthma, must also be ruled out.

Many of these diagnoses can be excluded on the basis of careful history and physical examination. Elements of history that suggest the diagnosis of hyperventilation syndrome include chronicity of the complaint, unrelatedness of symptoms, references to breathlessness, and expressions of anxiety. Assessing whether voluntary hyperventilation reproduces the patient's symptoms is also helpful. This provocation test is currently the best diagnostic method, and is accomplished by asking the patient to hyperventilate for at least 3 minutes, enough to bring the $PaCO_2$, below 50% of baseline. Termination of symptoms on rebreathing into a paper bag is another suggestive finding. When the syndrome is recognized, extensive laboratory evaluation is rarely required in the pediatric population and may add to the child's overwhelming anxiety. However, as in many clinical situations that result in a diagnosis with psychological or psychiatric implications, the emergency physician may elect to order laboratory data to support the diagnosis. The specific tests obtained should be determined by the patient's symptoms, but will usually be selected from among chest radiograph, ECG, serum calcium and electrolytes, and blood gas determinations.

Treatment

The classic remedy for hyperventilation is breathing into a paper bag. The patient rebreathes his or her own expired air and thus inhales air enriched with CO_2.

Most experts recommend that the patient understand the mechanism by which the symptoms are produced. Patients need to be reassured in specific terms relevant to their fears. For adolescents in particular, emphasizing that the patient has control over the production of symptoms is important. This understanding is often accomplished by voluntary overbreathing, attribution of cause of symptoms to hyperventilation, and breathing training with emphasis on diaphragmatic breathing and slowing of expiration. Counseling and supportive therapy are necessary to discover the sources of the psychological disturbance experienced by the child. These efforts should start in the ED, but psychiatric consultation and/or referral is often required. Propranolol has been used successfully in children with chronic refractory hyperventilation; however, its use should be limited to patients who fail to respond to educational and counseling therapy.

SLEEP DISTURBANCES

Background

Childhood sleep disturbances are common, and most reflect the particular stresses of certain developmental stages in infants and toddlers. These stresses include resistance to being put down for the night, frequent nighttime awakenings in infancy, and nightmares in the toddler. Anticipatory guidance on the part of the primary care physician can minimize the degree of such disturbances. Such problems are chronic and are rarely seen in the ED.

Some sleep disorders are based on disturbed sleep processes and may appear with dramatic, often paroxysmal clinical features that are frightening to the family and are likely to be seen in the ED (Table 131.2). The differential diagnosis and management of these disorders is discussed more fully in this chapter. Before such discussion, however, briefly reviewing current knowledge of sleep processes may be helpful.

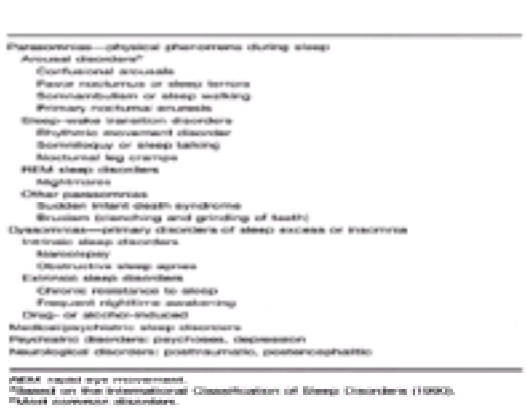
Parasomnias—physical phenomena during sleep
Arousal disorders[a]
Confusional arousals
Pavor nocturnus or sleep terrors
Somnambulism or sleep walking
Primary nocturnal enuresis
Sleep–wake transition disorders
Rhythmic movement disorder
Somniloquy or sleep talking
Nocturnal leg cramps
REM sleep disorders
Nightmares
Other parasomnias
Sudden infant death syndrome
Bruxism (teeth grinding and grinding of teeth)
Dyssomnias—primary disorders of sleep excess or insomnia
Intrinsic sleep disorders
Narcolepsy
Obstructive sleep apnea
Extrinsic sleep disorders
Chronic resistance to sleep
Frequent nighttime awakening
Drug- or alcohol-induced
Medical/psychiatric sleep disorders
Psychiatric disorders: psychoses, depression
Neurological disorders: posttraumatic, postencephalitic
REM, rapid eye movement.
[a]Based on the International Classification of Sleep Disorders (1990).
[b]Most common disorders.

Table 131.2. Sleep-Related Disorders [a]

Sleep Processes

Sleep consists of two non-wakeful states that are best distinguished by the presence or absence of rapid eye movements (REMs). REM and non-REM states have vastly different process characteristics alternating in an orderly cycle of 90 minutes, with progressive increases in the REM sleep across the night. Non-REM sleep is what is commonly thought of as sleep and, after 6 months of age, is typically the first type of sleep entered from the awake state. Non-REM sleep is divided into stages, ranging from stage 1, drowsiness, to stage 4, deep sleep. During non-REM sleep, the pulse and

respiratory rates are slow and regular and some baseline muscle tone with minimal body movements is present. Mentation is light, and imagery is not vivid or easily recalled by a subject awakened from this stage.

REM sleep, conversely, is characterized by facial and body movements and the occurrence of bilateral, synchronous, rapid eye movements. REM sleep represents a period of heightened central nervous system activity, although arousal threshold (the level of stimulus necessary to awaken the subject) is comparable with the deeper non-REM stages. Dreaming occurs during REM sleep. Heart and respiratory rates are faster and more irregular than in non-REM sleep, whereas resting muscle tone is suppressed.

Clinical Manifestations and Management

Parasomnias

Arousal Disorders. All arousal disorders share common features. They tend to occur in the transition from deep non-REM to light non-REM sleep. The various disorders may occur at different times in the same child. A positive family history is often present. Males outnumber females in incidence. The disorders are paroxysmal, associated with activation of the autonomic nervous system and skeletal muscles, unresponsiveness to the environment, and amnesia for the episodes. Fever, physical or mental fatigue, sleep deprivation, and emotional distress can trigger these parasomnias in susceptible individuals.

Confusional arousals consist of disorientation in time and space, slow speech and mentation, and bizarre behavior, such as placing a piece of cloth in the refrigerator. These episodes may last from minutes to hours. They gradually decrease in frequency with age.

Pavor nocturnus (night terrors) is usually seen in preschool-age children. The child abruptly awakens 15 to 90 minutes after sleep onset, with a piercing scream or cry, sits up in bed with wide-open eyes, extreme anxiety, and many autonomic phenomena (sweating, flushing, racing heart-beat, and rapid breathing). The child may be inconsolable for 10 to 15 minutes, but then relaxes suddenly and falls back to quiet sleep. Night terrors usually occur so rarely that treatment is not necessary. Their frequency decreases with age.

Somnambulism appears in school-age children. The child sits up suddenly with eyes glassy and staring, and may then arise and walk around clumsily for 15 seconds to 30 minutes. The child often walks toward a light or a noise, and, after returning to bed, falls asleep uneventfully. Temporal lobe epilepsy may be difficult to distinguish from somnambulism but can be differentiated by adequate sleep EEG studies. Management consists of “sleep-proofing” the home because sleepwalkers can hurt themselves. As opposed to the incidence in adult sleepwalkers, significant psychopathology is not present in school-age children. Parents can be reassured that most children outgrow sleep-walking over several years.

Primary nocturnal enuresis is defined as enuresis occurring in a child older than 3 years of age, who is otherwise well, and has never been dry at night, although he or she can stay dry all day. This condition is the most common non-REM disorder, with an incidence ranging from 5 to 17% of all children between 3 and 15 years of age. The enuretic episode typically occurs during the first cycle of the night. It is characterized by tachycardia, tachypnea, penile erection in males, increased intravesical pressure, and spontaneous bladder contraction. The differential diagnosis includes organic problems such as diabetes mellitus, diabetes insipidus, and urinary tract infection, although these symptoms would be rare if the definition just given is adhered to. Treatment starts with conditioning modalities. Medications such as imipramine that have both anticholinergic effects on the bladder and stimulant effects on sleep stage patterns, or anti-diuretic hormones (DDAVP) are considered a last resort to be prescribed only in severe cases under the supervision of the primary care physician.

Sleep–Wake Transition Disorders. *Rhythmic movement disorder* starts around 8 months of age, and generally stops by age 4. Boys outnumber girls 3:1. The usual pattern is body rocking followed by head rolling or banging against the crib sides, immediately before sleep onset and throughout light sleep, lasting several minutes to an hour. Head banging is typically not associated with crying. No significant injuries are incurred, although callus formation and contusions may be observed. Management consists of reassurance and advice to pad the crib.

Somniloquy appears in school-age children. The speech can be spontaneous or induced by conversation from another person. Sleeptalking may be vivid and revealing, but is also usually outgrown with time.

REM Sleep Disorders *Nightmares* are unpleasant dreams from which the child is usually awake and responsive by the time the parents arrive, and for which substantial recall can occur. Nightmares usually occur in the last third of the night when REM sleep predominates. The child who just had a nightmare should be reassured with embraces and soothing words, and the parent should stay until the child is calm. Parents of children with occasional nightmares should be reassured about the benign nature of these episodes. Frequent nightmares may be a sign of distress that merits a psychological evaluation.

Dyssomnias

Narcolepsy is a rare syndrome of abnormality of REM sleep and excessive sleepiness. Although narcolepsy can begin before the age of 10, gradual onset between 15 and 35 years of age is usual. The two most important symptoms of narcolepsy are daytime sleepiness (that cannot be fully relieved by any amount of sleep) and cataplexy (sudden loss of muscle tone with preservation of consciousness). Other complaints include attacks of daytime sleep, sleep paralysis (inability to move during the onset of sleep or on awakening), hypnagogic hallucinations (vivid imagery at the onset of sleep or awakening), and disturbed nighttime sleep. The occurrence of REM sleep at the onset of sleep is the most characteristic and striking abnormality observed in narcolepsy. Management includes a regular schedule of naps and stimulant drugs, such as a low dosage of dextroamphetamine or methylphenidate for daytime sleepiness, and tricyclic antidepressants for cataplexy, sleep paralysis, and hypnagogic hallucinations. The degree of sleepiness rarely lessens

but cataplexy, sleep paralysis, and hypnagogic hallucinations improve or disappear with age in one third of patients.

Obstructive sleep apnea syndrome refers to upper airway obstruction, frequent apneic spells during sleep, and disordered sleep stages with increased stages 1 and 2 and decreased stages 3 and 4 of non-REM sleep. Children may present with loud snoring, excess daytime somnolence, morning headaches, hypertension, cardiac arrhythmias, cor pulmonale, enuresis, anoxic seizures, and poor school performance. Management involves otolaryngologic consultation for thorough airway evaluation and appropriate measures to relieve obstruction, maintenance of continuous positive airway pressure (CPAP), and initiation of weight loss in obese children (see Chapter 95).

Miscellaneous

A few additional sleep-related disorders are noted in Table 131.2, including nocturnal leg cramps, bruxism, sleep disorders related to substance abuse, neuropsychiatric conditions, and sudden infant death syndrome. The latter is discussed in detail in Chapter 10.

Suggested Readings

GENERAL APPROACH

Altmeier WA. The fun of the chase. Pediatr Ann 1997;26:397–401.

Barron T. The child with spells. Pediatr Clin North Am 1991;38:711–724.

BREATH-HOLDING SPELLS

DiMario FJ. Breath-holding spells in childhood. Am J Dis Child 1992;146:125–131.

Evans OB. Breath-holding spells. Pediatr Ann 1997;26:410–414.

Hannon DW. Breath-holding spells: waiting to inhale, waiting for systole, or waiting for iron therapy? J Peds 1997;130:510–512.

Lombroso CT, Lerman P. Breathholding spells (cyanotic and pallid infantile syncope). Pediatrics 1967;39:563–581.

Zelnik N, Kahana L, Rafael A, et al. Prolactin and cortisol levels in various paroxysmal disorders in childhood. Pediatrics 1991;88:486–489.

TICS AND MOVEMENT DISORDERS

Singer HS. Tic disorders. Pediatr Ann 1993;22:22–29.

Vedanarayanan VV. Paroxysmal movement disorders. Pediatr Ann 1997;26:402–408.

HYPERVENTILATION

Hanna DE, Hodgens JB, Daniel WA Jr. Hyperventilation syndrome. Pediatr Ann 1986;15(10):708–712.

Herman SP, Stickler GB, Lucas AR. Hyperventilation syndrome in children and adolescents: long-term follow-up. Pediatrics 1981;67(2):183–187.

Holder EL, Cohen DJ. Repetitive behavioral patterns in childhood. In: Levine MD, Carey WB, Crocker AC, eds. Developmental-Behavioral Pediatrics. Philadelphia: WB Saunders, 1992.

Saisch SG, Wessely S, Gardner WN. Patients with acute hyperventilation presenting to an inner-city emergency department. Chest 1996;110(4):952–957.

SLEEP DISORDERS

Adair RH, Bauchner H. Sleep problems in childhood. Curr Probl Pediatr 1993;23(4):147–170.

Aldrich MS. Narcolepsy. N Engl J Med 1990;323(6):389–394.

American Sleep Disorders Association. International Classification of Sleep Disorders: Diagnostic and Coding Manual. Lawrence, KS: Allen Press, 1990.

Ferber R. Sleep disorders. In: Levine MD, Carey WB, Crocker AC, eds. Developmental-Behavioral Pediatrics. Philadelphia: WB Saunders, 1992.

Holder EL, Cohen DJ. Repetitive behavioral patterns in childhood. In: Levine MD, Carey WB, Crocker AC, eds. Developmental-Behavioral Pediatrics. Philadelphia: WB Saunders, 1992.

Loughlin GM. Obstructive sleep apnea in children. Adv Pediatr 1992;39: 307–336.

Wise MS. Parasomnias in children. Pediatr Ann. 1997;26(7):427–433.

CHAPTER 132

The Emergency Department Response to Incidents of Biological and Chemical Terrorism

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BACKGROUND—THE THREAT OF BIOLOGICAL AND CHEMICAL TERRORISM

Emerging Patterns of Terrorism

At 8:00 AM on March 20, 1995, at the height of a Monday morning rush hour, members of the Japanese Aum Shinrikyo religious cult placed 11 containers of the deadly nerve agent sarin in various downtown locations of the Tokyo subway system and unleashed on their fellow citizens, and the world, a new era in terrorism. An earlier sarin attack (targeting several judges involved in trials of Aum cult members) on a residential neighborhood of Matsumoto in June 1994 had caused hundreds of exposures, seven deaths and grim notice by experts in counter-terrorism. This same group also possessed, and attempted to deploy, anthrax and botulinum toxin weapons on several occasions.

However, the chemical warfare attack on downtown Tokyo, one of the world's largest and most cosmopolitan capitals, resulting in more than 5000 victims and 12 deaths, brought the potential of terrorist use of weapons of mass destruction on large civilian populations to international attention. Of course, the prior several decades had already witnessed enormous tragedy caused by terrorists of various ethno-nationalist and separatist groups, with attacks in most cases targeted at military installations, government officials or representatives, or on occasion at relatively small groups of ordinary civilians in highly visible circumstances.

Still, the 1990s has brought us across a new threshold, as detailed by Hoffman in his thoughtful analysis, *Inside Terrorism.* The rise of religious terrorism has allowed violence to become a seemingly justifiable means to accomplish a sacred theological imperative. This rationalization of violence by placing it in subservience to unquestioned religious ends has the effect of removing normal moral and political constraints to violence in the minds of many religious fundamentalists. Their deeply held convictions serve to legitimize in their own eyes the indiscriminate and large-scale killing of often broadly defined infidel enemies, who may include most of a population. Several terrorist attacks in the 1990s have highlighted this new willingness to kill large numbers of innocent people indiscriminately: witness the Oklahoma City bombing, and the bombing of the World Trade Center in New York City where the perpetrators were attempting to topple one of the twin towers into the other with hopes of killing up to 250,000 people. The threshold to attempt killing mass numbers of persons has already been crossed.

The 1980s and 1990s saw several additional examples of religious and political terrorism in the United States. In 1984, the Rajneeshee cult poisoned 750 citizens of The Dalles, Oregon by intentionally contaminating restaurant salad bars

with *Salmonella typhimurium.* Recently, additional persons linked to white-supremacist militias have obtained anthrax, stockpiled the highly toxic substance ricin, a biotoxin extracted from castor beans, and have been arrested for transporting the bacterium *Yersinia pestis,* the causative agent of bubonic plague. Such incidents further illustrate the potential for biological or chemical terrorist attacks.

Finally, the Aum Shinrikyo sarin attack in Tokyo has opened Pandora's box not only by demonstrating the willingness to use chemical or biological warfare agents but also by proving that it can be done successfully. The stakes have been raised for other terrorists to outdo the Aum, and the coming of the millennium, with its mystical, apocalyptic interpretations to so many such cults, may portend even worse disasters. Thus, health care workers, particularly first-responders and emergency department (ED) personnel, must be familiar with the expedient diagnosis and management of chemical and biological terrorist events.

Weapons of Mass Destruction

The term *weapons of mass destruction* (WMD) has been used to denote weapons using *nuclear* (radiologic), *biological* or *chemical* (NBC) agents in devices intended to intentionally kill and injure large numbers of victims. Biological and chemical weapons are currently believed by U.S. military and counterterrorism experts to constitute a plausible and favored mode of WMD attack on civilian populations by terrorist groups. These agents are relatively inexpensive and technically far less difficult to produce and deploy than nuclear weapons, and the raw materials for their production are less highly regulated (radiologic exposure is discussed in Chapter 90). In recent years, state sponsorship of terrorism has risen, and access to biological and chemical agent information has increased via the Internet.

Chemical and biological weapons could be deployed in several ways readily accessible to terrorist groups. Relatively simple explosive devices can be combined with these agents, and additional simple mechanical devices such as garden sprayers, aerial crop dusting equipment, insect control vehicles, or aerosol spray devices, might all be used. In addition, a conventional attack on factories, chemical production facilities, or tank cars might result in the release of large quantities of toxic industrial chemicals, with resulting effects similar to an attack by military chemical warfare agents. Biological agents would likely involve aerosol dispersal.

Medical Consequences and Epidemiology

Estimating the real probability of a chemical or biological terrorist attack is difficult. According to Tucker's recent overview, extreme scenarios, such as a massive state-sponsored attack, can be conceptualized as a “low-probability, high-consequence event” that could potentially kill thousands and result in unparalleled demands on health care facilities for treatment of casualties. On the other hand, events such as The Dalles, Oregon, salad bars incident, and/or numerous hoaxes, are much more likely but have a lower potential for severe consequences. Chemical events, in particular, would likely combine elements of traditional mass disasters (e.g., secondary to earthquake), in which large numbers of casualties occur almost immediately, and elements of traditional hazardous materials (HAZMAT) incidents. However, chemical events differ from HAZMAT incidents in several important ways: 1) the intent is to inflict mass casualties; 2) the “hazardous materials” in these cases are of particularly high lethality; 3) the “HAZMAT” environment created would be extremely toxic to rescue workers; 4) considerable ambiguity might initially occur regarding the exact nature of the agent(s) involved; 5) the numbers of patients requiring relatively urgent treatment might be overwhelming to existing emergency medical services (EMS); 6) an even larger number of patients who were minimally affected, if at all, might self-transport to receiving EDs and place yet additional demands on the health care system; and 7) mass hysteria and panic would almost certainly ensue, further compromising the health care system's ability to respond optimally.

Okumura et al. reported many of these features in the Tokyo sarin attack. At about 8:00 AM, five subway cars were attacked simultaneously. By 8:28 AM, an ambulatory victim arrived to one area ED a short distance from the affected subway stations. At 8:43 AM, the first ambulance arrived, and the next hour brought an additional 500 patients, including 3 in cardiopulmonary arrest. City wide, 5510 persons sought emergency medical treatment at more than 200 facilities within a few hours of the attack, and of these, about 25% required hospitalization. Of note, most of the victims went to hospitals by taxi, bus, or private vehicles of good Samaritans, rather than by formal EMS transport, further compounding the understandable initial chaos. Until the identity of the agent was known, significant efforts at patient decontamination were lacking, resulting in several occurrences of illness among hospital staff, although most were of a mild degree. The morbidity of the attack was actually thought to have been substantially mitigated by several fortuitous factors. The sarin was dispersed via evaporation from plastic bags left open on the subway cars. Although considered a “nonpersistent” agent, the volatility of sarin at room temperature is only about the same as water, and thus the sarin had not disseminated widely throughout the cars when the incident was discovered. In addition, the Tokyo subways are considered to have an excellent ventilation system. The fact that the exposure to patients was via vapor, and that consequently their skin and clothing were minimally contaminated, also lessened secondary exposure to hospital personnel.

In a biological attack, it is important to emphasize that, because of their incubation periods, biological agent releases must be thought of in a different context than conventional and chemical terrorist attacks. Biological agent attack should be thought of as a public health emergency, when patients present remote in time and space from the point of exposure. Without an announcement by a terrorist group, an unusually large number of persons presenting with clinical findings of the disseminated disease would likely be the first indicator of a biological agent attack. The situation is further complicated because early symptoms resulting from biological agents are non-specific. Thus, patients may present to various clinics and EDs in piecemeal fashion, complaining, for example, of flu-like symptoms. The clinician in such settings must maintain a high index of suspicion if a biological attack is to be diagnosed in time for useful measures to be undertaken.

Moreover, as stressed by Franz et al., all such personnel must have a grasp of the fundamental principles of epidemiology and be able to apply these principles in working up an unexpected outbreak of unusual illness. These principles include 1) careful documentation of who is affected, 2) possible routes of exposure, 3) clinical findings of the

disease, 4) efforts at rapid identification of the causative organisms, 5) formulation of a case definition, 6) quantifying the number of cases, and 7) calculating the attack rate. The epidemic can then be described in terms of timing, place, routes of exposure, and other clinical characteristics of ill patients. The disease pattern that evolves is critical to recognizing a biological agent attack. Most natural epidemics evolve with a gradual rise in disease incidence because persons are progressively exposed to increased numbers of infectious patients, fomites, or vectors that spread the organism.

In contrast, after a biological agent attack, most persons would initially be exposed at the same time, and thus become ill and present in a relatively compressed time frame. Variations in the incubation period may occur, however, even with a point source exposure, possibly because of differences in dose of the agent received, immunologic status, and other factors. Diseases that are rare, not endemic in the area of exposure, or that are normally spread by vectors that are not indigenous to the relevant geographic area would of course also be suspect, especially if numerous cases developed simultaneously. Additional clues to a biological agent attack might include especially high infection rates among exposed persons, more respiratory forms of disease than usual, particularly high morbidity or mortality, several epidemics at once, attack rates lower in persons sheltered from the suspected route of exposure, infected or dying animals, and the discovery of suspicious actions or potential delivery systems. The earliest suspicion of such an outbreak should be reported at once to appropriate public health authorities.

DECONTAMINATION AND HEALTH CARE WORKER PROTECTION

The approach to patient decontamination after a WMD attack, similar to any significant hazardous materials incident, poses complex logistical challenges, involving both pre-hospital and ED-based operations. The on-scene and pre-hospital aspects of this process are of critical importance, but are beyond the scope of this discussion. From the hospital perspective, prioritization is to 1) protect current patients, staff, and facility; 2) provide optimal care to the patients presenting as victims of the incident; and 3) consider protection to the environment external to the hospital. In many communities, decontamination activity for most HAZMAT incidents is expected to be carried out on the scene by specially trained HAZMAT teams within the EMS system.

As previously noted, this scenario may be overly optimistic for a WMD event. Thus, although decontamination of the most severely affected patients would likely occur in the field, EDs should have a plan in place to enable decontamination of patients who arrive unexpectedly or on their own without prior decontamination. Although some distinctions can be made between necessary and sufficient health care worker protection from radiologic, chemical, and biological exposures, the predicted rarity of need for such a response suggests the utility of devising a single, universal ED decontamination protocol, and keeping this protocol as simple, practical, and inexpensive as possible. The following paradigm, adapted from the recent review by Barbera et al. in 1999, offers overall guidelines to such an approach.

Decontamination capability must be available on a short set-up time basis. Many models have been proposed, but most authorities recommend an outdoor facility with multiple patient stations, arranged so that parallel lines of ambulatory and non-ambulatory patients may be processed simultaneously. An outdoor facility is more capable of handling multiple patients and may make the use of copious water irrigation easier; however, it may end up exchanging a direct indoor airborne-dissemination hazard for the problem of defining and containing areas of outdoor vapor or aerosol hazard.

Optimally, an elevated platform would allow drainage, minimizing risk of patients slipping and falling, and of further exposure of victims to contaminated rinse water. Medical personnel in personal protective equipment (PPE) should staff an initial triage station at the entrance to the decontamination structure. Triage at this point facilitates rapid identification of patients requiring immediate antidotal or other life-saving intervention, as well as diversion of non-ambulatory patients to the appropriate area with medical assistance. Ambulatory patients are instructed in self-decontamination. Obviously, young children require assistance, or may be accompanied by parents if present. The outdoor facility must provide adequate water, temperature control during environmental extremes, and measures to maintain personal modesty, such as curtains or other barriers separating shower lines for males from lines for females.

A universal decontamination process has been proposed that simplifies staff education and uses easily accessible, inexpensive components. The sequential approach begins with complete disrobement and containment of contaminated clothes in impervious plastic bags. This step is followed by a dilute bleach wipe-down with 0.5% sodium hypochlorite solution made from a 1:10 dilution of standard household bleach (1 part bleach added to 9 parts water). This step is expected to disinfect almost all biological agents and may inactivate many chemical agents, including mustard and organophosphates. Little field testing has occurred under actual WMD circumstances, however, and even less discussion has taken place regarding applicability to infants and young children. The bleach solution is not recommended for open thoracic or intra-abdominal wounds, exposed nerve tissue, or in the eyes. Because as much as 15 to 20 minutes of contact time may be necessary to optimal inactivation of some chemical agents by bleach solutions alone, decontamination efforts should stress physical and mechanical removal over chemical decontamination for chemical warfare agents. In fact, the evolving consensus on biological agent decontamination is that clothing removal combined with soap and water washing/showering will be adequate in most cases of biological agent exposure.

Far more important than choice of dilute bleach or water for casualties exposed to these agents is gentle but thorough rinsing of affected skin, eyes, and wounds. Following the dilute bleach wipe-down, or in lieu of it, patients undergo a copious warm water rinse, and then thorough washing with a hypoallergenic soap and soft-bristle brushes or sponges (stiff brushes or abrasives should be avoided), followed by another rinse, and then a final rinse once they have moved past other in-use patient decontamination stations. A few hazardous materials, such as reactive metals (sodium, potassium, lithium) and strong corrosives in powder or particulate form are rare exceptions to the “universal” approach in that application of water is best avoided until all visible particles are removed with forceps, gentle brushing or vacuuming.

Riot-control agents are better not treated with dilute bleach because the reaction of bleach with these compounds creates equally irritating byproducts that could worsen skin burns. Persistent or thickened agents may require more prolonged soap and water washing. After passing through the decontamination process, patients are triaged again, with treatment categories assigned as appropriate. The matter of wash/rinse water run-off, and its containment, has been the

subject of some controversy. The potential for significant environmental damage in the context of ED-based patient decontamination after a WMD attack is likely limited, and notification of appropriate water-department authorities is probably sufficient.

Appropriate PPE for ED staff is an important consideration. The amount of chemical or biological agent believed to contaminate patients who would arrive alive at the ED after a WMD attack would consist essentially of that on their skin and clothing, and would thus be of far lower concentration than rescue workers would face at the scene of exposure. Most authorities believe adequate protection in this context would be afforded to ED staff garbed in level C PPE, which consists of a non-encapsulated chemically resistant body suit, gloves, and boots, with a full-face air purifier mask containing a cartridge with both an organic-vapor filter for chemical gases and vapors and a HEPA filter to trap aerosols of biological and chemical agents. Such PPE is much less cumbersome to work in than level A or B outfits (which use self-contained breathing apparatus) and is much less expensive.

Choices regarding specific materials used in level C PPE options are difficult because few such barrier materials have been tested against WMD agents. At least one such material, DuPont's Tyvek F, has been found effective against mustard and organophosphate agents, but again, given the predicted low concentration and short contact times relevant to the ED decontamination process, less expensive fabrics may be adequate. Biological agents require a considerable degree of energy to re-aerosolize from contaminated skin or clothing, and they are not (in contrast to chemical agents), either volatile or (with the exception of trichothecene mycotoxins) dermally active (i.e., they neither cause skin lesions nor penetrate intact skin to cause disease). Thus, surface decontamination of biological agents, although still necessary, is nevertheless considered less critical than decontamination of chemical agents.

BIOLOGICAL AGENTS

For biological threats in particular, considering the aims of the potential perpetrator is useful. Organized nation states may envision biological agents as either strategic or tactical weapons. When the strategic threat is considered, few agents possess the necessary stability to serve as large-scale weapons; the list of potential threats in this regard might include anthrax, plague, and smallpox. From a tactical perspective, toxin agents such as botulinum toxin and staphylococcal enterotoxins might be added to this list, although in reality, biological agents would seldom make good tactical weapons because these agents possess an important characteristic not shared by conventional or nuclear weapons; namely, an incubation period (of note, several chemical agents also exhibit a delay between exposure and the onset of clinical signs and symptoms; for chemical agents, this interval, analogous to the incubation period for biological agents, is called the latent period). Typically, these incubation periods are in excess of 24 hours. Many belligerents would not want to deploy a weapon on the battlefield and then wait several days for its effects to be realized.

When the terrorist threat is considered, however, the problem becomes immense. The aim of a potential terrorist often may simply involve publicity for a cause. In this sense, anthrax, plague, botulism, and smallpox still make excellent vehicles for terrorism. In addition, diseases not considered viable military weapons, such as those caused by human immunodeficiency virus (HIV), *Ebola, Shigella, Escherichia-coli* O157:H7, and the like, may be employed by terrorists for their “headline-grabbing” potential. This chapter limits discussion to a short list of the most viable agents typically considered in a weapons context (Table 132.1). A concise but comprehensive overview of biological warfare agents and their management is available in the U.S. Army Medical Research Institute of Infectious Diseases' (USAMRIID) monograph *Medical Management of Biological Casualties—Handbook.* An encyclopedic treatment of these agents, edited by Sidell, *Medical Aspects of Chemical and Biological Warfare* in Part 1 (Warfare, Weaponry and the Casualty) of the series *Textbook of Military Medicine,* has also been recently published.

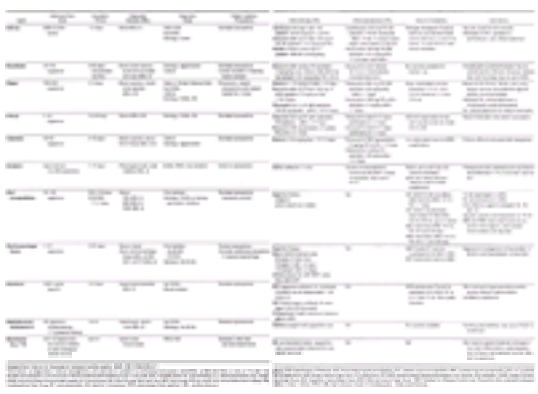

Table 132.1. Summary of Biological Warfare Agents

Anthrax

Background

Most planners agree that anthrax clearly represents the single greatest military and terrorist threat from biological warfare agents. Moreover, a World Health Organization (WHO) report estimated that 3 days after the release of 50 kg of anthrax spores along a 2-km line upwind of a city of 500,000 people, 125,000 infections would occur, producing 95,000 fatalities. This statistic represents far more fatalities than were predicted with any other agent considered. Furthermore, anthrax was made into a weapon by the United States in the 1950s (stockpiles were subsequently destroyed in 1969 when President Nixon renounced the use of biological weapons), by the Soviet Union, and by Iraq, attesting to its utility as a strategic weapon. In fact, an accident at a Soviet military compound in Sverdlovsk in 1979 resulted in at least 66 deaths caused by inhalational anthrax, an inadvertent demonstration of the viability of this weapon. None of these 66 known

victims were children. Whether this outcome is because of differences in susceptibility between pediatric and adult patients is unclear. A recent report by Elkin et al. from Canada noted a similar phenomenon in bison calves as compared to mature animals.

Pathophysiology

Anthrax is caused by infection with the Gram-positive sporulating rod-shaped bacterium *Bacillus anthracis.* The spore form of this organism is extraordinarily hardy and can survive long periods without nutrients or moisture. It lends itself well to aerosolization, drifts readily on air currents, and resists environmental degradation. Moreover, an anthrax biological weapon can be manufactured to contain particles of the ideal size (2 to 6 microns in diameter) for impinging on human lower respiratory mucosa, optimizing the chance for infection.

Most cases of endemic anthrax are cutaneous in nature and are contracted by close contact with the hides, wool, bone, and other byproducts of infected herbivores, principally cattle, sheep, and goats. Cutaneous anthrax is amenable to therapy with any number of antibiotics and is thus rarely fatal. Although common in parts of Asia and sub-Saharan Africa, anthrax is rare in the United States, the last case having been reported in 1991. Gastrointestinal (GI) anthrax is contracted via the consumption of contaminated meat and is also rare. Inhalational anthrax, also called woolsorter's disease, has been an occupational hazard of abattoir and textile workers; immunization has all but eliminated the hazard for these workers in western nations. However, this inhalational form of the disease poses a significant threat when anthrax is used as an aerosol weapon.

Upon inhalation of the necessary inoculum (estimated to be 8,000 to 10,000 spores), infection begins with the uptake of spores by pulmonary macrophages that carry the spores to lymph nodes located in the mediastinum. Here, *B. anthracis* finds a favorable milieu for growth, vegetates, and begins to produce proteins known as edema factor (EF) and lethal factor (LF). A third protein, termed protective antigen (PA), serves as a necessary carrier molecule for EF and LF and permits penetration into cells. Edema toxin results from the combination of EF and PA, whereas LF and PA together constitute lethal toxin. These toxins result in necrosis of lymph nodes and adjacent tissues, releasing more anthrax organisms. The organisms gain access to the circulation, and an overwhelming fatal septicemia rapidly ensues. At autopsy, widespread hemorrhage and necrosis, involving multiple organs, is seen.

Clinical Manifestations

Inhalational anthrax has an incubation period of 1 to 6 days. The ensuing flu-like illness is characterized by fever, myalgia, headache, cough, and chest “tightness.” A brief intervening period of improvement sometimes follows 1 to 2 days of these prodromal symptoms, and a rapid deterioration then occurs. This second phase is indicated by high fever, dyspnea, cyanosis, and shock. In some cases, chest wall edema (thought to be caused by the effect of EF) may be seen. Terminal hemorrhagic meningitis is present in up to 50% of cases. Chest radiographs obtained late in the course of illness may demonstrate a widened mediastinum and peripheral blood smears at this stage may reveal the characteristic Gram-positive spore-forming bacilli. Death is universal in untreated cases and may occur in as many as 95% of treated cases if therapy is begun more than 48 hours after the onset of symptoms.

A diagnosis of anthrax should be suspected upon finding Gram-positive bacilli with terminal spores in skin biopsy material (in the case of cutaneous disease) or in blood smears. Chest radiographs exhibiting a widened mediastinum in the proper setting of fever and constitutional signs and, in the absence of another obvious explanation (e.g., penetrating trauma), should also lead the clinician to consider a diagnosis of anthrax. Confirmation is obtained by culturing *B. anthracis* from blood.

Management

Ingelsby et al have recently published consensus-based recommendations for the management of anthrax used as a bioweapon. Endemic strains of *B. anthracis* are typically sensitive to a wide variety of antibiotics, including penicillin G, which remains the preferred drug for endemic anthrax. Because antibiotic-resistant strains of *B. anthracis* can be readily produced in laboratories, however, many experts consider ciprofloxacin (adult dosage: 400 mg intravenously every 12 hours; pediatric dosage: 20 to 30 mg/kg per day intravenously divided in two daily doses) the preferred drug for treating victims of intentional anthrax exposure. Doxycycline (adult dosage: 100 mg intravenously every 12 hours; pediatric dosage: 2.5 mg/kg intravenously every 12 hours) is considered an acceptable alternative, although doxycycline-resistant strains of *B. anthracis* are known. In cases of endemic inhalational anthrax or in which organisms are known to be susceptible, penicillin G (adult dosage: 4 million units intravenously every 4 hours; pediatric dosage: 200,000 units/kg per day intravenously in divided doses every 6 hours) is recommended.

Post-exposure prophylaxis against anthrax is achieved with oral ciprofloxacin (adult dosage: 500 mg orally every 12 hours; pediatric dosage: 20 to 30 mg/kg per day orally divided in two daily doses) or doxycycline (adult dosage: 100 mg orally every 12 hours; pediatric dosage: 2.5 mg/kg orally every 12 hours). All potentially exposed persons should be administered one of these regimens at the earliest possible opportunity. In addition to such chemoprophylaxis, victims must be immunized; based on animal data, chemoprophylaxis should be continued until a victim has received at least three doses of vaccine (thus, for a minimum of 4 weeks). The available vaccine is fully licensed, and consists of a purified preparation of protective antigen. It is given in a pre-exposure regimen at 0, 2, and 4 weeks, and at 6, 12, and 18 months. Persons at continuing risk of exposure receive yearly boosters. Exposed victims of bioterrorism would receive at least three doses, assuming no further exposure is likely, before discontinuing chemoprophylaxis. Postexposure chemoprophylaxis with antibiotics should be administered for up to 60 days if no vaccine is available for exposed individuals.

Several unique problems confront those managing the pediatric patient. First, anthrax vaccine is approved only for those 18 to 65 years of age. Second, both the quinolones and tetracyclines have relative contraindications to their use in children, although the dental staining seen with tetracyclines usually requires multiple courses and the risk of cartilage

problems associated with quinolones is theoretical. The extreme danger posed by anthrax exposure seems to warrant use of any or all of these modalities in the exposed child.

Anthrax has little potential for person-to-person transmission; standard precautions are thus adequate for health care workers managing anthrax victims. Moreover, given the 1- to 6-day incubation period, decontamination of victims presenting days after exposure would rarely be necessary.

Plague

Background

Plague, caused by infection with the Gram-negative bipolar-staining bacillus, *Yersinia pestis,* is usually transmitted in nature via the bite of fleas. This era is the plague's third global pandemic, and disease is still seen in areas of South and Southeast Asia, as well as in South America and Africa. Plague has long appeared attractive to bioweaponeers. Testimony to its extreme lethality and infectivity can be obtained by considering that the “Black Death” eliminated one-third of the population of Europe during the Middle Ages. Even then, its potential as a weapon was noted when Tatar invaders, in 1346, catapulted infected corpses over the walls of Kaffa (now Fyodosia) in the Crimea in an attempt to infect defenders within the city. Plague was extensively studied by the Japanese Unit 731 in occupied Manchuria in the 1930s and was suspected to have occupied a prominent place in the Soviet bioweapons arsenal.

Pathophysiology

Y. *pestis* is a facultative intracellular pathogen that is able to survive temporarily within the macrophage, thus aiding its dissemination to distant sites following inoculation or inhalation. It is lymphotrophic, and significantly tender regional lymphadenopathy (e.g., in the distribution of a flea bite) is often a prominent feature of bubonic plague. A plasminogen activator and coagulase produced by the organism contribute to the spread of the organism throughout the body. Pneumonic plague (along with smallpox) is one of the few bioterrorist threats readily transmissible from person-to-person via the respiratory route, and coughing patients are often highly contagious.

Clinical Manifestations

Bubonic plague is characterized by the classic bubo, a tender, enlarged, fluctuant lymph node in the distribution of the infected flea bite. Fever and malaise are usually present. Bubonic plague may progress to septicemia as bacteria gain access to the circulation; 80% of bubonic plague victims have positive blood cultures. Petechiae, purpura, and overwhelming disseminated intravascular coagulation (DIC) may develop.

Pneumonic plague may arise secondarily after seeding of the lungs or may be seen primarily after aerosol exposure. Symptoms include high fever, chills, malaise, fatigue, headache, and cough. Chest radiographs may reveal a patchy or consolidated bronchopneumonia, and the classic clinical finding is one of blood-streaked sputum; DIC and an overwhelming sepsis may develop as the disease progresses. Meningitis develops in 6% of cases. Untreated pneumonic plague has a mortality rate approaching 100%.

A presumptive diagnosis of plague can be made by observing the classic bipolar-staining “safety-pin”–like bacilli in Gram or Wayson stains of sputum, aspirated lymph node material, or cerebrospinal fluid. Confirmation is obtained via blood, sputum, or aspirate culture; the organism grows on standard blood or MacConkey's agars but is often misidentified by automated systems.

Management

Traditionally, streptomycin (15 mg/kg intramuscularly every 12 hours) has been the preferred drug for treating all forms of plague. Because of difficulties in obtaining this drug, however, clinicians have turned to alternatives, such as gentamicin (1.75 mg/kg intravenously every 8 hours) or doxycycline. Chloramphenicol (75 to 100 mg/kg per day in four divided doses) should be used in cases of plague meningitis. To be effective, therapy for pneumonic plague must be initiated within 24 hours after the onset of symptoms.

Post-exposure prophylaxis should be administered to asymptomatic victims of a bioterrorist attack, as well as to close contacts (including medical personnel) of pneumonic plague victims. Prophylaxis consists of tetracycline (25 to 50 mg/kg per day orally in four divided doses for 7 days) or doxycycline (2 to 4 mg/kg per day orally in two divided doses for 7 days). Again, although tetracyclines are relatively contraindicated in children, the potential severity of plague would seem to warrant their pediatric usage in this context. Trimethoprim—sulfamethoxazole may be used in tetracycline-intolerant patients. Droplet precautions should be employed in cases of suspected pneumonic plague. Such precautions should be continued in confirmed cases until sputum cultures are negative. Standard precautions are adequate in managing bubonic plague victims. Given the incubation period, decontamination would rarely be necessary in a clinical setting. A licensed plague vaccine exists, but production difficulties have made it problematic to obtain. This vaccine was developed to prevent bubonic plague in endemic regions, and animal data suggest that it is unlikely to protect against the pneumonic form of the disease.

Smallpox

Background

The global eradication of smallpox represents one of the great success stories of public health, with the last endemic case occurring in Somalia in 1977. Since then, research stockpiles of variola virus have been consolidated into two WHO-approved stores: at Centers for Disease Control and Prevention (CDC) in Atlanta, and at a Russian institute in

Koltsovo, near Novosibirsk. This achievement would seem to make terrorist use of this virus impossible; however, several factors give cause for concern. First is the fear that other stockpiles already exist in the hands of belligerent nations unbeknownst to WHO. Second, the entire viral genomic sequence is known and published; therefore, it is likely only a matter of time before technology permits reconstruction of the virus. Finally, although the virulence factors of variola virus are poorly understood, it may be possible for someone to manipulate related orthopoxviruses such as monkeypox to enhance their virulence in humans and create a disease like smallpox, as noted recently by Bremen and Henderson.

Smallpox, like plague, has a history of use in warfare. In 1763, during the French and Indian War, Sir Jeffery Amherst of the British Army was assigned to capture Fort Carillon from its Indian defenders. He obtained the scabs from smallpox victims, laced blankets with material from these scabs, and passed the blankets as “gifts” to the Indians. These Indians did, in fact, succumb to smallpox and Fort Carillon fell to the British.

Several factors might make smallpox an attractive weapon to potential belligerents. First, immunity after vaccination lasts only 3 to 5 years. Vaccine is no longer in production, stockpiles are dwindling and losing potency, and susceptibility to the disease is becoming nearly universal. Second, effective therapy is lacking. Third, modern health care providers are unfamiliar with the disease. Finally, the potential for rapid spread potentially permits a terrorist to cause widespread disease and panic with a minimum of infectious material.

Pathophysiology

Smallpox has an incubation period of 7 to 17 days, permitting wide dispersal of disease by exposed persons before clinical symptoms appear. During this time, the virus replicates in upper respiratory tract mucosa, giving rise to a primary viremia. The liver and spleen are then seeded, further amplification of the virus occurs, and a secondary viremia ultimately develops. The skin is seeded with this secondary viremia, and the classic exanthem of smallpox develops.

Clinical Manifestations

Clinical illness begins rather abruptly during the phase of secondary viremia, and is characterized by fever, malaise, rigors, vomiting, headache, and backache. The classic exanthem typically begins 2 to 4 days later as macules on the face and extremities. These lesions progress in synchronous fashion to papules, then to pustules, finally forming scabs. As scabs separate, survivors are left with disfiguring depigmented scars. The rash spreads centrally to the trunk but remains more abundant at the periphery. This centrifugal distribution and synchrony distinguish smallpox from the principal differential diagnostic consideration, chickenpox, which has a centripetal distribution of lesions in varying stages of development. An enanthem usually accompanies the characteristic exanthem, and internal organs become viral targets as well. Death occurs in 30% of variola major (the predominant form of smallpox in the past) patients and typically results from visceral organ involvement. Eye involvement leads to blindness in a small number of victims. Uncommon variants with lesser (variola minor) or greater (hemorrhagic, flat-type smallpox) mortality also existed.

Management

Even a single case of smallpox would represent a grave public health emergency, as detailed recently by Henderson et al. Strict quarantine (with both airborne and contact precautions) should be instituted immediately (preferably at dedicated sites away from hospitals) for victims (until all scabs separate) as well as for close contacts (for 17 days from last exposure). Based on past experience, vaccination of victims within the first several days after exposure should be efficacious in preventing disease; vaccine may be obtained through the CDC.

Although successful at eradicating smallpox, vaccination with live vaccinia virus is fraught with complications. Formation of a “pock” at the vaccine site is believed to be necessary for development of adequate immunity. This pock contains infectious material, and scratching the lesion can lead to autoinoculation of the skin or eye. Generalized vaccinia, a self-limited, immune-mediated phenomenon, occurs in a significant minority of vaccine recipients, and more serious complications may arise if vaccine is given to immunocompromised patients, patients with atopic dermatitis (eczema), or pregnant women. To manage these complications, vaccinia immunoglobulin (VIG) must be kept on hand when contemplating a vaccination effort. VIG (0.6 mg/kg intramuscularly) may be given to vaccine recipients who experience severe complications or to significantly immunocompromised individuals exposed to smallpox. An additional concern is that current stocks of VIG are no longer FDA-approved at this time. In the case of a bona fide exposure to smallpox, however, pregnancy and atopic dermatitis would not constitute absolute contraindications to vaccination.

No licensed antiviral drugs are currently available to treat smallpox. Cidofovir has shown promise in treating other orthopoxvirus infections in animal models.

Botulism

Background

Botulism's potential as a weapon has long been recognized. Pancho Villa developed recipes for the manufacture of botulinum toxins using decaying meat and beans; he may have used these preparations as crude biological weapons against Mexican federal troops during the period from 1910 to 1912. Botulinum toxin was included in the U.S. biological arsenal in the 1950s and 1960s, and was weaponized by the Soviet Union and by Iraq. The Aum Shinrikyo cult in Japan tried unsuccessfully to disseminate botulinum toxin before deciding to release sarin in the Tokyo subway system.

Botulism occurs as a result of exposure to one of seven botulinum neurotoxins (A through G). Only types A, B, E, and rarely, F appear to cause human botulism in nature. Therefore, a licensed antitoxin having activity only against types A, B, and E is produced. This fact has potential implications to the terrorist attempting to weaponize this agent.

Pathophysiology

Botulinum toxins are produced by certain strains of *Clostridium botulinum,* a strictly anaerobic spore-forming Gram-positive bacillus commonly found in soil. Recently, a few cases of type F neonatal botulism have been described; in these cases, *Clostridium baratii* was believed to be the source of toxin. Botulism has additional unique epidemiologic considerations in infants and neonates; more extensive discussion of infant botulism, and of botulism in general, may be found elsewhere in this text (see Chapter 83 and Chapter 88). The botulinum toxins are the most toxic substances known to man, with an LD50 of 0.001 µg/kg. These toxins function at the presynaptic nerve terminal, preventing the release of acetylcholine, thereby leading to a generalized flaccid paralysis and autonomic symptoms.

Clinical Manifestations

Following a latent period ranging from 24 hours to several days, victims begin to experience cranial nerve dysfunction, manifesting as bulbar palsy, ptosis, photophobia, and blurred vision owing to difficulty in accommodation. Symptoms progress to include dysarthria, dysphonia, and dysphagia. Ultimately, a descending, symmetric, flaccid paralysis ensues. The mucous membranes are dry; this fact, along with mydriasis, the nature of the paralysis (lack of initial fasciculations), and the latent period, all differentiate botulism from nerve agent intoxication. A solitary case of botulism must also be differentiated from myasthenia gravis, Guillain-Barré syndrome, tick paralysis, and a few other uncommon neurologic disorders. The presence of multiple casualties with similar symptoms makes the diagnosis of botulism rather straightforward. Botulism deaths result finally from paralysis of respiratory muscles.

Management

Supportive care, with meticulous attention to ventilatory support, remains the mainstay of botulism management. Patients may require such support for several months, making the management of a large-scale botulism outbreak especially problematic in terms of medical resources.

A trivalent (types A, B, and E) botulinum antitoxin is licensed and available through the CDC. Although administration of antitoxin is unlikely to reverse disease, it may be useful in preventing progression when administered to exposed persons. The antitoxin is prepared from horse serum and requires that a test dose be administered before therapy; patients reacting to this test dose require desensitization prior to treatment. An investigational heptavalent despeciated (Fab2) antitoxin, also produced in horses, is available through USAMRIID on a compassionate use protocol, and has been used on rare occasion to treat type F botulism in neonates. Administration of a test dose is also required with this product because it still contains approximately 4% whole antibody. A human botulinum immunoglobulin is being used on an investigational basis by the California State Health Department to treat infant botulism cases in California (see Chapter 83 and Chapter 88).

Other Agents

Numerous other agents may present bioterrorist threats of varying degrees. In addition to previously discussed incidents, terrorists and belligerents have attempted to use *Salmonella, Shigella,* Glanders, ricin, Cholera, Typhus, and probably many other organisms or toxins to induce disease. Many of these agents are discussed adequately elsewhere in this and other texts; several others warrant additional comment (see Table 132.1).

Venezuelan equine encephalitis makes an attractive weapon because of its high infectivity; virtually all non-immunes contracting the virus become symptomatic. In adults, this disease is usually self-limiting, with few patients developing encephalitis. In infants and young children, however, the disease can be severe, with as many as 4% developing overt encephalitis, often leading to permanent sequelae and death. In nature, the disease is transmitted via the bite of *Culex* mosquitoes. When delivered intentionally via aerosol, access to the olfactory bulbs may produce more rapid and/or more severe neurologic disease. Treatment is supportive.

Tularemia is another disease with high infectivity; as few as 10 organisms constitute an infectious dose in humans. Given this high infectivity, clinical laboratories should be alerted to the possibility of tularemia before attempting to culture the causative agent, *Francisella tularensis.* Several clinical forms of tularemia are known, but inhalational exposure in a terrorist attack would presumably lead to pneumonia or to the typhoidal form of the disease and would manifest as a variety of non-specific symptoms, including fever, prostration, weight loss, and abdominal pain. Treatment is accomplished with streptomycin or gentamicin at dosages similar to those recommended for plague. Post-exposure prophylaxis consists of doxycycline, tetracycline, or ciprofloxacin given for 2 weeks. Given the potential problems with these drugs in young children, prophylaxis should not be undertaken without adequate cause.

Staphylococcal enterotoxins are often mentioned as biological threat agents, and staphylococcal enterotoxin B (SEB) has been weaponized in the past. Although familiar to many clinicians as a common cause of food poisoning, SEB would also be a potent toxin if delivered by aerosol. Symptoms produced in this manner would begin 3 to 12 hours after exposure and consist of fever, headache, chills, myalgias, and non-productive cough. Dyspnea and chest pain accompanies high dosages of inhaled toxin. Nausea, vomiting, and diarrhea may occur as a result of inadvertently swallowed toxin. Treatment is supportive; meticulous attention should be paid to fluid management. Patients may be ill for as long as two weeks with aerosol exposure.

Various fungal toxins, such as the trichothecene mycotoxins, have been mentioned in a biowarfare or bioterrorism context. After the Vietnam War, the U.S. government accused the Soviets of using a trichothecene toxin, T-2 (otherwise known as “yellow rain”) against Hmong tribesmen. The Iraqis are known to have weaponized another fungal toxin, aflatoxin, which in addition to acute clinical effects is a potent hepatic carcinogen. Trichothecene toxins are of particular importance to those clinicians treating children because young infants may possess a unique susceptibility to the effects of these toxins. From 1993 to 1996, a cluster of acute pulmonary hemorrhage cases occurred among infants in

Cleveland, Ohio. In a case control study, homes of affected infants were found to have greater amounts of *Stachybotrys atra,* a fungus that appears to thrive on damp building materials, than did control homes. *S. atra* produces trichothecene toxins, which were implicated in these infants' illnesses. Symptoms produced by various mycotoxins are variable and depend on the route of exposure. The trichothecene mycotoxins are different from virtually all other bioterrorist agents in that they are dermally active. Treatment is supportive.

Chemical Agents

Background

The history, toxicology, and medical management of chemical warfare agents have recently been reviewed in an excellent monograph prepared by the U.S. Army Medical Research Institute of Chemical Defense (USAMRICD) at Aberdeen Proving Ground, Maryland. An encyclopedic discussion of these agents can be found in the Army's recently released volume, *Medical Aspects of Chemical and Biological Warfare,* in Part 1 (Warfare, Weaponry, and the Casualty) of its series *The Textbook of Military Medicine.* Chemical warfare dates back to antiquity, when the Chinese used arsenical smokes in about 1000 BC and Spartan allies used burning sulfur and coal smoke to attack the Athenian-held fort of Delium in 423 BC, during the Peloponnesian war. The 19th and early 20th centuries saw early but unsuccessful attempts to limit chemical warfare on ethical grounds via the Brussels Convention of 1874, and the Hague conventions of 1899 and 1907. World War I became the venue for the first large-scale use of such agents, particularly the gases chlorine and phosgene and the vesicating agent sulfur mustard. Thousands of casualties occurred because of pulmonary, cutaneous, and ocular effects.

The Geneva Protocol of 1925 proscribed the first use, although not the possession or retaliatory use, of biological and chemical weapons, and has since been signed by most nations. In the 1930s, the German chemical industry synthesized potent organophosphate compounds in the search for improved insecticides. These compounds were weaponized during World War II as tabun (GA) and sarin (GB), the first of the nerve agents. Neither side is believed to have used chemical agents in battle during World War II, with the possible exception of poorly documented attacks by Japan against China. However, both Allied and Axis powers produced and stored enormous amounts of both vesicants and (after the war) nerve agents, leaving a heritage of chemical-weapon stockpiles throughout the world today. Iraq is known to have used chemical agents against Iran and also its own Kurdish population during the Iraq–Iran war in the 1980s. During the Gulf War, the existence of the Iraqi chemical agent armamentarium was well known, and led to an increased determination to prepare and defend American military forces against such an attack. Finally, as previously noted, religious terrorists used sarin against civilians in Japan in 1994 and 1995.

Most chemical warfare agents are liquids at room temperature, but may exist as aerosols of tiny droplets after dispersal by munitions. Liquids are also volatile to varying degrees and thus may vaporize into a gaseous phase, particularly in conditions of high temperature, strong wind, and deposition onto relatively non-porous surfaces. A few agents, for example chlorine, phosgene, and hydrogen cyanide, exist primarily as gases in typical summertime conditions. Chemical agents may also be characterized by their environmental persistence, which is inversely related to volatility. Persistent agents, such as mustard and the nerve agent VX, pose a greater secondary contamination hazard from exposed terrain or material, or via contact by rescue and/or health care workers with a victim's clothing or skin than do the nonpersistent agents chlorine, phosgene, hydrogen cyanide, and the G nerve agents.

Toxic effects from chemical agents may occur via topical injury to the skin, the eyes, and the respiratory epithelium of the respiratory tract and via systemic absorption. Although chemical agents are volatile and dermally active, biological agents in general possess neither of these properties. Most of the modern medical literature on the clinical effects of chemical warfare agents centers on the nerve agents, which are emphasized in this chapter. Clinical syndromes related to vesicants, pulmonary agents, cyanide, and riot-control agents are also briefly summarized (Table 132.2). General principles of supportive care for poisoned patients are detailed in Chapter 88, and apply largely to the general support of chemical warfare agent victims as well.

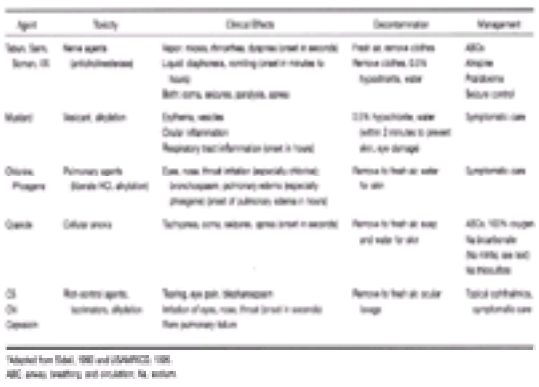

Table 132.2. Principal Chemical Warfare Agents*

Nerve Agents

Nerve agents are organophosphorus compounds, and, similar to organophosphate insecticides, are potent and essentially irreversible inhibitors of acetylcholinesterase (see Chapter 88). Certain oximes can dissociate bound nerve agents from acetylcholinesterase but only initially; after a variable period (depending on the structure of the nerve agent), a portion of the organophosphate is cleaved (in a process called aging) and the resulting nerve-agent–cholinesterase complex becomes refractory to oxime action. The “G” (for “German”) nerve agents, developed in Germany just before and

during World War II, include GA, or tabun; GB, or sarin; and GD, or soman (“GC” was allegedly not used to avoid confusion with the common designation for gonorrheal infection); VX (reportedly “Venom” X) was developed by Great Britain and the United States in the late 1940s and early 1950s. All four nerve agents are liquids at temperate conditions, but may be aerosolized by spraying or during an explosive detonation.

The G agents are moderately volatile, and relatively nonpersistent; the most volatile, sarin, evaporates at almost exactly the same rate as does water. Although VX is minimally volatile, potentially lasting weeks or longer on contaminated surfaces, at temperatures above 100°F (37.8°C) it can cause inhalational toxicity. The time required for these agents to undergo aging varies from a few minutes for soman to 48 hours for VX. The nerve agent vapors are all heavier than air, and would thus affect persons closer to the ground (e.g., those in trenches and basements, and perhaps young children) disproportionately. Although all these agents are hazardous by ingestion, inhalation, and cutaneous absorption, the primary danger from the G agents is inhalation of vapor, whereas VX is primarily a skin-contact hazard except when it is explosively aerosolized.

Toxicology

Nerve-agent–induced inhibition of acetylcholinesterase causes the neurotransmitter acetylcholine to accumulate in cholinergic synapses and in neuromuscular and neuroglandular junctions; this excess of acetylcholine initially causes end-organ stimulation which may then lead to end-organ failure. Cholinergic sites are found in the central nervous system (CNS), in the neuromuscular junctions of somatic nerves, and in several autonomic nervous system sites, including parasympathetic nerve endings, some sympathetic nerve endings (e.g., sweat glands), and both parasympathetic and sympathetic ganglia. The cholinergic syndrome thus produced is classically divided into CNS, nicotinic (neuromuscular junction and sympathetic ganglia), and muscarinic (smooth-muscle and exocrine-gland) effects. CNS effects include altered mental status progressing through lethargy to coma, ataxia, convulsions, and respiratory depression (central apnea). Although seizure initiation is probably a result of excess cholinergic stimulation, other mechanisms, including excitatory glutamate receptor stimulation and antagonism of inhibitory gamma-aminobutyric acid receptors, may also play important roles in seizure propagation. Neuropathologic changes observed in animal studies also suggest that prolonged seizure activity further disturbs excitatory amino acid and *N*-methyl-D-aspartate receptor function, ultimately leading to neuronal calcium influx and neuronal injury, as recently reviewed by McDonough and Shih.

Nicotinic effects include muscle fasciculations and twitching, and then weakness progressing to flaccid paralysis. Nicotinic effects on sympathetic activity may also result in tachycardia, hypertension, and metabolic aberrations (e.g., hyperglycemia, hypokalemia, metabolic acidosis). Muscarinic toxicity is manifested by 1) ocular findings (miosis, visual blurring, eye pain, and lacrimation); 2) respiratory findings (watery rhinorrhea, bronchospasm, and increased bronchial secretions causing cough, wheezing, dyspnea, and cyanosis); 3) dermal findings (flushing and sweating); 4) GI findings (salivation, nausea, vomiting, and diarrhea progressing to fecal incontinence and abdominal cramps); 5) genitourinary findings (frequency, urgency, and incontinence); and 6) cardiovascular findings (bradycardia, hypotension, atrioventricular block). Because muscarinic effects on the heart are opposed by the cardiovascular effects of nicotinic hyperstimulation at autonomic ganglia, heart rate and blood pressure in nerve-agent victims may be either elevated or depressed and are not reliable indicators of the severity of nerve-agent intoxication.

Clinical Presentation

The clinical presentation in a given patient depends on dose and route of exposure. For vapor exposures, mild toxicity would be suggested by miosis, rhinorrhea, mild dyspnea, and wheezing—all local effects caused by contact of vapor with epithelial surfaces. As the dose increases, and systemic distribution of the agent occurs, the victim might experience increased respiratory secretions and dyspnea, nausea, vomiting, and muscle weakness. In the Tokyo experience with sarin vapor exposure, miosis (99%), dyspnea (63%), nausea (60%), and headache (74%) were particularly common among moderately symptomatic patients at hospital admission. In severe cases with exposure to high vapor concentrations, rapid onset of paralysis and seizures leading to death from respiratory arrest may occur within minutes. In the Tokyo sarin incident, 3 patients of 640 presented to one ED in cardiopulmonary arrest. One patient was unresponsive to initial resuscitation, one patient experienced severe hypoxic damage and died on hospital day 28, and one patient was extubated after one day and recovered fully.

With vapor inhalation, affected patients do not typically deteriorate once they are removed from the exposure. In contrast, with dermal exposure, symptoms may progress even after the agent is removed from the skin surface. Initial findings after a small dose might include localized sweating, followed by localized fasciculations of underlying muscle. Systemic effects from larger doses of liquid usually begin with GI signs and symptoms but then progress to generalized fasciculations, muscle weakness, paralysis, convulsions, and death resulting from respiratory failure from CNS depression and respiratory muscle paralysis. Eye findings and obstructive respiratory effects tend to be less prominent in these patients. Because of the time (up to 18 hours for a small drop of VX) needed for liquid nerve agent to penetrate the skin, dermal exposures have a longer latency, and patients may not become symptomatic for several hours after exposure, even after decontamination. However, a pin-head-sized droplet (10 mg) of VX may cause sudden collapse with paralysis, apnea, and death after a latent interval of only 10 to 30 minutes.

Management

The diagnosis of nerve-agent poisoning is primarily by clinical recognition and response to antidotal therapy. Routine toxicologic studies do not identify organophosphate compounds or their metabolites in blood or urine. Measurements of acetylcholinesterase in plasma or in erythrocytes has traditionally been used to confirm organophosphate insecticide poisoning, and the activity of these enzymes is decreased after significant nerve agent toxicity as well. Erythrocyte acetylcholinesterase activity is a more accurate guide to acute toxicity, whereas measurements of plasma cholinesterase (pseudocholinesterase, or butylcholinesterase) are more useful for monitoring patient recovery during the weeks after exposure. However, correlation between cholinesterase levels and clinical effects is poor in mild to moderate exposures, and the test is rarely available on a stat basis. Treatment for symptomatic patients is indicated without awaiting

cholinesterase levels, but antidotal therapy is not needed for exposed asymptomatic patients, even if cholinesterase levels are depressed.

Treatment considerations begin with appropriate decontamination strategies and ED staff personal protection, as previously reviewed. Triage is based on patient prognosis and availability of resources. In cases of exposure to nerve agent vapor, in particular, maximum toxicity is manifested rapidly, and patients presenting with severe symptoms obviously require emergent therapy. Patients similarly exposed with milder effects will not worsen after arrival to the ED (unless they have continuing exposure from vapor trapped in and then released from clothing) and pose little threat of secondary contamination. The primary decontamination necessary for these individuals is clothing removal and perhaps hairwashing to remove agent vapor absorbed to scalp or facial hair. In contrast, patients with liquid dermal exposure require meticulous decontamination. Their skin and clothing pose a serious risk to ED personnel, and these patients may develop late-onset effects, or worsen, even after decontamination. Patients with ocular exposure should have copious eye irrigation with saline or water.

The overall approach to specific interventions for these agents focuses on airway and ventilatory support, and aggressive use of antidotes, particularly atropine. If a rapid sequence intubation is necessary, most authors discourage the use of succinylcholine because its effect is significantly prolonged (as a result of depressed cholinesterase activity). After intubation, positive-pressure ventilation may prove difficult because of bronchospasm and copious airway secretions. Because prompt administration of atropine may be life-saving in a patient with otherwise intractable bronchospasm, atropine should be administered in such a patient even before intubation and ventilation are attempted. If the patient presents with seizures, and in any patient with signs of severe intoxication, immediate administration of benzodiazepines beginning in usual anticonvulsant doses is indicated as well (e.g., diazepam, 0.1 to 0.3 mg/kg intravenously in children, 5 to 10 mg intravenously in adolescents or adults). Additional doses of diazepam may be required in such patients. Ongoing seizure activity may be missed if the patient is paralyzed by prolonged iatrogenic neuromuscular blockade or by the paralytic effects of the nerve agent toxicity, and subsequent bedside electroencephalographic monitoring may be necessary. Rapid, aggressive treatment of seizure activity is considered vital to prevent nerve agent–induced neuropathology, and prophylactic administration of benzodiazepines in severely poisoned patients at high risk for seizures is also crucial.

Antidotal therapy includes atropine for its anti-muscarinic effects and pralidoxime chloride to reactivate acetylcholinesterase. By acting as a competitive inhibitor of acetylcholine at postsynaptic and postjunctional cholinergic receptors, atropine antagonizes the effects of excess acetylcholine on smooth muscle, the heart, secretory glands, and the CNS, thus alleviating bronchospasm and increased bronchial secretions, bradycardia, and the GI effects of nausea, vomiting, diarrhea, and cramps. However, atropine does not reverse skeletal muscle paralysis. The dose is 0.02 mg/kg in children with mild to moderate exposure (minimum dose 0.1 mg), but may be increased to 0.05 mg/kg in more severely affected patients. Comparable doses in adolescents or adults are 2 and 5 mg, respectively. This treatment may be repeated every 2 to 5 minutes as needed to reverse muscarinic effects.

Miosis is not a good clinical sign for gauging atropinization, and although atropine may elevate an already increased heart rate, tachycardia by itself is definitely not a contraindication to continued use of atropine. Conversely, bradycardia by itself is not an indication for additional atropine, and an underlying cause such as hypoxia should be sought. The two major clinical endpoints for atropine administration are 1) decrease in secretions, and 2) decrease in airway resistance, manifested by decreasing resistance to ventilation in an apneic patient or decreasing dyspnea in a conscious patient. In nerve-agent poisoning, the total dose of atropine (usually no more than 20 mg during the first 24 hours in adults) needed has been observed to be considerably less than the massive quantities (sometimes more than 1000 mg) necessary in severe intentional pesticide overdoses.

Pralidoxime (2-PAM) is an oxime that cleaves the bound nerve agent from the acetylcholinesterase, and regenerates the intact enzyme if aging has not yet occurred. Although the action of this antidote is on the bound enzyme/nerve-agent complex, the observed clinical effects are predominantly nicotinic, with improved muscle strength. Pralidoxime thus complements the actions of atropine and should be given with atropine in all serious cases of nerve-agent poisoning. The dosage is 25 to 50 mg/kg intravenously over 30 minutes in children (1 to 2 g for adolescents or adults). Side effects of pralidoxime include nausea, vomiting, transient diplopia, headache, and most important, hypertension, all of which are minimized by the recommended slow infusion rate. Too rapid intravenous delivery of 2-PAM has also been reported to temporarily worsen cholinergic findings.

For severe cases without initial improvement, a repeat dose may be given immediately after the first. If some improvement is noted, a second dose 60 to 90 minutes later is warranted. Patients with nerve agent poisoning who have undergone adequate decontamination are not expected to require either numerous repeated doses or continuous infusion of pralidoxime, as has been advocated with severe insecticide overdose. Both atropine and pralidoxime may be administered intramuscularly at similar dosages if intravenous access is not readily available. In fact, the U.S. military has issued intramuscular autoinjector kits of atropine and 2-PAM to soldiers for in-the-field buddy and self-administration and is developing a combination atropine–2-PAM autoinjector. Intramuscular injection of 2-PAM in normotensive patients results in therapeutic blood levels within 10 minutes, and may hasten the action of concomitantly administered atropine.

Many authorities, including military medical specialists, have recommended routine use of anticonvulsant doses of benzodiazepines in significant cases even without observed convulsive activity, because animal studies have indicated some amelioration of subsequent seizures and morphologic brain damage with such use. However, ED personnel administering benzodiazepines prophylactically should anticipate the need for airway support because benzodiazepine-induced respiratory depression may exacerbate that caused by nerve-agent toxicity. Last, recent work by Lallement et al. suggests that a glutamate antagonist, GK-11 (gacyclidine), has synergistic anti-convulsive and neuroprotective effects with atropine and pralidoxime in a primate-soman model.

Pediatric Considerations

Little experience is available to comment on differences between pediatric and adult patients in the dose–response curve or the toxic-effect spectrum with exposure to nerve agents, or in response to therapy. The thinner skin of children might make them more susceptible to dermal absorption on a mg/kg basis in comparison to adults. Likewise, the immature blood–brain barrier in infants might increase the relative risk of CNS toxicity. One case series of anticholinesterase pesticide poisoning in children found that depressed sensorium and muscle weakness/flaccidity were more prominent than muscarinic findings. Nevertheless, more than half of these patients did demonstrate miosis (80%), tearing and excess salivation (60%), and GI findings (52%).

Also, severe organophosphate pesticide poisoning in children may certainly manifest by dramatic muscarinic findings, including respiratory compromise, in many cases (see Henretig, 1994, Chapter 88). It seems doubtful that the nerve agent toxidrome, and hence the appropriate management approach, would differ significantly in children from that in adults. During the Gulf War, 240 Israeli children were evaluated for accidental autoinjection of atropine, most commonly in the hand. None of these patients had been exposed to nerve agent. Administered doses were up to 17 times the recommended dose for age. Systemic effects occurred in 48% of victims, but no seizures, severe arrhythmias, or deaths were observed. Thus, atropine, even at high dosages, and without organophosphate nerve agent toxicity, seemed to be fairly well tolerated in this pediatric cohort.

Disposition and Prognosis

The disposition of exposed patients depends on severity of symptoms and route of exposure. Most patients presenting after vapor exposure manifest peak toxicity by the time of hospital arrival, and when their symptoms have either resolved, or abated to only mild eye findings (miosis from exposure to nerve-agent vapor may persist for up to 6 weeks), they may be discharged. After dermal exposure, symptom onset may lag up to 18 hours, and most experts recommend a 24-hour observation period even in initially asymptomatic victims.

The prognosis for apparently full recovery from even severe nerve agent poisoning appears to be good with timely life support interventions and adequate antidotal therapy. Apneic patients have recovered ventilatory function within 3 hours, and once consciousness was regained, muscle weakness and obtundation have resolved over a few days, whereas miosis and subtle mental status effects have persisted for several weeks. Nerve agents, unlike some pesticides, have not been implicated in delayed peripheral neuropathy.

Vesicants

The major vesicants, or blistering agents, are cellular poisons and include the mustards (sulfur mustard and nitrogen mustards) and Lewisite. The mustards are believed to act primarily as alkylating agents, whereas Lewisite is an organic arsenical thought to affect the thiol groups in critical cellular enzymes. Both nitrogen mustards and sulfur mustard were used as chemical-warfare agents in World War I, but only sulfur mustard is regarded as a significant concern today (one of the nitrogen mustards, HN_2 or mechlorethamine, was introduced in the 1940s as the first cancer chemotherapy drug). Sulfur mustard was used extensively in World War I, by several countries after that war, and recently by Iraq in the Iraq–Iran war of the 1980s. Lewisite is not known to have been used in battle, but concern about the potential of Lewisite as a chemical-warfare agent led British scientists to develop an antidote, British Anti-Lewisite (BAL), at the end of World War II. This compound is recognized today as an important chelating medication, finding use in the treatment of poisoning by lead and mercury, as well as by its original target metal, arsenic. However, because little clinical experience with Lewisite exposure exists, this discussion focuses on mustard.

Mustard exists as an oily, yellow to dark brown liquid with a garlic or mustard odor. It has relatively low volatility and is considered persistent, although at high temperatures vapor hazard is considerable. An estimated 80% of mustard casualties during World War I were caused by mustard vapor exposure (although 80% of World War I *fatalities* from chemical warfare agents were caused by pulmonary agents). The lethality of mustard to well-protected troops on the battlefield in World War I was less than 5%, but this agent would be far more deadly against unsuspecting and unprotected civilians, as mustard use against Abyssinia (now Ethiopia) in the 1930s and in the Iraq–Iran War of the 1980s amply demonstrated. An amount as little as one teaspoon may kill a 70-kg adult.

Mustard causes injury to rapidly reproducing cells (i.e., is “radiomimetic”), and its clinical effects are most evident on the skin, in the eyes, and in the respiratory tract. With severe exposures, the bone marrow, GI mucosa, and the CNS may also be damaged. Although mustard-induced cell injury begins within the first few minutes after exposure, clinical effects of mustard usually follow a latent period that is inversely related to dose but that is often 4 to 6 hours. Skin lesions after liquid contact begin with erythema, followed by blister formation, or if the dose is large enough, skin sloughing without blister formation. The burns are usually partial-thickness (second-degree). Blister fluid does not contain active mustard and is not hazardous. Vapor exposure results in later, and usually milder, skin injury.

Ocular lesions from vapor include conjunctival inflammation, corneal damage, and often severe lid edema. Permanent blindness is a rare complication, but many patients presenting for treatment may be functionally blind because the pain and blepharospasm induced by mustard renders them unwilling to open their eyes. Vapor-induced pulmonary effects begin with upper respiratory tract irritation, and may progress through dyspnea and a productive cough to a severe necrotizing tracheobronchitis with pseudomembrane formation. Patients may succumb to secondary bacterial bronchopneumonia. Bone marrow damage may occur in severe cases on about the third to fifth days after exposure, and manifest as progressive pancytopenia. Low leukocyte counts (less than $500/mm^3$) or a precipitous decrease in leukocyte count portend a serious risk of sepsis and death. An accident involving the explosion of a mustard-containing shell caused a heavy exposure to three children. These patients presented acutely with altered mental status and muscle activity, and two of them died 3 to 4 hours after exposure. A case series of Iranian children and adolescents exposed to mustard during the Iraq–Iran War found that compared with adults, the younger victims exhibited a shorter onset and more severe dermal lesions, attributable to the more delicate skin in this age group.

Because mustard penetrates tissue rapidly, and binds to cellular components within the first 2 to 5 minutes, the most important early intervention is immediate (i.e., at the scene) decontamination as soon as possible after exposure. Skin and eye decontamination are accomplished similarly as discussed for nerve agents. Additional, or delayed, decontamination at the time of ED arrival may still be of value in preventing further skin damage, systemic absorption, and secondary contamination of the patient and health-care workers, although it must again be pointed out that blister fluid from mustard casualties does not pose a contamination threat. No specific antidotes to mustard poisoning are available. Supportive care for skin lesions is analogous to that provided for burn injury, although fluid requirements are usually far less than with comparable body-surface-area thermal burns. Additional treatment of respiratory tract inflammation, ocular injury, and immunosuppression associated with leukopenia may be required (see Chapter 114,Chapter 95,Chapter 111, and Chapter 87, respectively).

Pulmonary Agents

Toxic inhalant agents may cause injury in several ways, including simple asphyxia by displacing oxygen, topical damage to airways or alveoli, systemic absorption through the pulmonary capillary bed, and allergic hypersensitivity reactions. Both chlorine and phosgene were used in battle in World War I, are commonly used for industrial purposes today, and are reviewed briefly in this section.

Chlorine is considered a gas with relatively low to intermediate water solubility and chemical reactivity, whereas phosgene is considered to have low solubility and reactivity. Because the initial irritant symptoms of gas exposure tend to correlate with water solubility and chemical reactivity, low-dose exposures to chlorine, and even moderate exposures to phosgene, might cause either no symptoms at all or only mild irritation of eyes, nose, and upper airways during exposure. Victims could easily dismiss these effects, thus prolonging exposure and the severity of the ultimate lung injury. Chlorine lung injury is probably mediated by both hydrochloric acid generation in the upper airway and by free oxygen radical cascade at the alveolocapillary membranes in the lower airway.

Phosgene (carbonyl chloride) is also thought to generate hydrochloric acid, contributing particularly to upper airway, nasal, and conjunctival irritation, as well as a carbonyl group that participates in acylation reactions at the pulmonary alveolocapillary membranes; the resulting leaking of fluid across damaged membranes eventually leads, after an asymptomatic period, to pulmonary edema. Phosgene lung injury may also be mediated in part by an inflammatory reaction associated with leukotriene production.

Chlorine is a dense, acrid yellow–green gas of intermediate water solubility and tends to settle close to the ground. Initial effects after mild to moderate exposure include ocular and nasal irritation, followed by cough, and progressing to a choking sensation and substernal chest tightness. Bronchospasm often occurs, especially in patients with a history of reactive airway disease. Pulmonary edema may follow significant exposures within 2 to 4 hours. Severe exposures result in the rapid onset (within 30 to 60 minutes) of pulmonary edema, in addition to the initial irritation.

Mild to moderate exposures to phosgene may be initially asymptomatic, with only the perception of a pleasant odor of newly mown hay. Thus, lung exposure time may be significant before the victim removes himself or herself from the affected area. Pulmonary edema occurs after a considerable delay, typically 4 to 6 hours, but with lower exposures as late as 24 hours after exposure. In these cases, dyspnea precedes objective clinical or radiologic findings. With higher exposures, early lacrimation may be followed by cough and dyspnea, and pulmonary edema, although still delayed, supervenes earlier than with a low-dose exposure. The pulmonary edema may be so severe as to result in hypotension from hypovolemia. The onset of dyspnea within the first 4 hours after exposure to phosgene portends the eventual development of massive pulmonary edema and a grave prognosis.

Management of exposure to pulmonary agents is primarily supportive (see Chapter 95). Decontamination is primarily removal to fresh air. Careful attention to control of pulmonary secretions, bronchospasm, and pulmonary edema, as well as to aggressive treatment of secondary bacterial infection (often occurring 3 to 5 days after exposure) is required. Enforced bed rest may delay the onset and reduce the eventual severity of phosgene-induced pulmonary edema. Steroids have not been of significant benefit and may increase risk of secondary infection; however, they may be warranted in patients with severe bronchospasm and a history of asthma. In chlorine exposures, some symptomatic relief has been reported with nebulized 2% sodium bicarbonate therapy, but the impact of this regimen on pulmonary damage is unknown. Animal models have suggested a benefit of anti-inflammatory agents, including ibuprofen and *N*-acetylcysteine, to ameliorate phosgene-induced pulmonary edema, but these interventions have not yet been reported in clinical trials.

Cyanide

Compounds containing the cyanide ion (CN^-) have a long history as favored agents for homicide and suicide, but their efficacy as chemical warfare agents is somewhat limited by their volatility in open air and, on the battlefield, by their flammability. However, if released nonexplosively in a crowded, closed room, they could have devastating effects. Chemical agents containing cyanide include the liquids hydrocyanic acid (hydrogen cyanide, HCN) and cyanogen chloride (ClCN), both of which rapidly vaporize after release.

Cyanogen chloride may cause some initial eye, nose, and airway irritation from its chlorine moiety, but its systemic effects are the same as those of hydrocyanic acid and result from toxicity of its cyanide anion. Hydrogen cyanide dissociates only minimally to hydrogen ions and cyanide, but the intact molecule (HCN) appears to act by the same mechanism as the cyanide anion itself.

Toxicology

Some cyanide is normally present in human tissues and several pathways exist for its metabolism. Cyanide reacts

reversibly with metals such as ferric ion (Fe^{3+}) and cobalt; in the body, the reaction of hydroxocobalamin with cyanide yields cyanocobalamin, or vitamin B_{12}. Cyanide also reacts with sulfur-containing compounds. The enzyme rhodanese detoxifies cyanide by catalyzing its reaction with a sulfur donor to form the relatively nontoxic thiocyanate and sulfite ions, which are then renally excreted. The ability of the body to metabolize small quantities of cyanide given sufficient time accounts for the dependence of cyanide toxicity on conditions of concentration and exposure time. The same amount of cyanide that will kill when given over a few minutes may be successfully metabolized by the body if administered over several hours.

Doses of cyanide large enough to overwhelm normal metabolism inhibit electron transport at the cytochrome aa_3 complex (cytochrome oxidase) of the mitochondrial cytochrome chain. The inactivation of this enzyme site, critical to aerobic adenosine triphosphate (ATP) production, results in cellular anoxia and a decreased arteriovenous oxygen difference (from inability of cells to utilize delivered oxygen), as well as in lactic acidosis (from attempts to generate energy anaerobically).

Clinical Manifestations

Clinical manifestations of cyanide poisoning relate to cellular anoxia, and thus those organs that are metabolically most active, particularly the brain and heart, are most severely affected. The carotid body chemoreceptors, which receive the highest relative blood flow and oxygen delivery of any tissue in the body, are rapidly stimulated by the presence of high concentrations of cyanide, and mediate a pronounced gasping reflex, which increases rate and depth of respiration. They also indirectly stimulate the adrenal medulla to release epinephrine, with resulting initial tachycardia and hypertension. Thus, high concentrations of cyanide vapor initially produce tachypnea, hyperpnea, and hypertension within 10 to 15 seconds. Anoxic injury to the CNS and myocardium soon follow, with unconsciousness and seizures (30 seconds after exposure), opisthotonus, trismus, decerebrate posturing, bradycardia, arrhythmias, hypotension, and eventually cardiac arrest (as soon as 4 to 8 minutes after exposure).

Exposure to low concentrations of vapor produces nonspecific effects such as headache, light-headedness, nausea, and ataxia. “Classic” signs of cyanide poisoning are said to include severe dyspnea without cyanosis, or even with cherry-red skin (because of lack of peripheral oxygen use), and a bitter almond odor to breath and body fluids. However, some cyanide-poisoned patients develop cyanosis, and only about half the population is genetically capable of detecting the cyanide odor. Noteworthy laboratory abnormalities in cyanide poisoning include an abnormally high mixed venous oxygen saturation (because of decreased peripheral oxygen utilization), a decreased arteriovenous oxygen content difference, and a high anion gap metabolic acidosis (because of increased lactic acid production with the shift to anaerobic metabolism).

Sidell has emphasized that in a chemical attack, the observation that people are convulsing or dying within minutes of exposure implies the weapon is either cyanide or a nerve agent. With high concentrations of cyanide, seizures begin within seconds and death ensues within minutes, usually with little cyanosis or other findings. Exposure to lethal concentrations of a nerve-agent liquid or vapor may also lead to sudden collapse with pre-terminal apnea and convulsions. Cyanosis in such cases is more common than in cyanide casualties. Miosis and increased naso-ocular secretions indicates exposure to nerve-agent vapor rather than to cyanide but could be absent in a victim exposed only to nerve-agent liquid.

Management

Management of cyanide poisoning begins with removal to fresh air. Dermal decontamination is unnecessary if exposure has been only to vapor, but wet clothing should be removed, and the underlying skin should be washed with soap and water, or with water alone if liquid on the skin is a possibility. Attention to the basics of intensive supportive care is critical, and includes 1) provision of 100% oxygen to all significantly symptomatic patients (regardless of arterial P_{O_2}), 2) mechanical ventilation as needed, 3) circulatory support with crystalloid and vasopressors, 4) correction of metabolic acidosis with intravenous sodium bicarbonate, and 5) seizure control with benzodiazepine administration. The cyanide-induced inhibition of cellular oxygen use might lead to the expectation that supplemental oxygen would not be of use in cyanide poisoning, but in fact, administration of 100% oxygen has been found empirically to exert a beneficial effect, possibly by directly displacing cyanide from cytochrome oxidase binding sites.

Symptomatic patients, especially those with severe manifestations, may further benefit from specific antidotal therapy, which is provided in a two-step process. First, a methemoglobin-forming agent such as amyl nitrite (available as crushable ampoules for inhalation in the pre-hospital setting, or when intravenous access is not immediately available) or sodium nitrite (for intravenous use) is administered. The ferric ion (Fe^{3+}) in methemoglobin has an even higher affinity for cyanide than does cytochrome aa_3. The equilibrium of this reaction causes disassociation of bound cyanide from the cytochrome oxidase and restores aerobic energy production.

Nitrites may also have therapeutic efficacy independent of methemoglobin formation, possibly via conversion to nitric oxide with subsequent beneficial vasodilatory effects. However, nitrite administration is potentially hazardous because too rapid intravenous infusion may cause or exacerbate hypotension, and overproduction of methemoglobin may compromise oxygen-carrying capacity. Thus, nitrite is probably not indicated for conscious patients with minimal symptoms, and is relatively contraindicated in patients whose cyanide toxicity is complicated by existing impaired oxygen delivery (e.g., smoke inhalation victims from a house fire, with likely concomitant lung injury and carbon monoxide poisoning).

These potential adverse effects of nitrites would obviously be less compelling in the context of a severely intoxicated, prostrate casualty of a terrorist cyanide vapor attack, and careful attention to proper dosing and rate of administration should allow safe use of this antidote. Pediatric nitrite dosing depends on body weight and hemoglobin concentration. The recommended initial pediatric dosage, assuming hemoglobin concentration of 12 g/dL, is 0.33 mL/kg of the standard 3% sodium nitrite solution, given slowly intravenously over 5 to 10 minutes; the initial adult dosage is 10 mL, equivalent

to one of the two sodium nitrite vials in the standard Pasadena (formerly Lilly) Cyanide Antidote Kit. Dosing may be adjusted for patients with significant anemia, although this knowledge would rarely be available in the context of emergent treatment of a critically poisoned child.

The second step is provision of a sulfur donor, typically sodium thiosulfate, which is used as a substrate by rhodanese for its conversion of cyanide to thiocyanate. Thiosulfate itself is efficacious, relatively benign, and also synergistic with oxygen administration, and thus may be used without nitrites in situations such as smoke inhalation. The initial thiosulfate dose for children is 1.65 mL/kg of the standard 25% solution intravenously, and the initial adult dose is 50 mL, equivalent to one of the two large bottles in the Pasadena kit. Second treatments with each antidote may be given at up to half the original dose if needed. Several alternative therapies and experimental antidotes are used in Europe (DMAP, or 4-dimethylaminophenol, a methemoglobin former; and cobalt compounds such as dicobalt edetate or hydroxocobalamin, as cobalt itself binds cyanide) or undergoing clinical trials (other aminophenol derivatives, also methemoglobin formers; aldehydes, which are cyanohydrin formers).

Riot-Control Agents

Riot-control agents, also called lacrimators (“tear gas”), include several compounds, the most important of which are CS (*o*-chlorobenzylidene malononitrile), CN (1-chloroacetophenone, “Mace”), and pepper spray (containing capsaicin).

CS and CN are solids and are typically dispersed as an aerosol of fine particles. Although the United States does not consider riot-control agents to be official chemical-warfare weapons, these agents are widely available, cause significant incapacitating effects in closed spaces, and could conceivably be used in a terrorist attack; therefore, they are outlined briefly in this section. As a group, these agents are often used by law enforcement agencies to cause temporary incapacitation for riot control and related situations. Their mechanism of action after low-level exposure is unclear but, in the case of pepper spray, appears to be related to the release of the pain-modulating neurotransmitter substance P. All of these agents cause 1) transient ocular effects, including burning sensation, tearing, blepharospasm, and photophobia; 2) irritation of the nose, throat, and upper airway; and 3) skin burning, erythema, and vesication (at high concentrations and high ambient temperatures and humidity). A few riot-control agents, such as Adamsite (DM), cause pronounced vomiting in addition to irritating the eyes and the upper airway and are referred to as vomiting agents. Most victims, under usual circumstances of exposure, become symptomatic within seconds, but remain so for only 20 to 60 minutes. However, high concentrations in closed spaces or discharge of agent close to the victim's face have been associated with serious medical complications, including severe ocular toxicity, dermal burns, and pulmonary failure. A few lethal cases have been described in which death was caused by severe tracheobronchitis with pseudomembrane formation and pulmonary edema.

Management includes careful ocular and dermal decontamination. The skin should be washed with soap and water, although this may cause transient increased pain. Hypochlorite solution should not be used because it may exacerbate dermal burns via the creation of toxic byproducts. The eyes should be thoroughly lavaged after a single dose of topical anesthetic if necessary. Respiratory complications must be managed supportively, as previously described for mustard and pulmonary agent toxicity. Severe respiratory effects may not manifest for 12 to 24 hours; therefore, patients with dyspnea or any objective findings should probably be observed in the hospital. Severe respiratory complications from exposure to riot-control agents have been described in at least two young infants, one of whom was in a house into which CS was sprayed. A canister of pepper spray was accidentally discharged directly into the face of the other infant. Both survived with prolonged care, the latter requiring ventilatory support including 5 days of extracorporeal membrane oxygenation. A few cases of children ingesting CS powder are known, which resulted only in transient diarrhea and abdominal cramping.

EMERGENCY DEPARTMENT AND EMERGENCY MEDICAL SERVICES PREPAREDNESS

Overview

The appropriate ED and EMS preparation for a medical response to terrorist incidents involving WMD is currently the subject of intense activity by the U.S. government, including the military and numerous civilian federal agencies. In addition, preparation is being actively pursued at state and local levels by regional EMS and HAZMAT systems, police and fire departments, and individual hospitals, poison control centers, and physicians with various backgrounds, including public health, toxicology, infectious disease, and emergency medicine. The 1996 Centennial Olympic Games in Atlanta provided a recent opportunity to conduct a highly orchestrated “disaster drill” preparation for a possible terrorist incident involving chemical or biological weapons. A comprehensive review of this experience, highlighting the coordinated efforts by local, state, federal, and military resources, was reported by Sharp et al. The preparedness of various cities and localities to manage the medical consequences of a terrorist chemical or biological attack varies from well prepared to totally unprepared. The pre-hospital and community-wide responses to terrorist chemical or biological attacks are obviously of critical importance, but a detailed analysis is beyond the scope of this chapter.

In brief, local first-responders, typically police, fire department, and EMS personnel, would be first at the scene of a chemical attack, and would be responsible for on-scene extrication, decontamination, triage, emergent field treatment, and evacuation of patients to area hospitals. They would also be at greatest risk of personal injury. Local hospital and health care providers would provide care to the first wave of casualties on short notice, and, based on the Tokyo experience, would likely receive numerous unannounced patients transported by private vehicle or other non–EMS-based means. Many such health care workers would be exposed initially to chemical agents if patients arrive before an accurate understanding of the event has occurred. If the consequences of the attack overwhelm local resources, mechanisms are in place for regional and state authorities to activate the federal response system, although depending on the locality involved this response may take several hours to activate.

Emergency Department Issues

Hospital EDs will likely be involved in WMD incidents in one of two characteristic scenarios, as recently reviewed by Barbera et al. A “sudden” presentation would result from a chemical attack with resulting large numbers of symptomatic victims presenting almost immediately and with little advance warning. Similarly, an announced biological agent release, or discovery of such a release, might result in numerous patients seeking health care services, although in most cases victims would initially be asymptomatic. The second scenario involves an “insidious” presentation (not unlike a natural epidemic in some ways) with sub-acute onset of unusual clinical symptoms in a number of patients presenting to diverse health care facilities, most likely as a result of biological agent attack. In this latter scenario, health care providers must recognize the pattern as a potential WMD exposure, as detailed previously under the discussion regarding epidemiologic aspects. In both presentations, many persons who have sustained either minimal exposure, or no exposure at all, will experience stress-related symptoms (the “worried-well”), and likewise present for services, placing further demands on EDs already likely to be overwhelmed. In either scenario, initial suspicion of the WMD incident should trigger ED protocols for initiation of the previously noted community-wide response, notification of public health authorities, and rapid deployment of the already-established decontamination structure and process.

The ED response to WMD incidents needs to be integrated into the hospital's standing disaster plan. Appropriate protocols for calling in extra help, using hospital security for patient direction and diversion at the ED entrance and around the decontamination site, and handling the dissemination of information to the public and news media all should be anticipated. Hospital spaces that are not routinely used for patient care, such as cafeterias, may be used as holding areas for large numbers of exposed but minimally symptomatic patients. Routine hospital supplies such as gowns and towels may be depleted rapidly in the face of mass casualties. Demands for hospital beds, and particularly intensive care unit beds, are likely to be overwhelming. Alternative care facilities staffed by outside help may be needed in mass casualty situations (e.g., warehouses or other such buildings might need to be converted to temporary care sites). Pre-disaster planning for both “natural” disasters and WMD incidents must take into account such factors. As previously noted, a framework exists for activating medical assistance plans at the federal level through the Federal Response Plan, and the National Disaster Medical System, augmented if necessary by military medical assets from the Department of Defense.

The issue of stocking specific antidotes, medications, and vaccines in the context of planning for a WMD event involving the potential for mass casualties poses additional challenges. Many hospitals do not routinely stock adequate amounts of such pharmaceuticals for even one critical patient. WMD incident planning should establish some mechanism for local or regional stockpiling of these critical medications, and/or a means to rapidly acquire them. For some medications, such as atropine, lower-cost alternatives such as rapid bulk preparation from pharmaceutical-grade powder may be an attractive option. Table 132.3 offers an attempt to quantify the magnitude of antidotal medications that might be needed in one ED for the management of a nerve agent or cyanide attack involving both pediatric and adult victims, on a scale of the Tokyo sarin attack. A biological agent attack would place similar enormous demands on the hospital pharmacy for antibiotics, vaccines, antitoxins, and so on. Funds have been allocated for a federal stockpile of pharmaceuticals, but in some cases (such as with smallpox vaccine) no current production capability exists.

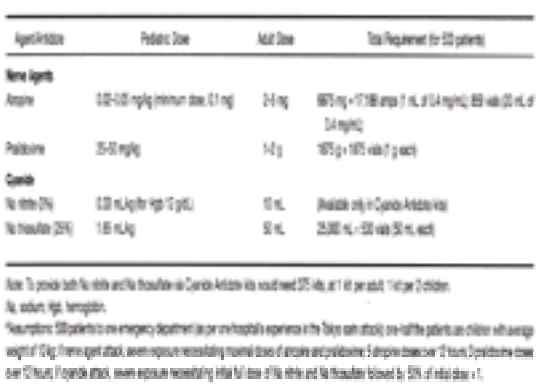

Table 132.3. Pharmaceutical Stocking Estimates for One Emergency Department in a Hypothetical Chemical Agent Attack[a]

In the context of a biological agent attack, concern arises regarding possible transmission of infection to health care workers and other ED patients. Most such biological agents pose minimal risk of person-to-person transmission to ED staff and other patients if standard barrier protection techniques are practiced. Nevertheless, patients with smallpox or pneumonic plague (both of which are easily transmissible by the respiratory route) and patients with viral hemorrhagic fevers should be placed into appropriate isolation as soon as possible after their identification. Further issues in such a context would include the need for isolation rooms with anterooms and special air-handling precautions (e.g., negative pressure and HEPA-filtered exhaust), large isolation wards, potential quarantine, and chemoprophylaxis. In the case of smallpox, pneumonic plague, and hemorrhagic fevers, health care worker respiratory and mucous membrane protection may have to be upgraded.

The earliest suspicion of these infections would obviously necessitate expert consultation with infectious disease and public health specialists. CDC, USAMRIID, and (through Domestic Preparedness resources) USAMRICD have 24-hour hot-line services available to aid physicians and local public health authorities in this context (CDC 770-488-7100; USAMRIID 1-888-USA-RIID; CB HelpLine [for non-emergency planning and information]: 1-800-368-6488; CB HotLine [for emergencies]: 1-800-424-8802).

Obviously, many unanswered questions exist regarding ED preparedness for a WMD attack, including 1) optimal decontamination techniques, especially for young children; 2) optimal PPE for ED staff; 3) issues regarding safety of

decontamination water run-off into public drainage systems; 4) the community-wide needs for education and training; and 5) financial considerations for individual hospital and regional planners. Continued activity on an expert consensus basis, as well as new research, should help to address many of these issues.

CONCLUSION

The prospect of a terrorist WMD incident with resulting mass casualties is unfortunately more likely now than ever before. Although the impact of such an event is almost unimaginable, at the same time, efforts must be made to prepare for the "unthinkable." Such preparedness requires highly coordinated responses involving local and regional EMS systems, HAZMAT teams, police and fire departments, hospital EDs, local and federal public health agencies, and military medical specialists. In particular, EDs must consider important issues, including 1) the early recognition, triage, decontamination, treatment, and disposition of multiple casualties of such an attack, 2) protection of health care workers and existing patients, and 3) the integrity of the ED itself to provide on-going care to later arriving casualties and to continue to meet normal patient demands.

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[1]The views, opinions, assertions, and findings contained herein are those of the authors and should not be construed as official U.S. Department of Defense or Department of the Army positions, policies, or decisions unless so designated by other documentation.

Suggested Readings

Agency for Toxic Substances and Disease Registry (ATSDR). Hospital Emergency Departments: A Planning Guide for the Management of Chemically Contaminated Patients. Atlanta: US Department of Health and Human Services, 1992.

Amitai Y, Almog S, Singer R, et al. Atropine poisoning in children during the Persian Gulf crisis. A national survey in Israel. JAMA 1992;268:630–632.

Barbera JA, Macintyre AG, Christopher GW, et al. Weapons of mass destruction events with civilian casualties: effective planning for health care facilities. 1999 (in press).

Billmore DF, Vinocur C, Ginda M, et al. Pepper-spray-induced respiratory failure treated with extracorporeal membrane oxygenation. Pediatrics 1996;98:961–963.

Breman JG, Henderson DA. Poxvirus dilemmas—monkeypox, smallpox, and biologic terrorism. N Engl J Med 1998;339:556–559.

Carus WS. Bioterrorism and biocrimes: the illicit use of biological agents in the 20th century. Center for Counterproliferation Research, National Defense University, 1998:116.

Cox RD. Decontamination and management of hazardous materials exposure victims in the emergency department. Ann Emerg Med 1994;23:761–770.

Dearborn DG, Infeld MD, Smith PG, et al. Update: pulmonary hemorrhage/hemosiderosis among infants—Cleveland, Ohio, 1993–1996. MMWR 1997;46:33–35.

Elkin BT, Gates CC, Ellsworth T, Dragon DC. Meso- and macro-ecology of anthrax in bison in northern Canada: the 1993 MacKenzie outbreak as a case study. Third International Conference on Anthrax, University of Plymouth, 1998 [abstract].

Franz DR, Jahrling PB, Friedlander AM, et al. Clinical recognition and management of patients exposed to biological warfare agents. JAMA 1997;278:399–411.

Henderson DA, Inglesby TV, Bartlett JG, et al. Smallpox as a biological weapon—medical and public health management. JAMA 1999;281: 2127–2137.

Henretig FM. Special considerations in the poisoned pediatric patient. Emerg Med Clin North Am 1994;12:549–567.

Hoffman B. Inside Terrorism. New York: Columbia University Press, 1998.

Holstege CP, Kirk M, Sidell FR. Chemical warfare: nerve agent poisoning. Crit Care Clin 1997;13:923–942.

Ingelsby TV, Henderson DA, Bartlett JG, et al. Anthrax as a biological weapon—medical and biological health management. JAMA 1999;281:1735–1745.

Lallement G, Clarencon D, Masqueliez C, et al. Nerve agent poisoning in primates: antilethal, antiepileptic and neuroprotective effects of GK-11. Arch Toxicol 1998;72:84–92.

McDonough JH Jr, Shih TM. Neuropharmacological mechanisms of nerve agent-induced seizure and neuropathology. Neurosci Behav Rev 1997;15:559–579.

Meselson M, Guillemin J, Hugh-Jones M, et al. The Sverdlovsk anthrax outbreak of 1979. Science 1994;266:1202–1207.

Momeni AZ, Aminjavheri M. Skin manifestations of mustard gas in a group of 14 children and teenagers: a clinical study. Int J Dermatol 1994;33:184–187.

Okumura T, Suzuki K, Fukuda A, et al. The Tokyo subway sarin attack: disaster management. Part 2: Hospital response. Acad Emerg Med 1998;5:618–624.

Okumura T, Takasu N, Ishimatsu S, et al. Report on 640 victims of the Tokyo subway sarin attack. Ann Emerg Med 1996;28:129–135.

Sharp TW, Brennan RJ, Keim M, et al. Medical preparedness for a terrorist incident involving chemical or biological agents during the 1996 Atlanta Olympic Games. Ann Emerg Med 1998;32:214–223.

Sidell FR. What to do in case of an unthinkable chemical warfare attack or accident. Postgrad Med 1990;88:70–84.

Sidell FR. Chemical agent terrorism. Ann Emerg Med 1996;28:223–224.

Sidell FR, Borak J. Chemical warfare agents: II. nerve agents. Ann Emerg Med 1992;21:865–871.

Sidell FR, Takafuji ET, Franz DR, eds. Textbook of Military Medicine. Part 1: Medical Aspects of Chemical and Biological Warfare. Washington DC: Office of the Surgeon General, Walter Reed Army Medical Center, 1997.

Sofer S, Tal A, Shahak E. Carbamate and organophosphate poisoning in early childhood. Pediatr Emerg Care 1989;5:222–225.

Tucker JB. National health and medical services response to incidents of chemical and biological terrorism. JAMA 1997;278:362–368.

US Army Medical Research Institute of Chemical Defense. Medical Management of Chemical Casualties. 2nd ed. Aberdeen Proving Ground, MD: US Army Medical Research Institute of Chemical Defense, 1995.

US Army Medical Research Institute of Infectious Diseases. Medical Management of Biological Casualties—Handbook. Ft. Detrick, MD: US Army Medical Research Institute of Infectious Diseases, 1998.

Vinsel PJ. Treatment of acute chlorine gas inhalation with nebulized sodium bicarbonate. J Emerg Med 1990;8:327–329.

World Health Organization (WHO). Report of a WHO group of consultants. Health aspects of chemical and biological weapons. Geneva: WHO, 1970:97–99.

APPENDIX A

Pediatric Emergency Medicine Equipment

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Emergency Department
Office or Clinic

EMERGENCY DEPARTMENT

1. **Airway**

    1.1 Tongue blades
    1.2 Suction catheters 6, 8, 10 Fr (2 of each)
    1.3 Yankauer suction tips (4)
    1.4 Magill forceps
    1.5 Oxygen catheters for suction 10, 14 Fr (2 of each)
    1.6 Oropharyngeal airways 0–5 (2 of each)
    1.7 Nasopharyngeal airways 12, 16, 20, 24, 28, 30 Fr. (2 of each)
    1.8 Humidivent
    1.9 Meconium aspirator
2. **Breathing**

    2.1 Oxygen supply
    2.2 Oxygen flow meter
    2.3 Oxygen tubing
    2.4 Cylinder key
    2.5 Oxygen masks
    2.6 Nasal cannula
    2.7 Non-rebreathing mask
    2.8 Nebulizer and administration equipment
    2.9 Self-inflating bags with oxygen reservoir (adult, infant)2.10 Mapleson D bags with reservoir (0.5, 1, 5 L)
    2.11 Laryngoscope handle with knurled finish
    2.12 Laryngoscope blades

        2.12.1 Miller 0, 1, 2, 3
        2.12.2 Wis-Hipple 1.5
        2.12.3 MacIntosh 2, 3

    2.13 Extra “C” batteries (2)
    2.14 Endotracheal tubes

        2.14.1 Uncuffed sizes 2.5–8.5 (2 of each)
        2.14.2 Cuffed sizes 7–9 (2 of each)

    2.15 Stylets (1 adult, 1 infant)
3. **Circulation**

    3.1 Stat IV tray
    3.2 Central venous pressure tray 5, 10, 11 (2 of each)
    3.3 Cutdown tray (2)
    3.4 Umbilical catheterization tray
    3.5 Intra-osseous needles—16, 18 (2 of each)
    3.6 Radial artery tray (2.5F 5 cm, 2.5F 2.5 cm)
    3.7 Drugs prepackaged

        3.7.1 Epinephrine 1:1000
        3.7.2 Dextrose (D 25%)
        3.7.3 Atropine
        3.7.4 Sodium bicarbonate
        3.7.5 Calcium chloride

3.7.6 Xylocaine 2%

3.8 Drugs

3.8.1 Adenosine
3.8.2 Afrin®Pr Nasal Spray
3.8.3 Benadryl
3.8.4 Bretylium
3.8.5 Calcium gluconate
3.8.6 Chloramphenicol
3.8.7 Cyanide kit
3.8.8 Diazepam
3.8.9 Diazoxide
3.8.10 Digoxin
3.8.11 Dilantin
3.8.12 Dobutamine
3.8.13 Gastrografin
3.8.14 Gentamicin
3.8.15 Glucagon
3.8.16 Heparin Vial
3.8.17 Hydralazine
3.8.18 Hydrocortisone
3.8.19 Isuprel
3.8.20 Ketamine
3.8.21 Furosemide
3.8.22 Lidocaine 1%
3.8.23 Lidocaine 2%
3.8.24 Mannitol
3.8.25 Flumazenil
3.8.26 Naloxone (1 mg/mL)
3.8.27 Neostigmine
3.8.28 Norepinephrine (Levophed)
3.8.29 Procainamide
3.8.30 Solu-Medrol
3.8.31 Thiopental
3.8.32 3% saline
3.8.33 Vasopressin
3.8.34 Vecuronium
3.8.35 Verapamil

3.9 IV fluids

3.9.1 Tubing
3.9.2 Stopcocks
3.9.3 Normal saline solution 1 L (10 bags)
3.9.4 Lactated Ringer's solution 1 L (10 bags)
3.9.5 Albumin 5%, 25%

3.10 Syringes
3.11 Alcohol pads
3.12 Needles
3.13 Broselow tape or wall chart
3.14 Infusion pumps (3)
3.15 Cardiac board
3.16 Arm boards
3.17 Tape
3.18 Tincture of benzoin

4. **Monitoring**

4.1 Sphygmomanometer Doppler and aneroid
4.2 Blood pressure cuffs (neonate, child, adult, large adult, thigh)
4.3 ECG/leads
4.4 Pulse oximeter
4.5 End-tidal CO_2 monitor
4.6 Doppler (hand-held)
4.7 Defibrillator/defibrillator paste
4.8 Temperature probe
4.9 Hypothermia thermometer
4.10 Blood pressure–monitoring lines
4.11 Intracranial pressure–monitoring lines

5. **Laboratory testing**

5.1 Syringes
5.2 Needles
5.3 Alcohol pads

5.4 Betadine
5.5 Tubes including culture media
5.6 Blood gas kit
5.7 Glucometer, test strips
5.8 Hemoccult cards
5.9 Sterile basins, bedpans, urinals
5.10 Evidence bags
5.11 Shroud, autopsy permits, related supplies

6. **Trauma care**

6.1 Cervical collars: Baby no-neck®Pr, Pediatric no-neck®Pr, Short, Regular
6.2 Nasogastric tubes, 10, 16 (2 of each)
6.3 Feeding tubes, 3½–8 Fr (2 of each)
6.4 Foley catheters, catheterization tray
6.5 Tracheostomy tray (2)
6.6 Thoracentesis tray
6.7 Chest tube insertion (2)
6.8 Chest tubes 12, 16, 20, 24, 28, 32, 34, 40 Fr (2 of each)
6.9 Pleurovac
6.10 Thoracotomy tray
6.11 Minor procedure tray (3)
6.12 Peritoneal tray (11Fr Dialysis set)
6.13 Obstetric pack
6.14 Blood administration sets
6.15 Blood warmer
6.16 Pressure bags
6.17 Garder-Walls tongs
6.18 Hare traction splint
6.19 Scalpels No. 10, 11, 15
6.20 Suture Material—1 box of each:

2.0 silk ties
3.0 silk
4.0 vicryl
Tevdek cardiovascular 2.0, 4.0, 5.0
TFE polymer pledgets (8677-01)
TFE polymer pledgets (8675-01)

7. **Other**

7.1 Protective supplies

7.1.1 Gloves
7.1.2 Gowns
7.1.3 Masks
7.1.4 Shoe covers
7.1.5 Protective eye shields
7.1.6 Needle receptacles

7.2 Stat worksheet on clipboard
7.3 Key phone numbers
7.4 Drug formulary
7.5 Drug labels
7.6 Scissors, heavy gauge
7.7 Flashlight
7.8 Ophthalmoscope
7.9 Otoscope
7.10 Overbed warmers
7.11 Blankets

8. **Transport equipment**

8.1 Portable suction
8.2 Portable monitors
8.3 Infusion pump
8.4 Airway box
8.5 Drug box

OFFICE OR CLINIC

1. **Airway equipment**

1.1 Oxygen tank with flow meter
1.2 Face masks
1.3 Oxygen reservoir masks
1.4 Nasal cannula

1.5 Oxygen tubing
1.6 Oropharyngeal airways—(all sizes)
1.7 Nasopharyngeal airways—(all sizes)
1.8 Suction machine, portable
1.9 Suction catheters
1.10 Yankauer suction tips (4)
1.11 Magill forceps

2. **Breathing equipment**

2.1 Bag-valve-mask with O_2 reservoir (adult, pediatric)
2.2 Masks—(infant to adult sizes)
2.3 Pulse oximeter

3. **Circulation equipment**

3.1 Cardiac board
3.2 Butterflies—25, 23, 21 (3 of each)
3.3 Medicuts—22, 20, 18 (3 of each)
3.4 Intraosseous needles (4)
3.5 Normal saline 5% dextrose, 500 mL (2)
3.6 Normal saline solution, 500 mL (2)
3.7 Solusets (2)
3.8 Sphygmomanometers—cuffs (4 sizes)
3.9 Drug box—prepackaged syringes
3.9.1 Epinephrine 1:10,000
3.9.2 Sodium bicarbonate, full strength
3.9.3 Sodium bicarbonate, half strength
3.9.4 Dextrose, 25%
3.9.5 Atropine, 0.4 mg/0.5 mL
3.9.6 Naloxone
3.9.7 Diazepam/lorazepam
3.9.8 Phenobarbital
3.9.9 Activated charcoal
3.9.10 Tourniquets
3.9.11 Betadine swabs
3.9.12 Alcohol swabs
3.9.13 Tape
3.9.14 Syringes
3.9.15 Armboards

4. **Other equipment**

4.1 Resuscitation cart checklist
4.2 Semirigid cervical collars—(adult, pediatric)
4.3 Sand-bags (3)
4.4 Splints, inflatable
4.5 Nasogastric tubes
4.6 Rubber gloves
4.7 Protective eyewear
4.8 Broselow tape or wall chart

APPENDIX B

Emergency Drug Compendium

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The purpose of Appendix B is to provide an easily accessible resource for dosages and side effects of medications included in the main text. The dosages included here are based on literature available at the time of preparation of the text. For medications that are relatively new to pediatrics (e.g., cisapride, sumatriptan, parenteral labetalol), further refinement in drug dosage recommendations may occur after publication. Other frequently updated drug information resources such as the American Hospital Formulary Service (updated quarterly) or Facts and Comparisons (updated monthly) may provide additional information. Finally, for added safety, prescribers should check the package insert and one or two additional resources for dosage information when prescribing an unfamiliar drug.

A list of the abbreviations used and their definitions is presented at the end of this appendix.

APPENDIX C

Parental Instruction Sheets

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ANIMAL BITES

Your child was bitten by an animal. This can be frightening to some children and your child may need some extra comfort to feel better.

The examining doctor cleaned the bite wound. The doctor may have given you instructions to clean the wound at home. They are:

[ ] Your child was given a **tetanus** shot in the emergency department (ED). Show this to your regular doctor to update your child's records. _____

[ ] Your doctor has decided that **anti-rabies** treatment is needed. Your child was given the first injection (shot) today. He or she will need to return to the ED for the rest of the shots.

3rd day __ 7th day __ 14th day __ 28th day__
If your child gets a fever or the needle site is red or swollen, call your doctor. All the shots must be given to protect your child.

[ ] Your doctor has given your child a prescription for **antibiotics.** Not all bites need antibiotic treatment. Your child must take all the medicine as directed. Your dose is: _____

Signs of Infection

Call your doctor or return to the ED if any of these signs develop after your visit:

1. Increased redness around the bite
2. Pain
3. Discharge or pus from the bite
4. Increased swelling
5. Bad smell
6. Fever

WOUND CARE

Your child hasan injury in which the skin was broken. These injuries can be fixed in different ways depending on the age of your child and the size and location of the injury. However, all wounds heal with a scar. This scar will remodel itself in the first 6 months.

[ ] *Stitches (Sutures) or Staples*

Your child had ___ stitches or staples placed for an injury to his or her _____.

___ These stitches must be removed in ___ days. Call your doctor for an appointment to remove the stitches. If you do not have a regular doctor_____

___ Your child's wound should be rechecked in ___ days. Call your doctor for an appointment or return to the ED.

___ The stitches do not need to be removed. They will come out by themselves. Follow up with your regular doctor in _____ days.

Washing with a washcloth at the sink is OK. Do not take a bath, soak the stitches/staples, or allow your child to swim with them. If the stitches should loosen or if the wound pops open, bring your child back to the ED. If the wound starts bleeding, apply direct pressure for 15 minutes. If it continues to bleed, call your doctor or return to the ED.

___ Apply an antibiotic ointment to the stitches ___ times per day.

___ Keep the area covered with a clean bandage.

___ No cover is necessary after the first 48 hours.

Apply sunscreen to wound area when outdoors to protect new skin.

[ ] *Steri-Strips*

Your child had paper tapes (Steristrips) used to fix the wound. Allow these to fall off by themselves (usually in 5 to 7 days). Do not soak in water or allow your child to swim with steristrips. Keep clean and dry.

___ Your child received a **tetanus** shot in the ED. Show this to your regular doctor to update your child's records.

___ Your doctor has given your child a prescription for **antibiotics.** Not all wounds need antibiotic treatment. Your child must take all the medicine as directed. Your dose is: _____

Signs of Infection

Call your doctor or return to the ED if any of these signs develop:

1. Increased redness around the wound
2. Pain
3. Discharge or pus from the wound
4. Increased swelling
5. Bad smell
6. Fever

ASTHMA

Children with asthma have a reactive airway. The tubes that carry air to the lungs and the small passages in the lungs are sensitive to many things. When triggered, the airways react by getting smaller, swelling, and forming mucous plugs. This reaction can happen with colds and viruses; with exposure to pets, dust, odors, or allergens; or with exercise and emotional stress. The treatment your child received in the ED helps open the airways and reduce the swelling.

Asthma is a condition that can affect your child for many years. Identifying something that triggers wheezing in your child may help avoid future episodes. **No smoking** should be allowed in the house because smoke irritates the airways of all asthmatics. Your child can return to school if he or she is feeling better. If medicine needs to be taken at school, talk with your doctor and the school nurse. If an older child uses an inhaler, get permission for him or her to carry it. Your child can participate in gym class but may need to be excused during a cold if coughing or mild wheezing is present. If your child has multiple episodes of asthma, he or she should wear a medical alert bracelet.

Home Treatment

1. If your child starts wheezing, keep him or her calm or playing quietly. Excitement and physical activity can make the wheezing worse.
2. Give the medicine as prescribed. Talk with your doctor about restarting medicine when your child gets a cold to prevent future episodes.
3. Do not run out of the medicine. Make sure you always have refills.

4. Over-the-counter medicines often do not work in asthma. Call your doctor before giving your child a nonprescription medicine.

Your doctor has prescribed the medicine below:

Nebulized Medicine (Aerosol)

[ ] Give _____ by aerosol ___ times per day at the following times_____.

[ ] Give _____ by aerosol mixed with _____times per day at the following times _____.

Inhaler

[ ] Use ___ puffs _____ inhaler ___ times per day at the following times _____.

Other Medicines

[ ]

Steroids (Used for a Few Days to Decrease Airway Swelling)

[ ]

Call Your Doctor or Return to the Emergency Department If:

1. Your child has increasing shortness of breath or trouble breathing.
2. Your child is breathing fast.
3. Your child looks blue or passes out **(call an ambulance immediately, do not drive yourself).**
4. Your child looks sick or anxious.
5. You have any questions or concerns.

BRONCHIOLITIS

Children with bronchiolitis have an infection with a virus that produces wheezing and trouble breathing. The tubes that carry air to the lungs and the small passages in the lungs themselves are infected by the virus. This condition causes the most problems in young babies (less than 2 months), children with heart or lung problems, and babies who were premature or have other medical problems. The most common virus causing bronchiolitis is the respiratory syncytial virus (RSV). No medicine or antibiotic can cure this infection. Because it is a virus, antibiotics do not help. We do try to make children more comfortable and help them breathe easier. Your doctor has decided that your child can be treated at home, but we know that this infection can sometimes worsen. If you think your child is getting worse you need to return to the ED.

Home Treatment

1. If your child starts wheezing, keep him or her calm or playing quietly. Excitement and physical activity can make the wheezing worse.
2. Run a vaporizer in your child's room. (We prefer a cool mist vaporizer to avoid the risk of burns.)
3. If your child's nose is stuffy you may use a bulb syringe and salt water drops to suction the mucus.
4. Encourage fluids.
5. Over-the-counter medicines often do not work in bronchiolitis. Call your doctor before giving your child a nonprescription medicine.

Sometimes breathing treatments that open the airways help children breathe easier. Sometimes these treatments do not help. If your doctor prescribes breathing treatments, he or she must think they will help. Use them as instructed. However, if you do not think they are helping and your child has trouble breathing, please return to the ED.

Your doctor has prescribed the medicine below:

Nebulized Medicine (Aerosol)

[ ] Give _____ by aerosol ___ times per day at the following times _____.

Other Medicines

[ ]

Call Your Doctor or Return to the Emergency Department If:

1. Your child has increasing shortness of breath or trouble breathing.
2. Your child is breathing fast (faster than 60 times per minute).
3. Your child stops breathing (even if he or she does not turn blue).
4. Your child is unable to drink (often a sign of trouble breathing).
5. Your child looks blue or passes out **(call an ambulance immediately, do not drive yourself).**
6. Your child looks sick or anxious.
7. You have any questions or concerns.

BURNS

Your child was treated for a burn. Burns occur when the skin is injured by contact with heat, fire, chemicals, or electricity. Your child has a clean dressing on the burn to protect it and prevent infection. We ask that you have the dressing changed for the first time by a doctor or nurse. Your doctor has arranged the following follow-up:

If the dressing falls off before this change, replace it with a clean, dry bandage. Your child may have received a tetanus booster in the ED. If so, notify your regular doctor to update your child's records.

How to Change the Dressing at Home

1. Wash your hands thoroughly with soap and water.
2. Remove the old bandage. If it sticks, you can soak it for a few minutes in warm (not hot) water.
3. Wash the burn with warm, soapy water.
4. Rinse and pat dry with a clean towel.
5. With a sterile tongue depressor, apply the antibiotic cream _____ to the burn area in a thin layer. Do not put a dirty tongue depressor back in the container of antibiotic cream.
6. Carefully rewrap the burn with a sterile bandage as directed by your doctor.

Signs of Infection

1. Increasing redness or red streaks around the burn
2. Swelling
3. Pain
4. Yellow pus or discharge
5. Fever

If you notice any signs of infection, call your doctor immediately or return to the ED.

Pain

Acetaminophen (e.g., Panadol, Tylenol) or ibuprofen (e.g., Advil, Motrin) can be used for pain. If your child was given a prescription for a different pain medication, you should use that medication as prescribed. Speak with your doctor about timing the dose with the dressing changes to relieve pain. Your medication is:

Exercise

Your doctor may have directed your child to perform certain exercises to help regain use of the burned area. Please ask your doctor if you have questions about the exercises or if you think your child is becoming stiff or tight around a burned area.

Long-Term Care

Once the skin has healed, apply a lubricating cream or lotion to the burned area. This treatment will keep it soft and decrease itching. Avoid extremes of heat or cold for 1 year after the burn. Avoid direct sunlight for 1 year after the burn. Apply a sunscreen to any burned areas to protect the new skin.

CAST OR SPLINT CARE

Your child has an arm or leg cast that will keep an extremity quiet and immobile after a serious injury.

What to Do in the First 48 Hours

1. Keep the cast elevated as much as possible to prevent swelling.
2. If your child has a splint and there is a lot of pain or the fingers or toes are cold and pale, unwrap the ace wrap to relieve the pressure from swelling. If this helps, rewrap it a little looser. If this does not help, rewrap the splint and return to the ED.
3. Give ibuprofen (Motrin, Advil) every 6 hours as needed for pain. Your child's dose is: ___

[ ] Your child may need additional pain medicine. Your doctor has written a prescription for:___

_____

General Cast or Splint Care

1. Do not allow your child to walk or put weight on the cast unless your doctor specifically tells you to do this.
2. Keep long arm casts in a sling at all times except when sleeping.
3. Do not get the cast wet unless you are told this is OK.
4. Do not allow your child to place objects inside the cast.
5. Do not use devices such as knitting needles, coat hangers, and so forth to scratch underneath the cast.
6. Your child can take a bath if the cast is covered with a plastic bag and kept above the water.
7. Keep the skin around the cast edges clean and dry. You can put rubbing alcohol on the skin near the cast edge to prevent irritation.
8. If the cast edge feels rough, you can put adhesive tape around it or “petal” around the edge with moleskin. Ask your doctor or nurse how to do this.
9. If your child is unable to go to school, have his or her teacher provide homework assignments and ask for a tutor if necessary.

Return to the Emergency Department or See Your Orthopedic Doctor If:

1. Your child's fingers or toes feel numb or cold, look blue or pale and unwrapping the splint does not help.
2. Your child complains of tingling, tightness, or pain in the injured arm or leg.
3. There is pain under the cast in one spot, or pain anywhere for no apparent reason.
4. It hurts your child to move the fingers or toes.
5. Your child has a fever.
6. You smell a bad odor coming from the cast.
7. The skin around the cast edge is red or irritated.
8. The cast gets soft or cracked.
9. The pain medication does not make your child feel better.

[ ] *Use of Crutches*

1. Help your child walk with crutches as demonstrated. Do not allow him or her to put weight on the cast unless told to do so.
2. Help your child go up and down stairs until you are comfortable he or she can do it well.
3. Do not have your child rest his or her underarms on the crutches. Putting weight on the underarms can cause nerve damage.
4. Always use crutches with rubber tips, and wipe the tips dry if they get wet so they are not slippery.

CHICKENPOX

Chickenpox (Varicella) is a viral infection. It causes fever, tiredness, loss of appetite, and a rash. The rash starts out as small red bumps that then develop into clear, fluid-filled blisters that break and crust over. New blisters keep forming for 3 to 4 days and a child is contagious from 2 days before the rash appears until all the blisters are crusted over. You should keep your child away from people who cannot fight infection well, including pregnant women, small babies, people taking steroid medication, and those with cancer, leukemia, or acquired immunodeficiency syndrome (AIDS). Your child cannot return to school or day care until all the blisters have crusted over.

Treatment

1. Give acetaminophen (e.g., Tylenol, Panadol) for fever. **DO NOT USE ASPIRIN.** Some doctors do not like to use ibuprofen with chickenpox. Ask your doctor's opinion.
2. Encourage your child to drink plenty of fluids. He or she may have blisters inside the mouth, and citrus drinks or sodas might be irritating. Other cool liquids (e.g., milk, apple juice, Kool-Aid) may help soothe the blisters.
3. Chickenpox is very itchy. Bathe your child in lukewarm water. Add ½ cup baking soda to the water and allow your child to bathe for 30 minutes. You can also tie oatmeal flakes in an old stocking and swirl this in the bath water or use a commercial oatmeal product (Aveeno).
4. Keep your child's fingernails cut short to prevent scratching.
5. Have your child wear socks on his or her hands at night to keep from scratching.
6. Calamine lotion can be applied to the chickenpox blisters.
7. Your doctor may prescribe a medicine for itching. This medicine can make your child sleepy. If you notice a change in your child's behavior or if you think he or she is too sleepy, stop the medicine and call your doctor. Your medication is: _____

_____

[ ] Sometimes acyclovir is used for chickenpox in those children who are at risk for a serious infection. This medicine may shorten your child's illness and decrease the number of chickenpox blisters your child develops. Your doctor has decided to use this medicine for your child. Your dose is: _____

_____

Call Your Doctor or Return to the Emergency Department If:

1. The blisters or surrounding skin look infected, are red and swollen, or have yellow pus.
2. Your child is sleeping too much or has a change in behavior.
3. Your child has blisters in his or her eyes.
4. Your child has trouble walking or severe headache.
5. Your child has trouble breathing.
6. The fever persists after the third day or if the fever was gone and then came back.
7. Your child looks sick or you have any questions or concerns.

COMMON COLD (UPPER RESPIRATORY INFECTION)

The common cold is an infection of the nose and throat that is usually caused by a virus. It can make your child have sneezing, coughing, fever, and not feel well. No medicine can cure a cold, and it can last as long as 7 to 10 days. Colds spread from person to person by coughing or breathing and when people do not wash their hands well after blowing their nose or sneezing. Your child can return to school or day care when he or she feels well and does not have a fever. Small amounts of coughing and sneezing should not keep your child out of school.

Home Treatment

1. Home treatment is aimed at keeping your child comfortable. If your child is uncomfortable or looks sick, he or she needs to see a doctor for a reexamination.
2. Encourage your child to drink plenty of fluids such as juice, soda, or Kool-Aid. Do not force him or her to eat because it may cause vomiting. Your child may not feel like eating, but it is important that he or she drinks to prevent dehydration.
3. Warm liquids sometimes ease a sore throat and help open a clogged nose. Grandmother's chicken soup may be the perfect meal for a child with a cold!
4. For nasal congestion (runny nose), you can use salt water drops to loosen the mucus. These drops should be used before feeding your child and at bedtime, but they can be used in between if you think your child's nose is clogged and he or she has trouble breathing.

   To make salt water drops:

   Mix ¼ teaspoon salt with ½ cup warm water. If you have a clean medicine bottle, you can store this solution for 24 hours (label it salt water). If you do not have a clean bottle, throw away the salt water after each use.

   Using a medicine dropper, put two drops of the salt water in one nostril. Have your child lying flat when you do this. You may want to support his or her neck or shoulders with a rolled-up towel. Wait 30 to 60 seconds before suctioning the mucus with a rubber bulb syringe. Squeeze the air out of the bulb, put the tip of the bulb into the nostril. Let the air come back into the bulb, and the suction will pull the mucus out of the nose. Squeeze the mucus out of the bulb onto a tissue.

   Repeat this process with the other nostril. Do not do this more often than six times a day. Wash the bulb syringe in warm soapy water after each use. Squeeze it in the water to clean the inside.
5. Do not use over-the-counter cold medicines without discussing this with your doctor first. Those medicines can have dangerous side effects and often do not help the cold symptoms.
6. Use a cool mist vaporizer in your child's room. This will moisten the air and help loosen your child's nasal secretions. Warm mist vaporizers can cause burns so we recommend cool mist. Do not add medicine to the vaporizer; use plain water. Wash the vaporizer as instructed after each use.
7. Give acetaminophen (Panadol, Tylenol) or ibuprofen (Advil, Motrin) if your child has a fever. Your dose is:

Call Your Doctor or Return to the Emergency Department If:

1. Your child has trouble breathing or starts wheezing.
2. Your child gets a new fever higher than 102°F (38.5°C) (or higher than 100.5°F [38.0°C] if your baby is less than 6 months old).
3. Your child has trouble swallowing.
4. Your child is not drinking well or you think he or she is dehydrated (dry).
5. Your child complains of ear pain or tugs at his or her ears.
6. Your child is sleepy or lethargic.
7. Your child looks sick.
8. You have any questions or concerns.

CONJUNCTIVITIS (PINK EYE)

Conjunctivitis is an infection or irritation of the outer part of the eye. It makes the “white” of the eye appear pink or red and is commonly called pink eye. It can be caused by allergies or infections from viruses or bacteria. Pink eye is very contagious. All members of your household should wash their hands carefully, and other children should not touch your child's eye. Your child should have his or her own washcloth and towel. Whenever you touch his or her eye, you must wash your hands.

Treatment

1. Clean any pus or drainage with a warm, wet washcloth or cotton ball.
2. Your doctor may have prescribed antibiotic drops or ointment. Place this medicine in your child's eyes as directed by the doctor. Do not use the same medicine for other people in the house. Have them examined by a doctor and given their own medicine. Your medicine is: _____
3. Try not to get the medicine in the other eye if it is not affected.
4. Do not use the medicine longer than directed. If the infection persists, see your doctor.

Call Your Doctor If:

1. The eyelids get red or swollen.
2. Your child has trouble seeing or blurry vision.
3. Your child gets a fever or looks sick.
4. The infection is not better in 2 to 3 days.
5. You have any other questions or concerns.

CORNEAL ABRASION

Your child has a scratch on the cornea, which is the outer surface of the eye. The doctor who examined your child put some drops in the eye. These drops ease the pain and prevent the scratch from getting infected. We placed a patch over your child's eye. This will help protect the eye from further injury and allow it to rest while it heals. If possible, please keep this patch in place. We understand that some children try to remove the patch; try to distract your child so this does not happen.

Your child must be seen **tomorrow** to see how the scratch is healing. An infected scratch or one that does not heal well can cause a permanent scar on the surface of the eye. Your doctor has arranged the following follow-up:

_____

Call us if you have any questions or concerns.

CONSTIPATION

Constipation is hard, dry stools that usually hurt or cause pain when they are passed. There are no rules about the number of stools a child needs to have in a day or week. As long as your child has soft stools, he or she probably is not constipated. It can be normal for babies or small children to grunt, strain, and even cry while they are having a bowel movement. It can also be normal for a baby to go a few days without a bowel movement. Each child develops a pattern of his or her own.

The doctor who examined your child decided that he or she is constipated. Most constipation can be treated by some **dietary changes.**

1. If your baby is less than 4 months old, add 1 to 2 ounces of apple or prune juice to your baby's diet each day until stools are soft and regular. Constipation is very rare in breast-fed babies; talk with your doctor if you breast-feed. Do not continue the juice for more than 1 week without asking your doctor. You can also move your baby's legs in gentle bicycle motions if you think he or she is having trouble passing a hard stool.
2. If your baby is more than 4 months old, give him or her one 4-ounce bottle of juice per day, and encourage fruits and vegetables if he or she has begun taking baby food.
3. If your child is eating table food:
   a. Increase the amount of fruits and vegetables he or she eats. Adding raw ones provides increased “roughage.”
   b. Increase the bran content of his or her foods. Feed your child bran cereal, bran muffins, oatmeal, and whole wheat bread if possible.
   c. Increase fluids such as juices and water.
4. Some children have severe constipation or a long-term problem that needs additional help with medication. Your doctor has written any additional instructions below.

_____

_____

Additional Tips

If your child complains of pain with a bowel movement, he or she may have a small tear or fissure in the rectal area. You can put vaseline or a protective diaper cream in this area to allow it to heal.

Give older children protected time at least twice each day for toilet sitting. Your child should be able to sit on the toilet for about 15 uninterrupted minutes two times per day. After meals works best.

Exercise also helps improve constipation, and you and your child may want to participate in activities together.

Call Your Doctor If:

1. The constipation does not improve in 2 weeks.
2. Your child has severe abdominal pain.

3. You see more than a few drops of blood in the stool.
4. Your child begins losing control of bowel movements or soiling his or her underwear.

Final Note: Although you may reward your child for a successful bowel movement, never punish your child for not having a bowel movement or for soiling his or her underwear.

CROUP

Croup is a swelling of the upper airway in the area commonly called the windpipe and voice box, or more technically, the trachea and larynx. Most children with croup have a virus, and some children are likely to get croup more than once. Croup is worse at night and better during the day. It usually lasts about 5 days.

Children with croup may have a fever and cold symptoms. The cough is harsh like a barking dog or seal. The noisy breathing is called stridor and is made by the narrowing of the airway from the swelling.

Treatment

[ ] Some doctors like to use a steroid medicine (Decadron), which can help with the airway swelling in croup. Your doctor has decided to use this medicine for your child. Your dose is: _____

Home Treatment

1. Stay calm and keep your child calm. It can be frightening when your child has trouble breathing, but if he or she is anxious or crying, it will make things worse.
2. Use a cool mist vaporizer in your child's room to humidify the air. Do not use a hot steam vaporizer because it could burn your child if he or she gets too close.
3. Prop your child's head up with a few pillows, or sit up with him or her. Your child may find his or her own position that makes breathing easier, but sitting up often helps.
4. Give acetaminophen (e.g., Tylenol, Panadol) for fever. Use a suppository if your child has trouble breathing or is throwing up.
5. Encourage your child to drink clear liquids. Do not force your child to eat if he or she does not want to eat or has difficulty breathing.
6. If your child's breathing sounds worse or the noisy stridor is louder, turn on the hot water in the bathroom shower or sink. Close the door and let the room steam up. Take your child in the bathroom with you and sit down for about 15 minutes. Keep your child occupied by reading to him or her or playing with toys. STAY CALM!
7. Sometimes cool air will also help your child. If the steamy bathroom does not help, you can dress your child for the outdoors and then take him or her outside for 10 minutes.

Return to the Emergency Department Immediately If:

1. Your child has trouble breathing, is using his neck or abdominal muscles to breathe, is pulling in his chest to breathe.
2. Your child looks pale or has blue areas around the mouth/lips, fingernails, or toenails.
3. Your child passes out.
4. Coughing is continuous, or there is no improvement after mist or cool air is tried.
5. Your child is uncomfortable or unable to sleep.
6. Your child is drooling or has trouble swallowing.
7. Your child has a high fever (greater than 102°F [38.5°C]).
8. Your child seems to be getting tired.
9. You have any concerns or the child is rapidly getting worse.

Transport your child yourself if you have your own car, the drive is less than 15 minutes, and your child seems comfortable and stable. When driving with a sick child, have two people in the car, one in the back with the child and one in front driving. Keep the child in his or her car seat. Open the windows and allow some cool air inside. **Call an ambulance if your child has trouble breathing, is passed out, looks blue, or is getting worse.**

DIAPER RASH

A diaper rash is usually caused by irritation of the baby's skin from contact with urine or bowel movements. Sometimes the skin can also be infected with yeast or bacteria, and your doctor will let you know if your baby has an infection.

Treatment of Diaper Rash

1. Keep the baby's bottom as clean and dry as possible. Change the diapers often. Wash the diaper area gently with soap and water at each change and pat dry. Avoid premoistened wipes while the rash is present because they may sting.
2. Leave the diaper off, if you can, to allow the air to dry and heal the rash. This can best be done at naptime with your baby lying on his or her back on an open diaper.
3. If your doctor ordered a protective ointment, you should apply this in a thin layer with each diaper change. Medicated ointments or creams should be used as prescribed.

4. Do not use talcum powder or baby powder because it can injure your baby's lungs if inhaled.
5. Avoid plastic pants or tight-fitting disposable diapers if possible. They trap in moisture and make the rash worse.

Call Your Doctor or Return to the Emergency Department If:

1. Your baby develops a fever.
2. The rash does not appear to be improving after 5 to 7 days.
3. The rash is getting worse or pimples or blisters develop in the diaper area.
4. You are concerned about your baby.

EAR INFECTIONS

Your child has an ear infection, which is an infection of the middle ear, the space behind the eardrum. Ear infections can be caused by viruses or bacteria and are more common in the winter. An ear infection is not contagious, but it can start as a cold. The germs in the throat or nose migrate into the middle ear. Fluid also can build up in the middle ear and swelling from a cold prevents it from draining into the throat. The combination of germs and fluid becomes an infection and pus is produced, putting pressure on the eardrum and causing pain. If your child has a drainage from the ear, this may mean the ear drum has torn. The small tear in the eardrum will heal itself if the infection is treated. However, if fluid stays behind the eardrum for long periods, your child may have hearing problems. Therefore, you should follow up with your doctor to make sure the infection has been treated and the fluid is cleared. Your child can return to school or day care as soon as he or she is feeling better.

Treatment

1. **Antibiotics:** Ear infections are treated with antibiotics. The fever and pain may continue for 1 to 2 days after starting the antibiotic. Some antibiotics cause diarrhea. If the diarrhea is severe or you think your child is getting dehydrated (not urinating well, not drinking well, no tears when crying), see your doctor. If your child gets a rash, he or she may be allergic to the antibiotic. See your doctor.
   Your dose of medicine is:

   Give your child all the antibiotic prescribed, even if your child feels better. The full course is needed to kill the infection. Keep all medicines out of reach of small children.
2. **Ear Drops:** Your doctor may give you ear drops to treat an infection in the ear canal or external ear. Have your child lie on your lap with the infected ear up. Put two drops in the ear canal and massage in front of the ear to help the drops fall into the canal. Do not put anything else in the ear canal. Use the drops _____ times a day for _____ days.
3. **Pain and Fever:** Your child may have a fever or some ear pain. Acetaminophen (e.g., Tylenol, Panadol) or ibuprofen (Motrin, Advil) will treat this pain. Your child's dose is:_____
4. **Follow-up:** Follow up with your regular doctor after 2 weeks to make sure the infection is gone.

Call Your Doctor or Return to the Emergency Department If:

1. Your child looks sick or fever continues more than 2 days after starting antibiotics.
2. Your child has a new drainage from the ear.
3. Your child gets a rash.
4. Diarrhea becomes severe or you think your child is dehydrated (dry).
5. Your child is sleepy or lethargic, or if you have any questions or concerns.

ECZEMA

Eczema is common. Children with eczema usually have a family history of allergies, hayfever, or asthma. Eczema cannot be cured, but it can be controlled with skin care and some changes in your child's surroundings. Eczema usually improves as your child gets older.

Changing Your Child's Environment

1. Avoid wool clothing because it can be irritating. Dress your child in cotton when possible, and try to keep his or her arms and legs covered (tights or long pants, long-sleeve shirts).
2. Keep your child's room free of dust, and keep the air moist with a cool mist vaporizer or humidifier.
3. Wash your child's clothes in a mild detergent and avoid fabric softeners that can be irritating.

Skin Care

1. Do not overdo bathing. Wash your child when he or she is obviously dirty, but cut down on routine bathing to two times per week. Use a mild soap with moisturizer. Avoid deodorant soaps. Do not use a washcloth, and bathe in warm, not hot, water. Do not use bubble baths.
2. Your doctor may have prescribed a steroid cream for specific areas. Apply this cream as directed to those areas.
   Apply _____
3. One hour after applying the steroid cream, apply a general moisturizing cream to your child's entire body. Apply this moisturizing cream two times per day, *every day.*

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4. If itching is a problem, your doctor may prescribe a medicine to be used for a short period.

These medicines can make your child sleepy and teenagers should not drive a car while taking this medicine. Keep your child's fingernails cut short if scratching is a problem. Some children need to wear socks on their hands when they go to sleep to keep them from scratching.

Call Your Doctor If:

1. The eczema is red or irritated looking, your child develops sores or scabs, or your child gets a fever.
2. You think the eczema is not under good control.
3. You have any questions or concerns.

FEBRILE SEIZURE

A febrile seizure is a “fit” or “convulsion” that occurs with a fever. Most children who have febrile seizures outgrow them by 4 to 5 years of age. Having a febrile seizure does not mean that your child has brain damage or will be retarded. There is a chance, however, that your child may have another seizure when he or she has a fever.

When your child becomes sick, the suggestions below will help you control the fever and prevent a seizure.

1. Give acetaminophen (e.g., Tylenol, Panadol) or ibuprofen (Advil, Motrin) in the correct dose for your child's age. Acetaminophen can be given every 4 hours, ibuprofen every 6 hours while your child's temperature is 101°F (38.0°C) or higher.
2. Do not bundle or overdress your child. The body loses heat through the skin, and if you bundle him or her the excess heat cannot escape.
3. Sponge your child with lukewarm water or put him or her in a shallow tub containing 2 to 3 inches of water and drip water over his or her body. Do *not* use alcohol or cold water to bring your child's fever down. If your child begins shivering or shaking, stop sponging and remove him or her from the bath water.
4. While your child has a fever, give plenty of fluids to prevent dehydration.
5. Give any medications prescribed by your doctor.

Your child may have another febrile seizure because they can happen before you realize your child is ill.

If Your Child Has Another Seizure:

1. Stay calm!
2. Do not put anything in your child's mouth.
3. Place your child on his or her side to help drain secretions.
4. Loosen clothing.
5. Do not try to hold your child still. Move objects away from your child so he or she does not get hurt.
6. Support your child's head with a pillow or soft object.

Call an ambulance if the seizure is lasting longer than 5 minutes or if your child has difficulty breathing or looks blue. Otherwise, once the seizure stops, call your doctor or bring your child to the ED for further instructions and a physical examination. Your child may be sleepy after the seizure and needs to be checked by a doctor.

SEIZURE

Your child has had a seizure. A seizure happens when the brain cells send electrical discharges that cause the arms and legs to jerk or twitch, and the eyes to stare or blink. Usually a child is sleepy or confused after having a seizure.

What to Do If Your Child Has a Seizure

1. Stay calm!
2. Do not put anything in your child's mouth.
3. Place your child on his or her side to help drain secretions.
4. Loosen clothing.
5. Do not try to hold your child still. Move objects away from your child so he or she does not get hurt.
6. Support your child's head with a pillow or soft object.
7. Do not try to give your child any medicine during a seizure. It may cause him or her to choke.
8. Try to observe what happens during the seizure: which arm or leg twitches, how long the seizure lasts, and so on. This information may help your doctor decide how to treat the seizure.

Call for Help If:

1. Your child has trouble breathing or looks blue.
2. The seizure is lasting longer than 5 minutes.
3. You cannot wake your child 30 minutes after the seizure.

After the Seizure

Your child may be sleepy and should be allowed to rest. Continue to give any medications prescribed for the seizure disorder. Do not give extra medicine or change the dosage without calling your doctor. Make sure you do not run out of

the medication. Give all medicine as scheduled.

Follow up with your regular doctor when a seizure occurs. This can be a good time to review your child's medical care and make changes if necessary. Your child may want to participate in sports or activities such as bicycle riding, swimming, or driving a car or motorcycle. Discuss this with your doctor before you allow your child to participate.

FEVER

Your child has a fever. This means the body temperature is above normal. In the mouth, normal temperature is 98.6°F (37°C); under the arm, normal is 98°F (36.6°C); and by rectum, it is 100°F (37.7°C). A fever is the body's way of fighting an infection and is not always a bad thing. You only need to treat the fever if it is high (greater than 102°F [38.5°C]) or if your child is uncomfortable. How high the temperature is does not indicate how severe the illness is that causes the fever. Your child should see a doctor if you have any concerns.

What to Do to Keep Your Child Comfortable

1. Dress your child lightly to be comfortable in your home's temperature. Do not overbundle or use heavy blankets because this will raise your child's temperature further. A T-shirt and underwear or diaper with a light sheet or blanket is fine for sleeping.
2. Encourage plenty of liquids. Your child may not be hungry but it is important that he or she continues to drink and does not get dry.
3. Keep the room around 70°F if possible. In the winter, do not overheat, and in the summer, use a fan or air conditioner if available.
4. Acetaminophen (e.g., Panadol, Tylenol) lowers fever. Your pharmacy may sell a generic fever product with acetaminophen. These are just as effective and may cost less. Check your child's temperature before giving the medicine. The dose can be repeated every 4 hours.

   If your child is vomiting or does not like to take medicine, you can buy acetaminophen suppositories at your pharmacy without a prescription. Ask your doctor or nurse about how to use a suppository.
5. Ibuprofen (Advil, Motrin) is also used for fever control in children 6 months and older. The dose can be repeated every 6 hours.
6. Do not use aspirin for fever control in children.
7. Sponging your child with lukewarm water will also lower his or her temperature but is not as helpful as fever medicine. Do not use alcohol or add alcohol to a bath. Never leave your child alone in a bath. If your child starts shivering, take him or her out of the bath and dry off. Shivering can raise the body temperature.

Call Your Doctor or Return to the Emergency Department If:

1. Your child is less than 6 months of age and has a fever greater than 101°F (38.2°C).
2. The fever continues for more than 2 additional days without other symptoms.
3. Your child has other symptoms—rash, trouble breathing, ear pain, headache, stiff neck, vomiting, diarrhea, joint swelling, or pain.
4. Your child acts sick, is irritable, sleeps a lot, stops playing, or does not eat or drink.
5. You have any questions or concerns.

FEVER LESS THAN 2 MONTHS

Your baby has a fever. This means the body temperature is above normal. We worry about babies with fever because they can have more trouble fighting a serious infection, and it is harder for parents and doctors to tell when they are getting sick. Your baby was evaluated in the ED and the doctor decided he or she could be cared for at home. You should follow up as instructed.

What to Do to Keep Your Baby Comfortable

1. Dress your child lightly to be comfortable in your home's temperature. Do not overbundle or use heavy blankets because this will raise your child's temperature further. A T-shirt and diaper with a light sheet or blanket is fine for sleeping.
2. Encourage plenty of liquids. It is important that he or she continues to drink and does not get dry.
3. Keep the room around 70°F if possible. In the winter, do not overheat, and in the summer, use a fan or air conditioner if available.
4. Acetaminophen (e.g., Panadol, Tylenol) lowers fever. Your pharmacy may sell a generic fever product with acetaminophen. These are just as effective and may cost less. Check your baby's temperature before giving the medicine. The dose can be repeated every 4 hours.
   Your baby's dose is:_____

   Use the dropper that comes with the acetaminophen (e.g., Tylenol, Panadol) bottle. Other droppers may be a different size.
5. Follow-up is usually arranged with your physician or the ED within 24 hours of this visit. **Your follow-up is:**

Return to the Emergency Department If:

1. The fever continues for more than 2 additional days without other symptoms.
2. Your baby has other symptoms—rash, grunting or trouble breathing, stiff neck, vomiting, diarrhea, or pain.

3. Your child acts sick, is irritable, sleeps a lot, or does not drink.
4. You have any questions or concerns.

HEAD INJURY

The doctor who examined your child determined that he or she can safely be observed at home. You will need to watch your child for the next 24 to 72 hours and bring him or her back to the ED if necessary. Please tell your doctor before leaving the ED if you do not think you can do this.

Normal Behaviors in the First 8 Hours After a Head Injury

1. Your child may be **sleepy.** It is OK to let him or her sleep, but you need to wake your child every 1 to 2 hours initially. Your child should be able to wake up and behave normally, recognize people and things, and speak clearly.
2. **Vomiting,** or throwing up, is also normal in the first few hours following a head injury.
3. Your child may complain of a **headache.** You can give acetaminophen (e.g., Tylenol, Panadol).

What to Do

1. Have your child rest or play quietly for the first 24 to 72 hours.
2. Wake your child every 1 to 2 hours for the first 8 hours.
3. Feed your child a lighter than normal diet.
4. Give acetaminophen for a headache.

Return to the Emergency Department Immediately If:

1. **Vomiting** continues after the first 8 hours or begins later than the first few hours after the injury.
2. Your child is **difficult to wake up** or is **not acting normally** when awakened.
3. Your child's **headache worsens,** changes your child's behavior, or is not relieved by acetaminophen.
4. Your child has **trouble seeing** or **walking** or **acts clumsy** or uncoordinated.
5. Your child has **bleeding or clear drainage** from his or her **nose** or **ears.**
6. Your child has a **convulsion** or **seizure.**
7. Your child is unusually **sleepy** or has any **unusual behavior** or **change in behavior.**

HIVES

Urticaria, or hives, are red blotches on the skin. They can be many different sizes and are very itchy. Most hives are an allergic reaction to something your child touched, ate, or put on his or her skin. Hives can also be a reaction to cold, heat, emotional stress, or a viral infection. Some common substances that cause hives are peanuts, strawberries, shellfish, grass, bushes, perfumes, medicines, pets, insect bites, soaps, and detergents. Hives can last only a few hours or several weeks. Often, the cause is unknown, but if you think you know what caused the hives, you should try to avoid reexposing your child to this substance.

Treatment

1. Sometimes no treatment is necessary, and the hives go away on their own.
2. Warmth makes the itching worse, so use a cool washcloth or cool bath to make your child more comfortable.
3. Your doctor may prescribe a medicine for itching. These medicines can make your child sleepy. Teenagers should not drive while using this medicine.
   Your dose is:_____
4. The best treatment is to avoid whatever caused the hives, so try to play detective and figure out what caused them.

Call the Doctor If:

1. Your child has **trouble breathing** or feels a **tightness in his or her throat or chest.**

   Call an ambulance immediately and get to the nearest ED.
2. Your child has **lip or tongue swelling. You need to seek emergency treatment immediately.**
3. The itching is not relieved by the medicine prescribed.

IMPETIGO

Impetigo is a skin infection caused by bacteria. You can get impetigo by scratching and infecting insect bites and dry skin or by touching sores on other people. It is easily spread to other parts of the body and to other people, so your child should not return to day care or school until the crusts are gone.

Things to Do If Your Child Has Impetigo

1. Wash your hands before and after caring for your child.
2. Gently wash the crusty areas three times each day with soap and water. You may need to soak the area in warm water to remove all the crusts.
3. Blot the areas dry.
4. If your doctor prescribed an ointment, apply this to the sore and the area around it. Rub it in well.

5. If your doctor prescribed a medication by mouth, give it to your child as directed until it is all gone. Your dose is: _____
6. Carefully wash the bathtub or bathroom sink your child used with soap and water. Do not use the kitchen sink if possible. Wash your child's towel, washcloth, and bed linens after each use. Do not allow your child to share these.
7. Keep your child's fingernails cut short and try to keep him or her from scratching.

Call Your Doctor or Return to the Emergency Department If:

1. Your child gets a fever.
2. The infection is not improving in 4 to 5 days.
3. The sores appear to be spreading.
4. The sores are not cleared up after 10 days.

LICE

Head lice are small, gray insects that live on human beings. They can be spread by direct contact and by shared combs, hats, and clothes. They live in the hair and lay tiny white eggs called nits that stick to each hair shaft. Lice can cause itching.

Treatment

Your child was given a prescription for either cream rinse or shampoo to treat the lice. **To use the shampoo:**

1. Apply to dry hair until thoroughly coated (Wear rubber gloves when applying the shampoo).
2. Leave the shampoo on the hair for 10 minutes, but not longer.
3. Add a small amount of water to get a lather and shampoo the hair.
4. Do not get shampoo in the eyes or mouth. If you do, rinse immediately with water.
5. Rinse the hair with water and towel dry. Use a fine-tooth comb to remove all nits. They may stick to the hair shaft and be difficult to remove. If so, rinse hair with a dilute vinegar and water solution (dilute vinegar with an equal amount water). This will make nits easier to remove.

To Use the Cream Rinse:

1. Wash the hair with your regular shampoo and towel dry.
2. Apply the cream rinse to coat the hair thoroughly.
3. Leave the cream rinse on the hair for 10 minutes.
4. Rinse the hair with water and towel dry. Remove nits as previously described.

   Re-treat with the shampoo or cream rinse in 7 days if lice or nits are still present. Itching may continue for a few weeks even though the lice are gone.

To Eliminate Lice from Your Home:

1. Vacuum all surfaces thoroughly.
2. Clean combs and brushes in hot water with some antilice shampoo or cream rinse.
3. Wash all pieces of clothing worn in the last 3 days and any sheets, blankets, and pillow cases your child used in hot water (more than 130°F) and dry in a hot dryer for at least 20 minutes.
4. Any items not washable—stuffed toys, coats, hats—must be set aside in airtight plastic bags for 2 weeks.

NOSEBLEEDS

Nosebleeds are common in children. They are usually caused by dryness inside the nose plus some irritation from rubbing, picking, or cold symptoms. They can begin suddenly and sometimes happen during sleep.

How to Stop a Nosebleed

1. Have your child sit up and lean forward. You may need a container so that your child can spit out any blood that has drained into his or her throat.
2. Firmly pinch the soft part of the nostrils (not the tip) together for a **full** 5 minutes. Use a clock to time this and do not let go sooner. Tell your child to breathe through his or her mouth.
3. When you release the pressure, if the bleeding begins again, repeat Step 2 one time.
4. If the bleeding continues, call your doctor or take your child to the ED. If possible, have someone hold pressure on the nose while traveling to the ED.
5. Do not place anything inside the nose while it is bleeding (e.g., gauze, tissue).
6. Cold washcloths or ice to the face will not help stop the bleeding.
7. Swallowed blood can irritate the stomach, causing your child to vomit, and it can look bloody.

How to Prevent Nosebleeds

1. Using your finger, gently place a small amount of vaseline to the inside of the nose. This will help ease the dryness and irritation.
2. Use a cool mist vaporizer or humidifier in your child's room.
3. Discourage your child from picking his or her nose and keep fingernails cut short.

4. If your child has a stuffy nose, you can use saline nose drops to make the mucus easier to clear. Avoid vigorous nose blowing.
5. Do not give your child aspirin unless directed by your doctor. If your child has many nosebleeds and is on aspirin, tell your doctor.

Call Your Doctor If:

1. You cannot stop the bleeding or your child has many nose bleeds in one day.
2. You think a lot of blood was lost, or your child faints, or looks dizzy or pale.
3. You see blood elsewhere—in urine, stool—or your child has bruises or a rash.
4. Your child looks sick.

RINGWORM

Ringworm is a fungal infection that can cause a skin rash or infect the scalp and cause hair loss, scaling or pimples, and pus. Your doctor can usually diagnose this condition by looking at it. Sometimes he or she will scrape the rash and take a culture. Ringworm is spread from person to person, from animals, or from shared combs, towels, or hats.

Treatment

Skin Infection: Your doctor prescribed a cream. Put this cream on the rash as directed. The rash should improve in 7 to 10 days, but you should continue the cream for a full 2-week course. If it is not gone after 2 weeks, call your doctor.

Scalp Infection: Your doctor prescribed a medicine (griseofulvin) to be taken by mouth. Your child's dose is_____. This medicine should be taken for the full course and is best taken with milk or a fatty meal. If your child gets vomiting, diarrhea, abdominal pain, or a rash or if he or she looks ill while taking this medicine, call your doctor. Your doctor also prescribed a special shampoo (selenium sulfide 2.5%) to decrease the time that your child is contagious. This shampoo can be used twice a week or as directed by your doctor. If your child wears braids or ponytails, they should be undone so the shampoo can penetrate to the scalp. Your child's combs, brushes, and clothing should be cleaned with ordinary soap and water. Ask your doctor or school nurse about when your child can return to school.

Call Your Doctor If:

1. The rash is not gone after a full course of the medicine.
2. Your child gets ill while taking griseofulvin.
3. You have any questions or concerns.

SCABIES

Scabies are little bugs that burrow in the skin and cause severe itching and a rash. These bugs can only be seen with a microscope. They can spread easily from person to person by direct contact or by wearing clothes that have the scabies bug living in them.

Treatment

Your doctor has prescribed a special cream containing 5% permethrin (Elimite). Massage the cream into the skin from the head to the soles of the feet, including the scalp for an infant. Try to avoid the eyes because the cream can irritate them. If any of the cream gets in the eyes, wash them with cool water. Scabies like to live between the fingers and toes, under arms, and around the waist and genitals. Make sure to include all these areas. Leave the cream on for at least 8 hours, then give your child a bath to wash off the cream. The itching and rash may last for 2 to 4 weeks after treatment. Your doctor may be able to give your child some medicine that helps the itching. If it continues longer than this, return to your doctor. Everyone in your household should be treated at the same time because scabies spread easily from person to person. Even people without symptoms should be treated; ask your doctor about this.

Medication for itching:_____

Cleaning Your House

1. Scabies can live on clothing or bed linens for up to 1 week.
2. Using hot water (more than 120°F), wash all clothing, bed linens, towels, and washcloths used in the past week, and dry them with high heat for 20 minutes to kill the scabies.
3. Items that cannot be washed (e.g., toys, blankets) should be placed in a plastic bag and stored for 1 week.
4. Clean clothes and clean sheets should be used after applying the cream.

Your child can return to school after treatment with the cream. Remember to keep the cream stored out of reach of your child because it can be poisonous if swallowed.

Call Your Doctor If:

1. The itching persists longer than 4 weeks after using the cream.
2. You think the skin has become infected or looks red with blistering or crusting.

SORE THROAT

A sore throat happens when the tonsils or back of the mouth get infected by a virus or bacteria. The infection usually spreads from person to person by coughing or sneezing but can also spread by sharing drinking cups or eating utensils. Your doctor may have sent a swab of your child's throat for a culture. The culture will diagnose a strep throat caused by the streptococcus bacteria. The culture may take up to 2 days for a result, and sometimes a quicker test called a rapid strep is used.

[ ] **The rapid strep test was negative or not available at this time.** If the rapid test was negative, this does not absolutely mean your child does not have strep throat. Call _____ for the culture results on _____ from _____. Have your pharmacy phone number ready in case the doctor needs to phone in a prescription for your child.

[ ] **The rapid strep test was positive.** Your child has strep throat and needs antibiotic treatment. Your doctor may give you a prescription for an antibiotic, or your child can get a shot of long-acting penicillin in the ED. Both medicines will treat the infection. If you choose the antibiotic at home, you need to give your child all the medicine.
Your dose is:

Home Treatment:

1. The antibiotic will not make your child feel better right away. It may take 2 to 3 days to see improvement. Watch for a rash, trouble breathing, or swelling of the face, hands, or feet as signs of an allergic reaction to the antibiotic.
2. Encourage your child to drink plenty of fluids such as juice, soda, and fruit drinks. Soft foods like applesauce, pudding, and mashed potatoes may be less irritating to the throat.
3. Give acetaminophen (e.g., Tylenol, Panadol) or ibuprofen (Motrin, Advil) for fever. Your child's dose is:_____
4. Salt water gargles (½ tsp salt in 1 cup warm water) may make an older child's sore throat feel better. Do not let your child swallow the salt water. Have him or her spit it out.
5. Your child may go back to school 24 hours after starting the antibiotic as long as he or she feels well and does not have a fever.

Call Your Doctor or Return to the Emergency Department If:

1. Your child has drooling or difficulty swallowing.
2. Your child has a stiff neck.
3. Your child has trouble breathing.
4. Your child has a rash or swelling of the hands or feet.
5. Your child still has a fever 2 to 3 days after starting antibiotics.
6. You are unable to give your child the antibiotic.
7. Your child looks sick or you have any questions or concerns.

SPRAINS AND STRAINS

A *sprain* is an injury to the ligaments that hold your bones together. A *strain* is an injury to the muscle or muscle tendon from stretching or pulling.

Treatment

1. Keep the injured area quiet. If your doctor gave you a splint or crutches, have your child use these as instructed. Do not allow your child to put weight or stress on the area until your doctor tells you this is OK. Follow the doctor's instructions for exercise.
2. Keep the injured area elevated as much as possible. Prop up an arm or leg with pillows.
3. Use an ice pack for the first 24 hours. Do not put the ice directly against the skin; wrap it in a towel first.
4. After the first 24 hours, use a heating pad or hot water bottle (be careful not to burn the skin).
5. You may give your child ibuprofen (Motrin, Advil) for pain. Your dose is:_____
6. Follow up with your doctor or with an orthopedic surgeon in _____days. Do not let your child stop using the splint or crutches until you follow up with a doctor.

Call Your Doctor or Return to the Emergency Department If:

1. Your child has increased redness or swelling at the injury site.
2. Your child gets a fever.
3. Your child has no feeling in the injured arm or leg or it feels cold.
4. Your child is not feeling better in 3 to 5 days or is not making steady progress in 3 to 5 days.
5. You have any questions or concerns.

STOMATITIS

Stomatitis is a viral infection that can cause sores or blisters to develop on the gums, tongue, and other areas inside the

mouth. Your child may have a fever and the ulcers are painful. He or she may not want to eat or drink. Because this is a viral infection, it will usually clear up by itself within 5 days. However, some children may have sores in the mouth for 1 to 2 weeks.

To Keep Your Child Comfortable and Prevent Dehydration

1. Give acetaminophen (e.g., Tylenol, Panadol) or ibuprofen (Advil, Motrin) at the correct dosage for your child's age. This medicine will help with the pain and fever. Acetaminophen suppositories are available at your pharmacy without a prescription. Your dose is:_____
2. Encourage cold or cool liquids. These may be soothing to the mouth and help numb the pain. Avoid citrus and carbonated drinks (e.g., orange, grapefruit juices, lemonade, soda). Soft foods such as applesauce, yogurt, pudding, or mashed potatoes may be less irritating to the mouth.
3. Avoid salty or spicy foods.
4. If your doctor has given you a mouthwash or other medication, use as directed.

To Prevent Spread of this Infection

1. Wash your hands and your child's hands frequently and before eating.
2. Do not share your child's eating utensils or drinking cups while sick; wash after each use.
3. Wash any toys your child places in his or her mouth before and after your child plays with them.

Call Your Doctor or Return to the Emergency Department If:

1. Your child is refusing to drink or cannot swallow.
2. Your child appears dehydrated (no urine output in the last 8 hours, no tears when crying, lips are dry or cracked).
3. You think your child looks worse than when you were initially seen in the ED.
4. Your child is not getting better after 1 week.

URINARY TRACT INFECTION

Your child has been diagnosed with a urinary tract infection. This is an infection of the bladder or kidneys. It can cause symptoms of fever, abdominal or back pain, vomiting, or burning with urination, or it may have no symptoms in small babies. The diagnosis is made by looking at a clean sample of urine under a microscope and then growing bacteria with a urine culture.

Treatment

1. Antibiotics are used to treat the infection. Your child's dose is _____times per day. Make sure to give all doses for the full course to completely get rid of the infection. Diarrhea can be a side effect of antibiotics, so do not stop them if this happens. If your child does get a rash or severe diarrhea, call your doctor.
2. Encourage fluids to help clear the infection.
3. Allow your child to urinate as often as he or she desires, and encourage him or her not to “hold” the urine.
4. Follow-up is important to make sure the infection is cured. Your doctor should check another urine sample after the antibiotics are finished to confirm this.

Long-Term Follow-Up

Urinary tract infections can sometimes come back. Your doctor may want to follow your child more frequently and look at urine samples. Some children need to have their kidneys and bladder evaluated to make sure that there is not a problem that led to the infection. Your doctor may schedule tests to look for this.

Tips for Prevention

1. Teach your child to wipe from front to back and then throw away the toilet paper.
2. Have girls wear cotton underwear. Synthetics can irritate the genital area and lead to infection.
3. Avoid bubble baths, creams, or powders in the genital area.
4. Encourage your child to drink plenty of fluids each day.
5. Encourage him or her to empty the bladder completely every 3 to 4 hours during the day and to urinate before bedtime.

Call Your Doctor or Return to the Emergency Department If:

1. Fever lasts longer than 2 days with antibiotic treatment or your child develops a fever while taking the antibiotic.
2. Your child stops urinating or the urine becomes bloody.
3. Your child gets worse.
4. Your child refuses to take the antibiotic.
5. Your child gets a rash or has severe diarrhea while taking the antibiotic.

VOMITING AND/OR DIARRHEA

Vomiting or diarrhea happens when the lining of the stomach or intestines is irritated by an infection. Usually, the infection is a virus and needs to run its course, which may vary from 1 day to 1 week. The doctor who examined your child decided that you could treat this illness at home. To make your child feel better, he or she needs to rest the

stomach and intestines and help prevent more vomiting and diarrhea. This can be done by giving your child clear liquids and foods that are easily digested and by avoiding spicy or greasy foods that can further irritate the gastrointestinal tract. The goal is to keep your child from becoming dehydrated (dry). Your child can return to school or day care when the diarrhea or vomiting have resolved and he or she is feeling better.

Follow the Instructions Below:

For Babies 2 to 6 Months

1. Feed your baby an oral electrolyte solution (Pedialyte or Ricelyte). If severe vomiting or diarrhea is a problem, try a small amount at frequent intervals (example: 1 ounce every 1 hour). Babies who are breastfeeding may continue but still may need additional fluids such as Pedialyte or Ricelyte.
2. DO NOT GIVE PLAIN WATER.
3. If the vomiting and diarrhea improve in 24 hours, restart the baby's regular formula.
4. Some stooling may occur; keep feeding the formula unless your baby has very liquid stool, vomits more than five times, or has more than five stools in 1 day.
5. See your doctor or return to the ED if the vomiting and diarrhea continue. Do not give your baby Pedialyte or Ricelyte for more than 24 hours.

For Babies 6 Months to 1 Year

1. Feed your baby an oral electrolyte solution (Pedialyte or Ricelyte) for 24 hours. If your child does not like Pedialyte, you can also use Kool-Aid or flat soda (shake out the fizz), but only for 24 hours. DO NOT GIVE PLAIN WATER. Babies who are breast-feeding may continue but may still need additional fluids.
2. If the vomiting and diarrhea improve in 24 hours, restart formula. In addition, you can begin some soft foods such as rice cereal, bananas, or applesauce.
3. At 48 hours, you can add other fruits and vegetables that are in your child's diet.
4. At 72 hours, you can resume your child's regular diet.
5. If the vomiting and diarrhea return or continue at any stage, go back to the clear liquids and see your doctor. Do not give your baby clear liquids for more than 48 hours without talking to your doctor.

For Children Older than 1 Year

1. Give your child clear liquids only for 24 hours (clear liquids are ones you can see through), and allow the stomach to rest. Examples of clear liquids are flat soda (shake out the fizz), Kool-Aid, Gatorade diluted with an equal amount of water, Hawaiian Punch, juices (not apple, orange, grapefruit), tea with sugar, Jell-O, popsicles, water ice, sherbet, clear soups, or broth. DO NOT GIVE MILK.
2. At 24 hours, if the vomiting and diarrhea improve, add soft, bland foods. Examples include oatmeal, rice cereal, bananas, applesauce, dry toast, crackers, vanilla wafers, dry mashed potatoes, noodles, boiled chicken, and turkey. STAY AWAY FROM FRIED OR SPICY FOODS.
3. At 48 hours, add other fruits and vegetables and plain cooked meat.
4. At 72 hours, resume a regular diet and give milk as you would normally.
5. If the vomiting and diarrhea return or continue at any stage, go back to the clear liquids, and see your doctor. Do not give your child only clear liquids for more than 48 hours without calling your doctor.

Call Your Doctor or Return to the Emergency Department If:

1. The vomiting and diarrhea do not improve.
2. Your child is unable to take fluids or is weak, sleepy, or lethargic.
3. You think your child looks dry (eyes look sunken, soft spot is depressed, no tears when crying, mouth looks dry).
4. Your child has not urinated in 8 hours.
5. There is blood in the vomit or stool, or brown flecks like coffee grounds in the vomit.
6. Your child has abdominal pain.
7. Your child has a fever higher than 102°F.
8. You think your child looks sick or is getting worse.
9. You have any questions or concerns.

APPENDIX **D**

Practical Information

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VITAL SIGNS

Blood Pressure

Values:
Neonate Range: Systolic 40–80 mm Hg
Diastolic 20–55 mm Hg

	Percentile (Syst/Dias)	
Age (Yr)	50%	95%
2	96/60	112/78
6	98/64	116/80
9	106/68	126/84
12	114/74	136/88

Lower extremity pressures usually measure 10 to 40 mm Hg higher.

Pulsus Paradoxicus

Pulsus paradoxicus is defined as a drop in systolic blood pressure of greater than 10 mm Hg when taken during inspiration and then taken during expiration.

Respiratory Rate

Age	Per Minute
Neonate	30–50
1–6 mo	20–40
6 mo–2 yr	20–30
2–12 yr	16–24
Adolescent	12–20

Heart Rate

Age	Per Minute
Newborn	80–180
1 wk–1 mo	80–160
3 mo–2 yr	80–150
2–10 yr	75–110
10 yr–adult	50–100

Temperature

Conversion:
Fahrenheit vs. Centigrade:

$$°C = (°F - 32) \times 5/9$$

Example: $°C = (98.6 - 32)$ (or 66.6) $\times 5$ (= 333)/9 = 37

$$°F = (°C \times 9/5) + 32$$

Example: $°F = (39 \times 9 = 351)$ (/5 = 70.2) + 32 = 102.2
Extrapolating Points: 38°C = 100.4°F
39°C = 102.2°F
40°C = 104.0°F
41°C = 105.8°F

Weight

Conversion: Wt (lb)/2.2 = Wt (kg)
Wt (kg) × 2.2 = Wt (lb)

Weight

Conversion: Wt (lb)/2.2=Wt (kg)
Wt (kg)×2.2=Wt (lb)

	Percentiles (Using Kg)		
Age	5%	50%	95%
Neonate			
32 wk	1.3	1.8	2.8
40 wk	2.7	3.5	4.2
6 mo			
Female	5.9	7.2	8.6
Male	6.3	7.7	9.5
1 yr			
Female	7.8	9.5	11.4
Male	8.4	10.1	12.0
2 yr			
Female	9.8	11.8	14.0
Male	10.5	12.7	14.7
5 yr			
Female	14.1	17.9	22.2
Male	15.2	19.0	22.8
7 yr			
Female	15.5	19.5	29.0
Male	16.6	21.0	29.8
9 yr			
Female	21.7	28.0	40.6
Male	22.6	28.0	40.2

(See Fig. I.2 and Fig. I.3 in Chapter 1.)

Surface Area

Body surface area can be determined by connecting the height and weight numbers with a straight line. The point at which the line intersects the surface area abscissa is the reading for Surface Area in meters squared (Fig. D.1).

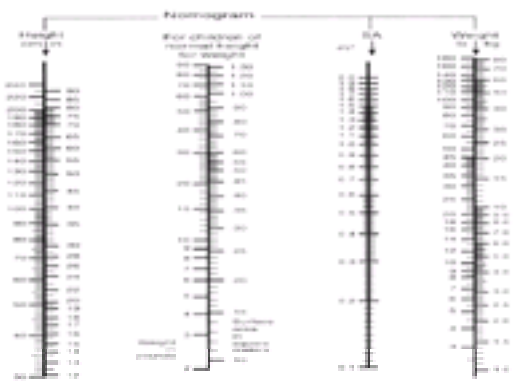

FIGURE D.1. Surface area. (Adapted from Harriet Lane Handbook. 14th ed., St Louis: Mosby, 1996.)

To determine the approximate body surface area for normally proportioned children, use either the second line from the left in the above nomogram or the following formula.

$$\text{Surface area } (m^2) = \text{Square root of:}$$
$$[\text{Ht (cm)} \times \text{Wt (kg)} \div 3600]$$

IMMUNIZATIONS

Recommended Schedule for Healthy Infants and Children

Age	Immunizations
Birth or 1 mo	Hep B-1
2 mo	DTaP or DTP, HIB, IPV, Hep B-2
4 mo	DTaP or DTP, HIB, IPV
6 mo	DTaP or DTP, HIB, HepB-3
(In the face of a measles epidemic, the first MMR can be given as young as this)	
12–15 mo	MMR, HIB
15–18 mo	DTaP or DTP, OPV, VAR
4–6 yr	DTaP or DTP, OPV
11–12 yr	MMR
14–16 yr	Td (repeat every 10 yr)
	VAR (if susceptible, 2 doses 1 month apart)

An oral rotavirus vaccine is to be added to the infant vaccine schedule at ages 2, 4, and 6 months.

Immunization Modifications for Children with HIV

1. Use only IPV; no OPV (same for household contacts).
2. Add PnV at age 2 years; booster 3 to 5 years later (a new conjugate PnV is in advanced stages of testing).
3. Varicella vaccine should not be used.
4. Severely immunocompromised patients should not receive MMR.

Children immunosuppressed for causes other than HIV should be immunized only in consultation with their primary or subspecialty physician.

Abbreviations:

OPV—oral polio vaccine.
IPV—inactivated polio vaccine.
DTaP—diphtheria, tetanus, acellular pertussis.
DTP—diphtheria, tetanus, pertussis.
Td—full tetanus toxoid dose; half diphtheria dose.
MMR—measles, mumps, rubella.
HIB—Haemophilus b conjugate vaccine.
PnV—pneumococcal vaccine.
Hep B— hepatitis B.
VAR—Varicella vaccine.

(Adapted from Report of the Committee on Infectious Diseases of the American Academy of Pediatrics. 24th ed. Georges P, ed. Elk Grove Village, IL. 1997.)

Recommendations for immunization upon exposure to tetanus, rabies, meingococcemia, hepatitis A and B, and varicella are defined in the Prophylaxis section of these Practical Information points (pp. 1946–1949).

BEDSIDE LABORATORY TESTING

Rapid Screening Test for Cold Agglutinins

Collect a few drops of blood in a purple-top tube (small test tube with about 0.2 mL of 3.8 NaEDTA). Place in ice water bath for about 60 seconds. Tilt tube and look for flocculation in the blood as it starts up the side of the tube. Warm the tube to room temperature and see if the flocculation disappears. Presence of flocculation when observed by the naked eye with subsequent disappearance on warming is a positive test for cold agglutinins and equates with about a 1:64 or greater cold agglutinin titer.

Apt-Downey Test for Fetal Blood

If a stool specimen passed by a neonate is grossly bloody, mix a small sample of the specimen in a test tube with an equal quantity of tap water. Centrifuge briefly or else filter out the solid material. The supernate should have a pink color due to the suspended blood. Add 1 part of 1.0% NaOH to 5 parts of the supernate. Read in 2 minutes. A persistent pink color indicates presence of fetal hemoglobin because fetal hemoglobin is resistant to alkali denaturation; if the supernate turns yellow, the hemoglobin is adult and therefore is probably swallowed maternal blood.

Stool Examination for Leukocytes

Place a small specimen of stool on a glass slide and smear a bit. Mix with a drop or two of methylene blue stain. Cover

with thin cover slip. Examine under microscope in about 3 minutes. Presence of a moderate to profuse number of polymorphonuclear leukocytes is suggestive of a specific bacterial cause of diarrhea.

Gram Stain

Make a thin smear of blood, spinal fluid, or vaginal or urethral secretion on a glass slide. Allow to air dry. Pour on gentian violet for 1 minute and wash with water. Then pour on iodine solution for 1 minute and wash. Decolorize by applying acetone/alcohol for a few seconds and wash. Stain with safranin for 15 seconds and wash. Examine under high-power microscope.

Tzanck Preparation

Denude a vesicular or bullous lesion; blot it and scrape the base with the scalpel edge. Spread the scraped material onto a glass slide and fix with methyl alcohol. Stain for 30 seconds with Wright stain. Wash. Look for multinucleated giant cells indicative of herpetic simplex or zoster lesions.

Pinworm Evaluation

1. Place a piece of cellophane over the end of a tongue stick with sticky side out. Press over the perianal mucosa with moderate pressure for about a minute. Spread the sticky side of the tape over a glass slide. Look through the microscope for ova.

or

2. Instruct the parents to turn over the child who has been asleep for about an hour and look carefully at the perianal area. Live, threadlike worms, about 1 cm in length can be seen wiggling out of the anus to lay their ova.

Methemoglobin Screening Test

To evaluate the apparently cyanotic patient who has no cardiac or respiratory impairment and does not respond to oxygen, place a drop of blood on filter paper. Wave it in the air for 60 seconds. Blood without methemoglobin will remain red or bluish, whereas blood with methemoglobin will turn chocolate-brown.

ELECTROCARDIOGRAPHIC CAVEATS

In young children, the right ventricle normally extends to the right of the sternum, as can be seen graphically on an anteroposterior chest radiograph. Because of this, an electrocardiogram in children less than 5 years of age must include a chest lead taken on the right side of the chest at a point analagous to the left-sided V4 lead. This lead is called V4R. Occasionally, if a heart is grossly enlarged and extends well to the right of the sternum, a V6R and even a V7R lead must be taken to make a complete tracing that properly displays right ventricular potentials.

The right ventricle is normally the dominant ventricle in young children. Right axis is normal, and aVR usually has a dominant R wave in its QRS complex. The QRS progression across the chest leads usually goes from dominant R wave in V4R through the transitional zone to dominant R wave again in the left-sided chest leads. This may be true until as late as 4 years of age.

On the right-sided chest leads and in extremity lead III, T waves are usually normally inverted in young children.

In determining whether a QT interval is prolonged, of particular importance in evaluating the patient with syncope and fainting, the formula to use is as follows:

$$\text{Corrected QT (QTc)} = \text{Measured QT (in fractions of a second)} \div \sqrt{\text{R-R interval (in fractions of a second)}}$$

QTc should not exceed:
- 0.45 in young infants
- 0.44 in older infants and children
- 0.43 in adolescents and adults

FLUID AND ELECTROLYTE AIDES:

A millimole is the atomic weight expressed in milli grams.
Equivalents are the number of electric charges per liter or the atomic weight divided by valence.
A milliequivalent is the equivalent weight expressed in milligrams.

Serum osmolality is calculated with the formula:

$$2(\text{Na}) + \text{Glucose (mg/dL)} \div 18 + \text{BUN (mg/dL)} \div 2.8$$

Normal range is about 285–295 mOsm/L.

Anion gap is the difference between measured cations and measured anions in the serum. Practically, it is:

$$(Na + K) - (Cl + bicarb)$$

Normal value is about 10–15 mEq/L.

Maintenance 24-hour fluid requirements for children:
For the first 10 kg, 100 mL/kg
For the second 10 kg, 1000 mL + 50 mL/kg
Beyond that, 1500 mL + 20 mL/kg

Maintenance electrolytes:
sodium, 2–3 mEq/kg/day
potassium, 1–2 mEq/kg/day
chloride, 2 mEq/kg/day

Total body water (as percentage of body weight):
80% at birth
70% at 6 mo
60% at 1 yr

Two-thirds is intracellular fluid and one-third extra cellular.

SENSORY NERVE DERMATOMES:

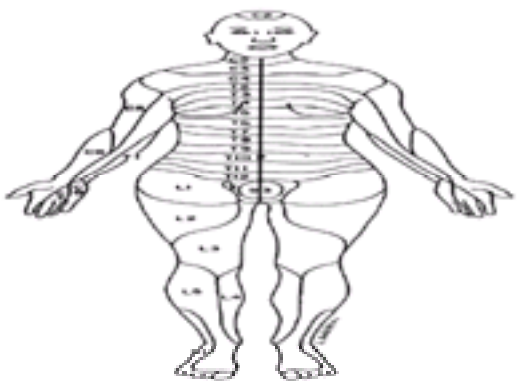

FIGURE D.2. Anterior aspect. (Adapted from Athreya BH, Silverman BK. Pediatric Physical Diagnosis. Norwalk, CT: Appleton-Century-Croft, 1985.)

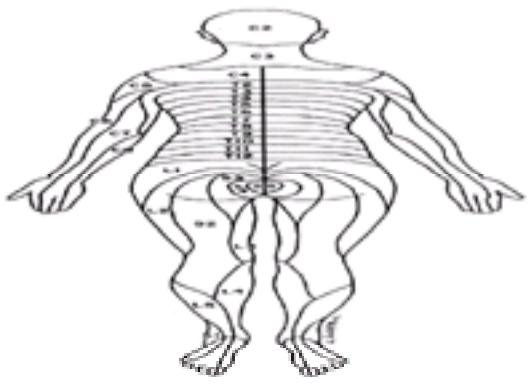

FIGURE D.3. Posterior aspect. (Adapted from Athreya BH, Silverman BK. Pediatric Physical Diagnosis. Norwalk, CT: Appleton-Century-Croft, 1985.)

PROPHYLAXIS AFTER DISEASE EXPOSURE

Tetanus Exposure

Exposure to *Clostridium tetani,* the causative organism for tetanus (lockjaw), occurs primarily through wounds incurred at a site contaminated by human or animal excreta. The first step in prophylaxis involves thorough cleaning and irrigation of potentially contaminated wounds. A decision about which, if any, immunization entity should be ordered is based on the nature and severity of the wound and on the prior tetanus immunization record of the patient. The following table serves as guide:

Prior Tetanus Toxoid Immunization (Doses)	Clean Minor Wound	All Other Wounds
Uncertain (or less than 3 doses)	DTaP or DTP or Td	DTP or Td & TIG or TAT
3 or more (most recent more than 10 yr ago)	Td	Td
3 or more (most recent within past 5 yr)	None	None

3 or more (most recent between 5–10 yr)	None	Td

DTP, diphtheria, tetanus, pertussis toxoid;*DTaP,* diphtheria, tetanus, acellular pertussis;*Td,* adult formulation of diphtheria, tetanus toxoid; *TIG,* tetanus immunoglobulin (dose: 250–500 units intramuscularly); *TAT,* tetanus antitoxin; should be used only if TIG is not available and only after testing (dose: 3000–5000 units intramuscularly).

Rabies Exposure:

Rabies exposure occurs on being bitten by an animal carrying the rabies virus in its saliva. The most common animal reservoirs include bats, skunks, raccoons, foxes, and woodchucks. Dogs and cats, when bitten by these animals, carry the virus in their saliva before becoming symptomatic and may transmit it to humans by biting or abrading. Rarely, small rodents and rabbits transmit the disease. In recent years, more than half the 36 cases of rabies diagnosed in the United States have been associated with bat variants of the rabies virus, in many cases with no clear history of a bite.

The decision about when to institute prophylaxis after an exposure must be guided by the location and severity of the wound, the status of the offending animal if known, and knowledge of the local epidemiology. In northeastern United States, for instance, rabies is prevalent in the raccoon population, with resultant fear of infection in the dog and cat population. If a patient has heavy exposure to bats in the home or workplace, even in the absence of a history of bite, prophylaxis with human diploid cell vaccine (HDCV), should probably be initiated.

Prophylaxis should begin with thorough cleaning and irrigation of the wound. HDVC, although expensive, is relatively free of side effects.

Active Immunization

Five doses of HDCV, 1 mL each, should be given intramuscularly on days 0, 3, 7, 14, and 28 after exposure. Persons who have been previously immunized can be given just two doses—one on day 0 and one on day 3.

Passive Immunization

In addition to HDCV, rabies immunoglobulin (human) should be given to patients who have had no prior immunization in a dose of 20 units/kg on day 0 of exposure. Half should be given IM and half infiltrated around the wound.

Pre-exposure prophylaxis for those traveling to or working in endemic areas consists of 3 injections of either HDCV or rabies vaccine absorbed (RVA) given on days 0, 7, and 28.

***Haemophilus influenzae* b Exposure**

Recommendations for use of rifampin on exposure to illness caused by *Haemophilus influenzae* b:

1. For all household contacts in which at least one of the contacts is less than 12 months of age and has not received the 12-month booster dose of Hib. For all household contacts in which those less than 48 months have not completed their Hib immunization series.
2. In day-care homes with children less than 2 years of age, in which contact is 25 hours a week or more, regardless of attendee's immunization status.
3. All attendees in day-care centers in which two or more cases have occurred within 60 days, regardless of the child's immunization status.
4. All household contacts in which an immunocompromised child lives, regardless of immunization status.
5. To ensure complete eradication of the organism from the pharynx, the index case should receive rifampin if the patient had been treated with ampicillin and chloromycetin.
6. Should not be given to pregnant women.
7. Provide a warning that urine may turn red and contact lenses may stain.

Dosage: rifampin, 20 mg/kg/day orally every day, to a maximum of 600 mg for 4 days.

Meningococcal Infection Exposure

All those who have had contact with the patient's oral secretions, including household, day-care center, and nursery school contacts and intimately exposed (intubaters, mouth-to-mouth) medical personnel, should receive antibiotic or chemical prophylaxis as soon as possible.

Drug Options and Dosage:

Rifampin—10 mg/kg/day to a maximum of 600 mg/day, divided in two doses for a total of 2 to 4 days (infants less than 1 month, 5 mg/kg/day); not for pregnant women; urine and contact lenses may turn red.

Ceftriaxone—has been evaluated for group A meningococcal strains only but is probably effective for all strains—single intramuscular dose of 125 mg for ages less than 12 years and 250 mg for ages 12 and older.

Ciproflaxin—for nonpregnant contacts 18 years and older. A single oral dose of 500 mg.

Sulfisoxazole—when the isolate is known to be sensitive to sulfa; 0.5 g/day for age less than 1 year; 0.5 g every 12 hours for ages 1 to 12 years; and 1 g every 12 hours for ages more than 12 years—total 2 days.

Meningococcal polysaccharide vaccine (groups A, C, Y, and W-135)—currently given to all military recruits, can be

considered in cases when ongoing exposure is likely. Has inconsistent effectiveness in infants but can be given to children 2 years and older in high-risk categories, including asplenia. Can be used as an adjunct to chemoprophylaxis. The vaccine does not include the B strain of meningococcus, which is the most common in the United States.

Measles (Rubeola) Exposure:

Within 72 hours of exposure: Live virus measles vaccine (to be repeated at appropriate age—15 months and again at 11 to 12 years for those between 6 months and 1 year; at 11 to 12 years for those older than 1 year). Children less than 6 months at the time of exposure should receive immunoglobulin (0.25 mL/kg intramuscularly).

More than 72 hours up to 7 days after exposure (or for those in whom time of exposure is unclear, as in some group home or shelter residents, when a shower of cases are appearing): Immunoglobulin (0.25 mL/kg) followed in 6 months by live virus measles vaccine.

Immunodeficient children: Immunoglobulin (0.5 mL/kg). Subsequent immunization with live virus vaccine will vary with the nature of the immunodeficiency and should be determined by the physician providing ongoing care.

Varicella Exposure

Varicella-zoster virus (VZV) is highly contagious. The vaccine is not currently recommended for preventing varicella in the exposed individual. Its use, however, is essentially free of risk and may or may not prevent clinical illness if given within 3 days of exposure.

The only guaranteed protection for the contact at this time is provided by varicella-zoster immunoglobulin (VZIG). Its use should be confined to those who have been exposed and who are at high risk of developing complications of varicella, particularly premature infants weighing less than 1000 g and others who are immunosuppressed or immunodeficient. Pregnant women with no history of varicella can be evaluated on exposure and considered for VZIG. Larger prematures and term neonates born to mothers who develop the disease within 5 days before or 2 days after delivery should also be given VZIG. Infants born to mothers given VZIG during the last few days of pregnancy should be given VZIG, even if the mother did not develop the disease.

With the advent of acyclovir as an effective treatment for varicella, it is not necessary to give VZIG to the healthy susceptible adolescent or adult who is exposed to the disease. Acyclovir therapy can be initiated shortly after the disease erupts. However, acyclovir has no role in prevention of varicella in an exposed individual.

The dosage of VZIG is 125 units (1.25 mL intramuscularly) per each 10 kg or less, up to a maximum of 625 units (6.25 mL).

Rubella Exposure

Rubella is a mostly benign disease except to the fetus of the pregnant woman. Antibody status should be determined before pregnancy and vaccine administered at that time to those women at risk. Immunoglobulin is of questionable value in rubella but should be administered to those who have serologic evidence of having developed the disease during pregnancy and who choose to continue the pregnancy. Congenital rubella may occur in the fetus despite administration of immunoglobulin to the mother. No other indication for prophylaxis against rubella exposure exists.

Mumps Exposure

Immunoglobulin is of no value. Live virus vaccine should be administered to susceptible adults when exposed to mumps.

Hepatitis A Exposure

Hepatitis A is foodborne and waterborne and is also transmitted by face-to-face personal contact. The hygienic measure of careful handwashing for all medical, day-care center, and restaurant personnel should be followed at all times. Potentially contaminated food and water should be avoided.

The hepatitis A vaccine is not recommended for postexposure prophylaxis. Household and sexual contacts, institutional residents, and day-care workers in contact with an index case can be protected with low-dose immunoglobulin (0.02 mL/kg intramuscularly) given within 14 days of exposure.

Individuals traveling to potentially endemic areas can be protected by immunoglobulin doses of 0.02 to 0.06 mL/kg, with the higher dosages recommended for longer stays.

Hepatitis B Exposure

Hepatitis B virus (HBV) is transmitted by exposure to contaminated blood through wounds, needle exposure, medical management, improperly prepared transfusion, and sexual activity and maternal–child transmission.

Universal administration of hepatitis B vaccine is begun in infancy. Medical and institutional personnel and others at high risk of exposure also should be vaccinated with hepatitis B vaccine. Four doses are given at intervals of 0, 1, 2, and 6 to 12 months. The dose is either 1.0 mL or 0.5 mL intramuscularly, depending on the patient's age and the vaccine used. The dose should be doubled for immunosuppressed patients.

Immunization can be started at the time of exposure, but previously nonimmunized individuals should receive

simultaneously with the first dose of vaccine a dose of hepatitis B immunoglobulin (HBIG) (0.06 mL/kg) (minimum dose: 0.5 mL). This treatment does not appear to interfere with the response to the vaccine.

Neonates born to hepatitis B surface antigen–positive mothers should receive HBIG (0.5 mL) and then begin a vaccination schedule by day 7.

Hepatitis C Exposure

Hepatitis C is parenterally transmitted. No vaccine is available. Immunoglobulin appears to be of equivocal value and is not recommended.

For additional information, refer to Report of the Committee on Infectious Diseases. American Academy of Pediatrics. Georges P, ed. Elk Grove Village, IL: American Academy of Pediatrics, 1997.

HIV Occupational Exposure:

Medical and dental personnel (MDP), particularly those working in emergency departments and in the various surgical and laboratory areas, are susceptible to inadvertent exposure to the human immunodeficiency virus (HIV). This may occur 1) percutaneously (needle stick or scalpel wound); 2) by contact of MDP skin or mucous membranes with potentially contaminated body fluid such as blood, semen, or vaginal secretions; or 3) by direct MDP skin or mucous membrane contact with laboratory specimens. If MDP skin is previously irritated or abraded, the risks under 2) and 3) are increased.

The first step in prophylaxis is always, when possible, prevention of exposure. This is best accomplished by MDP by carefully following the guidelines of appropriate handwashing, use of gloves and proper cleaning and draping for any procedure that may involve exposure, careful disposal of all needles and other disposable instruments and syringes, effective sterilization of all reusable equipment and devices, and careful monitoring of all the foregoing by trained infection control personnel in institutional and office settings.

When exposure does occur to MDP, careful cleaning of the wound area is essential. Relevant details of the exposure should be recorded in a confidential record, including date and time, job duty being performed, full details of the circumstances and nature of the exposure, and description of the source of the exposure material. Ongoing record should include details of counseling, postexposure management, and follow-up of the MDP and the individual who was the source of the exposure material.

Hepatitis B prophylaxis should be considered as described previously.

With permission, HIV testing should be performed on the source individual and, if possible, repeated periodically for 6 months. The exposed MDP should undergo HIV testing at the time of exposure. If the source individual for the exposure material is HIV positive, becomes HIV positive, or refuses to be tested, the MDP should be retested at periodic intervals up to 6 months. Seroconversion usually occurs within 3 months of exposure.

Consideration should be given to initiation of postexposure prophylaxis for the MDP when the source individual is known to be, or is at high risk for being, HIV positive. Two antiretroviral drugs, at least one of which should be zidovudine, should be included in the regimen. A protease inhibitor should also be added for the higher exposure risks. The potential toxicity of the drug regimens should be balanced against the degree of risk imposed by the exposure to the MDP. If prophylaxis is to be initiated, it should be done so within a few hours after exposure. A decision must be made by the MDP and personal physician after adequate and appropriate counseling and consideration of all the factors involved in weighing risk versus toxicity. (Adapted from the MMWR—Morbidity and Mortality Weekly Report—of the Centers for Disease Control and Prevention; 1996;45:468–472.)

SCORES

Glasgow Coma Scale

Eye Opening	
Spontaneous	4
To speech	3
To pain	2
None	1
Best Motor Response	
To verbal command	
Obeys	6
To painful stimulus	
Localizes	5
Flexion withdrawal	4
Flexion decorticate	3
Extension decerebrate	2
No response	1
Best Verbal Response[a]	
Oriented and converses	5
Disoriented; converses	4
Inappropriate words	3
Incomprehensible sounds	2
No response	1

[a]Children less than 2 years of age should receive full verbal score for crying after stimulation.

AVPU

A—alert

V—responds to vocal stimuli

P—responds to painful stimuli

U—unresponsive

Revised Trauma Score

Attribute	Coded Value
Respiratory Rate	
10–29	4
>29	3
6–9	2
1–5	1
0	0
Systolic Blood Pressure	
>89	4
76–89	3
50–75	2
1–49	1
0	0
Glasgow Coma Scale	
13–15	4
9–12	3
6–8	2
4–5	1
3	0
Unweighted Revised Trauma Score:	

Adapted from Champion HR, Sacco WJ, et al. A Revision of the trauma score. J Trauma 1989;29:623.
NOTE: Of value in determining whether to triage to a trauma center. Higher scores are generally associated with less urgency and better outcomes.

Pediatric Trauma Score

Component	+2	+1	-1
Size	>20 kg (40#)	10–20 kg	<10 kg
Airway	Normal	Maintainable	Unmaintainable
Systolic blood pressure	>90 mm Hg	50–90 mm Hg	<50 mm Hg
Central nervous system	Awake	Obtunded/loss of consciousness	Coma/decerebrate
Skeletal	None	Closed fracture	Open/multiple fractures
Cutaneous	None	Minor	Major/penetrating

Adapted from Tepas JJ, Mollitt DL, et al. The Pediatric Trauma Score as a predictor of injury severity. J Pediatr Sur 1987;22:14.

Assign a value to each of the six components. A score of 8 or less may suggest that care should be provided in a pediatric trauma center.

APGAR Score

Sign	0	1	2
Heart rate	Absent	Below 100	Over 100
Respiratory effort	Absent	Slow, irregular	Good, crying
Muscle tone	Limp	Some flexion of extremities	Active motion
Response to catheter in nostril (tested after oropharynx is clear)	No response	Grimace	Cough or sneeze
Color	Blue, pale	Body pink, extremities blue	Completely pink

To be checked on each newborn at 1 minute and again at 5 minutes after completion of birth.

Trauma Protocol

To be done simultaneously under direction of a leader and including a recorder if sufficient personnel are available.

Primary Survey and Resuscitation

“A” AIRWAY—positioning; stabilization of C-spine

“B” BREATHING—midline trachea; subcutaneous air; breath sounds; open pneumothorax; airway obstruction; tension pneumothorax; hemothorax; flail chest; gastric distension; oxygen; bag-valve-mask or intubation as indicated

“C” CIRCULATION—hemorrhage; peripheral pulses; heart sounds; capillary refill; jugular vein distension; vascular access (venous; intraosseous); bloods for lab, including type and crossmatch

“D” DISABILITY—pupils; level of consciousness; AVUP; Glasgow Coma Scale and/or Revised Trauma Score; other cranial nerves if possible

“E” EXPOSURE—open wounds, front and back

“F” FOLLOW—monitors; rhythm strip; catheter; G-tube

“G”—blood gases if indicated

“H” HISTORY—preliminary

Secondary Survey

- Complete physical examination, including cranial nerve check, fundal examination, careful neurologic screening, repeated check of vital signs, abdominal examination, check for blood from penis; rectal examination, and evaluation of all open wounds
- Complete history of the current episode and medical history if possible
- C-spine film, chest radiograph, abdominal and pelvic films; consideration of emergency ultrasound and/or computed tomography scan
- Careful splinting and wound dressing as indicated

SECTION II Signs and Symptoms

Fred M. Henretig, MD, Section Editor

Suggested Readings

In this section, the chapters focus on the problem-oriented approach to the rapid diagnosis and initial management of patients coming to the emergency department (ED) with specific signs or symptoms as chief complaints. The particular presenting complaints covered in the following chapters have been chosen because they may result from serious diseases, requiring urgent diagnosis and therapy, or because they are quite common.

In each chapter, the sign or symptom is defined, and in most cases, there is a brief review of underlying pathophysiology. The chapter then concentrates on a realistic diagnostic approach that is applicable in the ED. These chapters are supplemented in most cases by a table that lists the common causes of each sign or symptom and an algorithm that outlines the key steps in sorting out the differential diagnosis.

The chapters in this section mesh closely with those in the medical, surgical, and psychosocial sections that are organized around specific diseases. Thus, there will be many cross-references in Chapter 28, Fever, and to specific topics in Chapter 84, Infectious Disease Emergencies. These cross-references guide the reader to text that discusses details of the evaluation for the treatment of specific infectious diseases, once the likely diagnosis has been established. Brief comments on symptomatic therapy are included only if specific treatments are not discussed elsewhere in the text.

Entire textbooks have been devoted to the differential diagnosis of pediatric signs and symptoms; the spectrum of childhood disease demands such lengthy works. However, the few rapidly progressive, life-threatening causes of each complaint sometimes may become obscured in an all-inclusive detailed discussion by the information presented on less urgent conditions. Thus, the chapters in this section emphasize the serious conditions of greatest concern to the emergency physician or the pediatrician in an ED. Readers interested in pursuing broader coverage of particular pediatric signs and symptoms are referred to the invaluable works by Illingworth, Green, and Tunnessen.

Suggested Readings

Green M. Pediatric Diagnosis. 2nd ed. Philadelphia: WB Saunders, 1985.

Illingworth RS. Common Symptoms of Disease in Children. Oxford: Blackwell, 1979.

Tunnessen WW. Signs and Symptoms in Pediatrics. Philadelphia: JB Lippincott, 1988.